Original Article

Epithelioid hemangioma (ALHE) on the tongue of an infant treated with oral corticosteroids: A case report and review of the literature

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Abstract

Epithelioid hemangioma or angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare benign vascular lesion presenting as multiple nodules on the head and neck. Surgery had been considered to be the best treatment modality for ALHE. We report the case of a 6-month-old boy with ALHE on his tongue that was treated successfully with oral prednisolone.

Keywords: ALHE, angiolymphoid hyperplasia with eosinophilia, epithelioid hemangioma, mucosal, review, steroid, therapy, topical, treatment

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a benign vascular lesion that presents as single nodules (most commonly on the head and neck region in females in their third and fourth decades of life) in the dermis and the subcutaneous tissues.[1] It is most commonly seen on the head and neck region. Oral mucosal involvement is extremely rare in this disease.[2] Women in the third and fourth decades are most commonly affected.[3] We report a case of ALHE on the tongue of an infant.

Case Presentation

A 23-month-old boy was referred to our clinic with an extensive ulcerated lesion on his tongue and intermittent hemorrhage. The lesion had begun to develop on his tongue at 6 months and before that, he never had any problem [Figure 1]. He was born in the sixth month of gestation by cesarean section because of

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insufficient placental circulation (possible etiologic factors for ALHE). He was intubated in the neonatal intensive care unit (NICU) for the first month of his life and He was a bottle-fed infant. He did not have any significant problems up to the first 6 months, when he developed a small papule on the dorsal surface of his tongue, followed by intermittent hemorrhage. He had some degree of frontal bossing and club foot. No lymphadenopathy was detected in the physical examination. Blood examination showed no eosinophilia. A biopsy was done from the lesion, and the histopathological study showed denuded or ulcerated mucosa and mixed infiltration of inflammatory infiltrate, composed predominantly of lymphocytes, some histiocytes, some plasma cells, and frequent eosinophils. There were some extravasated erythrocytes and numerous vascular channels, lined by prominent large, pink, epithelioid endothelial cells, surrounded by inflammation [Figure 1]. These findings suggested the diagnosis of ALHE. The application of triamcinolone-acetonide in orabase for a month was useful in relative remission. Then, he was treated with 10 mg of prednisolone daily (1.5 mg/kg) for a month, which was tapered very slowly during 120 days (dose reduction was about 1.25 mg every 15 days). His response was good and the lesion decreased in size to about 25% of its initial size, and it was not ulcerative anymore. The ventral part of the lesion resolved near to complete remission; however, the parts on the lateral sides decreased in size to some extent without any complication for 6 months after treatment [Figure 1].

Discussion

ALHE is a rare benign vascular lesion presenting as multiple nodules in the dermis and subcutaneous tissues of the head and neck.[1,2] It affects women during early to mid-adult life more commonly.[4] The pathogenesis of ALHE is still unclear.[5] Males are more affected by ALHE in the oral mucosa (75%) with an average age of 35 years. About 40 cases of ALHE in the oral mucosa were reported (10 were on the tongue).[1,6] Lips and tongue were the first and second most frequent sites of involvement, respectively. The youngest patient who reported oral mucosal involvement was a 3-year-old boy with a nodule on his upper lip. Most lesions presented as nodules but ulcers, macules, and pruritus were other presenting signs and symptoms. Our patient was the youngest patient presenting with oral ALHE. Endothelial cells have a cobblestone appearance, also perivascular and interstitial lymphocytic and eosinophilic infiltrate is present.[7] Eosinophilic ulcer of the tongue is the most important clinical and histopathologic differential diagnosis of ALHE on the tongue but it is a self-limiting lesion.[8,9] In contrast, the lesion, in this case, was present on his tongue for about 18 months before treatment. Another important histopathologic differential diagnosis is Kimura's disease.[3,6] There are several treatment options for ALHE, among which surgery had been considered the best option.[4] Other treatment modalities for cutaneous ALHE include corticosteroids (topical, intralesional, and oral), isotretinoin, interferon alpha 2a, cytotoxic agents, diathermy, laser cauterization, cryotherapy, and pentoxifylline.[3] And other therapeutic approaches such as oral corticosteroids have not been used in mucosal ALHE. A 26-year-old woman with ALHE on her cheek and upper lip was treated successfully with an oral corticosteroid.[3] Although surgical excision was reported to be the treatment of choice in oral ALHE because of the low rate of relapse[3]; it is an invasive method to be done in infants, so our case was treated with oral corticosteroids. The ulcer healed completely. As the size of the patient's tongue was disproportionate to his

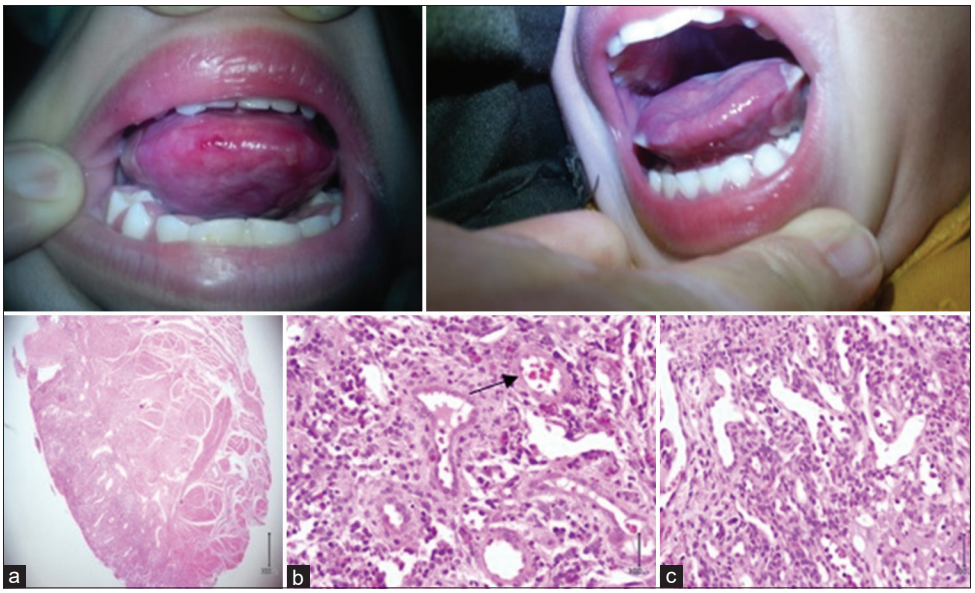

Figure 1: The top left picture presents an extensive ulcerated lesion on the tongue before treatment (The ulcerated area was present on the anterior third of the dorsal surface of his tongue with rather sharp edges and focal extensions to the ventral surface without active hemorrhage) and top right picture presents decrease in size after treatment with prednisolone. (a) Tongue muscle with overlying epithelioid hemangioma (Hematoxylin and Eosin (H&E), original magnification ×4). (b) Epithelioid vessels (in ALHE blood vessels are lined by plump epithelioid endothelial cells) with intracytoplasmic lumen containing Red Blood Cells (RBCs) (arrow) and frequent eosinophilic infiltration. (c) Vessels with hobnail endothelium (H&E, original magnification ×10)

mandible, trauma to the sides of his tongue continued; so, poor response of this part to our treatment can be explained. After discontinuation of prednisolone, tacrolimus 0.03% ointment was used twice daily for maintenance therapy, and the patient is still on this treatment for a month. In conclusion, using oral corticosteroids may be useful in the treatment of mucosal ALHE, especially in those patients where surgery can be a challenging method.

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Conflicts of interest

There are no conflicts of interest.

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