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PRIMARY IDIOPATHIC PSEUDOPELADE OF BROCQ: A CASE SERIES

Pathology

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ABSTRACT

Pseudopelade of Brocq (PPB) is a rare self-limiting hair disorder. The parietal scalp and vertex are commonly involved. The patient presents with small patches of alopecia with mild to moderate atrophy. There are no signs of folliculitis or marked inflammation or widespread scarring. PPB can be primary or secondary. Secondary PPB can occur as end stage of other alopecias such as lichen planopilaris or discoid lupus erythematosus. The diagnosis is based on clinical, histopatological and immunofluoresecence features. Here we report four such cases of Pseudopalade of Brocq.

KEYWORDS

Alopecia, foot print in snow, pseudopelade of Brocq

INTRODUCTION

Pseudopelade of Brocq (PPB) is a rare self-limiting hair disorder. The usual age of presentation is around 40 years and females are more commonly affected. The patient presents with small patches of alopecia with mild to moderate atrophy.[1] There are slightly depressed patches of irreversible alopecia occurring singly or in groups. These lesions have been described as 'footprints in the snow'.[2] The parietal scalp and vertex are commonly involved. PPB can be primary or secondary. Secondary PPB can occur as end stage of other alopecias such as lichen planopilaris or discoid lupus erythematosus. The diagnosis is based on clinical, histopatological and immunofluoresecence features.[3] Here we report four such cases of Pseudopalade of Brocq having no known underlying cause and final diagnosis was rendered on histopathological examination.

CASE STUDY

Case 1

A 21 year old female presented with complaints of patchy hair loss for last 3 years and it was progressive. It was not associated with any evidence of inflammation or trauma to the scalp. No family history of similar complaints was present. On dermoscopy, empty hair follicles were visible. Alopecia areata, lichen planopilaris and Pseudopalade of Brocq were kept as differential diagnosis and skin biopsy was done. On microscopic examination, section showed stratified squamous epithelium displaying unremarkable histomorphology. Underlying dermis showed absence of pilosebaceous unit with presence of normal arrector pili muscle and normal sweat glands.

Case 2

A 27 year old male presented with complaints of patchy hair loss for more than one month and it was progressive. It was not associated with any evidence of inflammation or trauma to the scalp. No family history of similar complaints was present. On examination, there was patchy hair loss over scalp giving a moth eaten appearance along with apparent epidermal and dermal scarring. Syphilitic alopecia, alopecia areata, lichen planopilaris and Pseudopalade of Brocq were kept as differential diagnosis and skin biopsy was done. On microscopic examination, section showed tissue lined by stratified squamous epithelium showing basket weave hyperkeratosis. Mild inflammatory infiltrates were seen in the dermis. Parafollicular fibrosis was present along with fibrotic bands in the dermis reaching upto the subcutaneous fat. There was absence of pilosebaceous unit with presence of normal erector pilli muscle and sweat glands.

Case 3

A 34 year old male presented with complaints of patchy hair loss over scalp since last 6 months. No associated complaints or similar history in family noted. No history of any trauma present. Alopecia areata, lichen planopilaris and Pseudopalade of Brocq were kept as differential diagnosis and skin biopsy was done. On microscopic examination, section showed tissue lined by stratified squamous epithelium showing basket weave hyperkeratosis. Mild inflammatory infiltrates were seen in the dermis. Parafollicular fibrosis was present along with fibrotic bands in the dermis reaching upto the subcutaneous fat. There was absence of pilosebaceous unit with presence of normal erector pilli muscle and normal sweat glands.

Case 4

A 45 year old male presented with complaints of patchy hair loss over scalp since 3 months.

Lichen planopilaris, Pseudopalade of Brocq and central centrifugal cicatricial alopecia were kept as differential diagnosis and skin biopsy was done. On microscopic examination, section showed tissue lined by stratified squamous epithelium showing basket weave hyperkeratosis and parakeratosis. Upper and mid dermis showed presence of fibrosis replacing hair follicles and normal sebaceous glands.

The histopathological findings are shown in figure 1.

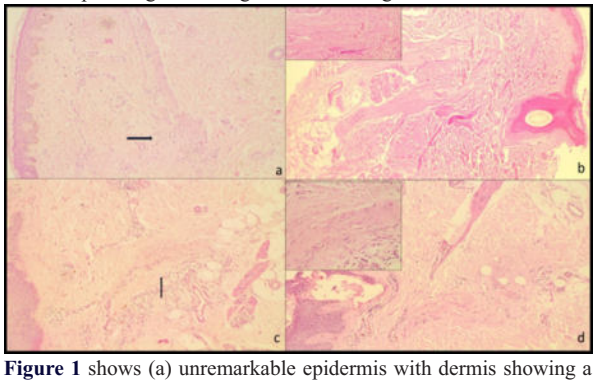

Figure 1 shows (a) unremarkable epidermis with dermis showing a follicular scar traversing the dermis longitudinally abutting the arrector pili muscle (arrow) (100 X, H & E)
(b)Linear scar running perpendicular to the epidermis (100X, H& E). Inset shows a scar aside the arrector pili muscle. (400 X, H&E).
(c) Column of scarring replacing hair follicle (arrow) (100X, H&E).
(d) Destruction of hair follicle with replacement by fibrous tract. (100, H &E) Arrectors muscle is spared. Inset shows follicular scarring (400X, H &E)

Based on the histopathological features, the final diagnosis of Pseudopelade of Brocq was rendered in all the four cases.

DISCUSSION

Pseudopelade of Brocq is a rare form of cicatricial alopecia. The term was first used by Louis-Anne-JeanBrocq, a French dermatologist, in 1888.[4] PPB is a diagnosis of exclusion. The differentials include

lichen planopilaris, alopecia areata, central centrifugal cicatricial alopecia, discoid lupus erythematous, tinea capitis, morphea, syphilitic alopecia, aplasia cutis congenital, and follicular degeneration syndrome.[5]

The characteristic histopathological feature of early stage of PPB is the absence of sebaceous glands and a sparse or moderate lymphocytic infiltrate around the infundibulum. The follicular epithelium becomes more and more atrophic in the later stages. These follicles are often surrounded by concentric lamellar fibroplasias[6,7] which is replaced by fibrous tracts in the end stage. Contracted dermis with dense collagen and loss of space between collagen bundles are morphologic hallmark for PPB. The elastin stain may show thickened elastic fibres, not seen in DLE and LPP.[8]

The criteria for diagnosis of PPB have been given by Braun-Falco *et al* [9] [Table 1].

Table 1: Braun-falco Criteria Of Pseudopelade Of Brocq

CLINICAL CRITERIA	HISTOLOGICAL CRITERIA
Irregular and confluent patches of hair loss	No marked inflammation
Moderate atrophy (late stage)	No widespread scarring
Mild redness around hair follicles (early stage)	No significant plugging of hair follicles
Female predominance (3:1)	No sebaceous glands
Long course of more than 2 years	Normal epidermis
Slow progression	Fibrotic streamers in the dermis
Spontaneous termination possible	
Direct immunofluorescence Negative (or only weak IgM on sun exposed skin)	

Histopathologic features of lichen planopilaris (LPP) or DLE are seen in 33-69% of PPB cases. However, no interface dermaitis are observed in PPB. Unlike PPB. aplasia cutis congenital do not show presence of inflammatory infiltrate or any reduction in number of sebaceous glands. Syphilitic alopecia is characterized by peribulbar lymphocytic infiltrate and hair miniaturization. Central centrifugal cicatricial alopecia can be differentiated from PPB by the different clinical presentation. It presents with central alopecia with some preservation of the frontal hairline. Tinea capitis shows presence of hyperkratosis, spongiosis alongwith dermal inflammatory infiltrate. The dermatophytes in tinea capitis is seen on PAS stain. The presence of reticular dermal fibrosis extending into subcutis is classically seen in morphea.[10]

The present study showed male: female ratio of 3:1 with age group from 21-45 years. The duration of disease varied from 1 month to 3 years. All the cases characteristically showed parafollicular fibrosis along with fibrotic bands in the dermis with absence of pilosebaceous unit and presence of normal erector pilli muscle and normal sweat glands. Based on histopathological findings, other differentials were excluded and the diagnosis of PPB was reported.

Theories of stem cell failure and sebaceous gland destruction have been described in the pathophysiology of scarring alopecias. Stem cells are present over bulge region of the hair follicle. Any damage to this leads to scarring alopecia. In a normal person, when the hair shaft exits from the skin there is degeneration of sebaceous glands. Hence, these sebaceous glands are also a key factor in pathogenesis of scarring alopecias. The pathogenesis of PPB is unknown. It is now suspected that acquired immunity, Borrelia infection and failure of follicular stem cell may play a role in development of PPB.[11]

There is no standard treatment for patients of PPB. Some of the options include topical corticosteroids, intralesional triamcinolone acetonide (10 mg/mL), prednisone, oralmini pulse, hydroxychloroquine, isotretinoin and mycophenolate mofetil.[11] Surgical treatment includes autologous hair transplantation and scalp reduction surgery. However, these are considered only when the disease is stable for at least 2 years.

CONCLUSION

Our patients showed clinical features in the form of atrophy, irregular patches of alopecia giving classical appearance of foot print in the snow. Slow progression, insidious onset, without any evidence of other disease established the diagnosis of primary idiopathic PPB.

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