

An Unusual Cause of Scalp Nodule in a Toddler

Smriti Gupta¹, Dipankar De¹, Sanjeev Handa¹, Debajyoti Chatterjee², Rahul Mahajan¹

¹ Department of Dermatology, Venereology and Leprology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

² Department of Histopathology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

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Corresponding Author: Rahul Mahajan, Associate Professor, Department of Dermatology, Venereology and Leprology, Post Graduate Institute of Medical Education and Research, Chandigarh, India-160012. Email: drmahulpi@yahoo.com

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE), also known as epithelioid hemangioma, is an uncommon benign proliferative disorder of vessel of uncertain origin. It presents as red-to-brown papules or nodules involving the head and neck area. It involves the scalp in up to 17% of cases. It is predominantly asymptomatic but can sometimes be pruritic, painful, or bleed. The mean age at presentation is 37 years, and there is no association with patients' sex [1]. ALHE is rarely reported in children, and the few reported sites include the lips and arms [2, 3].

Case Presentation

A three-year-old female was brought by her mother to the dermatology outpatient department with complaints of a single itchy red-colored lesion over the scalp for one year. There was no history suggestive of prior trauma and bleeding. On examination, a firm 2x2 cm erythematous nodule with no surface changes was present over the right parietal area (Figure 1a). Darier sign was negative. With the clinical

differentials of solitary mastocytoma, juvenile xanthogranuloma, and ALHE, a skin biopsy was performed. Histopathological examination revealed compact orthokeratosis in epidermis. The dermis showed moderate-to-dense mixed infiltrate comprising of lymphocytes and eosinophils along with proliferating endothelial cells. A few small lymphocytes aggregates were seen (Figure 2a-b). The final diagnosis of ALHE was made, and various treatment options were discussed with the parents. Owing to the age of the child, her parents did not opt for any surgical intervention, and after appropriate counselling, she was started on oral propranolol, which was slowly titrated to the dose of 2 mg/kg over two weeks. At eight-week follow-up, marked resolution of the lesion was observed (Figure 1b).

Conclusions

Scalp nodules can have various causes, and their diagnosis is primarily confirmed through histopathological examination. Solitary mastocytoma was one of the close differentials as the lesion was itchy and because it frequently occurs during childhood and often appears on face, scalp, and extremities [4].



Figure 1. A: Erythematous nodule over the right parietal area of scalp. B: Resolving erythematous nodule with crusting over the scalp.

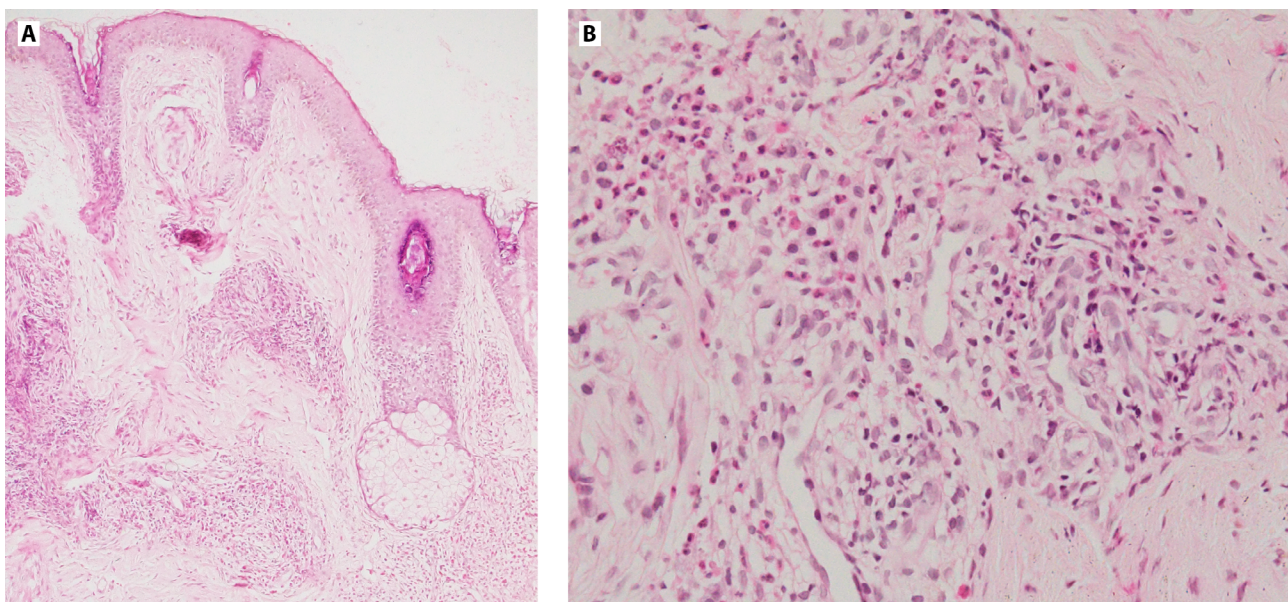


Figure 2. A: Histopathological image showing collection of inflammatory cells predominantly in the mid-dermis (hematoxylin and eosin, x10). B: Histopathological image showing proliferating blood vessel with perivascular lymphocytic and eosinophil infiltrate (hematoxylin and eosin, x40).

The other differential was juvenile xanthogranuloma, non-Langerhans cell histiocytosis, which affects children with a mean age of 3.3 years. It is a benign, self-limiting disorder characterized by asymptomatic nodules that are yellowish to brownish in color and typically found on the head and neck or trunk. Table 1 summarizes the differential diagnosis considered for the index case, along with clinical features, histopathological findings, and treatment options.

ALHE responds best to surgical excision, with the lowest recurrence rates. Other treatment options include CO₂ laser, pulsed dye laser, cryotherapy, and intralesional corticosteroids, all of which have varying rates of success. Several

oral medical therapies, such as dapsone, isotretinoin, and pentoxifylline, have been attempted, with very high rates of treatment failure [1]. Furthermore, there have been reports of propranolol showing positive responses in adults with ALHE, and we applied this treatment approach with similar favorable outcomes [5]. The probable mechanism by which propranolol acts in ALHE is by targeting the proliferative vascular endothelium [5]. In conclusion, ALHE should be considered as an important differential diagnosis in cases of scalp nodules in children, and propranolol can serve as an effective alternative medical therapy when surgical excision is deferred.

Table 1. Differential diagnosis of scalp nodule in index case.

	Angiolymphoid hyperplasia with eosinophilia	Solitary mastocytoma	Juvenile xanthogranuloma
Clinical features	Presents as solitary or multiple reddish-to-brown papules and nodules most commonly in head and neck area. Mostly asymptomatic but can be pruritic, painful, or can bleed [1].	Presents as asymptomatic solitary or multiple yellowish-to-brown papules and nodules. Commonly located on head and neck area, sometimes on trunk.	Solitary or multiple erythematous to yellowish nodules commonly involving head and neck area [6].
Histopathological findings	There is proliferation of blood vessels in the dermis which are lined by plumped endothelial cells containing eosinophilic cytoplasm and vesicular nucleus. Surrounding these proliferating vessels are lymphocytes and eosinophils which may infiltrate these vessels [2].	Epidermis shows increased melanization in the basal layer and dermis reveals infiltrates of mast cells mostly in upper part. These mast cells are best observed using special stains like toluidine blue and Giemsa stain [4].	The lesion consists of histiocytoid mononuclear cells, with oval-shaped clefted nuclei and intracytoplasmic lipid. The lipid component varies in the cytoplasm. Touton-type giant cells can be numerous. Variable numbers of lymphocytes and eosinophils can be present in the surrounding areas along with hyalinized collagen [6].
Treatment	Surgical excision: most effective. Others: CO2 laser, pulsed dye laser, cryotherapy, intralesional corticosteroids. Medical therapy: propranolol [5], isotretinoin, dapsone - variable success rate [1].	Avoidance of potential trigger factors like rubbing and certain drugs. Symptomatic treatment with oral H1 and/or H2 blockers [4].	Self-limiting and generally do not require treatment [6].

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