Angiolympyphoid hyperplasia with eosinophilia (epithelioid hemangioma) of the face: An unusual presentation

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Abstract

Angiolympyphoid hyperplasia with eosinophilia (ALHE) is a rare entity that usually occurs as 0.5–3 cm pink to brown nodules in the skin surface and subcutaneous tissues of the head & neck. Here we report an unusual occurrence in size and appearance of ALHE in the tissues of the cheek of an adult Asian male.

Keywords:
Angiolympyphoid hyperplasia
Eosinophilia
Epithelioid hemangioma
Face

1. Introduction

Angiolympyphoid hyperplasia with eosinophilia (ALHE, epithelioid haemangioma, inflammatory angiomatous nodule, atypical granuloma, pseudopagocytic granuloma, and histiocytoid hemangioma) is an uncommon, benign, reactive vaso-proliferative disease, presenting with painless, vascular nodules in the dermal and subcutaneous tissues of the head and neck, particularly around the ear.1 ALHE has also been reported in the scalp,2 lip,2 tongue,2 orbits2 and the conjunctiva.5 Although it is usually superficial in nature, some authors have reported muscular and bony involvement.1–5 ALHE is somewhat more common in females, however, a male predominance has been noted in selected Asian studies.6 It presents most commonly in patients aged 20–50 years, with a mean onset of 30–33 years.8 This condition is uncommon; it is rare in elderly patients and in the non-Asian paediatric population. ALHE can persist for years, but serious complications (e.g., malignant transformation) do not occur and have never been reported. It is often characterised by a distinctive infiltrate, composed of lymphocytes, eosinophils, and plasma cells. The condition is confirmed by the histopathological presence of abnormally proliferating, plump endothelial cells often in the vicinity of muscular arteries.1

ALHE lesions are typically small, pruritic, erythromatous nodules with diameter between 0.5 cm and 3 cm7 with larger presentations being a relatively rarer manifestation.8 We report an unusual presentation in size and appearance of ALHE in the tissues of the cheek of an adult Asian male.

2. Case presentation

An otherwise fit and healthy 51-year-old Indian male presented to our Maxillofacial clinic with a 8 cm × 6 cm solitary mass in his right cheek, lying deep to the skin, superior to his upper lip and lateral to the right nostril. It had slowly enlarged over the last three years. There was no obvious cutaneous ulceration, crusting or discolouration which are commonly associated with ALHE. The mass was non-tender, fixed, with a smooth surface possessing a solid consistency when palpated. No regional lymphadenopathy was observed. A full blood count (FBC) showed a white blood cell count (WBC) of 8.9 × 10⁹/l with eosinophilia of 1.74 × 10⁹/l (0.04–0.44 × 10⁹/l normal range). Cytology from an ultrasound-guided fine needle aspiration (FNAC) showed no malignant cells, but demonstrated loose connective tissue, lymphocytes, macrophages and eosinophils. A computerised tomography (CT) scan (Fig. 1) showed the mass lying in the subcutaneous tissues with no lytic involvement of underlying bone.

The lesion was fully excised under general anaesthesia via a modified Weber–Ferguson incision (Figs. 2 and 3) while maintaining the terminal branches of the facial nerve (CNVII) during dissection. This approach was chosen over a more conservative intra-oral vestibular approach following discussion in a Multi-Disciplinary Meeting with the radiologists and the histopathologists to ensure good access and full excision of the lesion. The approach would include the arterial and venous segments at the base of the lesion, while reducing the risk of facial...
nerve (CNVII) damage. A well vascularised fibrotic mass was found to extend deep in the facial tissues to within 5 mm of the oral mucosa. As expected, it also involved small branches of the facial nerve, which were successfully dissected free. Post-operative recovery was very good, and there was no clinical impairment of facial nerve function (Fig. 4). Histological examination showed proliferation of small blood vessels lined by plump endothelial cells and surrounded by a lymphoid infiltrate with formation of germinal centres. Large numbers of eosinophils were seen (Figs. 5 and 6). A diagnosis of ALHE was made. There has been no recurrence after a follow-up period of 18-months.
3. Discussion

Many theories exist to explain the aetiology of ALHE. The main contention remains controversial whether it is a neoplastic or an atopic hypersensitivity reaction (unusual reactive process) with the latter receiving more support. Several reports also indicate the possibility that ALHE may be secondary to infection or trauma. However, the exact relationship of these lesions to each other, including Kimura’s disease, which represents an allergic or autoimmune response, remains incompletely understood.

First described in 1969 by Wells and Whimster,¹ ALHE is a disfiguring lesion characterised by isolated or small numbers of pinkish to reddish-brown papules, frequently in the head and neck region. Although the range of diameters of lesions in ALHE is stated in dated literature between 1 and 10 cm,² the vast majority of reported presentations are within 0.5 cm and 3 cm in diameter.³ Larger lesions, falling into the upper end of the spectrum are relatively rare, and as far as the authors could find have not been reported in the last 30 years.

Although our patient exhibited all of the cardinal and distinguishing histopathological features of ALHE, the initial diagnosis of ALHE was difficult because the lesion was exceptionally large, with no obvious surface changes. The lesion’s clear demarcation, insidious growth, and sub-mucosal limitations indicated a benign neoplasia.⁴ Hence the differential diagnosis was primarily a mesenchymal tumour of the vessels, nerves or muscles in the head and neck, namely, neurilemoma, leiomyoma, hemangioma, and fibrosarcoma.⁵

Though there are several treatment options, including intralesional injections of isotretinoin, glucocorticoids, interferon alpha-2a, cytotoxic agents, and irradiation therapy,⁶ the most effective therapy is complete excision and follow-up.¹ Despite a 33% recurrence rate,⁹ our patient had a successful recovery with no recurrence in his 18-month follow-up.

ALHE has rarely been reported to exceed 3 cm in diameter, but based on our case report, it should be a viable differential diagnosis when encountering large subcutaneous tumours of the head & neck.

Conflicts of interest

No conflict of interest to declare by any of the authors.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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