INTRODUCTION

Pseudopelade of Brocq (PPB) is a rare, chronic, slowly progressive cicatricial alopecia that generally affects middle-aged women. Vertex and parietal scalp are commonly involved. It may be either a specific entity (primary) or the end of a variety of disorders of the scalp, all of which end in scarring (secondary). It presents with small patches of alopecia with mild-to-moderate atrophy and devoid of any sign of folliculitis or marked inflammation. We hereby report a case of a young female child with rapidly progressing cicatricial alopecia having no known underlying cause and the case was histopathologically confirmed as PPB.

CASE REPORT

A 10-year-old young girl presented with a complaint of patchy hair loss over the scalp for almost a year. She developed initial lesion 1 year back, but in the past 6 months, the disease was rapidly progressive. Her mother gave a history that continuously new lesions were developing and even the older lesions were increasing in size. There was no history of papules, pustules, or plaques preceding the alopecia. There was no history of any other cutaneous or systemic illness. There was no history of alopecia in any other family members. On cutaneous examination, irregular but well-defined confluent patch of hair loss over vertex giving a footprint in snow appearance was present. The patch was with minimal atrophy and wrinkling. Skin was shiny with lack of follicular ostia and only few normal hair follicles inside the lesion [Figure 1]. The lesions were asymptomatic with no signs of inflammation, scaling, crusting, or follicular papulopustules. Hair pull test was positive. It did not reveal any exclamation mark hair or any leukotrichia hair in the surrounding area. Rest of the facial and body hair were uninvolved, and there were no other cutaneous, mucosal, or nail findings. Potassium hydroxide examination of the adjacent hair was done just to rule out tinea capitis which was negative. Trichoscopy showed loss

Key words: Alopecia, cicatricial, pseudopelade of Brocq, scarring

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How to cite this article: Nair PA, Singhal R, Pariath K. Primary idiopathic pseudopelade of brocq in a young child. Int J Trichol 2017;9:113-5.
of follicular ostia, ivory white macules, and pigmentation at places [Figure 2]. Skin biopsy, with the differentials of PPB and lichen planopilaris (LPP), was done. The section showed atrophy of epidermis with the absence of viable pilosebaceous follicles, replaced by fibrosis. Fibrosis extended up to subcutaneous fat [Figure 3]. Changes were suggestive of PPB. We started her on oral prednisolone to arrest disease progression, along with multivitamins. Disease progress was halted after 3 months of therapy, but there was no regrowth of hairs in patches of alopecia.

**DISCUSSION**

According to Whiting, cicatricial alopecia constitutes 7.3% of all the hair loss cases. Females are more commonly affected (2.6:1), and primary cicatricial alopecia is more common than secondary cicatricial alopecia (4:1). Among causes of primary cicatricial alopecia, PPB (40.6%) is most frequent followed by LPP (12.6%) and folliculitis decalvans (FD) (11.2%). Tan et al. reported discoid lupus erythematosus (DLE) as the most common (33.9%), followed by pseudopelade (24.1%) and LPP (22.3%). Trachsler and Trueb studied 136 biopsy specimens of scarring alopecia and found that the most frequent diagnosis was LPP (26%), followed by DLE (21%), FD (20%), and PPB (10%).

The general pathogenesis of scarring alopecias includes:

- **Stem cell failure**: The bulge region of the hair follicle houses follicular stem cells. These cells are essential for hair growth. Direct damage to the bulge region may cause permanent scarring hair loss.
- **Sebaceous gland destruction**: The sebaceous gland connects to the hair follicle just superior to where the inner root sheath degenerates. This degeneration is required for the hair shaft to exit the skin normally, so sebaceous gland may have a crucial role in this process.

Pathogenesis of PPB is not completely understood as yet. Some of the suspected factors include acquired autoimmunity, *Borrelia* infection, and senescence of follicular stem cell reservoir. Few case reports and case series of familial PPB indicate that heredofamilial factors may be important in its pathogenesis.

In 1885, Brocq of Paris first described this condition. Pelade is the French word for alopecia areata. Pseudopelade means clinically resembling alopecia areata, but this patchy baldness is permanent as follicular ostia are absent. Pseudopelade can be recognized as primary idiopathic in which an autoimmune process sets in and perifollicular lymphocytic infiltrates damage stem cells that reside in the isthmus (bulge portion) leading to permanent hair loss. Idiopathic cases represent approximately 10% of patients.
with few having family history positive.[6] Secondary pseudopelade refers to burnt out or end stage of other scarring alopecias such as LPP, DLE, FD, and morphea.

It has an insidious onset and indolent course that generally affects a middle-aged woman which is contrary to our case who was a 10-year-old girl. There are very few reports of PPB in childhood. One such study of two brothers having PPB at 7 years of age was reported by Collier and James with family history positive suggesting some genetic factor playing a role in its occurrence.[7] Another case of childhood PPB was reported by Bulengo-Ransby and Headington.[8]

Vertex is the most common site for the initial lesion. It may affect the beard and eyebrows. There are irregular but well-defined confluent patches of alopecia giving footprints in a snow appearance. There is mild perifollicular erythema in early-stage and moderate atrophy in the late stage. In active lesions, hair pull test is positive. It has an unpredictable long course (>2 years) with spontaneous termination. The differential diagnoses of PPB are alopecia areata, LPP, DLE, central centrifugal cicatricial alopecia, morphea, secondary syphilis, tinea capitis, aplasia cutis congenita, and follicular degeneration syndrome.[9] Trichoscopy features of classic PPB are nonspecific which include loss of follicular ostia and ivory white areas[9] which can be appreciated in our case. Occasionally solitary dystrophic hairs at the periphery of the lesion may be seen. PPB is considered as diagnosis of exclusion both clinically and by trichoscopy.

Histologically, PPB has been classified into lymphocyte-mediated primary cicatricial alopecia.[10] Epidermis is normal. Predominant follicular scarring is characterized by columns of fibrosis replacing the hair follicles with no widespread interfolicular dermal scarring. There is loss or decrease of sebaceous glands. Perifollicular lymphocytic infiltrate is present that is limited to only upper two-third of the follicles. Dense elastic tissue is present cuffing around fibrous tracts.

Braun-Falco et al. proposed the diagnostic criteria for PPB on the basis of clinical, histopathological, and immunofluorescence features.[11]

There is no definite management to halt the disease progression. The response to intrallesional or systemic steroids (oral prednisolone 0.5 mg/kg/day) is also very poor. Hydroxychloroquine 200 mg twice daily, isoretinoin 1 mg/kg/day, and mycophenolate mofetil 1 g/day can be tried[12] with variable and poor results. Antimalarials, antifungals, or retinoids have also been tried with limited success. It can be surgically managed with hair transplants, scalp reduction or alopecia reduction surgeries, tissue expansion, and flap surgeries once the activity has subsided.

PPB still remains a challenge to the treating dermatologist as there is no definite management to halt the decrease in progression.

**CONCLUSION**

PPB can affect children though rarely. Proper evaluation with trichoscopy and biopsy at the right time can help reduce unnecessary treatment with topical steroids as well as the patient can be counseled about the progress of disease.

**Acknowledgment**

We would like to acknowledge Dr. Mustafa from department of pathology for the diagnosis and photographs of histopathology slides.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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