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Primary Idiopathic Pseudopelade of Brocq: Five Case Reports

Nilofar Diwan, Sneha Gohil, and Pragya A Nair

Department of Dermatology and Venereology, Pramukhswami Medical College, Karmasad, Gujarat, India
Address for correspondence: Dr. Pragya A Nair, Department of Dermatology and Venereology, Pramukhswami Medical College, Karamsad - 388 325, Gujarat, India. E-mail: drpragash2000@yahoo.com

Abstract

Pseudopelade of Brocq (PPB) is a rare, idiopathic self-limiting hair disorder resulting in progressive cicatricial alopecia primarily involving the parietal scalp and vertex. The general pathogenesis of scarring alopecias has been focused on theories of stem cell failure and sebaceous gland destruction. Acquired immunity, Borrelia infection and senescence of follicular stem cell reservoir plays suspected role. It classically presents as porcelain white hypopigmented and slightly depressed atrophic plaque. There is no standard treatment for PPB. Here, we present five cases which were labeled as primary idiopathic PPB, as on histopathology no specific changes of any cicatricial alopecia were seen.

Keywords: Alopecia, foot print in snow, pseudopelade of Brocq

INTRODUCTION

Pseudopelade of Brocq (PPB) is a rare, idiopathic self-limiting hair disorder resulting in progressive cicatricial alopecia. It is frequently described as discrete, asymmetrical, slightly depressed[1,2] alopecic patches primarily involving the parietal scalp and vertex.[3] It usually occurs in middle-aged white women. It can also be considered as a primary disorder where there are no clinical, histological or any other evidence for cause of the disease.[4] Here, we present five cases proven by histopathology as primary idiopathic PPB.

CASE REPORT

All of our patients presented with asymptomatic and gradual onset of hair loss [Table 1]. No evidence of any inflammation or trauma to the scalp in past. No other areas of the body affected. No history of similar complains in any family members. On examination, no exclamation mark hair or leucotrichia seen. Hair pull test was negative. No other cutaneous, mucosal, or nail findings were present. Histopathology of all cases showed predominantly follicular scarring with no other changes, hence were diagnosed as primary idiopathic PPB.

DISCUSSION

In 1888, the term pseudopelade was first used by a French dermatologist Louis-Anne-Jean Brocq, to distinguish this unique form of cicatricial alopecia from alopecia areata.[5] Pelade is the French term for alopecia areata, which is derived from a the word "*pelage*" meaning the fur, hair, wool, etc., of mammal.[6] PPB is considered end-stage permanent alopecia. In Germany, all types of inflammatory cicatricial alopecia are included in this group while American dermatologists have used this term as a diagnosis of exclusion.[7]

The general pathogenesis of scarring alopecias has been focused on theories of stem cell failure and sebaceous gland destruction.[8,9] Stem cells are essential for hair growth and are present over bulge region of the hair follicle. Damage to this region causes scarring alopecia. Sebaceous gland connects to the hair follicle just superior to where inner root sheath degenerates. The degeneration is required for the hair shaft to exit the skin normally; therefore, sebaceous gland also plays a crucial role. Pathogenesis of PPB is not completely understood. Acquired immunity, *Borrelia* infection and senescence of follicular stem cell reservoir plays suspected role.[10]

Mainly two types of PPB are recognized.

- Burnt - out or end-stage scarring alopecias (e.g., lichen planopilaris [LPP], discoid lupus erythematosus [DLE]) Pathophysiology corresponds to underlying disease process.[11]
- Primary idiopathic pseudopelade where pathophysiology is unknown.

Idiopathic cases represent approximately 10% of patients and have a different histology. In support of pseudopelade as a primary disorder, rare familial cases have been reported.[12]

Lesions of pseudopelade are distributed randomly. It classically presents as porcelain white hypopigmented and slightly depressed atrophic plaque. Lesions often are shaped irregularly, as opposed to the round or oval patches usually seen in alopecia areata.

Braun-Falco *et al*.[13] have given criteria for diagnosis of PPB [Table 2].

PPB often worsens in spurts with periods of activity, which may at sometimes followed by periods of dormancy. This is distinctly different from the slow but steady disease progression seen in several other forms of scarring alopecia.[14]

The differential diagnosis of PPB are alopecia areata, LPP, discoid lupus erythematous, central centrifugal cicatricial alopecia, morphea, secondary syphilis, tinea capitis, aplasia cutis congenital, and follicular degeneration syndrome.

As described by Pinkus[15] it is a histological entity and not a clinical entity. Idiopathic pseudopelade is characterized by contracted dermis with dense collagen and loss of space between collagen bundles. Elastic fibers are recoiled and appear thick. Broad fibrous tract remnants are seen with preservation of elastic sheath. Histopathologic features of lichen planopilaris (LPP) or DLE are seen in 33-69% of PPB cases.[16]

A sparse or moderate lymphocytic infiltrate around the infundibulum and the absence of sebaceous glands are pathologic hallmarks for an early PPB lesion. In later lesions, the follicular epithelium becomes more and more atrophic and follicles are often surrounded by concentric lamellar fibroplasias[17,18] until finally the follicle is replaced by fibrous tracts. Unlike in DLE and LPP, the elastic fiber network is preserved and elastin stain might show markedly thickened elastic fibers.[19]

Our cases showed male:female as 3:2. The age group ranged from 29 to 45 years with duration of disease varying from 8 month to 15 years. Along with the parietal and vertex area which are a common site of involvement in PPB, frontal and occipital area were also involved in our patients. All the patients presented with irregular, shiny and atrophic patches of alopecia with islands of normal hair, slow progression, insidious onset, no evidence of any disease over body, along with classical histological feature of follicular scarring, while two cases also showed the presence of lymphocytic infiltrate around appendages. All of our patients were started with intralesional triamcinolone acetonide (10 mg/ml) every 3-4 weekly given for 5-6 times, but gradually they were lost in follow-up owing to no response.

Disease progression eventually ends spontaneously. There is no standard treatment for PPB. Topical corticosteroids, intralesional triamcinolone acetonide (10 mg/mL), prednisone, oral mini pulse, hydroxychloroquine, isotretinoin and mycophenolate mofetil[10] are some of the options with almost nil results. Surgical options are considered when the disease is stable for at least 2 years and includes autologous hair transplantation and scalp reduction surgery.

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.

Footnotes

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Conflict of Interest: None declared.

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Figures and Tables

Table 1

Clinical and histopathological findings of the patients

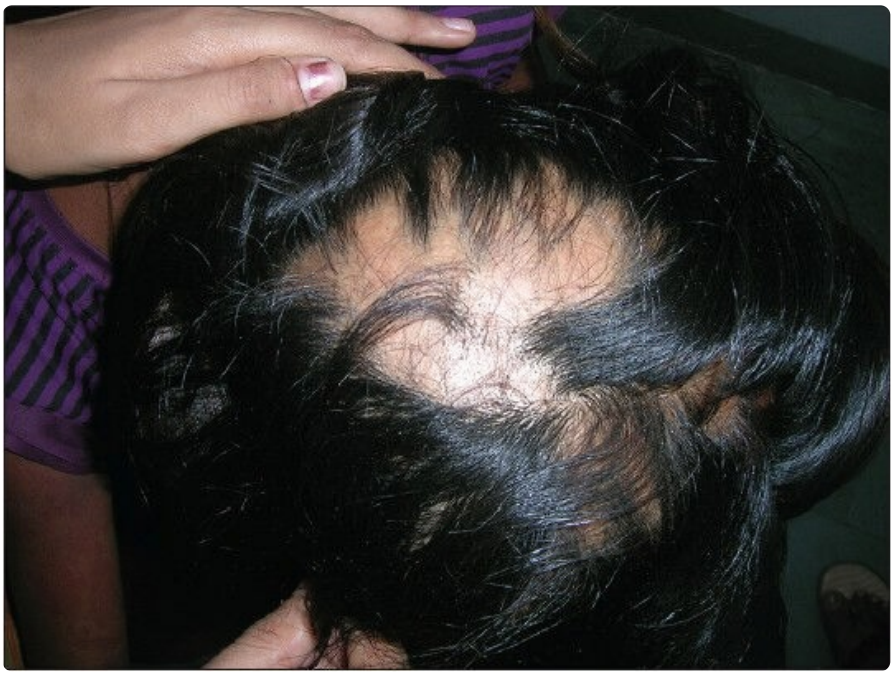
Age	Sex	Duration of disease	Site of involvement	Clinical feature	Histopathological examination
35	Female	1 year	Bilateral occipital region	Smooth, shiny atrophic patches of alopecia showing typical footprints in snow appearance. [Figure 1]	Epidermis is atrophic. Column of fibrosis replacing hair follicle extending into subcutaneous fat, accompanied by loss of sebaceous gland. [Figure 2]
33	Male	8 months	Vertex	Multiple randomly placed irregular smooth and shiny patches of alopecia. [Figure 3]	Dermis showed predominantly follicular scarring characterized by columns of fibrosis replacing hair follicles extending into lower dermis
29	Male	15 years	Frontal and vertex region of scalp	3-4 smooth, shiny patch of alopecia with atrophy. [Figure 4]	Mild acanthosis of epidermis. Replacement of pilosebaceous follicle by fibrosis with few perivascular lymphocytic infiltrations around the vessels in the upper dermis. [Figure 5]
45	Male	4 years	Left occipital region	Smooth, shiny patches of alopecia showing atrophy	Dermis shows absence of hair follicles and sebaceous glands along with thickened collagen and foci of fibrosis, with lymphocytic infiltrate
35	Female	15 years	Vertex, parietal and frontal areas of scalp	Irregular atrophic patches of alopecia with soft, wrinkled, glossy skin. [Figure 6]	Epidermis is atrophic. Column of fibrosis replacing hair follicle extending into subcutaneous fat accompanied by loss of sebaceous gland. No signs of inflammation

Table 2

Braun-Falco criteria of pseudopelade of Brocq

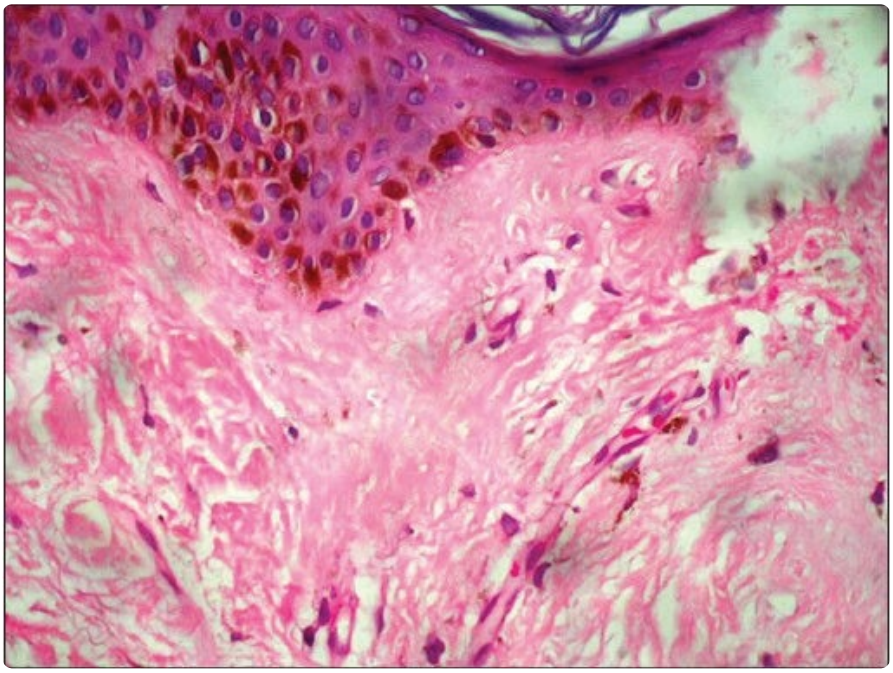
Clinical criteria	Histological criteria
Irregular and confluent patches of hair loss	No marked inflammation
Moderate atrophy (late stage)	No widespread scarring
Mild redness around hair follicles (early stage)	No significant plugging of hair follicles
Female predominance (3:1)	No sebaceous (oil) glands
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)	

Figure 1



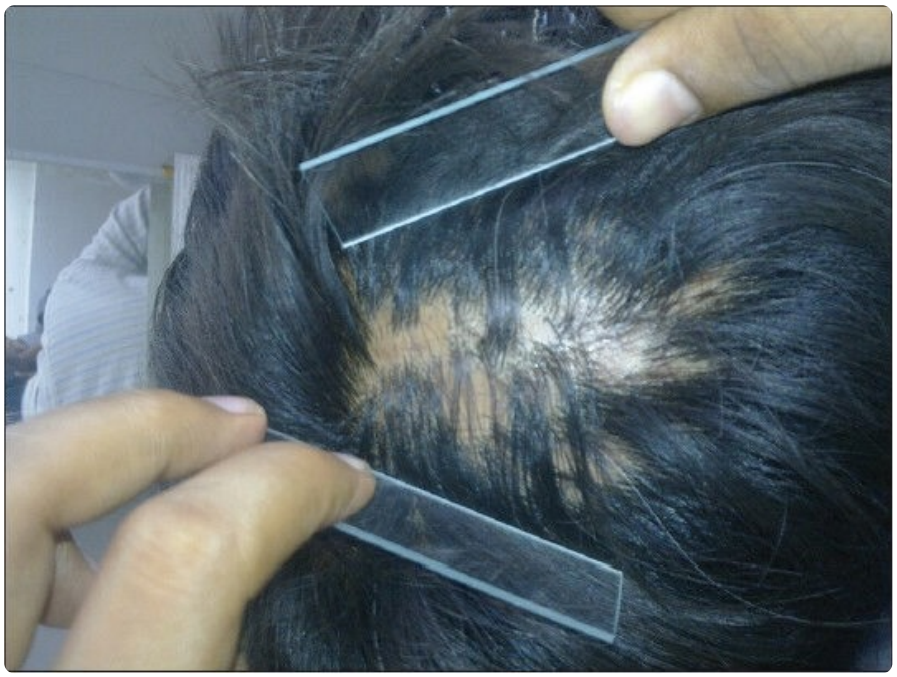
Shiny atrophic patches of alopecia over occipital regions

Figure 2



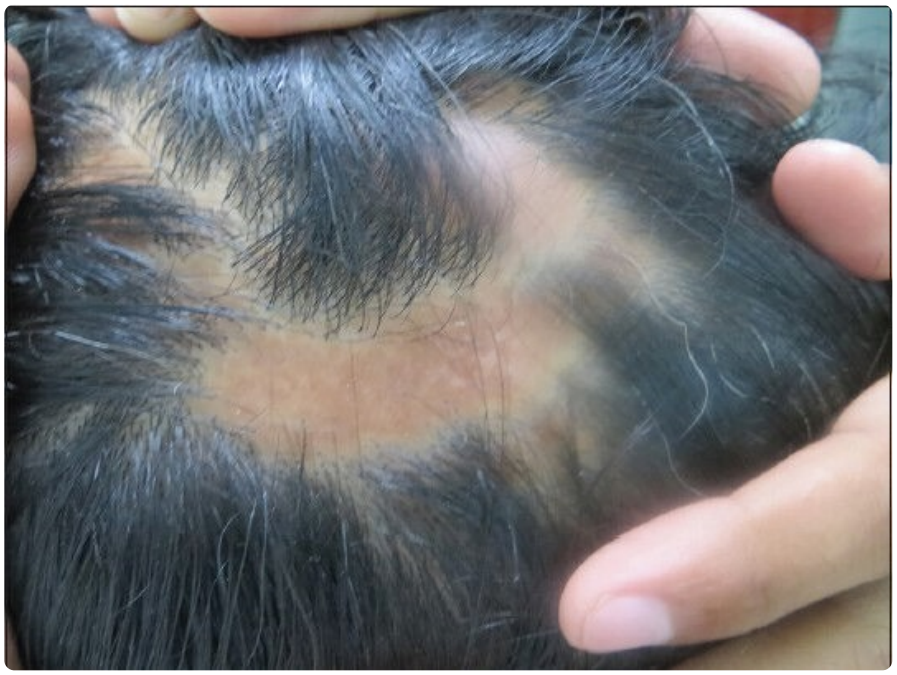
Atrophic epidermis with a column of fibrosis replacing hairfollicle extending into subcutaneous fat (H and E, ×40)

Figure 3



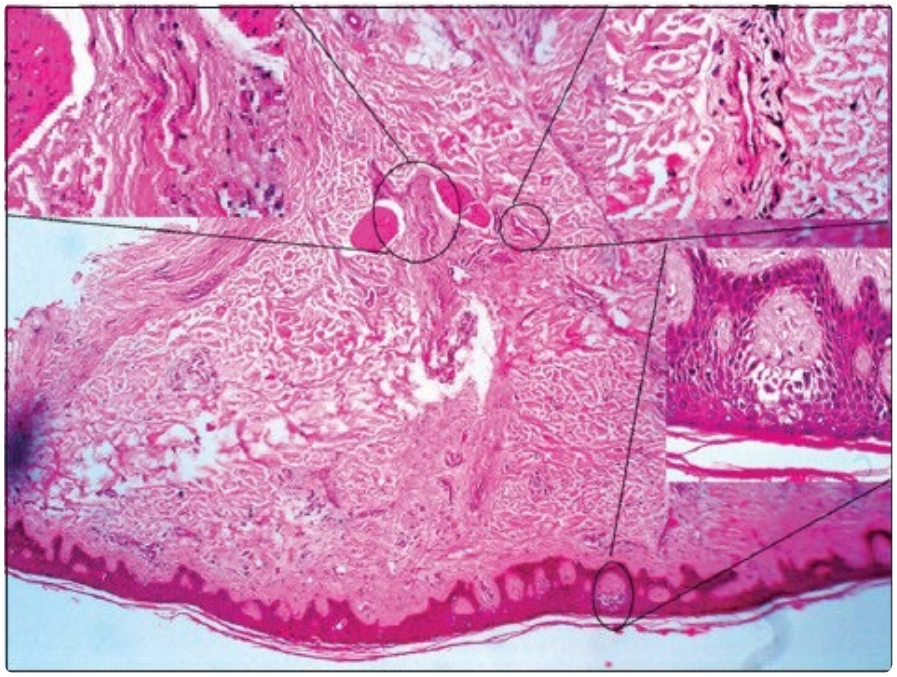
Multiple randomly placed irregular smooth and shiny patches of alopecia over vertex

Figure 4



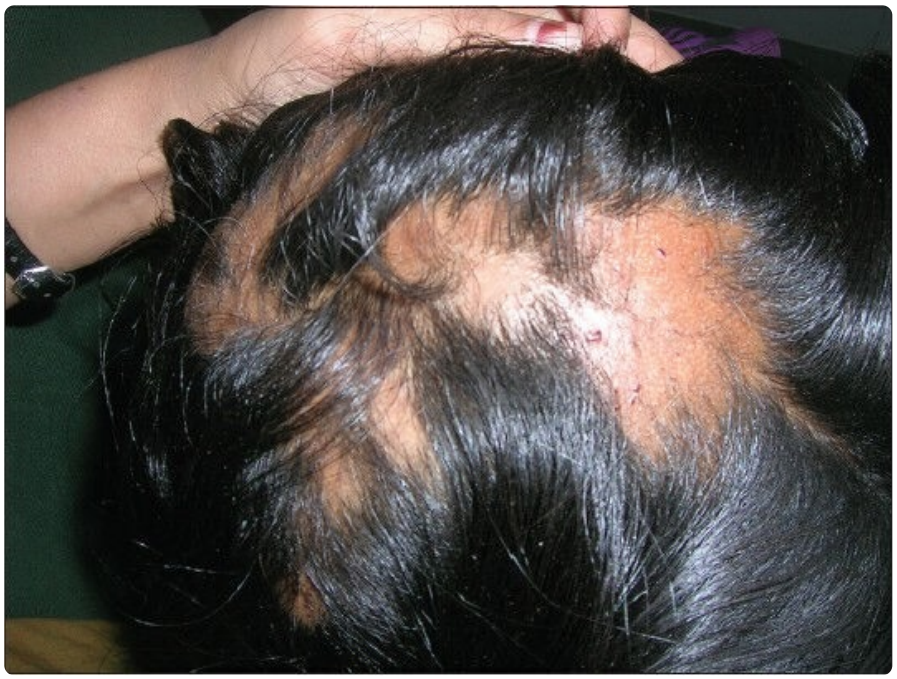
3-4 smooth, shiny patch of alopecia with atrophy over the frontal and vertex of scalp

Figure 5



Mild acanthosis of the epidermis with replacement of pilosebaceous follicle by fibrosis. Few perivascular lymphocytic infiltrations around the vessels in the upper dermis are seen. (H and E, ×10 image with inset ×40)

Figure 6



Irregular atrophic patches of alopecia with soft, wrinkled, and glossy skin over frontal, parietal and vertex of the scalp