

Thick, Adherent, Asbestos-Like and Overlapping Scales Encircling the Tufts of Hair Shafts in a 13-year-old Girl

A 13-year-old girl presented with thick, adherent and overlapping yellowish scales encircling the tufts of hair shafts for the last 4 years. Her lesions had started initially over the vertex and progressed to involve the whole scalp [Figure 1a]. It was associated with fissuring, itching, pain and mousy smell on occasions especially during summers and rainy seasons. It remained relentlessly progressive and repeated treatments in the form of shampoos and topical lotions did no benefit. During last 2 years, she also noticed asymptomatic, dry, hyperkeratotic, brownish-red asymptomatic papular lesions over left supraclavicular area, and over left [Figure 1b] and right

[Figure 1c] fronto-tempo-parietal scalp margins. Examination for mucous membranes, nails, palms and soles was normal. Her other medical history and systemic examination were unremarkable. She was the only child born to non-consanguineous otherwise healthy parents after an uneventful pregnancy. No other family members had similar problem. Microscopy showed no hair shaft abnormality or fungal elements in KOH mounts. The findings of hematoxylin-eosin-stained histology sections of a skin lesion are shown in Figure 2a and b.

What is your diagnosis?

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Figure 1: (a) Hyperkeratotic scalp lesions with classic matted hair and overlapping crusts in a tiled appearance. (b) Multiple brown hyperkeratotic papular lesions over left temporal, and (c) right temporal hair margins

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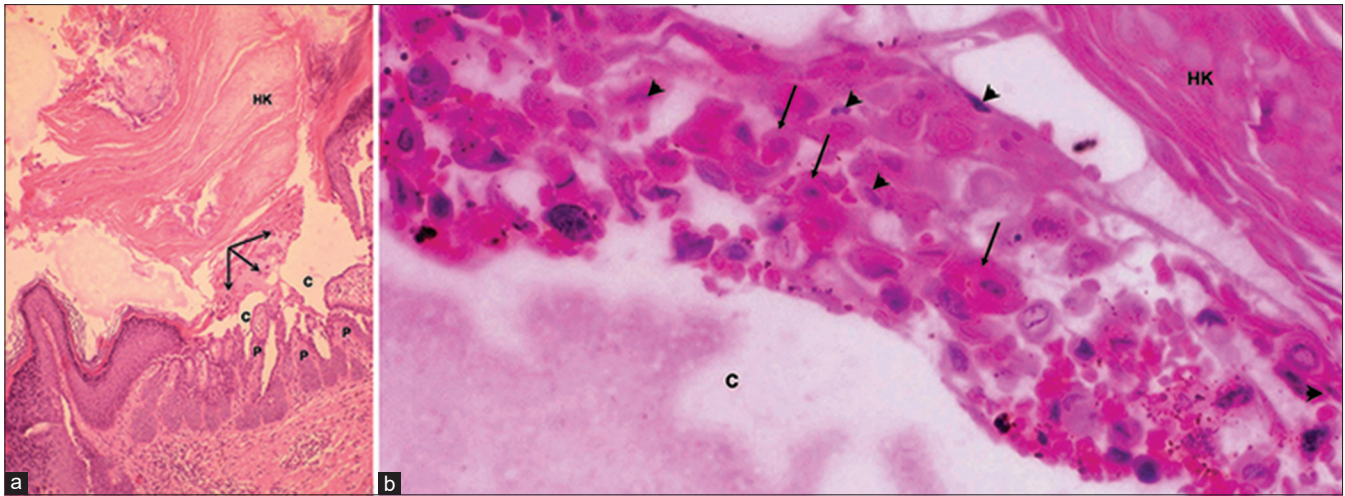


Figure 2: (a) Extensive hyperkeratosis (HK), irregular dermal papillae lined by single layer of basal cells (P) projecting into the suprabasal acantholytic intercellular cleft (C) within the central epidermis with no acantholytic cells. Arrows indicate dyskeratotic cells in the stratum granulosum (stain H and E, $\times 40$). (b) Dyskeratotic cells, corps ronds with small pyknotic nuclei, a perinuclear clear halo and eosinophilic cytoplasm (arrows) and grains, compressed cells with elongated nuclei (arrow heads) are visualized in stratum granulosum. Extensive hyperkeratosis (HK) and suprabasal acantholytic cleft (C) are also seen (stain H and E, $\times 100$)

Diagnosis: Darier's disease presenting as pityriasis amiantacea

Microscopic Findings

Histopathology revealed features consistent with Darier's disease comprising extensive hyperkeratosis, papillomatosis, suprabasal acantholysis and cleft formation, and villus-like dermal papillae lined by single layer of basal cells projecting into it [Figure 2a]. Corp ronds and grains were visualized in the stratum granulosum [Figure 2b]. Absence of acantholytic cells and minimal dermal infiltrate were other notable features.

Treatment

Her skin lesions resolved and scaling from scalp decreased 4 weeks after treatment with oral isotretinoin 20mg once daily, ketoconazole (2%) shampoo for alternate day scalp washing and topical clobetasol propionate 0.05%+salicylic acid 3% lotion for once daily scalp application. However, the isotretinoin dose had to be decreased due to severe cheilitis. Subsequent flare up was treated successfully with oral cefadroxil 500mg twice daily given for 10 days. She did not continue follow-up.

Comments

Pityriasis amiantacea is a distinct and usually localized inflammatory scalp disorder affecting young adults, adolescents and children. Clinically, adherent, thick and overlapping, asbestos-like silvery or yellowish scales encircling the hair shafts and binding down tufts of hair and occasional temporary alopecia are characteristic. Its etiopathogenesis remains unclear and is thought to represent a reaction pattern to inflammatory skin

disease. Scalp psoriasis was confirmed in 35.3% while features of seborrheic and atopic dermatitis were observed in 34.2% in series of 85 cases.^[1] Lichen planus, lichen simplex chronicus, pityriasis rubra pilaris, and superficial fungal or pyogenic infections were other notable causes. However, it is considered a manifestation of psoriasis by most workers. Darier's disease (syn. Keratosis follicularis, Darier-White disease) is an uncommon disorder of autosomal dominant inheritance with reported incidence of 1:30,000 to 100,000 population. A mutation in ATP2A2 gene has been mapped to chromosome 12q23-24.1 that encodes sarcoplasmic/endoplasmic reticulum Ca^{2+} -ATP isoform 2 protein (SERCA2) which impairs intracellular Ca^{2+} signaling and affects cell adhesion.^[2,3] However, genetic studies are rarely needed for diagnosis. Clinically, it usually presents at a mean age of 6-20 (range 4-70) years affecting both genders equally. Exacerbations occur from sunlight, heat, sweat, and humidity, and secondary bacterial infections. There is increased susceptibility to infections while short stature, mental retardation, epilepsy, mood disorders, schizophrenia occur rarely.^[4] Pemphigus foliaceus, Hailey-Hailey disease, acrokeratosis verruciformis of Hopf, and transient acantholytic dermatosis remain major differentials.

Darier's disease as comedonal lesions or localized to scalp as coalescing hyperkeratotic papules has been reported infrequently in adults.^[5] However, Darier's disease presenting as pityriasis amiantacea as in our patient is extremely rare.^[6] Their causal relationship is not clear, but an exaggerated inflammatory response to the Darier's disease or repeated pyogenic infection seems most plausible explanation.

The treatment of Darier's disease remains unsatisfactory and it tends to persist for life with partial remissions. Most cases have been treated with topical emollients, keratolytics, steroid, and/or retinoid creams during exacerbations. Systemic retinoids (isotretinoin, acitretin) are effective to resolve keratotic lesions but recurrences follow after treatment cessation and long-term use remains problematic due to various adverse effects and in child bearing age.^[7] Dermabrasion, electrosurgery, CO₂, Er: Yag, pulse-dye lasers or other fractional resurfacing devices, electron beam therapy, photodynamic therapy, and surgical excision have been used with variable success. The therapeutic efficacy of miglustat, an α -glucosidase inhibitor that increases adhesion strength by restoring mature adherens junctions and desmosomes in dyskeratotic keratinocytes, remains unevaluated.^[8] Oral vitamin A appears an attractive safe and low-cost therapeutic option.^[9]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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