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PHOTOQUIZ

Pediatric Dermatology WILEY

A single plaque on the chin

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CASE REPORT

A 16-year-old girl, without significant past medical history was evaluated for a painless skin lesion on her chin. The lesion had appeared 2 months prior and had shown no response to topical tacrolimus 0.1% ointment twice daily for 2 weeks. The patient did not recall any prior trauma.

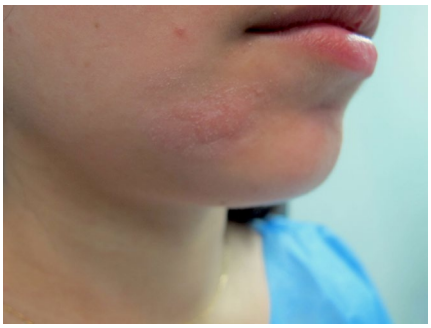

FIGURE 1

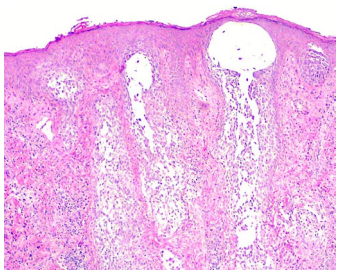

FIGURE 2

Physical examination revealed a solitary, pink plaque, measuring 2 cm in diameter with irregular borders, rough surface, and a raised center located on the right side of her chin (Figure 1). The rest of clinical examination was unremarkable. A skin biopsy was performed (Figures 2-4).

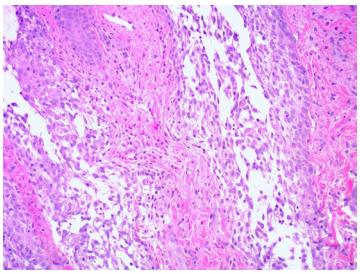

FIGURE 3

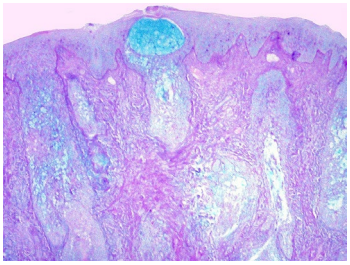

FIGURE 4

WHAT IS THE DIAGNOSIS?

The brightness and contrast of figures have been adjusted, following the ethical considerations of the journal.

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Diagnosis: Primary follicular mucinosis

DISCUSSION

Hematoxylin and eosin stain showed the epidermis with mild spongiosis and presence of dilated hair follicles with intrafollicular lymphocytes and a peripheral infiltrate composed of lymphocytes and eosinophils (Figures 2 and 3). Alcian blue stain revealed prominent intrafollicular mucin deposition (Figure 4). Treatment with one intralesional injection of betamethasone resulted in complete disappearance of the plaque.

Follicular mucinosis (FM) is a rare inflammatory disorder of unknown cause, characterized by mucin deposition within the follicular epithelium and sebaceous glands. Numerous classifications have been described for FM, but it is mainly categorized into two types. The benign idiopathic, or primary FM, typically occurs in patients from 11 to 35 years (mean 20.4 years).[1] Primary FM may appear as acute or subacute plaques composed by clustered follicular papules, sometimes showing scaling. It usually favors the face and neck and is associated with alopecia. Primary FM is usually self-limiting, and most cases heal spontaneously after 2 to 24 months.[2]

Secondary FM is associated with inflammatory conditions including lupus erythematosus, insect bites, eczema, alopecia areata, and hypertrophic lichen planus, or malignant diseases such as mycosis fungoides, Sezary syndrome, leukemia cutis, cutaneous B-cell lymphoma, and Hodgkin's disease.[3-5] In children, primary FM is more common than malignancy associated FM.[6]

It is important to differentiate primary FM from the follicular variant of mycosis fungoides (FMF).[2,7] The definitive diagnosis should take into account a correlation between clinical findings (age, location and number of lesions, and response to treatment), histopathology, and immunohistochemistry. The diagnosis of primary FM is favored by an early age onset, single mildly infiltrated plaques of confluent follicular papules, located on the head and neck, with or without alopecia, and fast improvement with treatment. In contrast, FMF is more frequent in adults, commonly presenting as multiple, large, hypo-, or hyperpigmented erythematous infiltrated plaques with overlying follicular papules and alopecia, on any location, without sustained improvement to conventional treatments.[8] Histopathology provides additional information, as it reveals mixed inflammatory infiltrate and mucin within hair follicles, with mild or absent fibrosis in FM, while in FMF the infiltrate may reveal atypical T lymphocytes, with epidermotropism and folliculotropism, sometimes forming intraepidermal clusters (Pautrier microabscess). Immunohistochemistry of the T lymphocytes in most FMF cases shows loss of CD5+ and CD7+ expression, and CD4+ cells outnumbering CD8+ cells.[2,7-9]

Other diagnoses in the differential of primary FM include keratosis pilaris, lichen spinulosus, alopecia areata, and allergic contact dermatitis and a skin biopsy is often required for the diagnosis.[10] Despite being a benign condition, monitoring patients with FM for cutaneous T lymphoma is advised, especially in persistent cases, although the progression toward MF has not been observed in some studies.[1]

There is no specific treatment for primary FM and treatment modalities are variable, but initial treatment consists of mild-to-moderate potency topical corticosteroids. Systemic and intralesional corticosteroids, topical pimecrolimus, PUVA, dapsone, hydroxychloroquine, minocycline, isotretinoin, and imiquimod have also induced improvement of lesions.[11]

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