**MEDICAL UNIVERSITY OF GRAZ**

**Master of Science in Dermoscopy and Preventive Dermato-Oncology**

# DERMOSCOPY OF POIKILODERMA OF CIVATTE

## THESIS

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## Statutory declaration

I declare that I have written this work independently and without assistance other than those specified sources, and have not used sources or means without declaration in the text. The Master Thesis was not used in the same or in a similar version to achieve an academic grading.“

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# Table of Contents

## Summary

**Introduction:** Poikiloderma of Civatte (PC) is a rather common, acquired, chronic dermatosis, most often affecting fair-skinned individuals, especially menopausal females. It is characterized by a combination of linear telangiectasia, mottled hyperpigmentation, and superficial atrophy in a reticular pattern. It is typically located on the V, the sides of the neck, and the peripheral face, invariably sparing the anatomically shaded areas. It runs a chronic, slowly progressive, but indolent course, producing only cosmetic disfigurement. Genetic predisposition; chronic cumulative sun exposure; hormonal changes of menopause (including iatrogenic menopause); and contact sensitization to perfumes and cosmetics, have all been incriminated in its obscure pathogenesis.

**Material and Methods:** Twenty-eight consecutive patients, clinically diagnosed with PC, were included in the study. All patients were evaluated by detailed history, clinical examination, and dermoscopic examination with hand-held dermatoscope and video-dermatoscope. Dermoscopic findings i.e. global patterns, local and additional features, have been recorded and analyzed.

**Results:** In total, 28 patients with PC were recruited. The median age was 55 years (range 26-73 years). There were 19 females (67.86%) aged 46-73 years (median 54 years), and 9 males aged 26-65 years (median 59 years). Six patients (21.43%) had erythemato-telangiectatic PC, two (7.14%) had the pigmented type, and twenty (71.43%) had the mixed type. On dermoscopic examination, the red-white polka dot pattern was observed in 10 cases (35.71%); the fishnet-like pattern in 15 cases (53.57%); the non-specific pattern in 9 cases (32.14%); the spaghetti and meatballs-like pattern in 4 cases (14.28%). In most cases (78.57%), the coexistence of more than one pattern at different sites was noted. As far as local dermoscopic features are concerned, we observed flying seagull-like vessels in 18 cases (64.29%); rhomboidal/polygonal vessels in 15 cases (53.57%); linear irregular vessels in 17 cases (60.71%); dotted/globular vessels in 10 cases (35.71%); white macules in 23 cases (82.14%); brown macules in 11 cases (39.29%); and whitish follicular plugs in 6 cases (21.42%). On videodermoscopy, we could identify the same local features, i.e. the various types of telangiectatic vessels and pigmented structures. Brown macules, in particular, could be readily recognized much more efficiently than with classic

dermoscopy. White macules could be appreciated as the holes of the vascular network or areas of sparing.

**Conclusions:** The dermoscopic picture of PC is highly characteristic and corresponds well to both clinical and histological findings. Although the diagnosis of PC is made on clinical grounds, dermoscopy may assist, in doubtful cases, the differentiation from other clinically important forms of poikiloderma, such as poikiloderma atrophicans et vasculare and poikiloderma associated with collagen-vascular disease, as well as from benign dermatoses of the face and neck. In addition, dermoscopy provides further support to the speculation that PC and erythemato-telangiectatic rosacea, may be related and may represent variants in the same nosological spectrum.

**Keywords:** poikiloderma, dermoscopy, videodermoscopy, epidemiology, histopathology, rosacea.

# Zusammenfassung

**Einführung:** Die Poikilodermie von Civatte (PC) ist eine ziemlich häufige, erworbene, chronische Dermatose, die öfter Menschen mit einem hellen Hauttyp betrifft, vor allem Frauen in der Postmenopause. Sie zeichnet sich durch eine Kombination linearer Teleangiektasie, punktförmiger Hyperpigmentierung und eine oberflächliche Atrophie in einem Netz-förmigen Muster aus. Typisch tritt sie in V-Form auf, anbeiden Seiten des Halses und in den seitlichen Gesichtsbereichen, wobei die anatomischen, nicht im Licht ausgesetzten Gebiete nicht betroffen sind. Sie hat einen chronischen, langsam progressiven Verlauf, indem sie nur ästhetische Probleme verursacht. Genetische Disposition, chronische, kumulative Strahlenexposition, Hormonveränderungen durch die Menopause (einschließlich einer iatrogene Menopause) und Kontaktsensibilisierung mit Parfüm und Kosmetika sind als die wahrscheinlichen Ursachen ihrer ungeklärten Pathogenese zu betrachten.

**Materialien und Methoden:** 28 Patienten mit Diagnose einer Poikilodermie von Civatte wurden in die Studie eingeschlossen. Alle Patienten wurden durch ausführliche Anamnese, klinische Untersuchung und dermatoskopische Untersuchung mit einem Hand-Dermatoskop und einem Video-Dermatoskop evaluiert. Die dermatoskopischen Befundewurden bezüglich Muster-, Lokal- und Zusatzmerkmaleanalysiert und ausgewertet.

**Ergebnisse:** 28 Patienten wurden in die Studie eingeschlossen. Das Durchschnittsalter war 55 Jahre (26 bis 73 Jahre). 19 Frauen (67,86%) im Alter von 46 bis 73 Jahre (durchschnittlich 54 Jahre) und 9 Männer im Alter von 26 bis 65 Jahre (durchschnittlich 59 Jahre) haben daran teilgenommen. Sechs Patienten (21,43%) hatten den erythematös-teleangiektatischen Typ der Poikilodemie von Civatte, zwei Patienten (7,14%) hatten den pigmentierten Typ und zwanzig (71,43%) den gemischten Typ. Bei der dermatoskopischen Untersuchung erschien das rot-weiße polka dot Pattern in 10 Fällen (35,71%), das «Fischnetz» Pattern in 15 Fällen (53,57%), das unspezifische Pattern in 9 Fällen (32,14%), das *"spaghetti and meatballs" Pattern in* 4 Fällen (14,28%). In den meisten Fällen (78,57%) wurden mehrerer Muster gleichzeitig beobachtet.In Bezug auf die lokalen dermatoskopischen Befunde haben wir "Seemöwe-artige" Gefäße in 18 Fällen (64,29%) beobachtet, rhomboide/polygonale Gefäße in 15 Fällen (53.57%), linienförmige,

irreguläre Gefäße in 17 Fällen (60,71%), gepunktete/ kugelförmige Gefäße in 10 Fällen (35,71%), weiße Makulae in 23 Fällen (82,14%), braune Makulae in 11 Fällen (39,29%) und weiße Follikel-Hornstacheln in 6 Fällen (21,42%). Bei der Video-Dermatoskopie haben wir die gleichen lokalen Befunde i.e. die verschiedenen Typen von teleangiektatischen Gefäßen und pigmentierten Strukturen beobachtet.Im Vergleich zu der klassischen Dermatoskopie waren besonders die braunen Makulae besser zu erkennen. Die weißen Makulae wurden als Löcher innerhalb des Gefäßnetzes in Gebieten, die nicht dem UV-Licht ausgesetzt sind, lokalisiert.

**Resultate:** Die dermatoskopischen Befunde der Poikilodermie von Civatte sind besonders typisch und entsprechen sowohl den klinischen als auch den histologischen Befunden. Obwohl die Diagnose anhand der klinischen Befunde durchgeführt wird, kann die Dermatoskopie bei zweifelhaften Fällen in der Differenzialdiagnose zu anderen, klinisch wichtigen Varianten der Poikilodermie helfen. Diese Varianten umfassen die Poikiloderma atrophicans et vasculare,diePoikilodermie, die mit Kollagen- und Gefäßerkrankungen und auch Poikilodermien, die mit anderen, gutartigen Hautkrankheiten des Gesichts- und Halsbereiches im Zusammenhang stehen. Zusätzlich bietet die Dermatoskopie Daten, auf die die Hypothese stärkt, dass die Poikilodermie von Civatte und die erythematös-teleangiektatische Rosazea vielleicht miteinander zusammenhängen und Varianten des gleichen, nosologischen Spektrums darstellen.

**Schlüsselwörter:** poikiloderma, dermoscopy, videodermoscopy, epidemiology, histopathology, rosacea.

# Introduction

Poikiloderma of Civatte (PC) is a rather common benign dermatosis of the neck and face, mainly affecting postmenopausal females. It is characterized by a combination in a reticular pattern of linear telangiectasia, mottled hyperpigmentation and superficial atrophy.[1,2] The aetiopathogenesis of PC is complex and it is incompletely understood. Exposure to ultraviolet radiation; hormonal changes of menopause; contact sensitization to perfumes and cosmetics; and normal ageing, have been incriminated. The course is slowly progressive and irreversible, often causing significant cosmetic disfigurement. The diagnosis is usually clinical and can be confirmed by histology, which is characteristic, but not pathognomonic. Differential diagnosis is wide and includes, among others, poikiloderma atrophicans et vasculare and poikiloderma associated with collagen vascular disease, i.e. conditions with guarded prognosis.[1,2]

Dermoscopy is an in vivo diagnostic technique that uses a light source of polarized or non-polarized light and a magnifying glass, thus permitting the visualization of colors and structures invisible with the unaided eye.[3] Following its successful use for the diagnosis of malignant melanoma, and other benign and malignant skin tumors, dermoscopy has been also applied in hair and nail disorders; inflammatory dermatoses; skin infections and infestations and, practically, in every disease in the context of General Dermatology.[4,5] Until the day of writing, very few have been published regarding dermoscopy of PC. Aim of the present study was to describe the dermoscopic findings of PC in an attempt to establish a dermoscopic confirmation of the usually clinical diagnosis, thus reducing the need for biopsy as a means of differentiation by histologic documentation.

## Definition

The term poikiloderma refers to a combination of linear telangiectasia, mottled hyperpigmentation or depigmentation, and superficial atrophy in a reticular pattern.[6] Poikiloderma may have various aetiologies and can be either acquired or congenital.[7] Poikiloderma of Civatte (PC) is a common, idiopathic, acquired poikiloderma of the neck and face that runs a chronic, benign and irreversible course, producing only significant

cosmetic disfigurement.[8]

## History

PC was originally described as a distinct entity in 1923 by the French dermatologist Achilles Civatte under the name «poikilodermie reticulee pigmentaire du visage et du cou» (reticular pigmented poikiloderma of the face and neck).[9] Earlier (1906) the American paediatrician A. Jacobi was the first to describe a form of acquired poikiloderma that he named «poikiloderma atrophicans et vasculare». In his original description, Civatte reported three female cases with reticular patches of hyperpigmentation, telangiectasia (erythema), and atrophy on parts of the face and neck. He differentiated this benign condition of poikiloderma from Jacobi's poikiloderma. Actually, one of Civatte's cases was initially described by Jacobi as poikiloderma atrophicans et vasculare. Civatte was the first to notice that this condition affects mostly menopausal females. He gave a detailed description of the histological findings, and suggested that this condition may be related to adrenal insufficiency, as his patients improved after treatment with adrenal extract. For this reason, he proposed the alternative name «adrenal poikiloderma» for his disease.[9] In 1928, E.M. Graham-Little, a British Dermatologist, published a study that was in agreement with the considerations made by A. Civatte, and proposed the disease to be named after the one who described it, i.e. «Civatte's disease».[10] Graham-Little was the first to highlight the distribution in sun-exposed areas of the face and neck, suggesting a pathogenetic role for solar radiation. During the 1920's and the 1930's, there were several cases of reticular pigmentation of the face and neck that were discussed during the meetings of Dermatological Societies. However, most of these cases do not correspond to what today is known as PC. Since 1917, another entity, Riehl's melanosis, had been described, adding confusion to the differential diagnosis.[11] In 1938, a detailed review of the literature was published by the Argentinians Pierini and Bosq.[12]They reviewed all 43 published cases in the international literature, and added 4 more cases. They discussed extensively epidemiology, clinical presentation, histology and differential diagnosis of Civatte's disease. Since then there was paucity of articles for PC in the literature. In 1989, PC was re-introduced by R. Graham in his classical article entitled «What is poikiloderma of

Civatte».[8] With the advent of laser therapies for vascular lesions, several articles appeared in the literature concerning the use of various light sources in the treatment of PC. In the 2000's, the pathophysiology, genetics, epidemiology, clinical presentation and histopathology of PC were revisited by Katoulis and co-workers.[13-16]

## Epidemiology

*Incidence and Prevalence*

All authorities agree that PC is a common skin condition. Nevertheless, data concerning the exact prevalence of the disease is lacking. In many case the disease is mild and patients do not seek medical advice. In Greece, an estimated prevalence of 1.4% among 3976 dermatologic patients has been reported.[14] However, the true prevalence must be higher, especially in white populations living in low latitudes with many sunshine hours yearly.

***Race***

PC has been reported only in white individuals. There are no reports of PC occurring in blacks or Asians.

***Age and Sex***

PC most often affects middle-aged or elderly individuals, usually in their 4$^{th}$ to 7$^{th}$ decade.[8,14] In the literature, there are two cases of PC adolescents diagnosed with PC.[12] In a Greek clinico-epidemiological study of 50 cases, the mean age at diagnosis was 47.8 years for females (range 39–68 years), and 61.7 years for males (range 48–74 years).[14] PC affects both sexes, but it is more common in females.[8] In Civatte's original description, the 'reticular pigmented poikiloderma of the face and neck' was observed in women about or during menopause.[9] In the review of all previously reported cases by Pierini and Bosq,[12] there were 44 females among 47 patients in total. In the most recent epidemiological study from Greece, women also predominated (68%).[14] However, men were not uncommonly affected (32%). Interestingly, PC was diagnosed at an older age among males (mean 61.7 years vs. 47.8 years for females), suggesting that cumulative chronic sun exposure may play a more important part in males.[14] It must be noted that PC remains under-reported in

males, because many do not seek medical advice for an only cosmetically disfiguring condition.

### *Skin phototype*

Patients are usually fair-skinned and most often have lighter skin phototypes (I-III). In the Greek study, 88% of the patients had skin phototype II or III.[14]

### *Occupation*

Males who are engaged in outdoor occupations are more commonly affected.[8] In Great Britain, this condition is called the «Berchshire's neck», due to its high prevalence among the inhabitants of this cattle-farming region.[8] In contrast, most female patients with PC have an indoor occupation.

A study from Norway suggested that a relationship might exist between video display terminal work and aggravation of seborrheic dermatitis, acne, rosacea, and probably PC.[17]

### *Associated conditions*

Menopause is the most commonly associated condition. Even in the earliest reports, PC was most often seen in association with menopause, physiologic or iatrogenic. In the Greek cohort, among 34 women with PC, 26 (76.4%) were at a peri-menopausal stage, including 3 cases of iatrogenic menopause (8.8%).[14]

Rosacea is the most commonly associated skin condition. In contrast, non melanoma skin cancer and dermatoheliosis occur much less frequently among patients with PC than one might expect.[14]

## Aetiology and Pathogenesis

The aetiopathogenesis of PC appears to be multifactorial and remains incompletely understood. Several causative factors have been implicated, including long-term exposure to ultraviolet radiation (UVR), hormonal changes of menopause, genetics, and contact delayed hypersensitivity, especially to fragrances and cosmetics.[1,2,18]

### *Sun exposure*

The typical distribution dictates a central role for the UVR. PC strictly affects the sun exposed areas of the neck and face, while the anatomically shaded areas remain invariably spared. The age distribution of the patients most of whom are middle-aged or elderly, indicates that chronic cumulative sun exposure is probably pathogenetically implicated.[1,2,8] The predominance of lighter skin phototypes, who are more vulnerable to the effects of UVR, is also consistent with this hypothesis. Solar elastosis is the sine qua non histological feature of PC, reinforcing the central role of UVR in the development of PC.[16]

### *Genetic factors*

Families with several of their members affected by PC have been reported.[13] In an epidemiological study of 50 patients, there were 4 cases (8%) with family history. The recognition of familial cases, as well as the occurrence of the disease in patients in whom all suspected causal factors are absent, can lead to the speculation that a genetic predisposition to the disease may exist.[13] This predisposition may be expressed as an increased sensitivity of the dermal connective tissue to normal doses of UVR.

Based on the analysis of 7 PC cases among the members of two unrelated Greek families, it has been suggested that, if a genetic basis does exist, the presence of the disease in successive generations but not in all siblings, as well as the occurrence in individuals of both genders, is consistent with an autosomal dominant mode of inheritance with variable penetrance.[13]

### *Hormonal factors*

The predominance of females, most often of peri-menopausal age, as well as the association with the menopause, physiologic or iatrogenic, suggests that hormonal factors, namely low estrogen levels, in combination with the normal ageing process may be involvel.[8,14-16] Hormonal changes of menopause have been associated with vasomotor reactions and flushing, as well as with rapid reduction of bone and dermal collagen.[19] It can be suggested that in a similar way, they may contribute to the development of PC.

***Contact and photo-contact hypersensitivity***

It has long been speculated that photodynamic substances in perfumes and cosmetics possibly induce a photoallergic or a photoxic reaction that triggers the disease process.[8] To test this hypothesis, Katoulis et al patch-tested thirty-two PC patients, 24 females and 8 males, aged 33 to 74 years,using the European standard series and the Fragrance series. Additionally, photo-patch testing with the Photo-allergens series, and phototesting with a monochromator, were performed.[15] Thirteen patients (40.62%) had one or more positive reactions to allergens of the standard series. Eight patients (25%) had positive reactions to fragrance mix and/or Balsam of Peru, or to allergens of the fragrance series. Nickel sulphate was the single most common cause of contact sensitization (18.75%). A statistically significant difference in the frequency of positive reactions to fragrances between the PC group and the control group was documented ($x^2$=3.91, $p$<0.05). In contrast, none of the PC patients had a positive photo-patch test for any of the allergens included in the photo-allergens series. The minimal erythemal dose for the PC group was in all cases within normal limits for all wavelengths of ultraviolet radiation examined.[15]

Based on the positive patch testing results in a post-menopausal woman with PC, Khunkhet et al suggested a a role for methylchloroisothiazolinone/methylisothiazolinone (Kathon) in the development of PC.[20] Interestingly, patient's symptoms of itching and burning, as well as cutaneous changes improved after abstaining from the use of personal care products containing Kathon. Katoulis et al reported the case of 48-year old woman practicing aroma-therapy with poikilodermatous changes in extensor aspects of the forearms, typical PC on the V of the neck, and erythemato-telangiectatic rosacea of the central face. Patch testing revealed positive reactions to Fragrance mix and Nickel sulphate.[21]

Vachiramon and Vattanakrai reported a 49-year-old woman with PC and a positive reaction to 6-methylcoumarin during photopatch-testing with standard photo-allergens.[22] In this case, 6-methylcoumarinwas found in the patient's perfume. This report is the first to provide evidence for a possible involvement of photo-allergic contact sensitization in the pathogenesis of PC. Contact sensitization, mostly to perfume ingredients, may develop in PC and, possibly play a pathogenetic part, at least in a subset of patients. Despite negative

results of photo-patch testing, an allergic photo-contact reaction cannot be definitely excluded.

***Poikiloderma of Civatte and Rosacea***

PC and erythemato-telangiectatic rosacea share common epidemiological, clinical and histological features. On this basis, it has been suggested that PC and rosacea may represent variants in the same nosological spectrum. According to Katoulis et al,[23] in both these conditions, the dermal connective tissue may be a possible primary target, resulting in solar elastosis and leading to telangiectasia. In PC, a secondary target appears to be the basal cell layer leading to melanin incontinence and subsequent mottled hyperpigmentation. In rosacea, the hair follicle may be the secondary target resulting in the inflammatory lesions of papulopustular rosacea. Given their different localization, it can be postulated that local factors, such as the infestation of the hair follicles by *Demodex folliculorum* in rosacea or the application of perfumes on the neck in PC, may determine the differing clinical evolution of these conditions.

Katoulis et al proposed the following histopathogenetic scenario for the development of PC.[16] In genetically predisposed individuals, chronic exposure to UVR produces dermal connective tissue damage (solar elastosis), leading to telangiectasia due to loss of vascular support. These changes of collagen and elastic tissue are most probably augmented in females by the menopausal decrease in the estrogens levels, which affects the rate of collagen production. Chrono-aging offers a favourable setting, contributing to the epidermal atrophic changes. A delayed contact hypersensitivity reaction, probably triggered by perfume and cosmetic ingredients, may cause basal layer changes, resulting in melanin incontinence, clinically expressed as reticulate hyperpigmentation. Thus, the classical triad of poikiloderma that is: telangiectasia, mottled hyperpigmentation and superficial atrophy, is integrated.[14-16]

It remains unclear why PC is preferentially located on the sun-exposed neck and upper chest and not in other sun-exposed areas. A possible explanation could be that these skin areas although they are intensely exposed to UVR, they are most commonly inadequately protected. Moreover, they are sites where perfumes and cosmetics are often applied. In

addition, the skin of the neck is thinner than the skin of the face with a paucity of pilosebaceous glands, and being intermittently exposed to the sunlight is protected less effectively than the face by intrinsic mechanisms.[16]

## Clinical Presentation

The onset is usually gradual and the V of the neck is the most common site of first involvement.[14] PC is manifested by pink to red to brownish, ill-defined, reticular patches, which are located over the V, the sides of the neck, the upper chest, and the peripheral face. Anatomically shaded areas of the head and neck, such as the submental region and the anterior aspect of the neck, are invariably spared.[8,14] In the past it was believed that PC is accentuated upon if not restricted to the face.[9,10,12] In a clinico-epidemiological study of 50 patients, involvement of the face was present in 38% of them; it was usually mild and it was limited to the pre-auricular region and/or the chin.[14] In our opinion, facial involvement is not as common and as severe as it was considered in the early reports. In the same study,[14] the most commonly affected sites were the V and the sides of the neck up to the retro-auricular region, and the upper chest. The areas shaded by the chin were unaffected in all cases (100%). Peripheral face, especially the parotid region, was involved in 38% of the patients. The distribution of the skin changes was symmetrical in all cases (100%). However, in four cases (8%) the severity of involvement differed between the two sides.[14] Patients are not uncommonly symptomatic, complaining of a mild pruritus or a burning sensation.[8,14] Some patients complain of a flushing-like reaction in the area of the neck, similar to that occurring in rosacea and triggered by the same factors as in rosacea.[14]Flushing had been described in the early reports of PC, but it was not given the appropriate attention.[10,12] In the Greek cohort, 46%of the patients complained of mild pruritus or a burning sensation, and 28% of them reported episodes of 'flushing' involving the affected area and triggered by exposure to the sun or to heat, alcohol drinking or stressful conditions.[14]

Based on the predominating clinical feature, i.e. erythema and telangiectasia or mottled hyperpigmentation, PC has been classified into three types: erythemato-telangiectatic, pigmented and mixedtype.[16] The erythemato-telangiectatic type was the most common clinical type (58%) in the Greek cohort.[14] Recognition of the clinical type may be important

for the selection of the most appropriate and effective treatment.

In accordance to what has already been described for rosacea (extrafacial rosacea),[24] extracervical PC may also exist. A48-year old woman who had been practicing aroma-therapy for several years, presented with poikilodermatous changes in extensor aspects of the forearms, typical PC on the V of the neck, and erythemato-telangiectatic rosacea of the central face.[21] Histopathology of the lesions on the forearms showed typical features of PC. Patch testing revealed positive reactions to Fragrance mix and Nickel sulfate. This case raises the question whether PC can develop in areas other than the face and neck, such as the forearms, that share a common pattern of sun exposure with the neck and also perfumes are commonly applied.

## Course and Prognosis

The course is chronic and slowly progressive. No spontaneous improvement is expected. The condition is benign. No significant co-morbidities have been recognized. Symptoms are usually mild, therefore morbidity is low. Cosmetic disfigurement, occasionally severe, is the only concern. It is noteworthy that many patients are disproportionately worried about the cosmetic disfigurement.[1,2]

## Laboratory Work-up

### *Histopathology*

The histologic picture of PC is very characteristic, but there are no pathognomonic features.[16] The most prominent and constant feature is solar elastosis of the papillary dermis, separated from the epidermis by a grenz zone. Katoulis at al studied lesional biopsies of 50 PC cases.[16] Histological findings included: solar elastosis (100%); flattened (84%) and atrophic (62%) epidermis; basket-weave orthokeratotic, hyperkeratosis (92%), with occasional follicular plugging (34%); mild or patchy vacuolar degeneration of the basal cell layer (46%); melanin granules irregularly distributed in the lower epidermis (94%); melanophages laden with melanin, as well as free melanin granules present in the dermis due to melanin incontinence (92%); dilated blood vessels (96%) with mild

perivascular lympho-histiocytic inflammatory infiltrate (78%) in the papillary dermis; increased numbers of mast cells perivascularly (22%).[16]

Histological findings reported in individual PC cases include: thinning of the epidermis with patchy hyperkeratosis; a well-marked stratum granulosum; thinning of stratum malpighi; degeneration of the lower epidermis with hyaline bodies; vacuolization of some of the basal cells; pigment granules irregularly distributed in the basal layer; absence of papillary processes; dilated superficial vessels; interstitial edema in the upper cutis; mild to marked chronic inflammatory infiltrate of round cells with perivascular and peri-appendageal distribution; band-like lymphonuclear infiltrate in the upper dermis; pigment granules and/or melanophages in the inflammatory infiltrate; a large cyst lined partly with epidermal cells and apparently derived from a hair follicle; and destruction of elastic tissue in the upper dermis.[9,10,12,20]

It is of interest that the histopathological findings of PC are very similar to those in erythemato-telangiectatic rosacea,[25] in terms of solar elastosis that results in erythema and telangiectasia due to loss of vascular support. However, it could be argued that solar elastosis in PC is a collateral finding, as it is virtually impossible to obtain biopsy specimens of patients of such age group that do not show solar elastosis. On the other hand, moderate or mild sun exposure was reported in 68% of PC patients and dermatoses associated with chronic sun exposure, such as actinic keratoses and/or non-melanoma skin cancer, were rather uncommon (8%) in one study.[14]As happens in rosacea, solar elastosis in PC may be related mostly to an inherent increased sensitivity to normal doses of ultraviolet radiation, rather than to a chronic excessive sun exposure.

### *Electron Microscopy*

In an ultrastructural study of 10 patients with PC,[16] the epidermis showed minor changes such as focal distention of the perinuclear pool in the basal cells and degenerative changes of the adjacent nuclear membrane. The dermo-epidermal junction was intact. In the papillary dermis, the collagen fibers were swollen and disrupted; there was focal degeneration of the collagen bundles and inactive fibroblasts in between them. At these sites, ill-defined foci of microgranular or microfibrular texture, containing degenerated

organelles, most probably mitochondria, could be seen. In two cases, several vacuolar spaces of varying size and shape were found just under the basal lamina. In most cases, melanin-laden macrophages (melanophages) and several electro-dense bodies (pigment granules) randomly scattered in the dermis, were detected. The endothelial cells of the dilated dermal blood vessels were normal.[16]

## Differential Diagnosis

PC should be differentiated from other conditions producing reticulate and mottled pigmentation of the neck. This differential diagnosis was systematically reviewed by Lautenschlager and Itin.[7]

PC must be differentiated, primarily, from Riehl's melanosis and erythromelanosis follicularis faciei et colli. In Riehl's melanosis, spotted brown pigmentation predominates, while telangiectasia is minimal or absent.[12] Lesions are less reticular and are commonly located on the face, being most intense on the forehead and temples. The clinical picture is greatly improved by the discontinuation of offending cosmetics.[26,27] Erythromelanosis follicularis faciei et colli most commonly affects young males. Clinically, it manifests itself by brownish-red hyperpigmentation, telangiectasia and follicular keratosis characterized by follicular papules. Lesions are localized on the peripheral face and/or the neck.[28]

Among acquired poikilodermas, most important is the differentiation of PC from poikiloderma of connective tissue disease and poikiloderma atrophicans et vasculare. This differentiation should be based: on the strict anatomic localization of PC; the absence of systemic manifestations; and the chronic benign course. Other conditions that should be considered in the differential diagnosis include chronic graft-vs.-host disease, berloque dermatitis, friction melanosis, etc.[1,2]

Clinical and histopathological differential diagnosis of PC is summarized in Table I.

## Treatment

Although a benign disease, as PC affects cosmetically important body areas, such as the face and neck, it worries patients who often ask for treatment. On the other hand, treatment of PC is difficult. Current therapeutic choices, mostly with various light sources, are costly when dealing with a merely cosmetic problem. The ideal treatment should be safe and

effective, addressing both the pigmented and the vascular component of the disease. In this context, identification of clinical type is important for the selection of the most appropriate therapy. Guidelines for the treatment of PC do not exist. Various modalities have been tried with differing results.

Classic methods of Dermatologic Surgery are of limited value for the treatment of PC. Electrosurgery may treat telangiectasia which is the basis of PC's clinical presentation, but it is time consuming and it is associated with side-effects, such as scarring or residual dyschromia.[29] Cryotherapy is not effective. Due to the pigmented component, depigmenting agents, such as topical hydroquinone 2-4%, topical azelaic acid 15 to 20% or kojic acid, can be used as an adjuvant in the treatment of the pigmented type of PC.[30] Topical retinoids and chemical peels may be used as they can improve photo-aged elastotic skin.[30] Topical flavonoids may reduce erythema, which most often is the predominating feature.

Lasers and light therapies are the mainstay in the treatment of PC. The disadvantage of these therapies is that they are costly, not widely available, and, most importantly, not consistently effective. This is of special significance especially when dealing with a merely cosmetic problem. The pulsed dye laser (PDL) at 585 nm has been effectively used for the treatment of vascular lesions, including PC and various types of telangiectasias.[30] Treatment with the first-generation flashlamp-pulsed dye laser (PDL) was employed in small numbers of patients with good, but varying results.[31,32] The response was better for the vascular component of PC, compared to the pigmented one, which remained relatively unaffected. Adverse events were common and included marked post treatment purpura, a mottled appearance, hyperpigmentation or occasional hypopigmentation, and atrophic or hypertrophic scarring. In addition, several sessions are needed.In a series of 7 PC successfully treated with the PDL, scarring that appeared 4 months after the treatment was noted in one patient.[33]Better early results have been reported with the new PDL devices, which have a longer pulse duration (1.5 ms) and 10-mm spot size. Recent advances in PDL technology enable 50% higher output energies and beam diameters of up to 15 mm with clinically relevant fluences. Bernstein et al treated 20 PC patients with a total of four treatments at monthly intervals, using the redesigned PDL.[34] Of the 17 patients who completed the study, 14 had an improvement greater than 40% and 10 had improvement

greater than 50%. The average improvement was 49%. Side effects were limited to mild edema and mild to moderate erythema and purpura.[34]

Although PDL is commonly used for the treatment of PC, no clear guidelines exist. Concerns have been raised about the occurrence of persistent depigmentation as a late adverse event in 6 of 8 patients with PC after treatment with PDL at intervals of 3 months, using a 585-nm wavelength, fixed pulse duration of 450 ms, fluence between 3.5 and 7 J/cm2, and 7- or 10-mm spot size.[35]Although significant improvement with respect to clearing of the vascular component was seen, six of them, treated with 5-7 J/cm2, experienced severe depigmentation 4-11 months after treatment. Therefore, it is advisable to use fluences as low as possible, not exceeding an upper limit of 5 J/cm2, on a 10-mm spot size.[33,35,36]

Quasi-continuous wave lasers, such as the APTDL, copper vapor, krypton, and KTP lasers, can be also used to treat PC, as they may prove more useful than other lasers in treating larger caliber vessels. However, they carry a high risk of complications. A major advantage of the quasi-continuous wave lasers is that they are not associated with post-operative purpura.[37]Some patients may prefer this option, although the PDL more often produces superior clinical results. A single case of PC successfully treated with the KTP laser at 532 nm has been reported.[38]

Fractional photo-thermolysis has been successfully used in the treatment of PC.[39] Its 1,550 nm wavelength largely targets tissue water and not melanin, creating thousands of microthermal treatment zones, but sparing the surrounding tissue. In this way, a nonspecific thermal injury of the epidermis that may induce scarring and hypopigmentation, is very unlikely, making this treatment modality a good option especially for dark-skinned individuals. Additionally, the "fractional" laser avoids bulk heating of the dermis. It is suggested that both these mechanisms work synergistically to produce reduction in telangiectasias with minimal side-effects. In a prospective pilot study in 10 subjects with PC, ablative fractional photothermolysis was both safe and effective for the treatment of the vascular, pigmentary and textural components of PC. The number of treatments required ranged from 1 to 3 at 6-8 week intervals, with an average of 1.4. For erythema/telangiecatasia, the mean score improved by 65.0%; for dyschromia, by 66.7%; for skin texture, by 51.7%; and for skin laxity, by 52.5%. Two months after treatment, the

mean score for the overall cosmetic outcome improved by 66.7%.[40] Intense pulsed light (IPL) systems are high-intensity light sources, which emit non-coherent polychromatic light in a broad wavelength spectrum of 515-1,200 nm. They allow a great variability in selecting individual treatment parameters thus permitting treatment of different skin conditions both vascular and pigmented in different skin types.[41]IPL sources have been used in PC with comparable efficacy and safety to the PDL.[42]In one study, 135 randomly selected patients with PC were treated IPL. After one to five treatment sessions, clearance of more than 75% for both telangiectasias and hyperpigmentation was observed. Additionally, patients reported subjective improvement in skin texture. Side effects, mostly dyschromic changes, were seen in 5% of the cases.[43] In another study, 66 patients with typical PC were treated with IPL at various settings every 4 weeks until the desired improvement occurred. After an average of 2.8 treatments, a 50 to 75% improvement was observed regarding both telangiectasias and hyperpigmentation. The incidence of hypopigmentation was 5%.[44] In a study of 175 patients with PC, three treatment sessions every 3 weeks with IPL at various settings resulted in clearance of more than 80% of vascular and pigmented components of PC. Side effects were minimal and transient and occurred in 5% of the patients. No scarring or pigment changes were noted.[45]

Photodynamic therapy (PDT) has also been tried in PC. In one report, significant clinical improvement with regards to pigmentary changes and telangiectasia, has been observed after two treatments with BF-200 ALA PDT.[46]

The use of light sources is associated with adverse effects that depend on the device used. These include scarring, irregular hypopigmentation, post-inflammatory hyperpigmentation, post-treatment purpura, mottled appearance, crusting, and erythema. Multiple sessions are usually necessary to obtain optimal clearing.

## Prevention

Strict photo-protection is fundamental. Sun protection measures include sun avoidance, use of hat and protective clothing, and regular application of broad spectrum sunscreen of SPF 50.[46] Additional measures, such as patch testing and avoidance of documented allergens are warranted and may integrate an effective management for PC.

# Material and Methods

The study was conducted at the 2nd Department of Dermatology and Venereology of the National and Kapodistrian University of Athens at "Attikon" General University Hospital in Athens Greece during a period of 18 months (January 2018 – June 2019). Patients with poikiloderma of Civatte (PC) visiting the outpatient clinics of our department for any reason have been recruited. The diagnosis of PC was made on clinical grounds. All entered patients were informed about the nature and the aim of the study and gave their consent. The study was approved by the Ethics Committee of the Hospital.

In all patients, a detailed history was obtained which included:

1. Demographic characteristics: age, gender, place of residence, occupation.

2. Family history with emphasis on the presence of a similar condition in other family members.

3. Medical history, including drug history. Patients were asked if they were suffering from any skin disease or if they had any skin manifestations or symptoms. Medical history of other systems for comorbidities, was also obtained. In females, a gynaecological history was taken and the menstrual status was recorded. In menopausal women, the cause of menopause either physiological or iatrogenic was noted.

4. Patients' skin phototype according to Fitzpatrick's classification and sun exposure habits. Patients were asked about their sun exposure both occupational and recreational. The level of exposure was classified as mild, moderate or severe for either intentional or unintentional exposure. Also patients were asked if they were using sunscreens regularly, sometimes, or never.

5. The use of fragrances or fragranced cosmetics, which were applied on the sides of the neck or the décolleté area of the chest, was assessed.

6. History of PC: site of first appearance, duration of disease, symptoms (burning sensation, itch) and occurrence of flushing episodes, as well as triggering factors for them.

A full body clinical examination followed. During clinical examination the following clinical parameters have been recorded: location (V of the neck, upper chest, sides of the neck, peripheral face); distribution (symmetric or asymmetric) of poikilodermatous lesions; clinical type (erythemato-telangiectatic, pigmented or mixed) based on the morphology and the color (pink to red to brown) of the lesions; predominating clinical feature (telangiectasia, mottled pigmentation, or both) to determine the clinical type of PC.The presence or not of clinical manifestations of coexisting rosacea, was investigated.

For the dermoscopic examination, a Dermlite DL200Hybrid (3Gen, San Juan Capistrano CA, USA) hand-held dermoscope was used. Examination employed polarized light and a x10 magnification, and was performed either without contact of the glass slide with the skin, or with contact but without applying pressure on the skin (for better visualization of the vascular component). Photographic documentation was made by the camera of iPhone 8. Photographs were taken from seven preselected sites, the same for all patients, on the upper chest and the sides of the neck, and the peripheral face, as shown in Figure 1. In each entered patient, a dermoscopy report was filled, in which the dermoscopic features and patterns that had been observed were recorded. As there was limited published experience with dermoscopy of PC, we attempted to describe the most often encountered dermoscopic features, namely vessel types and pigmented structures that predominated on dermoscopic examination among our patients.

***Vascular structures***

**Rhomboidal/polygonal vessels:** bright red, non-branching, linear telangiectatic vessels; they tend to connect with surrounding vessels forming interconnected rhomboidal or polygonal vascular structures, thus resembling parts of a fishnet with irregular holes (Figure 2).

**Linear irregular vessels:** bright red, non-branching, linear telangiectasias with irregular form and distribution.

**Dotted/globular vessels:** bright red, roundish, irregularly distributed dots or globules.

**Flying seagull-like vessels:** two curved red lines that meet at their neighboring end giving the impression of a seagull flying with the wings wide open; usually appear in small clusters giving the impression of a flock of seagulls flying in the horizon (Figure 3).

***Pigmented structures***

**Brown macules:** light brown structureless macules, 2-3 mm in diameter, with rather distinct but slightly irregular borders. They are better appreciated at higher magnifications (x20).

***Depigmented structures***

**White macules:** white roundish macules with rather distinct and regular borders, lighter than the normal skin color that are regularly distributed over a red background of erythema and linear telangiectasias.

***Patterns***

**Fishnet-like pattern:** areas with linear telangiectasias that are interconnected forming an irregular red network that is reminiscent of a fishnet (Figure 2). This network consists of thin red lines and quadrilateral or polygonal, irregular openings.

**Red-white polka dot pattern:** areas of bright red erythema produced by a network of linear telangiectasias, surrounding regularly distributed white roundish macules. It is reminiscent of a red-white polka dot print (Figure 4).

**Spaghetti and meat balls-like pattern:** a combination of linear irregular vessels and dotted/globular vessels giving the impression of spaghetti and meatballs. This pattern was described by Errichetti and Stingo.[47]

**Non-specific pattern:** areas of irregular linear telangiectasias that are irregularly distributed and do not correspond to any specific pattern.

In 10 non-selected patients, an additional digital dermoscopic examination was performed, using the FotoFinder (FotoFinder Systems GmbH, Bad Birnbach, Germany) video

dermatoscope. Photographs at x20 magnification were taken from the same sites as during dermoscopic examination (Figure 1).

***Statistical methods***

For continuous variables, the mean, standard deviation and range, or the median, 25$^{th}$ and 75$^{th}$ percentiles and range, were used after testing for normal distribution. For categorical variables the frequencies and percentages were used. The Shapiro-Wilk test for normality was applied.

Chi-squared and Fischer's exact tests were used for the comparison of categorical variables while unpaired t-tests and Mann-Whitney U tests were applied depending on the distributions of the continuous variables.

All statistical analyses were performed using Stata/IC version 15.

# Results

In total, 28 patients with PC were recruited. The median age was 55 years (range 26-73 years). There were19 females (67.86%) aged 46-73 years (median 54 years), and 9 males aged 26-65 years (median 59 years). Three patients (10.71%) were of skin photo-type I, seven (25%) of skin photo-type II, 14 (50%) of skin photo-type III, and 4 (14.29%) of skin photo-type IV. Of the 28 patients, only 3 (10.71%) reported occupational sun exposure, while 22 (78.57%) had significant recreational sun exposure. Two patients (7.14%) had a history of sunburn during childhood. Nineteen (67.86%) reported PC among other first degree relatives (family history). Sixteen patients (57.14%) were regularly using perfumes or perfumed cosmetics on the neck and upper chest. Among the 19 women, thirteen (68.42%) were in physiologic menopausal. There were no cases of iatrogenic menopause. Demographic characteristics and etiologic factors including sun exposure habits in our cohort are presented in Table II and Table III respectively.

Among our patients, six (21.43%) had erythemato-telangiectatic PC, 2 (7.14%) had the pigmented type, and twenty (71.43%) had the mixed type (Figures 5-7).The duration of disease was 1-5 years in 5 (17.86%), 6-10 years in 11 (39.29%), 11-15 years in 9 (32.14%),

and 16-20years in 3 (10.71%). The V of the neck and the upper chest was involved in 27 patients (96.43%); the sides of the neck in 24 (85.71%), and the peripheral face in 8 (28.57%). Most of our patients reported that the V of the neck was the most common site of first involvement. Only a minority of patients reported symptoms: burning sensation in eight (28.57%) and mild pruritus in five (17.86%).Six patients (21.42%) complained of a flushing-like reaction in the affected area of the neck and upper chest, triggered by environmental and emotional factors. The most commonly reported comorbidity was rosacea, most often of the erythemato-telangiectatic type, that was observed in 12 persons (42.86%). Thyroid disease was present in 8 patients (28.57%) including 2 cases of autoimmune thyroiditis, 2 cases of hypothyroidism; and 2 cases of thyroid nodules; hypercholesterolemia in 6 (21.42%); and hypertension in 5 (17.85%). It is of interest that 4 cases (14.28%) had a history of systemic lupus erythematosus. The clinical characteristics of our patients are summarized in Table IV. All reported comorbidities from the skin and other systems are listed in Tables Va and Vb respectively.

On dermoscopic examination, firstly the global pattern was assessed. In 22 cases (78.57%), the coexistence of more than one pattern at different sites was noted. The red-white polka dot pattern was observed in 10 cases (35.71%); the fishnet-like pattern in 15 cases (53.57%); the non-specific pattern in 9 cases (32.14%); the spaghetti and meatballs-like pattern in 8 cases (28.57%). When the global pattern was assessed by anatomic region, i.e. face, neck, and upper chest, no significant differences were observed. Examples of the dermoscopic global patterns among our patients are shown in Figures 8-11.

Regarding local dermoscopic features, we observed flying seagull-like vessels in 18 cases (64.29%); rhomboidal/polygonal vessels in 15 cases (53.57%); linear irregular vessels in 17 cases (60.71%); dotted/globular vessels in 15 cases (53.57%); white macules in 23 cases (82.14%); brown macules in 11 cases (39.29%); and whitish follicular plugs in 6 cases (21.42%). Examples of the dermoscopic local features observed among our patients are shown in Figures 12-18.

The global pattern and the local dermoscopic features as observed among our patients are shown in Tables VI and VII. Tables VIIIa-c show the frequencies of dermoscopic features

by PC clinical type.

On videodermoscopy and at x20 magnification, we could identify the same local features and the global patterns that we have described above. The predominant feature was linear telangiectasias as rhomboidal/polygonal, dotted/globular or linear irregular vascular structures. At sites, the highly characteristic flying seagull-like vessels could be seen. Brown macules could be readily recognized, mostly in the pigmented and mixed clinical type, as light brown spots. Videodermoscopy (x20 magnification) enables the visualization of brown macules much more efficiently than classic dermoscopy (x10 magnification). White macules can be seen as whitish or skin colored spots suggesting, that they correspond to the holes of the vascular network which is formed by the linear telangiectasias, representing areas of sparing. Three cases of PC as appeared on videodermoscopic examination are presented in Figures 19-21.

## Discussion

In the present study, we examined 28 patients with PC, aiming to describe the dermoscopic features of this skin disease. The epidemiologic and clinical characteristics in our cohort are consistent with those previously described in the literature.[1,2,14,18] As our results indicate, PC exhibits a rather characteristic dermoscopic picture. Although there are no pathognomonic features, dermoscopy can lead to the correct diagnosis with a high level of confidence. Furthermore, by combining history, clinical and dermoscopic examination, PC can be easily differentiated from other poikilodermatous states with guarded prognosis, such as poikiloderma atrophicans et vasculare and poikiloderma associated with collagen vascular disease, minimazing the need for a diagnostic skin biopsy and histologic examination.

The dermoscopic findings in PC consist of vascular and pigmented local features. Vascular structures are the most prominent feature. They are invariably present in all PC patients as they correspond to the linear telangiectasias, which are the cardinal feature of the clinical triad of poikiloderma. We distinguished four types of telangiectatic blood vessels:

rhomboidal/polygonal vessels; linear irregular vessels; dotted/globular vessels; and a distinctive type of short, double-curved vessels, named flying seagull-like vessels. In our patients, flying seagull-like vessels were the most common type (64.29%), followed by linear irregular (60.71%), rhomboidal/polygonal (53.57%), and dotted/globular vessels (32.67%). It must be made clear that these types of vessels coexisted on the same patient in varying combinations and at various sites.

Based on the presence of the aforementioned vessel types and their architectural distribution, we described four patterns that have been observed among our patients: the fishnet-like pattern resulting from the interconnection of the rhomboidal/polygonal vessels; the red-white polka dot pattern composed of regularly distributed white spots over a background of bright red erythema; the non-specific pattern of linear irregular vessels that are irregularly distributed; and the spaghetti and meatballs-like pattern resulting from the combination of linear irregular vessels and dotted/globular vessels. In our cohort, the fishnet-like pattern was the most common (53.57%), followed by the red-white polka dot pattern (35.71%), the non-specific pattern (32.14%), and the spaghetti and meatballs-like pattern (28.57%). There were no significant differences in the frequencies of global patterns among anatomic regions. Possibly with a larger number of observations, some differences may be documented.

Two of the local dermoscopic features we observed, were related to pigmentary changes. White macules were seen in 82.14% of our patients and in all clinical types of PC. They were either regularly distributed over an erythematous background, thus forming the red-white polka dot pattern, or irregularly distributed in the context of the fishnet or the non-specific pattern. The white macules exhibited a follicular or pseudo-follicular distribution. Brown macules were less often observed (39.29%) in our cohort. They were mostly associated with the pigmented and the mixed clinical type and they were rarely seen (16.67%) in the erythemato-telangiectatic type.

The dermoscopic diagnosis of PC should be based on the recognition of various types of telangiectatic vessels combined with pigmentary changes, i.e. white and brown macules, forming a reddish to brownish network. We were able to describe four distinct global

patterns (fishnet-like, red-white polka dot, spaghetti and meatballs-like, and non-specific) and four types of telangiectatic vessels (rhomboidal/polygonal, dotted/globular, flying seagull-like, and linear irregular). Of the vessel types, flying seagull-like vessels have not been described previously in any skin condition and, as they are frequently observed in PC, could be considered as highly characteristic for PC.

In our statistical analysis, we investigated possible correlations of the dermoscopic -- features and patterns with epidemiologic parameters, aetiologic factors and clinical characteristics of our PC patients. A major limitation of our study was the relatively small number of cases evaluated. Nevertheless, the following statistical correlations were documented: white macules were correlated with the mixed clinical type ($p$=0.015); flying seagull-like vessels were correlated with the erythemato-telangiectatic type ($p$=0.028); brown macules were associated with skin phototype IV ($p$=0.016) and with disease duration >5 years ($p$=0.009).

In the literature, there is little published experience on dermoscopy of PC. Errichetti and Stinco studied 8 consecutive cases (6 women and 2 men, aged 42–73 years, mean 51 years) of clinically diagnosed PC.[47] The authors described a combination of dotted/globular vessels and linear irregular vessels, giving the impression of “spaghetti and meatballs”, along with perifollicular whitish (spared) areas. In addition, they noted the presence of follicular keratotic plugs and delicate reticular or structureless brownish areas in 25% and 12.5% of the cases respectively. Based on these findings, they concluded that dermoscopy can confirm the diagnosis of PC in doubtful cases and can assist the differentiation from other dermatoses that enter in the differential diagnosis of PC.[47]Our findings are in line with the findings of Errichetti and Stinco. However, the presence of dotted/globular vessels was less common in our cohort (53.57%). Accordingly, the spaghetti and meatballs patternas described by Ericchetti and Stinco was not recognized by us as the predominant global pattern, but it was seen in 28.57% of the patients. Also, for the white macules or white areas of sparing, we noticed that they often have a pseudo-follicular distribution, i.e. they are not strictly related to the follicular openings.

The dermoscopic findings of PC are highly characteristic.To our knowledge, they have not

been described previously in any skin condition. By dermoscopy, PC can be easily differentiated from other skin conditions characterized also by telangiectasia, reticular pigmentation or true poikiloderma.

Rosacea that often coexists with PC, most commonly involves the central face. Vascular changes of erythemato-telangiectatic rosacea are highlighted by dermoscopy.[48] Linear telangiectatic vessels arranged in horizontal and vertical lines form polygons (polygonal vessels).[4,48,49] These vascular polygons should be differentiated from telangiectatic vessels in sun-exposed elastotic skin that typically lack this characteristic polygonal arrangement.[5] Poikiloderma presenting at sites of severely sun-damaged skin in the context of xeroderma pigmentosum displays white scar-like depigmentation and thin non-arborizing telangiectasia.[50] However, the vascular polygons are similar to the polygonal vessels observed in half of our patients with PC, providing further support to the theory that rosacea and PC are related and, possibly, belong to the same nosological spectrum.[23] In addition, rosettes, follicular plugs, white/yellowish scales, orange-yellowish areas, pigmentation structures, dilated follicles and follicular pustules (in papulopustular rosacea), have been observed.[4,48,49] The dermoscopic pattern of rosacea is considered as highly specific, allowing the differentiation of rosacea from other inflammatory facial dermatoses.[5]

In Riehl's melanosis, the pigmented dermoscopic featurespredominate,i.e.gray dots/granules and pigmented pseudo-network, combined with telangiectatic vessels.[51] In a recent study, these features were present in all 15 patients examined (100%), followed by slight flour-like scales (53.3%),follicular keratotic plugs and perifollicular whitish halo (46.7%).[51]

Dermoscopy of erythromelanosis follicularis faciei et colli in a 17 year old boy showed multiple round whitish areas with follicular plugs and occasionally centered by a hair, surrounded by blue-gray dots or peppering in a reddish-brown background. Also the presence of some white scales was noted.[52] According to Maouni et al,[52] the whitish areas with follicular plugs seems to correspond histologically to follicular hyperkeratosis, the grey blue spots/peppering to the pigmentary incontinence, and dermal melanophages, and the reddish-brown background to vasodilation and hyperpigmentation of the basal layer.

Pigmented type of PC involving the face may be confused with melasma. Dermoscopy in melasma reveals a light-to-dark brown background and brown granules and globules with perifollicular sparing.[53-55] The global pattern may be reticular or pseudoreticular, especially in deeper melasma.[53] It has been suggested that the pigment color may predict the depth of melasma,[55] although this has been argued against.[56]

Poikiloderma may appear in the context of collagen vascular disease. Early lesions of discoid lupus erythematosus exhibit on dermoscopy intense follicular plugging, perifollicular whitish halo, and white scale.[5] Later, signs of fibrosis and scarring predominate, associated with blurred telangiectatic, arborizing vessels; white structureless areas; and hyperpigmentation, but lacking the orange-yellowish pigmentation which characterizes granulomatous cutaneous diseases, such as sarcoidosis and lupus vulgaris.[5,57,59] Subacute cutaneous lupus erythematosus is characterized by whitish scales diffusely or peripherally distributed, and a vascular pattern consisting of at least two types of vessels among dotted, linear-irregular, linear, and branching vessels over a pinkish-reddish background with occasional orange-yellowish structureless areas.[4,57,58] Very few data has been published regarding dermatomyositis. Gottron papules on the fingers exhibit dotted vessels and scattered scales on a homogenous pink background.[60]

In chronic graft-versus-host disease, whitish scales associated with vessels of mixed morphology, namely dotted and linear can be seen by dermoscopy.[4] Kaminska-Winciorek et al studied 6 patients with acute GVHD after allogeneic hematopoietic stem cell transplantation. Dermoscopic findings included well-visible, multiple thin telangiectasias on a pinkish or reddish background. Moreover, in selected regions, serpentine vessels, multiple thick telangiectasias, and multiple dotted or globular vessels were present.[61]

Dermoscopy in mycosis fungoides reveals, most often, fine short linear vessels, irregularly distributed overorange-pinkish patchy areas.[62] Often a vascular structure resembling spermatozoon, composed of a dotted and a short curved linear vessel can be observed. Additional dermoscopic findings include fine white scales, dotted vessels and purpuric dots.Poikiloderma atrophicans et vasculare is a rare variant of early-stage mycosis fungoides or a premycotic condition, usually presenting as asymptomatic or moderately pruritic, slightly scaly, reddish-brown papule that coalesce into reticular lesions, which mainly involve the breast, abdomen, buttocks, and flexures.[63] Errichetti and Stingo reported

three females with poikiloderma atrophicans et vasculare located on the breast area in all three cases.[64] On dermoscopy, lesions exhibited a rather monomorphic pattern, consisting of relatively blurred branched vessels on a reddish or orangish-brown background and associated with sparse whitish scales. On this basis, the authors concluded that dermoscopy may assist the differential diagnosis over other similar papular dermatoses that might be considered in the differential diagnosis, such as guttate psoriasis, pityriasis lichenoides chronica, papular sarcoidosis, and lichen planus.[65] It can also help the differentiation from patch-stage mycosis fungoides, especially with regard to the vascular pattern.[62] Xu and Tan reported a 59-year-old man with widespread erythematous and scaly poikilodermatous patches for 30 years, who was histologically diagnosed as poikilodermatous mycosis fungoides.[66] On dermoscopy, multiple polygonal structures consisting of lobules of white storiform streaks, studded with fine red dots or hairpin vessels were noted. Between the lobules were septa of pigmented dots that were unevenly and intermittently distributed throughout. In addition, red and yellowish smudges were easily seen. Although a definitive diagnosis must be based on a combination of clinical presentation, histopathologic features, T-cell receptor gene analysis, and immunopathologic criteria, dermoscopy may also be of diagnostic help.

The dermoscopic differential diagnosis of PC is depicted in Table IX.

The dermoscopic findings correlate well with both the clinical and histologic features of PC, supporting the view that dermoscopy represents a bridge between clinical presentation and histology. The vascular dermoscopic structures (rhomboidal/polygonal vessels, flying seagull-like vessels, linear irregular vessels, dotted/globular vessels) are the dermoscopic correlates of linear telangiectasia of poikiloderma and represent the dilated hyperemic vessels of the papillary dermis that have been described in the histopathology of PC. When the course of the vessels parallels the skin surface, they appear on dermoscopy as rhomboidal/polygonal or linear irregular vessels, while when their course is perpendicular they appear as dotted/globular vessels. The mottled hyperpigmentation of poikiloderma is dermoscopically appreciated as brown macules, and results from the increased presence of melanin irregularly distributed in the basal layer of the epidermis, as well as the presence

of melanophages laden with melanin in the dermis. White macules correspond to the superficial atrophy that integrates the clinical triad of poikiloderma. White macules correlate histologically to a flattened and atrophic epidermis, overlying an elastotic papillary dermis at sites in between the reticulate telangiectasia. The clinico-dermoscopic and dermoscopic-pathologic correlation for PC is summarized in Table X.

## Conclusions

PC is a rather common chronic benign dermatosis that causes significant cosmetic disfigurement. Many aspects of this disease remain unclear and, most importantly, its aetiology and pathogenesis remain obscure. Also, treatment is difficult and its results often unsatisfactory. To our knowledge, this is the first study to systematically investigate the dermoscopic characteristics of PC. Although not pathognomonic, the dermoscopic picture is characteristic, leading to the clinical diagnosis with great confidence. We were able to describe patterns and features that are unique for PC, permitting the differentiation from other dermatoses of the face and neck, as well as from other forms of poikiloderma with guarded or serious prognosis. Dermoscopic findings correlate well with the clinical and histological features of PC. On this basis, biopsy and histologic examination is rarely necessary. Further studies in a larger number of patients are needed to verify our observations. Future research will, hopefully, better clarify the pathogenetic mechanisms of this disease and will provide more effective preventive and therapeutic approaches for PC.

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# Tables

**Table I.** Clinico-pathological differential diagnosis of poikiloderma of Civatte. Modified from Katoulis AC et al. Dermatology 2007; 214:177-182.

| Skin disorder | Etiology | Clinical Characteristics | Histopathology |
|---|---|---|---|
| **Riehl's melanosis** | Pigmented dermatitis possibly caused by perfumes or cosmetics | Brown spotted pigmentation, telangiectasia is minimal or absent Distribution: face, most intense on the forehead and temples | Liquefaction of the basal cell layer, basal membrane changes, melanin incontinence; solar elastosis and atrophic changes of the epidermis are absent |
| **Erythromelanosis follicularis faciei et colli** | Unknown; genetic predisposition | Young males; Brownish-reddish hyperpigmentation, telangiectasia, and minute follicular papules (follicular keratosis) of the peripheral face and/or neck; Shaded areas are not spared | Dilated hair follicles and hypertrophic sebaceous glands; solar elastosis and atrophic changes are absent |
| **Poikiloderma** | | Combination of linear telangiectasia, mottled hyperpigmentation, and superficial atrophy in a reticular pattern | Flattened, atrophic epidermis, hydropic degeneration of the basal layer, band-like infiltrate of the papillary dermis admixed with melanophages, which in places invades the epidermis |
| **Associated with collagen vascular disease** | Autoimmune disorder, sun-exacerbated | Dermatomyositis: heliotrope erythema and edema (periorbital) of the face, neck and chest, Gottron's sign, | Basal membrane is thick and/or interrupted, the upper dermis is atrophic and edematous with deposition of an |

| | | | |
|---|---|---|---|
| | | periungual telangiectasia, polymyositis, vasculitis Subacute lupus erythematosus: psoriasiform papulosquamous eruption on sun-exposed neck, trunk, upper extremities, telangiectasia, no atrophy, periungual telangiectasia Systemic lupus erythematosus: butterfly rash, discoid lupus plaques, palpable purpura, urticarial vasculitis, fever, arthritis, systemic involvement (renal disease, pneumonitis, pericarditis, CNS disease etc.) | alcianophilic mucoid substance and mild dermal infiltration Immunofluorescence of the skin: lupus band |
| **Associated with cutaneous T cell lymphoma (poikiloderma atrophicans et vasculare)** | Malignant transformation; precursor of mycosis fungoides | Telangiectasia, reticulate pigmentation, superficial atrophy with asymmetrical distribution on non-sun-exposed trunk Parapsoriasis en grandes plaques Patch stage mycosis fungoides | Epidermotropic large lymphoid cells with convoluted or cerebriform nuclei |
| **Chronic graft versus-host disease** | Immune reaction of histo-incompatible immunocompetent donor cells against immunocompetent host tissues | Lichenoid papules, sclerodermoid changes, poikiloderma; trunk, buttocks and extremities; mucosal involvement | Hyperkeratosis, acanthosis, basal vacuolization, mild perivascular inflammatory infiltrate, melanin incontinence. Late: sclerodermoid changes with loss of |

| | | | |
|---|---|---|---|
| | | | appendages |
| **Chronic radiation dermatitis** | Exposure to ionizing radiation as a result of therapy, occupational or accidental | History of exposure. Atrophy, hypopigmenation, telangiectasia, poikiloderma, loss of appendages, ulceration, necrosis at the portal of radiotherapy | Epidermal atrophy; loss of appendages; hyalinization, fusion of collagen and elastic tissue; vascular dilatation with fibrous thickening of the arterial wall |
| **Melasma** | Genetic factors, ultraviolet radiation, female sex hormones, drugs | Young adult females, macular brown, geographic hyperpigmentation, mostly on the central face | Increased melanin in the epidermis |
| **Berloque dermatitis** | Phototoxic reaction to 5-methoxypsoralen in bergamot oil contained in perfumes | Brown pigmentation following the pattern formed by the trickle of perfume over the skin | Hyperpigmentation of the basal layer, presence of dermal melanophages |

**Table II.** Demographic characteristics of patients with poikiloderma of Civatte (n=28).

| **Gender** | Male (%) | Female (%) |
|---|---|---|
| | 9 (32,14) | 19 (67,86) |
| **Age (Years)** | Median (Range) | Median (Range) |
| | 59 (26-65) | 54 (46-73) |

**Table III.** Sun exposure characteristics and etiologic factors among patients with poikiloderma of Civatte (n=28).

| **Skin phototype** | **I (%)** | **II (%)** | **III (%)** | **IV (%)** |
|---|---|---|---|---|
| | 3 (10.71) | 7 (25) | 14 (50) | 4 (14.29) |
| **Occupational sun exposure** | Yes (%) | No (%) | | |
| | 3 (10.71) | 25 (89.29) | | |
| **Recreational sun exposure** | Yes (%) | No (%) | | |
| | 22 (78.57) | 6 (21.43) | | |
| **Sunburns in childhood** | Yes (%) | No (%) | | |
| | 2 (7.14) | 26 (92.86) | | |
| **Use of perfumes** | Yes (%) | No (%) | | |
| | 16(57.14) | 12 (42.86) | | |
| **Menopause** | Yes (%) | No (%) | | |
| | 13 (68.42) | 6 (31.58) | | |
| **Family history** | Yes (%) | No (%) | | |
| | 9 (32.14) | 19 (67.86) | | |

**Table IV.** Clinical characteristics of patients with poikiloderma of Civatte (n=28).

| **Clinical type** | Erythemato-telangiectatic (%) | Pigmented (%) | Mixed (%) |
|---|---|---|---|
| | 6 (21.43) | 2 (7.14) | 20 (71.43) |

| **Duration** | 1-5 years (%) | 6-10 years (%) | 11-15 years (%) | 16-20 years (%) |
|---|---|---|---|---|
| | 5 (17.86) | 11 (39.29) | 9 (32.14) | 3 (10.71) |
| **Location** | V of the neck (%) | Sides of the neck (%) | Peripheral face (%) | |
| | 27 (96.43) | 24 (85.71) | 8 (28.57) | |
| **Symptoms** | Pruritus (%) | Burning (%) | Flushing (%) | |
| | 5 (17.86) | 8 (28.57) | 6 (21.42) | |

**Table Va.** Comorbid skin diseases among patients with poikiloderma of Civatte (n=28).

| Skin disorders | No of patients (%) |
|---|---|
| Rosacea | 12 (42.8) |
| Melanoma | 2 (7.14) |
| Hidradenitis suppurativa | 2 (7.14) |
| Atopic dermatitis | 2 (7.14) |
| Large number (>50) of nevi | 2 (7.14) |
| Psoriasis | 1 (3.57) |
| Lichen planus | 1 (3.57) |
| Polymorphus light eruption | 1 (3.57) |
| Folliculitis | 1 (3.57) |
| Genital herpes | 1 (3.57) |
| Dyshidrotic eczema (Pompholyx) | 1 (3.57) |
| Alopecia areata | 1 (3.57) |
| Albinism | 1 (3.57) |

**Table Vb.** Comorbidities from other systems among patients with poikiloderma of Civatte (n=28).

| Disorders | No of patients |
|---|---|
| Thyroid disease | 8 |
| Hypercholesterolaimia | 6 |
| Arterial hypertension | 5 |
| Systemic lupus erythematosus | 4 |
| Autoimmune thyroiditis | 3 |
| Thyreoidectomy | 3 |
| Osteoarthritis | 2 |
| Gastroesophageal reflux | 2 |
| Cardiovascular disease | 2 |
| Chronic respiratory failure | 1 |
| Diabetes mellitus | 1 |
| Hyperuricemia | 1 |
| Non-Hodgkin lymphoma | 1 |
| Idiopathic throbocytosis | 1 |
| Rheumatoid arthritis | 1 |
| Sjogren syndrome | 1 |
| Allerghic rhinitis | 1 |
| Bronchial asthma | 1 |
| Benign prostate hyperlasia | 1 |
| Trigeminal neuralgia | 1 |
| Multiple sclerosis | 1 |
| Migraine | 1 |
| Psychosis | 1 |
| Depression | 1 |
| Osteoporosis | 1 |

**Table VI.** Global dermoscopic patterns in patients with poikiloderma of Civatte (n=28).

| **Global pattern** | **No of patients (%)** |
|---|---|
| Fishnet-like | 15 (53.57) |
| Red-white polka dot | 10 (35.71) |
| Spaghetti and meatballs-like | 8 (28.57) |
| Non specific | 9 (32.14) |

**Table VII.** Local dermoscopic features in patients with poikiloderma of Civatte (n=28).

| **Dermoscopic feature** | **No of patients (%)** |
|---|---|
| Rhomboidal/polygonal vessels | 15 (53.57) |
| Dotted/globular vessels | 15 (53.57) |
| Linear irregular vessels | 17 (60.71) |
| Flying seagull-like vessels | 18 (64.29) |
| White macules | 23 (82.14) |
| Brown macules | 11 (39.29) |
| Follicular plugs | 6 (21.42) |

**Table VIIIa.** Local dermoscopic features in patients with erythemato-telangiectatic type of poikiloderma of Civatte (n=6).

| **Dermoscopic feature** | **No of patients (%)** |
|---|---|
| Rhomboidal/polygonal vessels | 2(33.33) |
| Dotted/globular vessels | 4 (66.66) |
| Linear irregular vessels | 3 (50) |
| Flying seagull-like vessels | 6 (100) |
| White macules | 3 (50) |
| Brown macules | 1 (16.67) |

**Table VIIIb.** Local dermoscopic features in patients with pigmented type of poikiloderma of Civatte (n=2).

| **Dermoscopic feature** | **No of patients (%)** |
|---|---|
| Rhomboidal/polygonal vessels | 0 |
| Dotted/globular vessels | 1(50) |
| Linear irregular vessels | 2 (100) |
| Flying seagull-like vessels | 1 (50) |
| White macules | 1 (50) |
| Brown macules | 2 (100) |

**Table VIIIc.** Local dermoscopic features in patients with mixed type of poikiloderma of Civatte (n=20).

| Dermoscopic feature | No of patients (%) |
|---|---|
| Rhomboidal/polygonal vessels | 13(65) |
| Dotted/globular vessels | 11 (55) |
| Linear irregular vessels | 13(65) |
| Flying seagull-like vessels | 12 (60) |
| White macules | 19 (95) |
| Brown macules | 9 (45) |

**Table IX**. Dermoscopic differential diagnosis of poikiloderma of Civatte.

| | **Dermoscopy** |
|---|---|
| **Riehl's melanosis** | Gray dots/granules and pigmented pseudo-network, combined with telangiectatic vessels; Less often flour-like scales, follicular keratotic plugs and perifollicular whitish halo |
| **Erythromelanosis follicularis faciei et colli** | Round whitish areas with follicular plugs, occasionally centered by a hair; surrounding blue-gray dots or peppering in a reddish-brown background |
| **Poikiloderma** | |
| **- Associated with collagen vascular disease** | Dermatomyositis (Gottron papules): dotted vessels and scattered scales on a homogenous pink background Subacute cutaneous lupus erythematosus: whitish scales diffusely or peripherally distributed; vascular pattern consisting of dotted, linear-irregular, linear, and branching vessels; pinkish-reddish background; occasional orange-yellowish structureless areas |
| **- Associated with cutaneous T cell lymphoma (poikiloderma atrophicans et vasculare)** | Fine short linear vessels irregularly distributed overorange-pinkish patchy areas; Spermatozoon- like vessels; fine white scales, dotted vessels and purpuric dots<br>**Poikiloderma atrophicans et vasculare:** blurred branched vessels on a reddish or orangish-brown background; |

| | |
|---|---|
| | sparse whitish scales. |
| **Chronic graft versus-host disease** | Whitish scales; Vvessels of mixed morphology, mostly dotted and linear |
| **Chronic radiation dermatitis** | |
| **Melasma** | Light-to-dark brown background;brown granules/ globules with perifollicular sparing;global pattern: reticular or pseudo-reticular |

**Table X.** Clinico-dermoscopic and dermoscopic-pathologic correlation in poikiloderma of Civatte.

| Clinical Feature | Dermoscopic Feature | Histopathologic Correlate |
|---|---|---|
| **Linear telangiectasia**<br>- transverse course<br><br><br><br>- perpendicular course | <br>Rhomboidal/polygonal vessels, linear irregular vessels, flying seagull-like vessels<br>Dotted/globular vessels | Dilated and hyperemic blood vessels in the papillary dermis |
| **Superficial atrophy** | White macules | Flattened and atrophic epidermis; solar elastosis of the papillary dermis |
| **Mottled hyperpigmentation** | Brown macules | Irregular distribution of melanin in the basal layer; melanophages in the dermis due to melanin incontinence |

# Figures

**Figure 1.** Preselected sites where dermoscopy and videodermoscopy were performed.

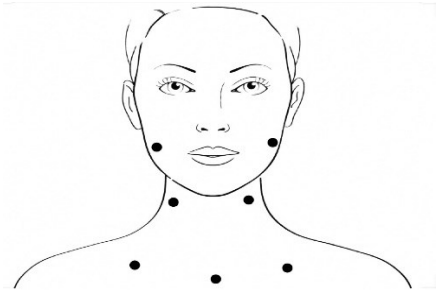

**Figure 2.** Schematic representation of a fishnet.

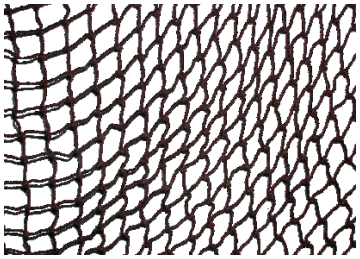

**Figure 3.** Schematic representation of flying seagulls.

**Figure 4.** The red-white polka dot print.

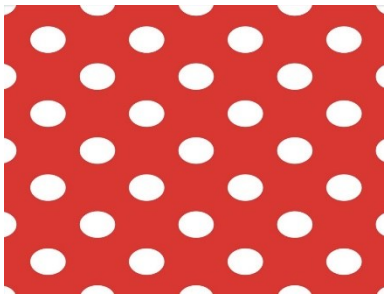

**Figure 5.** Cases with erythemato-telangiectatic type of poikiloderma of Civatte.

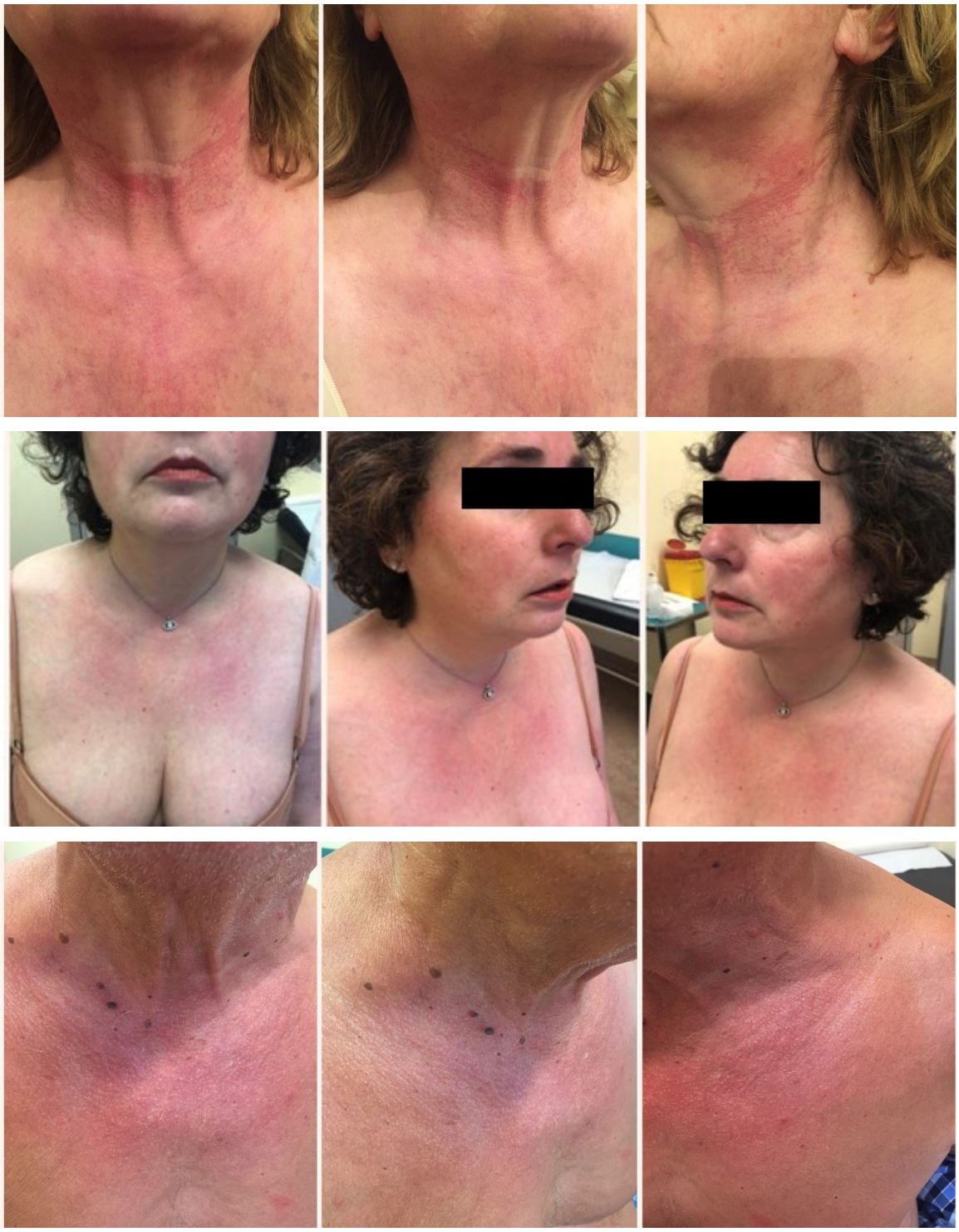

**Figure 6.** Cases with mixed type of poikiloderma of Civatte.

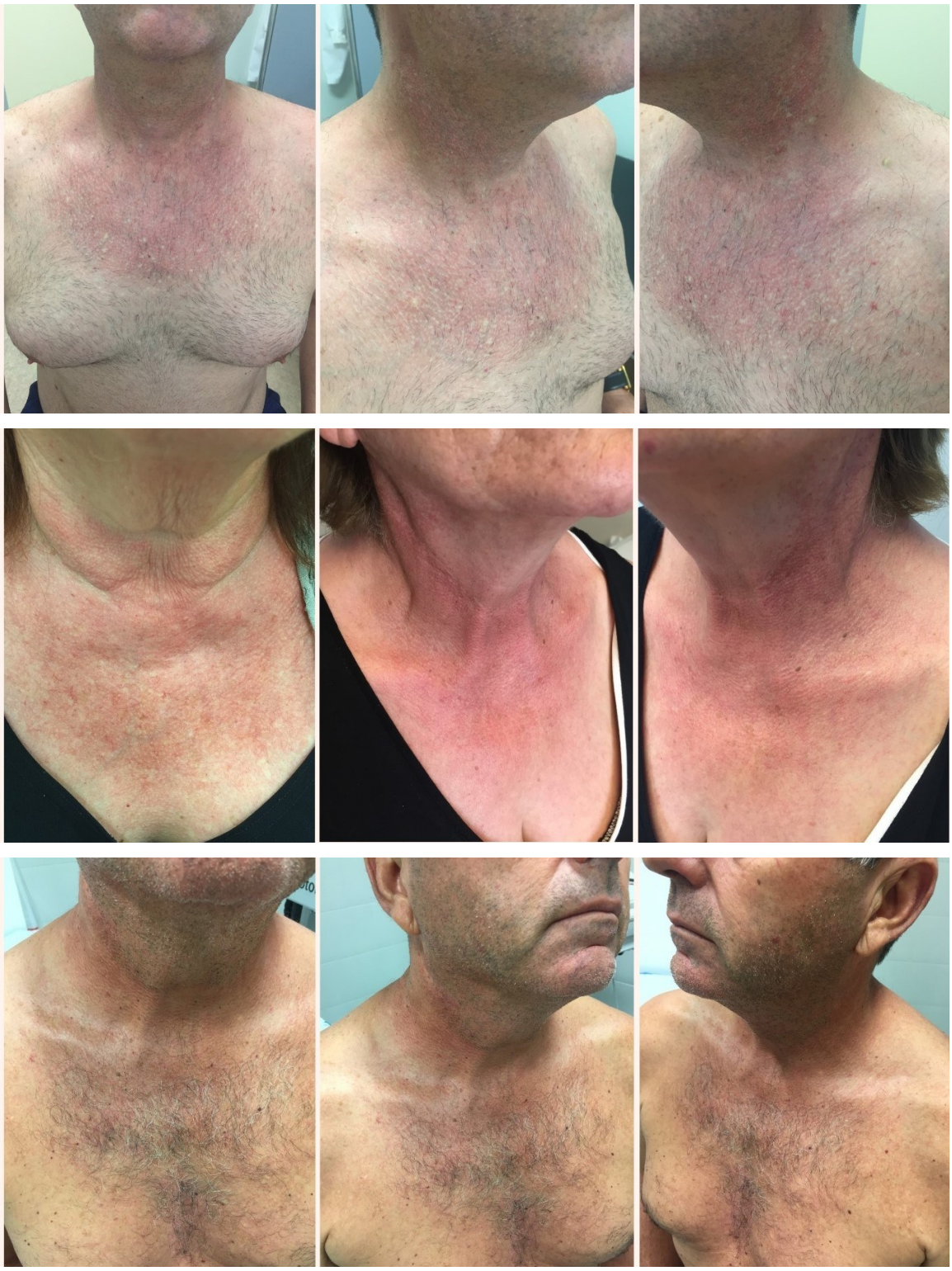

**Figure 7.** Cases with pigmented type of poikiloderma of Civatte.

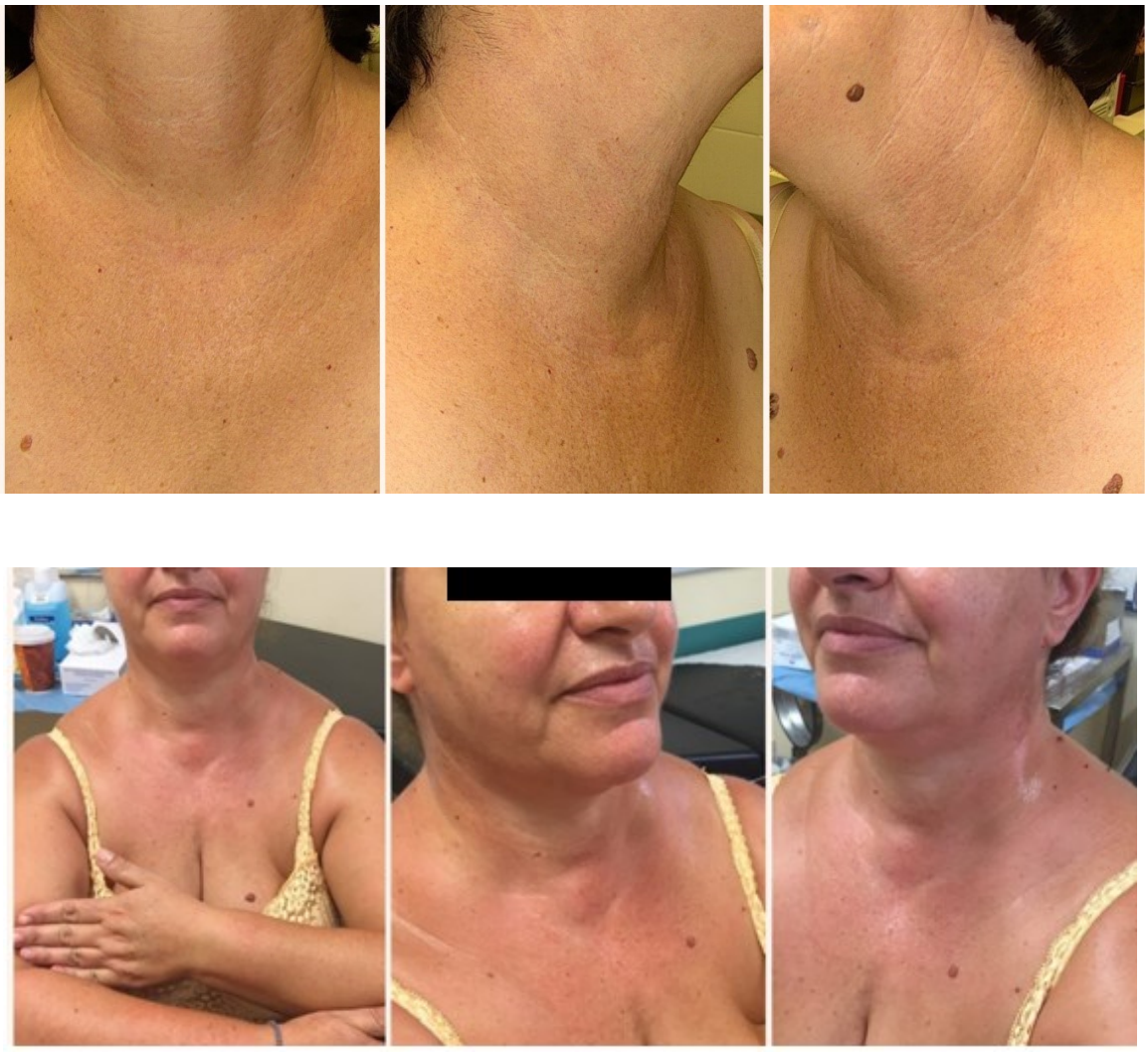

**Figure 8.** The fishnet-like pattern.

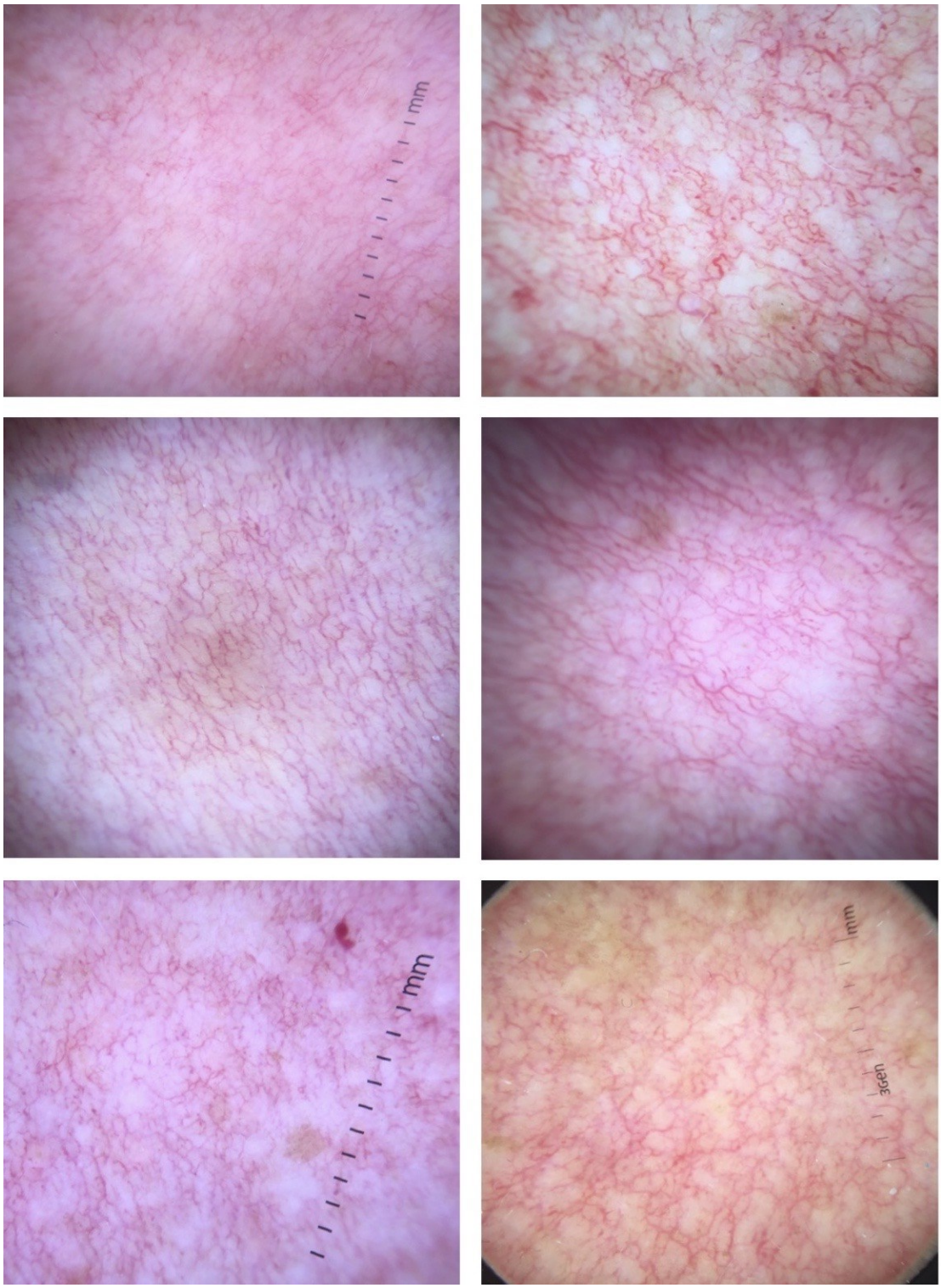

**Figure 9.** The red-white polka dot pattern.

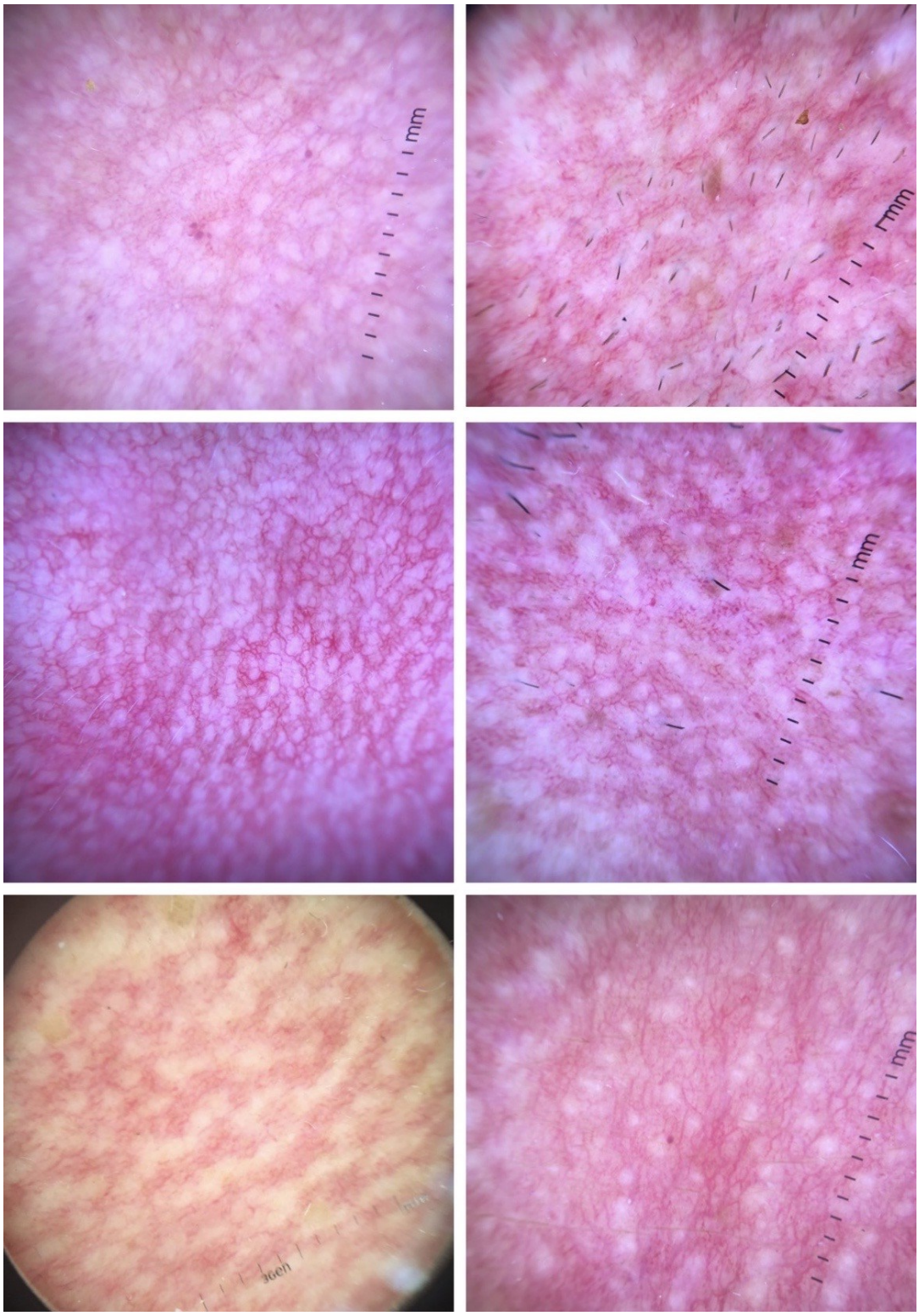

**Figure 10.** The spaghetti and meatballs-like pattern.

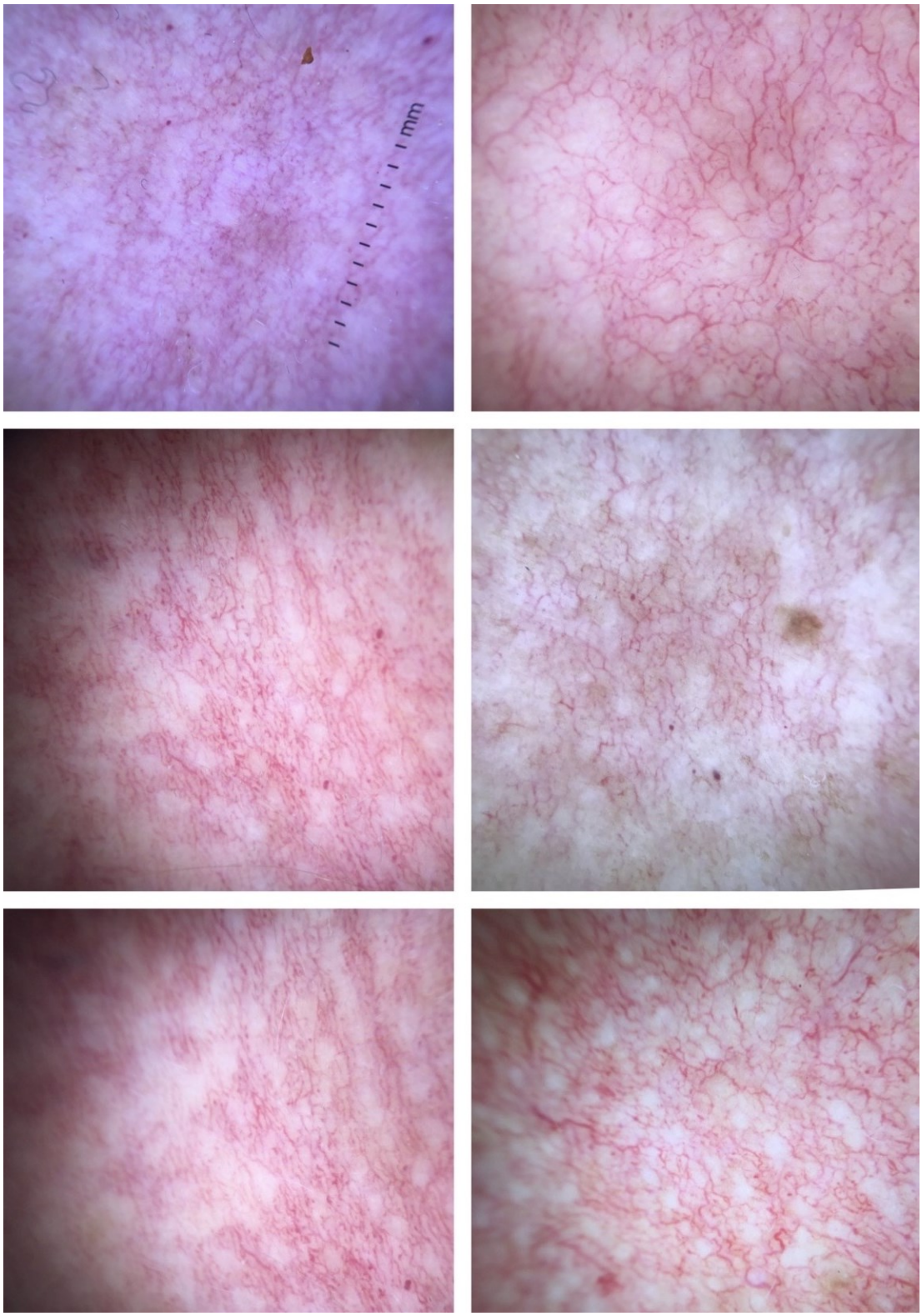

**Figure 11.** The non-specific pattern.

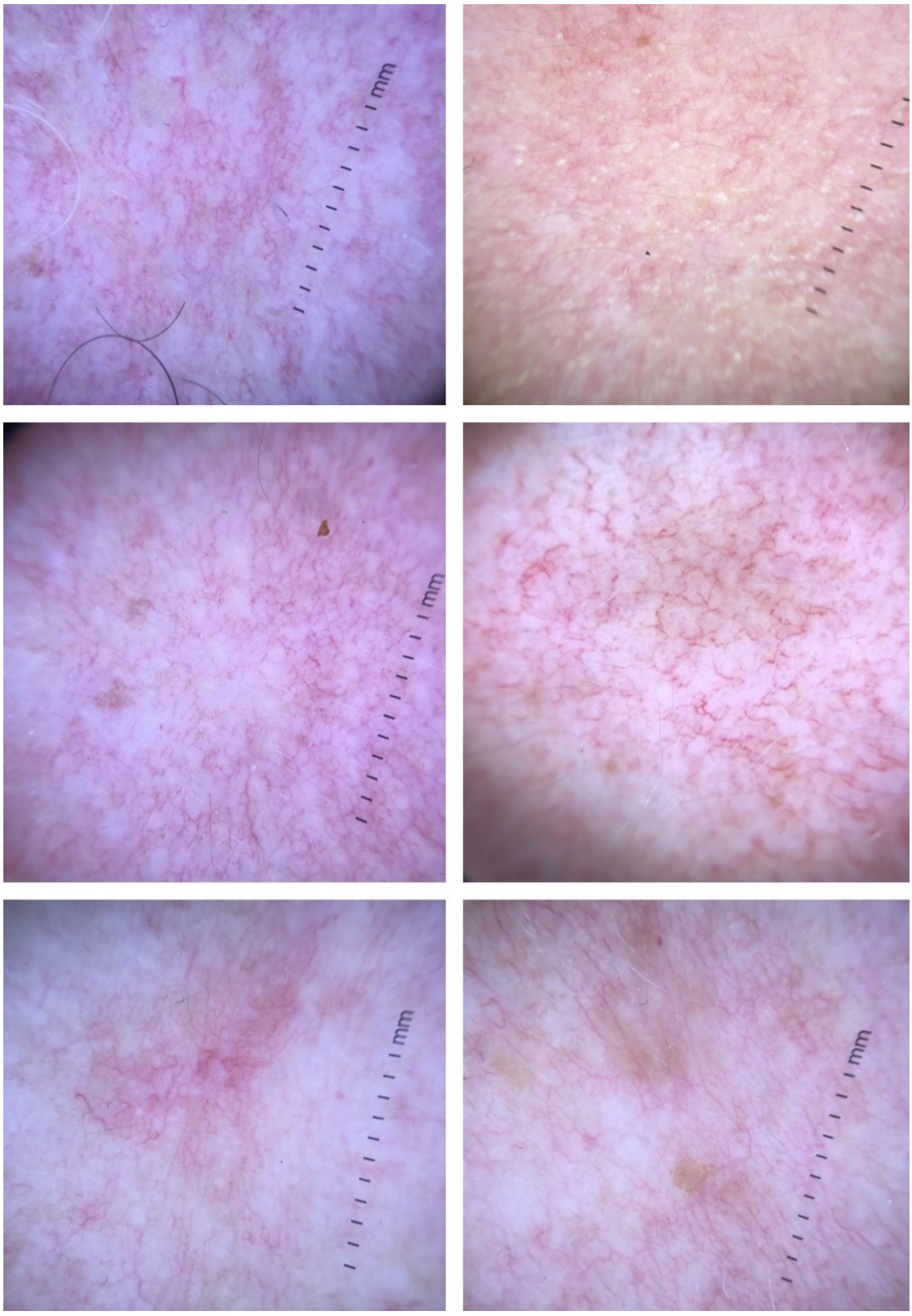

**Figure 12.** The rhomboidal/polygonal vessels.

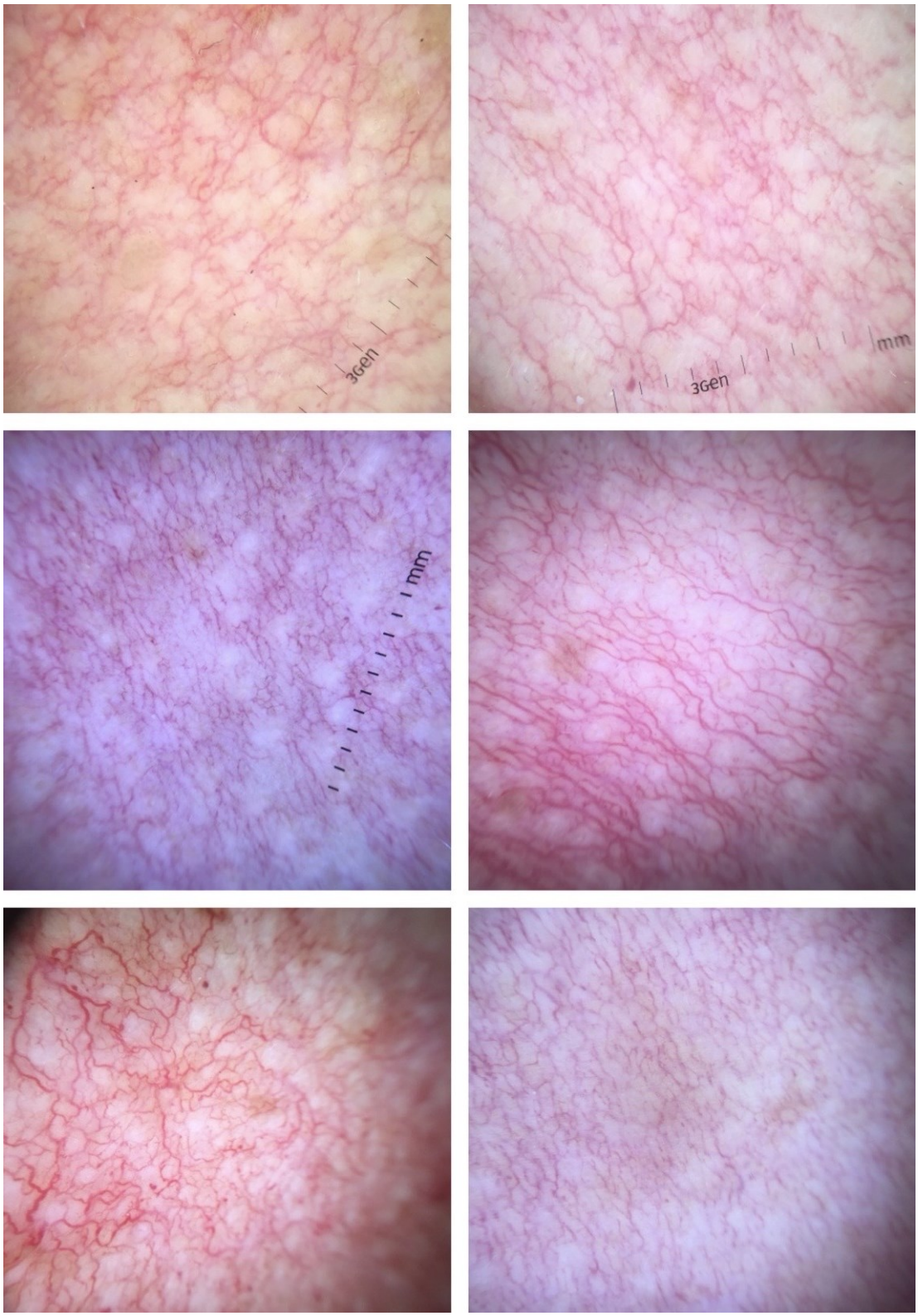

**Figure 13**. The flying seagull-like vessels.

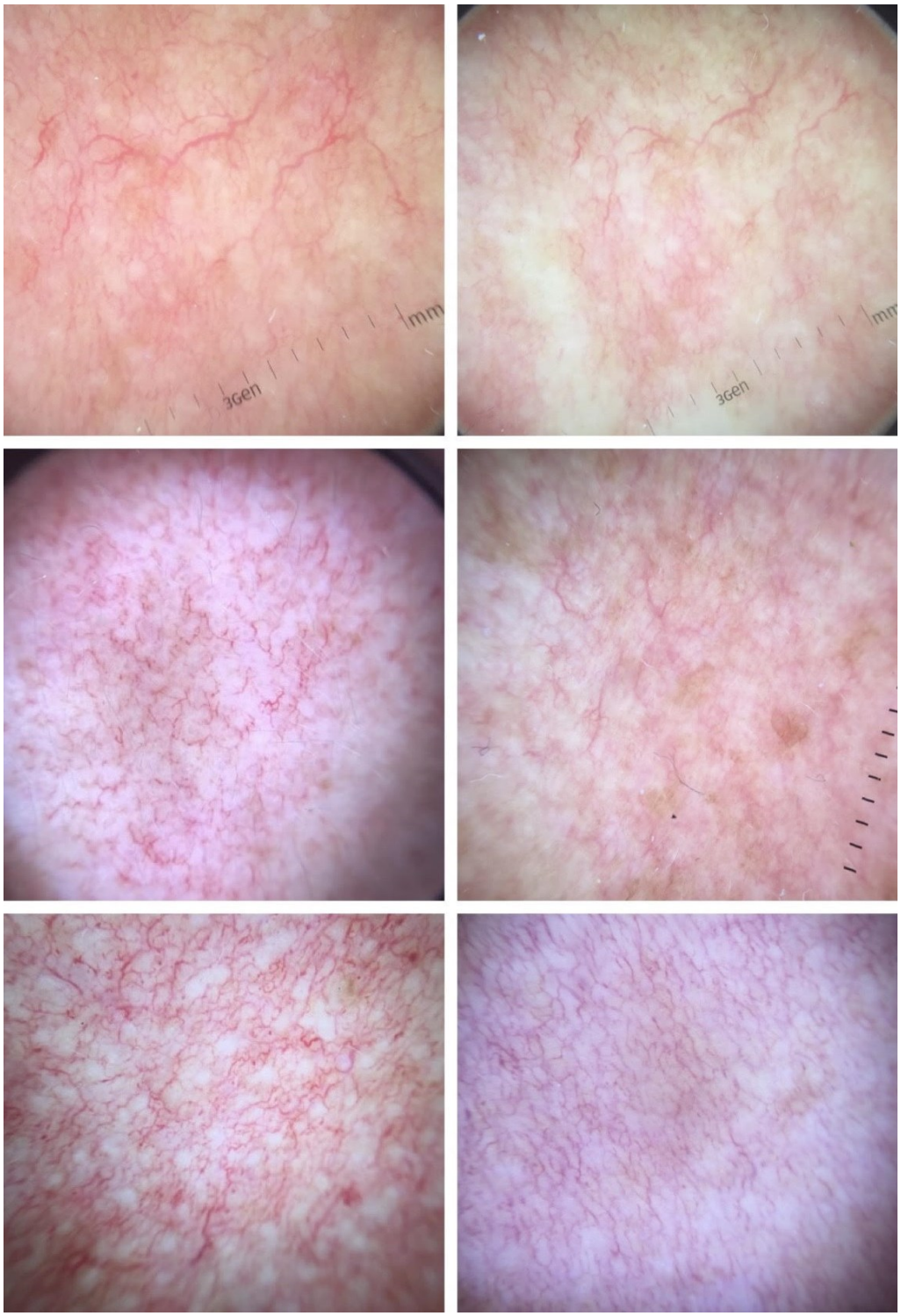

**Figure 14**. The dotted/globular vessels.

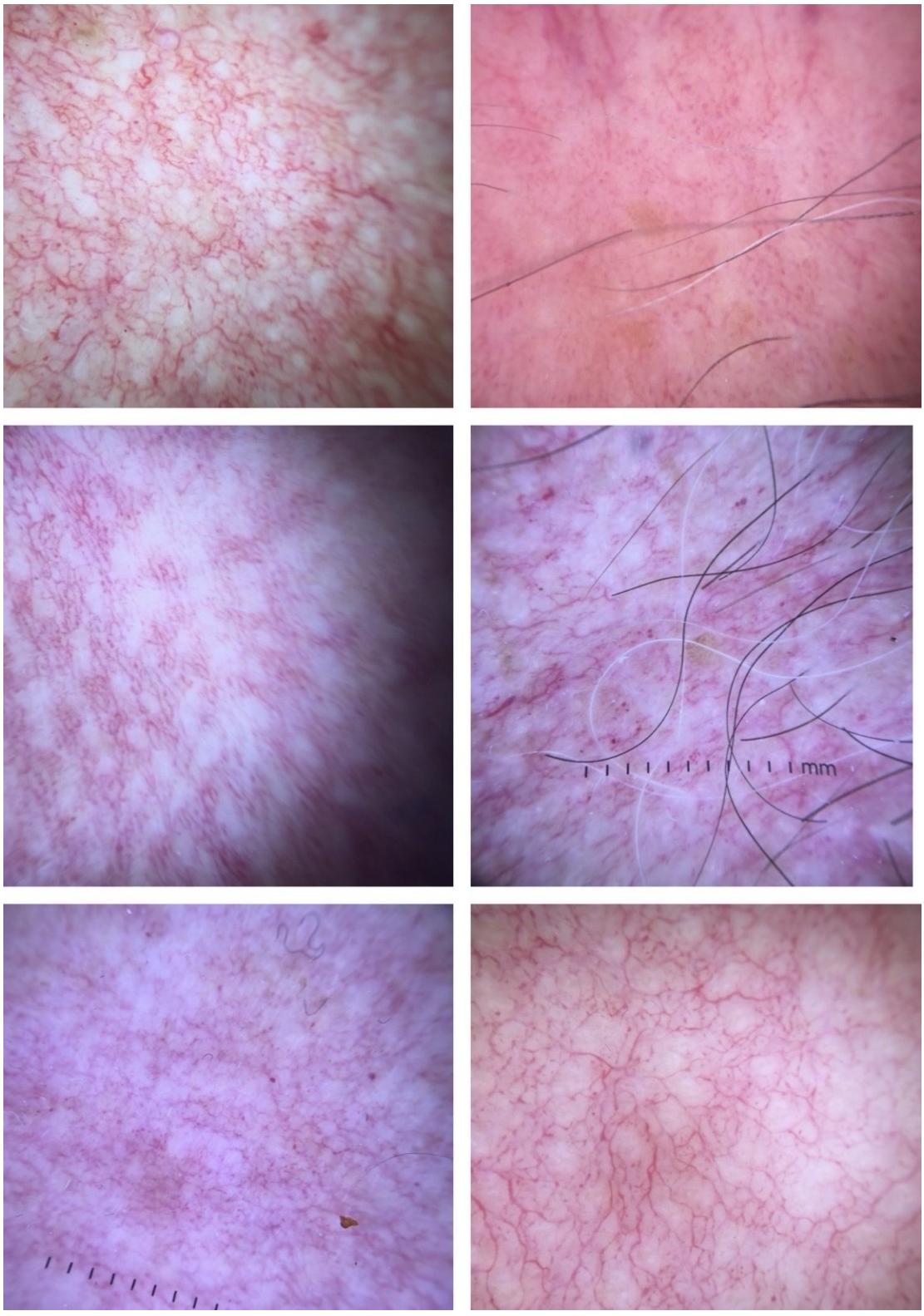

**Figure 15.** The linear irregular vessels.

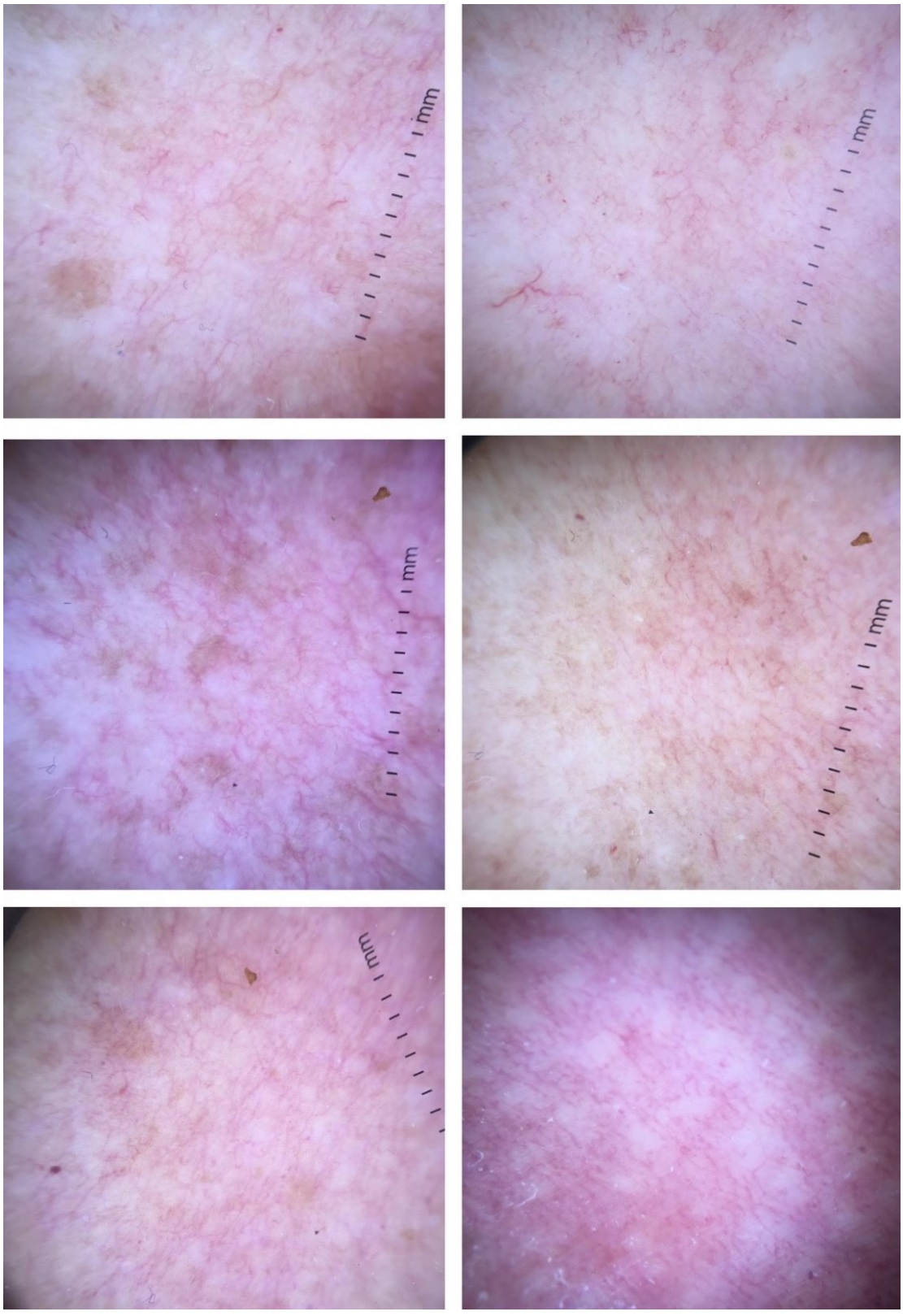

**Figure 16.** The white macules.

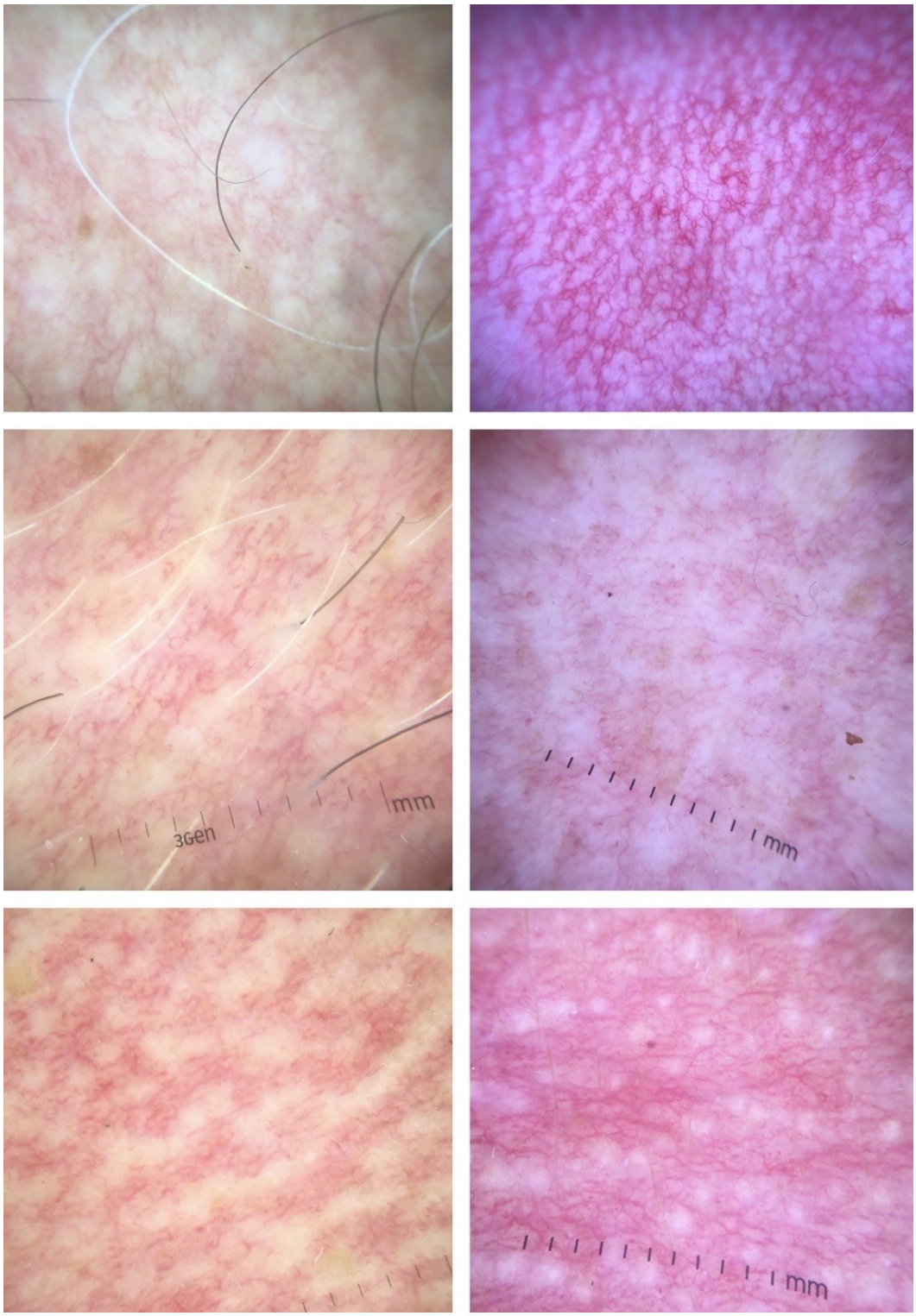

**Figure 17.** The brown macules.

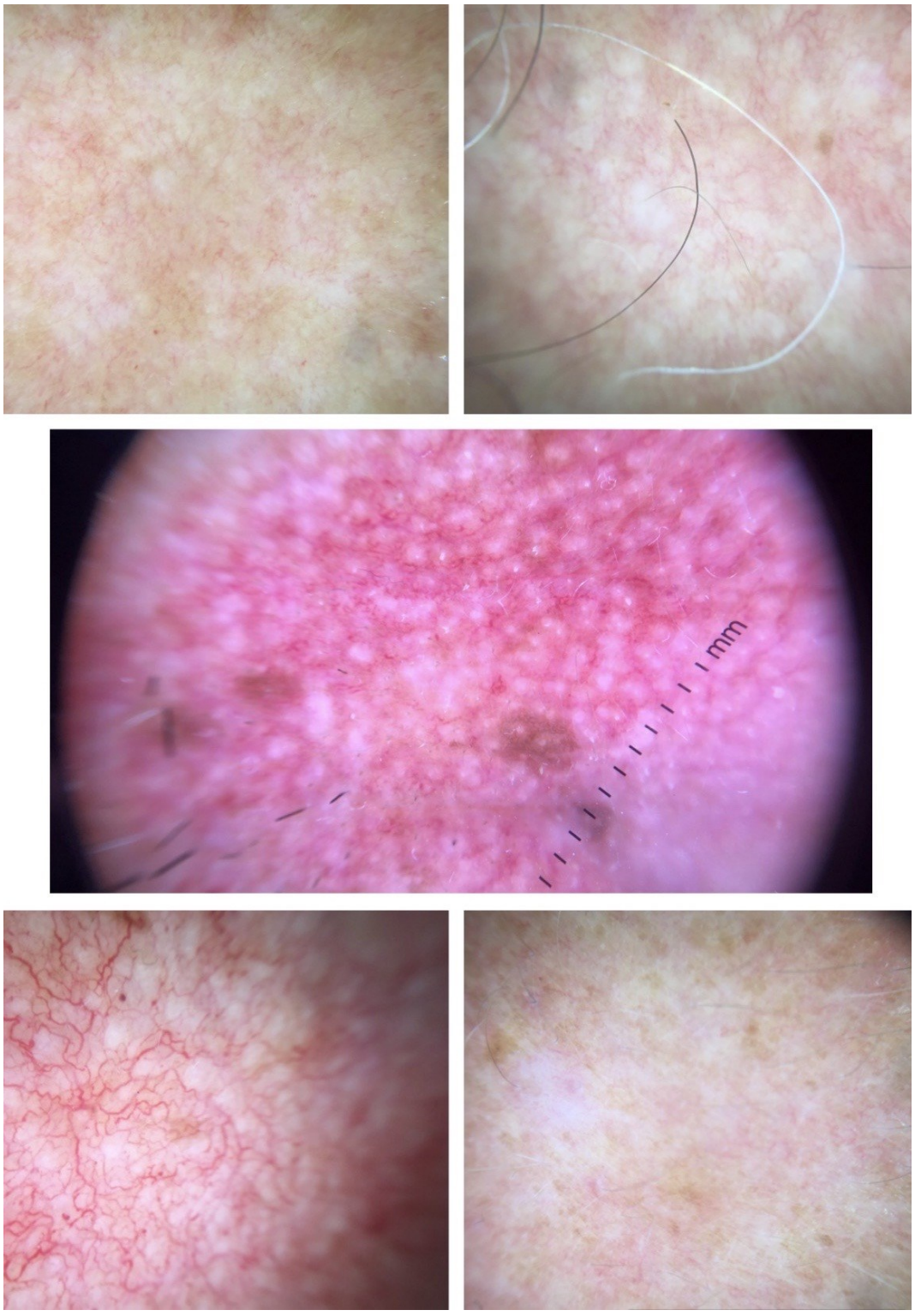

**Figure 18**. The follicular plugs.

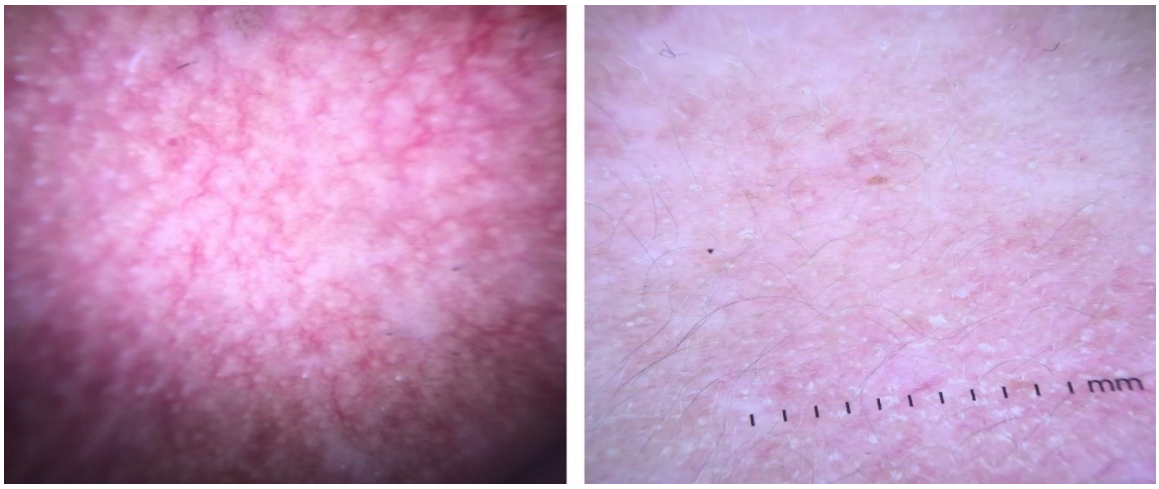

**Figure 19.** Videodermoscopy, Case 1: Male, 61 years old with mixed clinical type. Linear telangiectasias, including flying seagull-like vessels, white areas of sparing and light brown macules can be seen (magnification x20).

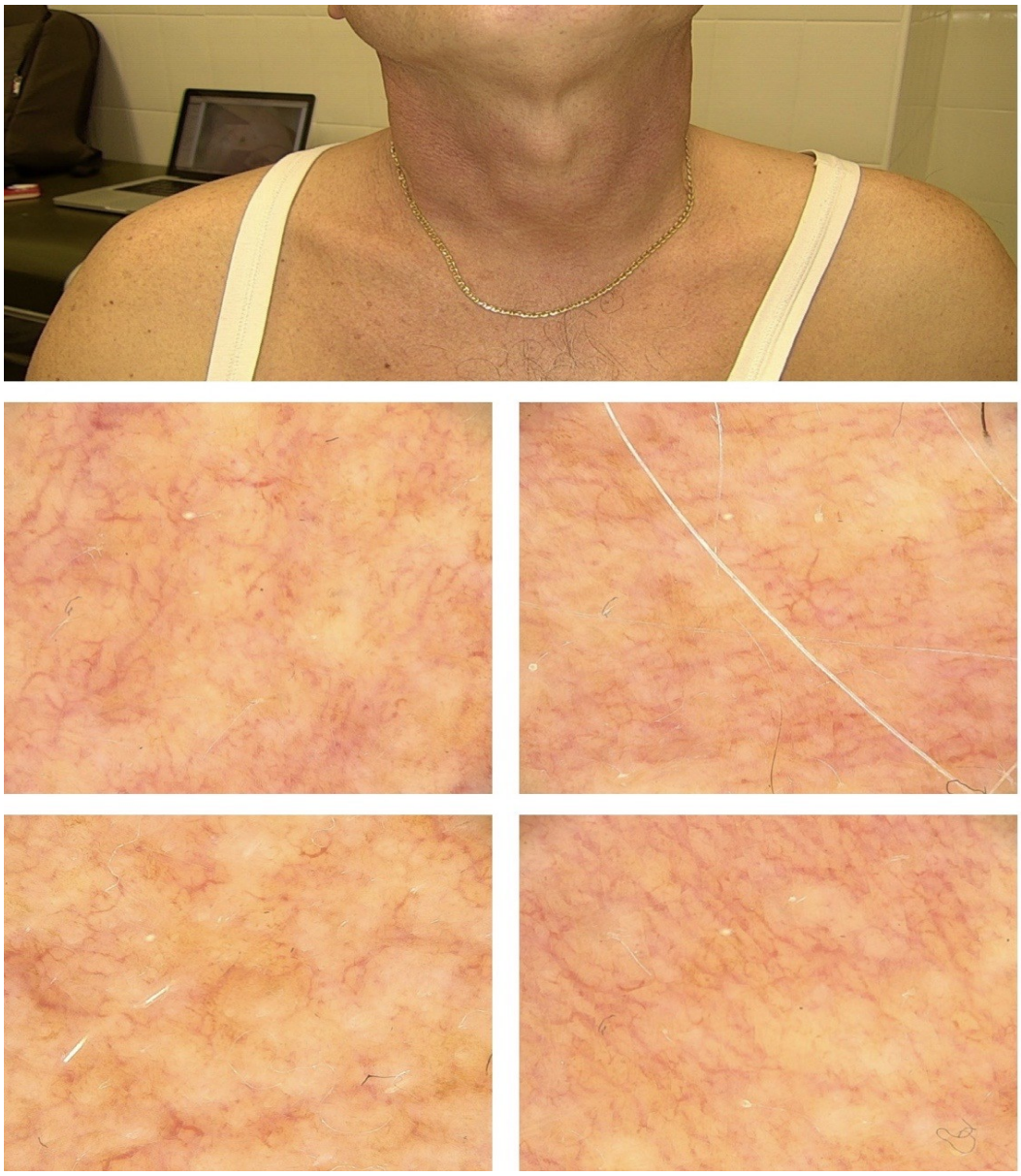

**Figure 20.**Videodermoscopy, Case 2: Female, 57 years old with erythemato-telangiectatic clinical type. Large linear irregular vessels and dotted/globular vessels, white macules and white spared areas can be seen (magnification x20).

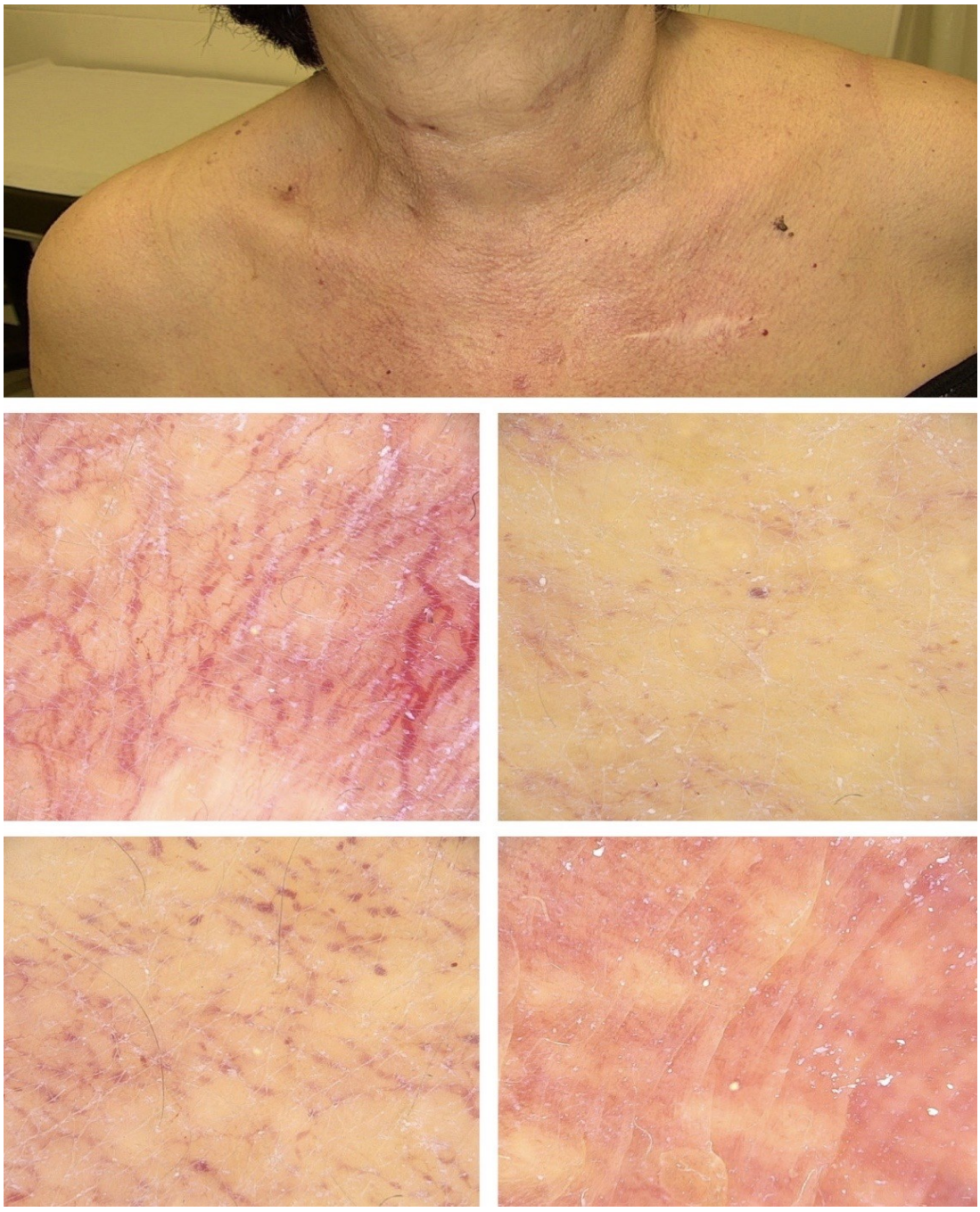

**Figure 21.** Videodermoscopy, Case 3: Female, 64 years old with pigmented clinical type. Linear telangiectasias appearing as polygonal, flying seagull-like and linear irregular vessels, white areas of sparing and light brown macules can be seen (magnification x20).