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Primary cicatricial alopecia subtypes: Clinical and histopathological patterns in a large Turkish cohort

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MAIN POINTS

- The largest series of PCA from Türkiye (n = 116, histopathologically confirmed).
- DLE was the most common (60.3%), followed by LPP (28.4%); FFA was rare (4.3%).
- DLE showed basement membrane thickening and dermal mucin; whereas LPP showedsebaceous gland atrophy and epidermal hypertrophy.
- Clinicopathological correlation is essential for accurate diagnosis.

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ABSTRACT

Aim: Primary cicatricial alopecias (PCAs) are a group of rare dermatological disorders characterized by irreversible destruction of hair follicles due to inflammatory processes, leading to permanent hair loss. The diagnostic process is often delayed because of nonspecific clinical presentation and overlapping histopathological features. This study aimed to analyze the clinical and histopathological characteristics of PCA subtypes diagnosed in a tertiary dermatology clinic in Türkiye over a 17-year period.

Materials and Methods: A retrospective analysis was conducted on 116 cases of histopathologically confirmed PCA between January 2008 and January 2025. Clinical diagnoses included discoid lupus erythematosus (DLE), lichen planopilaris (LPP), frontal fibrosing alopecia (FFA), pseudopelade of Brocq (PPB), and folliculitis decalvans (FD). Histopathological features were systematically reviewed and statistically analyzed using the Statistical Package for the Social Sciences version 25.0.

Results: The most frequent subtype was DLE (60.3%), followed by LPP (28.4%). DLE was associated with basal membrane thickening (82.9%) and dermal mucin deposition (71.4%), whereas LPP showed higher rates of sebaceous gland loss (51.5%) and interface dermatitis (54.5%). FFA exhibited overlapping features with LPP, including 100% basal cell degeneration. Statistically significant associations were found between specific histopathological patterns and clinical subtypes.

Conclusion: This study provides one of the comprehensive PCA datasets in Türkiye, highlighting distinct histopathological patterns associated with specific clinical subtypes. These findings underscore the importance of clinicopathological correlation in PCA diagnosis and contribute valuable data for improving diagnostic accuracy and patient management.

Keywords: Cicatricial alopecia, Lichen planopilaris, Discoid lupus erythematosus, Histopathology, Scalp diseases

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INTRODUCTION

Primary cicatricial alopecias (PCAs) are rare dermatological disorders that cause irreversible damage to the hair follicle and the surrounding structures due to inflammatory processes. The epithelial stem cells of the hair follicle are irreversibly destroyed, and their replacement by fibrous tissue leads to permanent hair loss. Psychological distress and reduced quality of life are associated with PCAs [1,2]. The prevalence of PCAs among all patients presenting to dermatology outpatient clinics is approximately 0.09% [1], and they constitute 2.1%–7.3% of all hair diseases [3–5]. This disease group exhibits a complex etiopathogenesis and a variable clinical course.

Although different classification systems have been proposed for PCAs, the North American Hair Research Society (NAHRS) classification is the most widely accepted model, which categorizes PCAs according to the predominant inflammatory infiltrate observed in histopathology. Ac-

cording to this classification, PCAs are classified into 4 main groups. The lymphocyte-predominant group includes patients with discoid lupus erythematosus (DLE), lichen planopilaris (LPP) and its subtypes, pseudopelade of Brocq (PPB), and central centrifugal cicatricial alopecia (CCCA). The neutrophil-predominant group includes folliculitis decalvans (FD) and dissecting cellulitis (DC). The third group, which shows mixed inflammatory cell infiltrates, includes acne keloidalis nuchae (AKN). Cases that do not show a specific inflammatory cell type are classified as nonspecific [6].

The diagnosis of PCAs can be challenging for both dermatologists and pathologists, particularly when patients present at advanced stages in which histopathological characteristics may no longer be specific. Therefore, early diagnosis largely depends on the combined evaluation of clinical findings and histopathological data [7]. Although many studies have investigated the distribution of PCA subtypes and their histopathological features in different geographical regions, case series from Türkiye remain limited. This retrospective study aimed to examine the demographic characteristics, clinical subtypes, and histopathological features of patients who presented to the dermatology outpatient clinic of a tertiary healthcare center and were definitively diagnosed with PCA based on clinical and histological findings. In addition, this study aims to provide local epidemiological data that may support diagnostic processes.

MATERIALS AND METHODS

In this study, the clinical and histopathological characteristics of patients diagnosed with cicatricial alopecia who presented to the dermatology outpatient clinic of a tertiary healthcare center between January 2008 and January 2025 were evaluated. A total of 119 patients were evaluated histopathologically; however, 3 patients were excluded due to missing histopathology slides and incomplete demographic data that prevented accurate classification. Consequently, 116 patients with confirmed histopathological diagnoses were included in the final analysis.

This was a descriptive, non-interventional study based on a retrospective review of existing patient records. In retrospective studies, the sample size depends on the suitability of the available records rather than the researchers' choice. Studies with similar designs can be conducted with smaller samples [8]. Therefore, in this study, the sample size was determined based on the accessible clinical records belonging to the study period.

The study was approved by the Atatürk University Non-Interventional Clinical Research Ethics Committee (date: 27.06.2025, decision no: 64). The principles of the Declaration of Helsinki were adhered to throughout the study; patient confidentiality was maintained, data were anonymized, and no identifying information was recorded. Clinical photographs were included with written informed consent and anonymized to ensure patient confidentiality. No age or sex restrictions were applied.

In our clinic, scalp biopsies are performed in patients with suspected cicatricial alopecia in the presence of clinical ambiguity, cases requiring differential diagnosis, such as suspected discoid lupus erythematosus, atypical or early-stage lichen planopilaris/pseudopelade of Brocq lesions, or lesions that are refractory to treatment.

The inclusion criteria were as follows: presentation to the dermatology outpatient clinic between January 2008 and January 2025 and confirmation of the clinical diagnosis of cicatricial alopecia by dermatopathological evaluation. Patients without a definitive diagnosis or with incomplete records were excluded from the study.

Dermatology clinic researchers retrospectively collected data using the hospital information management system, pathology reports, and clinical examination notes. For each patient, age, sex, year of diagnosis, clinical diagnosis (LPP, FFA, DLE, and PPB), and histopathological findings (epidermal atrophy and hypertrophy, follicular plugging, basal cell degeneration, basement membrane thickening, interface dermatitis, dermal mucin deposition, and sebaceous gland atrophy) were systematically recorded.

The study was conducted and reported in accordance with the STROBE guidelines.

Statistical analysis

Statistical analyses were performed using IBM SPSS Statistics version 25.0 (Armonk, NY: IBM Corp.). Descriptive statistics included mean, median, standard deviation, and percentage distributions. Pearson's Chi-square test was used for comparisons between subgroups for categorical variables. The likelihood ratio and linear-by-linear association tests were applied when appropriate. A p-value of <0.05 was considered statistically significant.

RESULTS

Among the 116 CAS included in the study, 51.7% were female (n = 60) and 48.3% were male (n = 56). most patients (52.6%) were aged between 31 and 50 years. When analyzed according to the year of diagnosis, 52.6% of the cases was diagnosed in 2020 or later.

Regarding clinical subtypes, the most common diagnosis was DLE with 60.3%, followed by LPP with 28.4%. Frontal FFA (4.3%), PPB (3.4%), FD (1.7%), and unclassified cases (1.7%) were observed less frequently. Figure 1 shows representative clinical photographs illustrating the characteristic features of the subtypes.

A statistically significant association was found between sex and clinical diagnosis (p = 0.048). LPP was more common in men, whereas FFA and PPB were detected only in women. No significant association was found between age groups and

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Table 1. Demographic characteristics of the participants and distribution of clinical diagnoses.

Variable	DLE (n=70)	LPP (n=33)	FD (n=2)	PPB (n=4)	FFA (n=5)	Unclassified (n=2)	p-value	χ^2	d
Sex							**0.048***	10.689	5
Male	34 (48.6%)	19 (57.6%)	0	1 (25%)	0	2 (100%)			
Female	36 (51.4%)	14 (42.4%)	2 (100%)	3 (75%)	5 (100%)	0			
Age Group							0.647	7.809	10
≤30	14 (20%)	6 (18.2%)	1 (50%)	2 (50%)	2 (40%)	0			
31–50	36 (51.4%)	18 (54.%5)	1 (50%)	1 (25%)	3 (60%)	2 (100%)			
≥51	20 (28.6%)	9 (27.3%)	0	1 (25%)	0	0			
Distribution by Year							**0.014***	22.211	10
2008-2013	9 (12.9%)	5 (15.2%)	0	1 (25%)	0	0			
2014-2019	32 (45.7%)	3 (9.1%)	0	3 (75%)	1 (20%)	1 (50%)			
≥2020	29 (41.4%)	25 (75.8%)	2 (100%)	0	4 (80%)	1 (50%)			

Table 2. Histopathological findings by clinical diagnosis groups.

Histopathological Features	DLE	LPP	Folliculitis decalvans	PPB	FFA	p	χ^2	d
Epidermal atrophy	39 (55.7)	18 (54.5)	0 (0.0)	4 (100.0)	2 (40.0)	0.286	6.212	5
Epidermal hypertrophy	2 (2.9)	7 (21.2)	0 (0.0)	0 (0.0)	0 (0.0)	**0.038***	11.789	5
Follicular plugging	38 (54.3)	13 (39.4)	2 (100.0)	0 (0.0)	3 (60.0)	0.073	10.073	5
Basal cell degeneration	54 (77.1)	25 (75.8)	0 (0.0)	0 (0.0)	5 (100.0)	**<0.001***	23.873	5
Basement membrane thickening	58 (82.9)	13 (39.4)	0 (0.0)	0 (0.0)	0 (0.0)	**<0.001***	40.943	5
Interface dermatitis	29 (41.4)	18 (54.5)	0 (0.0)	0 (0.0)	3 (60.0)	0.225	6.941	5
Mucindeposition dermis	50 (71.4)	1 (3.0)	0 (0.0)	0 (0.0)	0 (0.0)	**<0.001***	54.521	5
Sebaceous gland loss	14 (20.0)	17 (51.5)	2 (100.0)	4 (100.0)	3 (60.0)	**0.002***	24.630	5

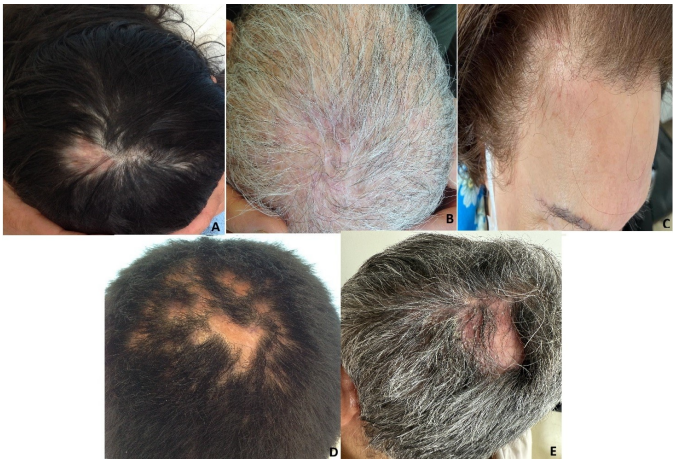

Figure 1. Representative clinical images of primary cicatricial alopecia subtypes: A) discoid lupus erythematosus: atrophic, scarring, erythematous alopecic plaques; B) Lichen planopilaris: perifollicular erythema and scaling; C) frontal fibrosing alopecia: frontal hairline recession and eyebrow loss; D) pseudopelade of Brocq: scarring alopecic patches lacking erythema; E) folliculitis decalvans: tufted appearance characterized by pustules, crusting, and multiple hairs emerging from single follicular openings.

clinical diagnoses (p = 0.647). Of the LPP cases, 75.8% was diagnosed in 2020 or later. A statistically significant association was found between the year of diagnosis and clinical subtypes (p = 0.014). Table 1 presents the demographic characteristics of the participants and the distribution of clinical diagnoses.

The most frequently observed histopathological findings were basal cell degeneration (72.4%), basement membrane thickening (61.2%), epidermal atrophy (55.2%), and follicular plugging (48.3%).

Differences were observed in the distribution of histopathological findings among PCA subtypes. Changes were present in both epidermal and dermal structures in DLE cases, including basement membrane thickening (82.9%), mucin deposition (71.4%), and basal cell degeneration (77.1%). Basal cell degeneration (75.8%), epidermal atrophy (54.5%), interface dermatitis (54.5%), and sebaceous gland atrophy (51.5%) were predominant in LPP. In addition, basement membrane thickening was less frequent in LPP (39.4%) than in DLE. In FFA cases, basal cell degeneration was present in all patients (100%). Other common findings included follicular plugging (60%), interface dermatitis (60%), and epidermal atrophy (40%) (Table 2).

Basal cell degeneration was significantly more frequent in both the LPP and DLE groups (p<0.001). Sebaceous gland atrophy and epidermal hypertrophy were also significantly more common in the LPP group (p = 0.002 and p = 0.038, respectively). In contrast, basement membrane thickening and mucin deposition were significantly more frequent in patients with DLE (both p<0.001). No statistically significant differences in epidermal atrophy (p = 0.286), interface dermatitis (p = 0.225), or follicular plugging (p = 0.073) were observed among the groups.

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DISCUSSION

This study constitutes one of the comprehensive series addressing the histopathological features of PCAs in Türkiye. The large sample of 116 patients provides significant support for correlating clinical subtypes with different histopathological patterns and for diagnostic differentiation.

In our study, DLE was the most frequently observed subtype, followed by LPP. FFA was detected in only 4.3% of the patients. PCA subtype prevalence has been reported at different frequencies in different populations. In addition to studies reporting FFA as the most common subtype and LPP as the second most common subtype [9], studies from Iran and Pakistan have reported that DLE is the most frequently observed subtype [10,11]. In other studies conducted in Türkiye, LPP has been stated as the most common subtype, whereas the frequency of DLE shows considerable variability [4,12]. The high rate of DLE in our cohort may be attributed to the frequent preference for biopsy in cutaneous lupus cases, whereas the low frequency of FFA may be explained by patients presenting to our clinic at advanced stages of the disease, at which point biopsy may no longer be considered necessary.

The frequency of FD in our study and in other series reported from our country (1.7%–2.1%) is significantly lower than that reported in studies conducted in China (40.7%) when the rarer subtypes are examined [3,4]. FD is usually clinically recognizable, and in our center, direct treatment is generally preferred over biopsy in inflammatory or infectious lesions. This clinical approach may explain the low frequency of FD observed in our study. In the literature, the prevalence of PPB has also been reported at highly variable frequencies (3.3%–40%) [1,5,7,10,13]. Among our patients, this rate was 3.4%, whereas it ranges between 5% and 8% in other Turkish case series [4,12]. This variability may be due to differences in diagnostic criteria and the conceptual ambiguity surrounding PPB. While some authors consider PPB as the end-stage manifestation of other PCAs [14], the North American Hair Research Society classifies it as a separate subtype [6]. Since biopsies in our center are generally obtained from active lesions, late-stage cases may have been excluded from histopathological evaluation.

When all findings regarding subtype distribution are evaluated, the factors influencing the distribution of PCA subtypes include not only disease prevalence but also patient presentation profile, geographical characteristics, clinical practice patterns, and institutional diagnostic approaches.

Regarding the relationship between sex and PCA subtypes, most studies report that LPP is more common in women [13,15]. Similar findings have also been reported in prevalence studies conducted in neighboring countries, such as Iran and Türkiye [4,11,12]. Contrary to the female predominance frequently described in the literature, LPP was more commonly observed in men in our study. This inconsistency may be associated with sampling characteristics specific to the center, differences in diagnostic practices, or the possibility that LPP variants observed in men may exhibit more prominent clinical features.

FFA and PPB were observed only in female patients in our study, which is largely consistent with the literature. FFA is particularly common in postmenopausal women, and this may be attributed to hormonal, immunological, or genetic factors [16,17]. Similarly, female predominance has also been reported in the literature [1,4,7,10].

No statistically significant relationship was found between age groups and clinical subtypes among the patients examined. However, FFA and LPP are more common in older adults, whereas DLE and PPB are more prevalent in younger adults [1,4,10].

FFA is considered a clinical variant or subtype of LPP [18], and several studies have emphasized a marked increase in FFA incidence over the past decade [5,9,16]. This increase may be attributed to the widespread use of noninvasive diagnostic tools, such as trichoscopy, increased awareness of hair disorders, earlier dermatological consultations, better clinical recognition of LPP, and improved sensitivity in histopathological evaluations [19,20]. In our study, 75.8% of patients diagnosed with LPP received their diagnosis in 2020 or later.

The most frequently reported histopathological findings were basal cell degeneration, basement membrane thickening, epidermal atrophy, and follicular plugging. Sardar et al. [1] reported that lymphocytic infiltration was the predominant feature and that basal cell degeneration and follicular plugging were the most frequently observed findings. Similarly, Hashmi et al. [10] described lymphocytic infiltration, perivascular infiltrates, sebaceous gland loss, perifollicular fibrosis, and epidermal atrophy. The histopathological features frequently observed in patients with DLE and LPP were largely consistent with the classical histopathological patterns described in the literature [1,10].

Basal cell degeneration was significantly higher in both the LPP and DLE groups. Follicular lichenoid inflammation and vacuolar degeneration are defined as characteristic findings in LPP [21]. Bernárdez et al. [22] reported that vacuolar interface dermatitis is commonly observed in DLE, whereas lichenoid interface changes are more typical in LPP, usually accompanied by basal cell degeneration.

In our study, basement membrane thickening and dermal mucin deposition were significantly more frequently observed in patients with DLE. Sardar et al observed these findings only in patients with DLE, whereas Hashmi et al. reported basement membrane thickening in all patients with DLE [1,9]. A PAS-positive thickened basement membrane has been defined as a classic feature of DLE [22], and this finding has been emphasized as being rarely observed in LPP [23]. Similarly, dermal mucin deposition is considered distinctive in lupus-related diseases [22,24].

In our study, sebaceous gland atrophy and epidermal hypertrophy were significantly more common in the LPP group

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than in the other cicatricial alopecia subtypes. However, the literature indicates that sebaceous gland loss is commonly observed not only in LPP but also in other PCA subtypes such as FFA, FD, and PPB [1]. Loss of sebaceous glands is commonly observed not only in LPP but also in other PCA subtypes, such as FFA, FD, and PPB [1]. Although Hashmi et al. [9] reported this finding most prominently in PPB cases, Inchara et al. [24] questioned its utility as a distinguishing diagnostic feature, as it can be seen in both LPP and DLE. Sebaceous gland atrophy may appear in the early stages of the disease [22].

Although epidermal hypertrophy is more commonly associated with DLE, FFA, and acne keloidalis nuchae (AKN), it has been reported in 20% of patients with LPP, similar to our study [1]. This rate was found to be statistically significant. Methodological factors such as sample size or variation in biopsy site selection may be responsible.

Limitations

This study has several limitations, such as its relatively small sample size, which included only histopathologically confirmed PCA cases in a tertiary healthcare center, and its cross-sectional design, which prevented the evaluation of the clinical course. Other important limitations include limited clinical data and the absence of trichoscopic evaluation. Methodologically, multiple comparison corrections were not applied because the comparison of more than two groups was descriptive in nature; this constitutes a limitation in the interpretation of the p-values. In addition, the small sample sizes in some subtypes partially restricted the chi-square test assumptions. Because the groups reflect the true distribution, they were not combined; therefore, Fisher's exact test would have been more appropriate in some analyses.

CONCLUSION

This retrospective study comprehensively analyzed the clinical and histopathological features associated with the subtypes of PCA. Findings such as basement membrane thickening and dermal mucin deposition in DLE and basal cell degeneration and sebaceous gland atrophy in LPP were identified as relatively specific features. Our findings emphasize that histopathological evaluation alone may not be sufficient for an accurate diagnosis and that diagnostic accuracy increases when histological findings are interpreted in conjunction with the clinical context. In this regard, our study represents one of the largest PCA case series reported from Türkiye.

Ethics Committee Approval: The study was approved by the Atatürk University Non-Interventional Clinical Research Ethics Committee (date: 27.06.2025, decision no: 64).

Informed Consent: Clinical photographs used in the manuscript were obtained after written informed consent from the patients. For the retrospective chart review component, informed consent was waived due to the anonymized nature of the data and the retrospective study design.

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