SHORT COMMUNICATION

Whorled Scarring Alopecia: A Rare Cutaneous Finding in Incontinentia Pigmenti or Overlooked Phenomenon? A Case Report of Incontinentia Pigmenti with Trichoscopic and Dermoscopic Findings

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Incontinentia pigmenti (IP) or Bloch-Sulzberger syndrome (MIM 308310) is a rare genetic disease that frequently affects the skin, hair, nails, teeth, eyes, and central nervous system. IP is an X-linked dominant inherited genodermatosis, caused by mutations in the NEMO gene (IKK-gamma) on Xq28 locus (1). It is generally recognized by cutaneous findings including linear erythematous, and pustular and vesiculobullous rashes in newborns. Cutaneous findings usually occur sequentially and manifest along the Blaschko lines and classically progress in 4 stages, which may show some overlap (2, 3).

IP can also involve the hair. Hair involvement has been reported in approximately 28% to 38% of patients (4). Scarring alopecia is the most common manifestation of hair involvement (4). Whorled scarring alopecia is a reported cutaneous finding in patients with IP; however; there are no certain data on its frequency. Here in we report a 5-year-old girl with whorled scarring alopecia on the vertex with trichoscopic signs and dermoscopic findings of Stage 3 of IP.

CASE REPORT

A 5-year-old female patient was referred to our dermatology department for hair loss on the scalp since birth and a hyperpigmented rash on the body. The wounds on the body started at birth and were in the form of fluid-filled blisters and pustules. After the age of 2, they turned into linear pigmented lesions in the groin and axillary areas. Genetic analysis was performed and revealed heterozygous deletion of exons 4–10 in the IKBG gene. She was diagnosed with IP and following up by the ophthalmology, dentistry, paediatric neurology, paediatric haematology, and paediatric cardiology departments. On dermatological examination there were reticular linear hyperpigmented macules in the axillary and inguinal areas (**Fig. 1**b), alopecic areas on the scalp (Fig. 1a), and hypoplastic teeth (Fig. 1c).

Trichoscopy was performed using Dermlite DL4 (x10 magnification in polarized mode) and revealed grey-brown globules, empty hair follicles, yellowish structureless areas, and thin hair (**Fig. 2**a). On dermoscopic examination, we showed linear distributed grey-brown dots on the brown pigmented background (Fig. 2b). The cranial MRI showed focal T2 high-signal foci located in the deep and subcortical white matter bilaterally. Follow-up is recommended by the paediatric neurology department for gliotic changes of sequelae nature.

DISCUSSION

Hair involvement (alopecia or abnormal hair [sparse hair, woolly hair, anomalies of eyebrows and eyelashes]) is one of the diagnostic criteria of IP (5). Alopecia occurs following vesiculation on the scalp area and may result in scarring alopecia (2). Wang et al. investigated 42 patients with IP and reported hair anomalies in 31%

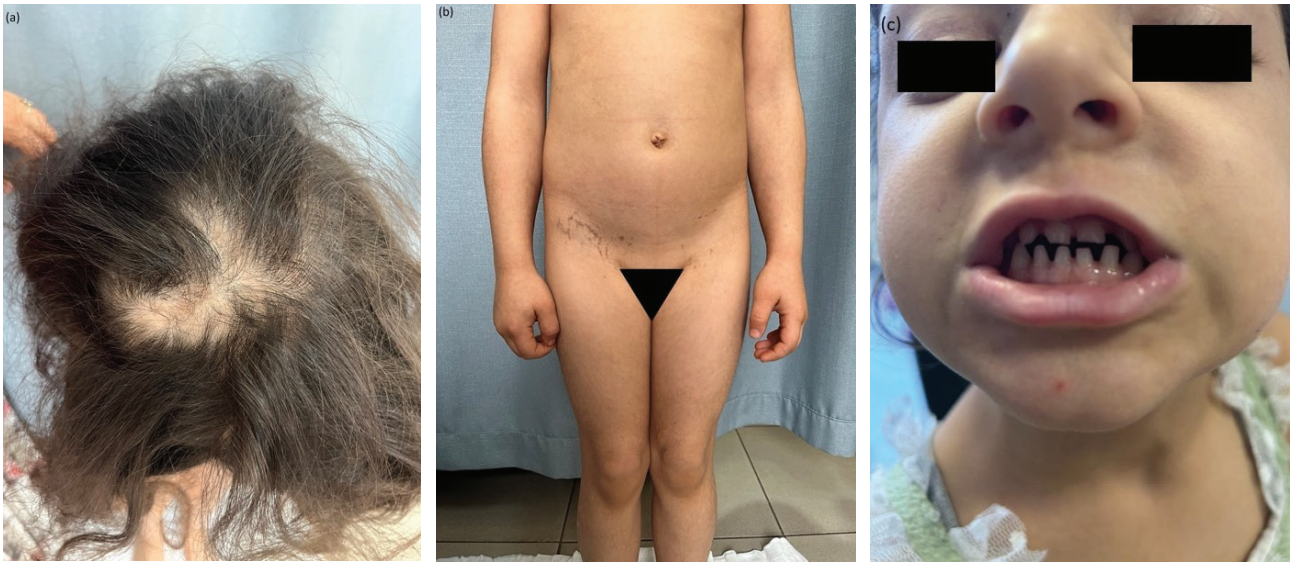

Fig. 1. (A) Scalp (scarring alopecia), (B) inguinal (Blaschkoid-distributed hyperpigmented lesions), and (B) teeth (hypoplastic and pegged teeth). Written permission has been given to publish these photos.

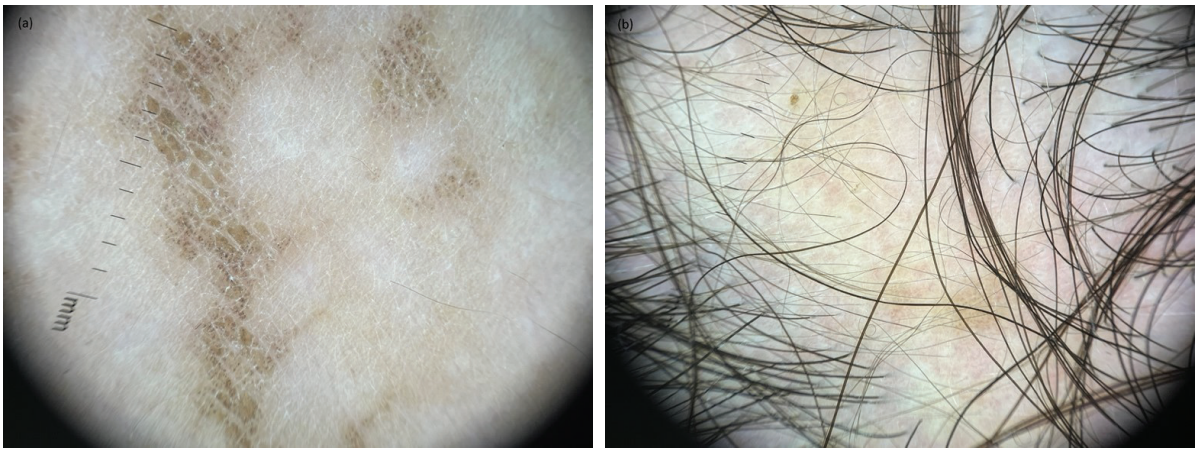

Fig. 2. (A) Dermoscopy of the skin shows linear distributed grey-brown dots on the brown pigmented background; (B) trichoscopy of the scalp shows grey-brown globules, empty hair follicles, yellowish structureless areas, and thin hair.

(vertex alopecia in 26%, thin hair in 12%, wiry hair in 5%, coarse hair in 2%) (6). Similarly, Carney reported diffuse vertex alopecia as the most common hair manifestation of IP and has shown this in 38% of patients (7). On the other hand, in a clinical study of 40 patients with IP, Hadj-Rabia et al. (4) found vertex alopecia in only 11 patients (28%). Similarly, Chan et al. (8) found whorled scarring alopecia in 4 of 26 patients with IP. However, it has also been suggested that whorled alopecia may be relatively more common and could be under-reported (9).

There are few reports on dermoscopic and trichoscopic findings in IP cases (10, 11). On trichoscopic examination of whorled alopecia in IP patient, Razmi et al. reported blue-grey pigment dots, empty hair follicles, and black dots (11). Similarly, we reported grey-brown globules and empty hair follicles, as well as yellowish structureless areas and thin hair.

Blaschkoid-distributed hyperpigmented lesions include linear and whorled nevoid hypermelanosis, hypomelanosis of Ito, and lichen planus pigmentosus, and may be confused with stage III of IP (12–14). On the other hand, dermoscopy can be a useful diagnostic tool in cutaneous lesions of IP. On dermoscopic findings in the pigment stage of IP, Bishnoi et al. reported linear brown to grey-brown dots (10, 11) and Minic et al. showed linear grey to grey-brown dots on the light-brown pigmented background that intermingled with normal skin and perifollicular depigmentation (14). Similarly, we showed linear distributed grey-brown dots on the brown pigmented background.

Scarring alopecia tends to be permanent and, in addition to dental findings, it may be the only cutaneous finding remaining in IP patients at older ages. However, it is not clear in how many of these patients the alopecia is distributed in the Blaschkoid pattern (8, 15). Dermoscopy and trichoscopy can be used on the differentiating cutaneous and hair findings of IP from other diseases. More studies are needed to determine the frequency of whorled scarring alopecia and its dermoscopic and trichoscopic clues in IP or its relationship with certain genetic mutations.

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