Angiolymphoid hyperplasia with eosinophilia on lip in a 10-year-old girl

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Abstract

Angiolymphoid hyperplasia with eosinophilia (ALHE) is an uncommon benign vascular lesion that is clinically characterized as papule or nodule and commonly found in the head-and-neck areas although rarely involve oral cavity. The etiology of ALHE is currently unknown, and various hypotheses include a reactive vascular proliferation, vascular malformation or neoplasm. ALHE is mostly occurs in middle-aged females and rare in children. Here, we are presenting a case of ALHE on lip in a 10-year-old female child.

Keywords: Angiolymphoid hyperplasia with eosinophilia, lip, oral mucosa

Case Report

A 10-year-old female child presented with a slow growing, painless soft lesion with smooth surface on labial surface of lower lip for 3 months [Figure 1]. The patient was of good built with no significant family and medical history. There was no history of trauma and insect bites. On intraoral examination, a solitary well defined, sessile and oval-shaped swelling measuring about 1.0 cm × 0.7 cm in size on mid-labial surface of lower lip with normal overlying mucosa. Rest clinical examination being within normal limit.

Routine preoperative biochemical reports were unremarkable, and no peripheral blood eosinophilia was noted. The lesion was completely excised under local anesthesia with primary closure done. Procedure was done on the Outpatient Department basis only, and the patient remained in follow-up till 1 year without any recurrence.

Microscopically, lesion showed well-circumscribed dermal nodule composed of proliferating blood vessels...
of varying sizes, around large vessels, lined by plumped epithelioid shaped endothelial cells (mostly cuboidal or occasionally hobnailed) with abundant cytoplasm and prominent nuclei. Perivascular infiltrate is primarily composed of eosinophils and lymphocytes. No cellular atypia and mitosis were evident. Lymphoid follicles were not noted in lesion. Superficial nonkeratinized stratified squamous epithelium with basilar hyperplasia was also evident [Figures 2-4]. Final diagnosis was consistent with ALHE.

Differential diagnosis includes mucocele, lipoma, fibroma, pyogenic granuloma, salivary gland tumors, Kimura’s disease (KD), angiosarcoma and epithelioid hemangioendothelioma.

DISCUSSION

ALHE presents most commonly in young to middle-aged adults, with a mean onset of 30–33 years, as single or multiple, flesh-colored papule or nodule. Although most commonly involved the head-and-neck areas, mainly ear, forehead and scalp, rarely involve the oral mucosa. ALHE has slightly more predilection for females.\[1-8\]

An etiology of ALHE is still unknown, but proposed pathogenesis includes a neoplastic process, a reactive process as hypersensitivity reaction or an inflammatory vascular reaction. Several studies support that ALHE may be associated with secondary to trauma, vaccination, elevated estrogen level and infections.\[1,6,8\] A mutation in TEK gene, encodes the endothelial cell tyrosine kinase receptor Tie-2, was to associated in pathogenesis of ALHE.\[1\] ALHE is mostly seen in Asians while lowest incidence in blacks.\[8,10\]

Characteristic histopathologic feature of ALHE shows proliferation of small blood vessels, lined by enlarged endothelial cells with large vesicular nuclei and abundant eosinophilic cytoplasm, surround central blood vessel. The perivascular inflammatory infiltrate consists of lymphocytes and eosinophils. Eosinophils typically comprise 5%–15% of an infiltrate and rarely may account for as much as 50%
of an infiltrate. Around 20% of the patients have blood eosinophilia. Diffuse infiltration of lymphocytes with or without follicle formation is evident.[1,8]

ALHE must be distinguished from KD because both are considered eosinophilic dermatoses and have morphological similarities. ALHE mainly presents in Caucasians and is clinically characterized by superficial papules or nodules, without lymphadenopathy and accompanied by a mild peripheral eosinophilia. However, KD is mostly observed in Orientals and presents as deep nodules with lymphadenopathy and a marked serum eosinophilia, hyperimmunoglobulinemia and sometimes may be associated with renal disease. The characteristic histopathological features of ALHE are proliferation of blood vessels of varying sizes lined by enlarged endothelial cells, whereas salient features of KD are florid lymphoid follicles with germinal center formation surrounded by eosinophilic infiltrate and fibrosis.[5,6,8,12]

ALHE must also be differentiated from angiosarcoma and epithelioid hemangioendothelioma. Angiosarcoma can be distinguished from ALHE by brisk mitotic activity, nuclear hyperchromasia and pleomorphism. However, epithelioid hemangioendothelioma is differentiated from ALHE by the presence of vacuolated endothelial cells arranged in streaks or cords without lymphoid and eosinophilic infiltrate.[1‑8]

ALHE rarely presents in children, and occurrence in oral cavity like on lip being rarer so it should always be considered in differential diagnosis of lip and only few cases have been published till now regarding this.[7,10]

Surgical excision is the first line of treatment and when completely excised rarely recurs. Medical treatment with intralesional corticosteroids, isotretinoin, topical imiquimod, tacrolimus, interferon α-2a and radiotherapy is generally not very effective. Hence, surgical excision is the most commonly used therapeutic intervention and has the lowest failure rate of 44.2% as compared to medical intervention with failure rate of 80%–100%. ALHE has chronic and benign course with good prognosis, and there are no reports of malignant transformation.[1,8,10,12]

**CONCLUSION**

Concerning the prognosis of ALHE, it tends to persist for years without regressing, which makes surgical or medical intervention necessary in most cases. Although rare, especially in pediatric patients, the differential diagnosis of ALHE should always be kept in mind. Each and every tissue dissected should always be sent for histopathological examination. The patient of ALHE should be well assured of benign nature of the disease along with chronic course and possibility of recurrence.

**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 initial(s) will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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