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Diagnosis and Differential Diagnosis of Poikiloderma of Civatte: A Dermoscopy Cohort Study

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ABSTRACT

Introduction: Poikiloderma of Civatte (PC) is a common, acquired, chronic, benign poikiloderma of the neck and face, most commonly affecting peri-menopausal females. At the time of writing, few studies have been published regarding the dermoscopy of PC.

Objective: To describe the dermoscopic picture of PC, so as to provide a clinico dermoscopic diagnosis and differential diagnosis for PC.

Methods: Twenty-eight patients with PC, aged 26-73 years, of whom 19 females (67.86%) were evaluated by detailed history, clinical examination, and dermoscopic examination with hand-held dermoscope.

Results: The reticular pattern was observed in 15 cases (53.6%); the white dot in 10 (35.7%); the non-specific in 9 (32.1%); and the combination of linear and dotted vessels in 8 (28.6%). Regarding local dermoscopic features, converging curved vessels were observed in 18 cases (64.3%); linear irregular vessels in 17 (60.7%); rhomboidal/polygonal vessels in 15 (53.6%); dotted/globular vessels in 10 (35.7%); white macules in 23 (82.1%); brown macules in 11 (39.3%); and whitish follicular plugs in 6 (21.4%).

Conclusions: The dermoscopic picture of PC is highly characteristic and corresponds well to both clinical and histological findings. Dermoscopy may assist clinical diagnosis, as well as the differentiation from other dermatoses of the neck and face, especially poikilodermas with guarded prognosis.

Introduction

Poikiloderma of Civatte (PC) is a rather common benign dermatosis of the neck and face, mainly affecting fair-skinned individuals, especially postmenopausal females. It is characterized by a combination of a reticular pattern of linear telangiectasia, mottled hyperpigmentation and superficial atrophy [1,2]. Clinically, it involves symmetrically sun-exposed areas of the face, the neck, and the V-shaped area of the chest, invariably sparing the anatomically shaded areas [1,2]. Depending on the prevalent clinical feature, PC can be classified into erythemato-telangiectatic, pigmented, and mixed clinical types [2]. The etiopathogenesis of PC is incompletely understood. Exposure to ultraviolet radiation, hormonal changes of menopause, contact sensitization to perfumes and cosmetics, and normal ageing have been incriminated. The diagnosis is usually clinical and can be confirmed by histology, which is characteristic, but not pathognomonic [3]. The course is slowly progressive and irreversible, often causing significant cosmetic disfigurement.

Dermoscopy is an in vivo diagnostic technique that has considerably increased our diagnostic skills. Its use has been extended, apart from Dermato-oncology, in almost every disease in the context of General Dermatology [4-6]. At the time of writing, very few studies have been published regarding the dermoscopy of PC.

Objectives

The aim of the present study was to describe the dermoscopic picture of PC. Furthermore, we sought to assist the differentiation of PC from other dermatoses of the face and neck and most importantly from acquired poikilodermas with poor or guarded prognosis (poikiloderma vasculare atrophicans and poikiloderma associated with collagen vascular disease), thus reducing the need for biopsy and histologic verification [1,2].

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 a clinical diagnosis of PC, visiting the outpatient clinic of our department, were recruited after they had given their informed consent. The study was approved by the Ethics Committee of the Hospital.

In all patients, a detailed history was obtained which included: demographic characteristics; family history with an emphasis on the presence of a similar condition in other family members; medical history, including drug history; in females, gynecological history and menstrual status; skin phototype according to Fitzpatrick's classification; sun exposure habits: occupational or recreational, and the level of exposure; use of sunscreens; use of fragrances or fragranced cosmetics, applied on the sides of the neck or the décolleté area of the chest; and history of PC.

During clinical examination, the following clinical parameters were recorded: location; distribution; clinical type; and presence of clinical manifestations of rosacea. The dermoscopic examination was performed by an experienced evaluator (A.C.K.). For the dermoscopic examination, a Dermlite DL200Hybrid (3Gen, San Juan Capistrano CA, USA) hand-held dermoscope was used. Examination employed polarized light and a 10-fold magnification 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 using an iPhone 8 camera. Photographs were taken from seven preselected sites, the same for all patients, on the upper chest, the sides of the neck, and the peripheral face in a symmetrical manner.

Firstly, we attempted to describe the local features, pigmented or vascular, that predominated on dermoscopic examination among our patients. The following structures were assessed: (i) rhomboidal/polygonal vessels, (ii) linear irregular vessels, (iii) dotted/globular vessels, (iv) brown macules, (v) white macules, (vi) follicular plugs. Secondly, we analyzed and described the global pattern.

Statistical Methods

For continuous variables, the mean, standard deviation and range, or the median, 25th and 75th 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

The demographics, etiologic factors and clinical characteristics of our patients are summarized in Table 1. 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 age 54 years), and 9 males aged 26-65 years (median age 59 years).

On clinical examination, the mixed type was the most prominent (71.43%), followed by the erythemato-telangiectatic type (21.43%) and the pigmented type (7.14%). The

Table 1. Demographics, etiologic factors and clinical characteristics of the patients with PC (N=28).

Demographics				
Gender	Females 19 (67.86)		Males 9 (32.14)	
Age	55 years		59 years	
Etiologic factors				
Phototype (Fitzpatrick's classification)	I	II	III	IV
	3 (10.71)	7 (25)	14 (50)	4 (14.29)
Occupational sun exposure	3 (10.71)			
Recreational sun exposure	22 (78.57)			
Sunburn in childhood	2 (7.14)			
Use of perfumes	16 (57.14)			
Menopause	13 (68.42)			
Positive family history	9 (32.14)			
Clinical characteristics				
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 of the lesions	V-shaped area of chest:	Sides of the neck:	Peripheral face:	
	27 (96.43)	11 (39.29)	8 (28.57)	
Clinical type	Erythemato-telangiectatic:	Pigmented:	Mixed:	
	6 (21.43)	2 (7.14)	20 (71.43)	
Symptoms	Burning:	Flushing:	Pruritus:	
	8 (28.57)	6 (21.42)	5 (17.86)	
Comorbidities	Rosacea	Thyroid diseases	Arterial hypertension:	Systemic lupus erythematosus:
	12 (42.8)	8 (28.57)	5 (17.85)	4 (14.28)

Numbers in parenthesis represent percentages

most common localization was the V-shaped area of the chest. Less than one-third of the patients complained of accompanying symptoms (burning, flushing and pruritus).

The most commonly reported comorbid skin condition was rosacea (mainly of the erythemato-telangiectatic type), observed in approximately half of the patients. Moreover, disorders of the thyroid gland were also common, involving 28.57% of the patients. It is of interest that 4/28 cases (14.28%) had a history of systemic lupus erythematosus.

On dermoscopic examination, the local dermoscopic features were assessed. We observed linear irregular vessels in 17 cases (60.7%); rhomboidal/polygonal vessels in 15 cases (53.6%); dotted/globular vessels in 15 cases (53.6%); white macules in 23 cases (82.1%); brown macules in 11 cases (39.3%); and whitish follicular plugs in 6 cases (21.4%). In 18 cases (64.3%), we identified a distinctive type of vessels, the converging curved vessels, presenting as two curved red lines that meet at their neighboring end, giving the impression of a flock of seagulls flying with their wings wide open ("flying-seagull-like" vessels). Examples of the local dermoscopic features observed among patients with PC are shown in Figure 1.

Based on the presence of the aforementioned vessel types and their architectural distribution, we described four patterns:

- Reticular (fishnet-like) pattern: areas with linear telangiectasias that are interconnected forming an irregular red network that is reminiscent of a fishnet. This network consists of thin red lines and quadrilateral or polygonal, irregular openings.
- White dot (red-white polka dot) pattern: white roundish macules, regularly distributed in areas of bright red erythema produced by a network of linear telangiectasias. It is reminiscent of a red-white polka dot print.
- Combination of linear and dotted vessels pattern: a combination of linear irregular vessels and dotted/globular vessels. This pattern was initially described in patients with PC by Errichetti and Stingo as "spaghetti and meatballs" pattern.[7]
- Non-specific pattern: areas of irregular linear telangiectasias that are irregularly distributed and do not correspond to any specific pattern.

The dermoscopic global patterns are summarized in Table 2. The reticular pattern was observed in more than

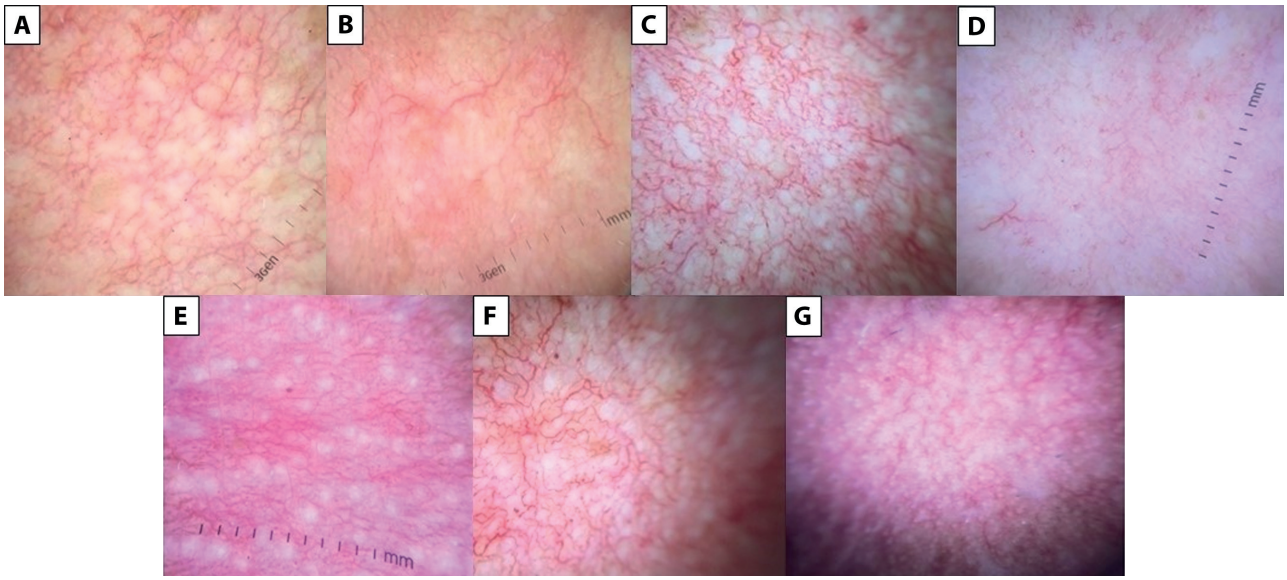
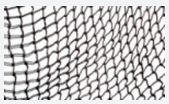
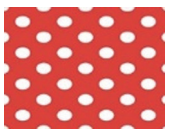


Figure 1. **"Local structures of poikiloderma of Civatte".** **A**) The rhomboidal/polygonal vessels, **B**) the converging curved vessels, **C**) the dotted/globular vessels, **D**) the linear irregular vessels, **E**) the white macules, **F**) the brown macules, and **G**) the follicular plugs.

Table 2. Dermoscopic features in patients with PC (N=28).

Dermoscopic patterns/structures	Description	Number of patients n (%)
Global pattern		
Reticular (fishnet-like) 	Areas with linear telangiectasias that are interconnected forming an irregular red network that is reminiscent of a fishnet. This network consists of thin red lines and quadrilateral or polygonal, irregular openings.	15 (53.57)
White dot (red-white polka dot) 	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.	10 (35.71)
Combination of linear and dotted vessels (spaghetti and meatballs-like[7]) 	A combination of linear irregular vessels and dotted/globular vessels.	4 (14.28)
Non-specific	Areas of linear telangiectasias that are irregularly distributed and do not correspond to any specific pattern.	9 (32.14)
Vascular structures		
Rhomboidal/polygonal	Bright red, non-branching, linear telangiectatic vessels; tend to connect with surrounding vessels forming interconnected rhomboidal or polygonal vascular structures, thus resembling parts of fishnet with irregular holes.	15 (53.57)
Linear irregular	Bright red, non-branching, linear telangiectasias with irregular form and distribution.	17 (60.71)
Dotted/globular	Bright red, irregularly distributed dots or globules.	10 (35.71)

Dermoscopic patterns/structures	Description	Number of patients n (%)
Converging curved vessels (Flying seagull-like) 	Double curved short red lines that meet at their neighboring end; usually appear in small clusters.	18 (64.29)
Pigmented structures		
Brown macules	Light brown structureless macules, 2-3 mm in diameter, with rather distinct but slightly irregular borders.	11 (39.29)
Depigmented structures		
White macules	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.	23 (82.14)
White keratotic follicular plugs	Follicular hyperkeratosis	6 (21.42)

Table 3. Dermoscopic differential diagnosis of poikiloderma of Civatte.[11-23]

	Dermoscopic findings
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 in dermatomyositis	• Enlarged linear irregular vessels; • Mixed features of hyperpigmentation and depigmentation.
Poikilodermatous mycosis fungoides	• Multiple polygonal structures consisting of lobules of white storiform streaks, studded with fine red dots or hairpin vessels; • Unevenly and intermittently distributed septa of pigmented dots, between the lobules; • Red and yellowish smudges.
Poikiloderma atrophicans et vasculare	• Blurred branched vessels on a reddish or orangish-brown background; • Sparse whitish scales.
Chronic Graft-versus-host disease	• Whitish scales; • Vessels of mixed morphology, mostly dotted and linear
Melasma	• Light-to-dark brown background; • Brown granules/ globules with perifollicular sparing; • Global reticular or pseudo-reticular pattern.

half of the patients, followed by the white dot pattern, and the non-specific pattern, while the combination of linear and dotted vessels pattern was the least frequently identified. In 78.57% of the cases, the coexistence of more than one pattern at different sites was noted. 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 observed among our patients are shown in Figure 2.

In our statistical analysis, we investigated possible correlations of the dermoscopic features and patterns with epidemiologic parameters, etiologic factors and clinical characteristics of our PC patients (Table 4). The following statistical correlations were documented: white macules were correlated with the mixed clinical type (p=0.015); converging curved 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

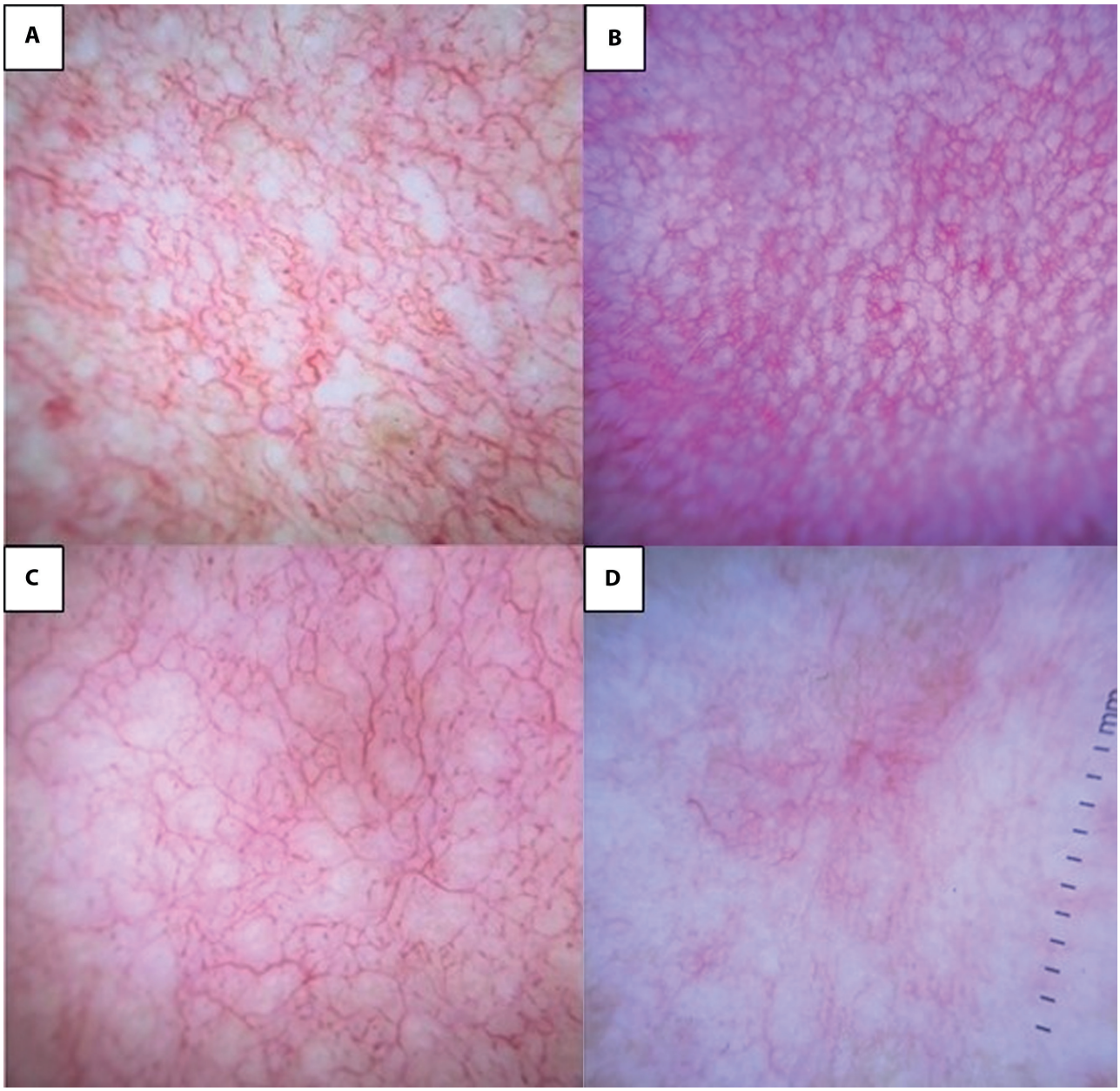


Figure 2. "Global patterns of poikiloderma of Civatte". **A**) The fishnet-like pattern, **B**) the red-white polka dot pattern, **C**) the combination of linear & dotted vessels pattern, and **D**) the non-specific pattern.

Table 4. Distribution of local dermoscopic features by clinical type among patients (N=28) with poikiloderma of Civatte.

	No of patients (%)		
Type of PC Dermoscopic Features	Erythemato-telangiectatic type (N=6)	Pigmented type (N=2)	Mixed type (N=20)
Rhomboidal/polygonal vessels	2 (33.33)	0	13 (65)
Dotted/globular vessels	4 (66.66)	1 (50)	11 (55)
Linear irregular vessels	3 (50)	2 (100)	13 (65)
Converging curved vessels	6 (100)	1 (50)	12 (60)
White macules	3 (50)	1 (50)	19 (95)
Brown macules	1 (16.67)	2 (100)	9 (45)

>5 years (p=0.009). Nevertheless, the number of cases in our study was relatively small to allow the extrapolation of solid conclusions.

Conclusions

As our results indicate, PC exhibits a rather characteristic dermoscopic picture. The dermoscopic findings in PC consist of vascular and pigmented local features, forming a reddish-to-brownish network. We were able to describe four distinct global patterns (reticular, white dot, combination of linear and dotted vessels, and non-specific). The reticular pattern was the most frequently identified (53.57%). In the majority of patients, the coexistence of more than one pattern was noted. We were able to describe four types of telangiectatic vessels (converging curved, rhomboidal/polygonal, dotted/globular, and linear irregular). Converging curved vessels have not been described previously in any skin condition and could be considered highly characteristic of the erythemato-telangiectatic type of PC (p=0.028). White macules were seen in the vast majority of our patients and in all clinical types, either regularly (white dot pattern) or irregularly (reticular or non-specific pattern) distributed and in a follicular or pseudo-follicular distribution. Brown macules were observed much less often, mostly associated with the pigmented and the mixed clinical type.

In the literature, there is little published experience on the 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.[7] The authors described, in all patients, 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.[7] Our findings are in line with the findings of Errichetti and Stinco. However, the combination of linear and dotted vessels pattern was not recognized by us as the predominant global pattern, but it was seen in as much as 28.6% of our patients. The presence of dotted/globular vessels was less common in our cohort as well. Accordingly, for the white macules, we noticed that they often have a pseudo-follicular distribution, i.e. they are not strictly related to the follicular openings. In our cohort, brown macules were mainly associated with skin phototype IV (p=0.016) and with a disease duration >5 years (p=0.009). Brown macules were mostly identified in the pigmented and mixed clinical types of PC.

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 (converging curved vessels, rhomboidal/polygonal vessels, linear irregular vessels, dotted/globular vessels) that we identified 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.[3] When the course of the vessels is parallel to 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 [3]. In our series, brown macules correlated with the mixed type of PC (p=0.015). 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 [3].

PC presents with a distinctive dermoscopic picture. By dermoscopy, PC can be easily differentiated from other skin conditions characterized also by telangiectasia and reticular pigmentation, such as rosacea, erythromelanosis follicularis faciei et colli and melasma, or true poikiloderma. Rosacea which often coexists with PC most commonly involves the central face. In rosacea, linear telangiectatic vessels arranged in horizontal and vertical lines form polygons (polygonal vessels) [5, 8, 9]. Vascular polygons are a characteristic dermoscopical feature of erythemato-telangiectatic rosacea [6]. 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 [10]. In rosacea, additional features such as rosettes, white/yellowish scales, orange-yellowish areas, pigmentation structures, dilated follicles and follicular pustules (in the papulopustular form), have been described that were not present in our cohort allowing differentiation between these entities [5, 8, 9]. The dermoscopic differential diagnosis of PC is depicted in Table 3.

Our study has several limitations. Due to the small number of patients, our results need further confirmation in larger prospective cohorts. Additionally, due to the lack of a control group, the sensitivity and specificity of the proposed criteria were not calculated.

To our knowledge, this is the first study to systematically investigate the dermoscopic characteristics of PC. Although not pathognomonic, the dermoscopic picture is highly 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 are rarely necessary. Future research will, hopefully, better clarify the pathogenetic mechanisms of this disease and will provide insights for more effective preventive and therapeutic approaches for PC.

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