Solitary Angiokeratoma of Tongue: A Case Report and Review of the Literature

Abstract
Angiokeratoma is a benign cutaneous lesion of capillaries. It is characterized by large dilated blood vessels in the superficial dermis and hyperkeratosis of extremities. It is mostly seen in generalized form affecting the extremity of the body, but we report this case of solitary angiokeratoma of the tongue which is a very rare type.

Keywords: Cutaneous, extremities, hyperkeratosis, solitary angiokeratoma

Introduction
Angiokeratoma circumscriptum is a rare benign cutaneous vascular lesion that usually occurs in the extremities.[1,2] It is usually asymptomatic and appears as well-demarcated lesion characterized by large dilated blood vessels in the superficial dermis and hyperkeratosis of extremities.[3,4] Mibelli (1989) first reported the case of angiokeratoma in the fingers and toes. The etiology of this disease is unknown, but the usual factors considered are congenital, pregnancy, trauma, itchy, and painful swelling on hand and foot, tissue asphyxia. These lesions are present in 70% of males and 39% of females who report with pinpoint, dark-red to blue-black, macular and papular lesions of around 4 mm, which do not blanch on pressure.[4,5] The oral cavity is usually involved with a generalized form of the lesion or with multiple lesions of the tongue, but the following case report is on the solitary form of the lesion on the tongue which is found to be a rare condition.

Case Report
An 11-year-old boy presented to the Department of Oral and Maxillofacial Surgery, Faculty of Dental Science, Sri Ramachandra Medical Centre with a chief complaint of difficulty swallowing and a burning sensation on consuming hot and spicy food. The patient gave a history of swelling on the right side of the tongue. The swelling was noticed by the patient 4 years back which gradually increased in size with time and was not associated with pain or pus discharge. On local examination, the swelling was found to be an erythematous, nontender, nonfluctuant, noncompressible, mass measuring 4 cm × 1.5 cm wide spread over the right dorsum of tongue [Figure 1]. The swelling extended anteriorly from the tip of the tongue to the posterior one-third of the tongue. Medially, it extends until the midline of the tongue and laterally about one-half cm from the lateral border of the tongue dorsally. On general examination, no other swelling or lesion was present in the extremities or body. As an investigatory finding, ultrasound of tongue was taken which revealed it to be a vascular malformation of the tongue.

A computed tomography (CT) angiogram plain and contrast was taken (80 ml of intravenous contrast) which further showed no hypertrophic changes of arterial or venous channels and no feeder noted from the lingual artery or other branches of the external carotid artery to the lesion [Figure 2]. CT revealed the swelling of the tongue as a nonvascular lesion. An excisional biopsy under general anesthesia was performed.

The excised specimen [Figure 3] was sent for histopathological examination which showed parakeratinized stratified squamous epithelium with elongated rete ridges [Figure 4]. Connective tissue revealed numerous large dilated blood-filled spaces lined by endothelial cells surrounded

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by chronic inflammatory infiltrate. Areas of extravasations of blood were also present, and a final diagnosis of solitary angiokeratoma of the tongue was made. Patient has been under follow-up for 2 years without any recurrence [Figure 5].

Discussion

The first case of angiokeratoma was reported by Mibelli in the year 1889 on fingers and toes.[4,6] Angiokeratoma circumscriptum was first described in 1915 by Fabyr as a localized lesion on the lower extremity or trunk.[5] The following are the variants of angiokeratoma out of which the third variant is very rare and was reported by us. Angiokeratoma is classified as:[7,8]

1. Angiokeratoma of Mibelli (also known as “Mibelli’s angiokeratoma, Telangiectatic warts) consists of 1–5-mm red vascular papules, the surfaces of which become hyperkeratotic in the course of time. The disease is named after Italian Dermatologist Vittorio Mibelli (1860-1910).[2-6]

2. Angiokeratoma of Fordyce (also known as “Angiokeratoma of the scrotum and vulva,” although not to be confused with Fordyce’s spots) is a skin condition characterized by red to blue papules on the scrotum or vulva[2,4,5]

3. Solitary angiokeratoma is a small, bluish-black, warty papule that occurs predominantly on the lower extremities[1,2]

4. Verrucous vascular malformation (also known as “Angiokeratoma circumscriptum naeviforme”) is a malformation of dermal and subcutaneous capillaries and veins. This is a congenital vascular malformation, on which, over time, a verrucous component appears

5. Angiokeratoma corporis diffusum of Fabry— The generalized systemic type.[5,6]

Angiokeratoma circumscriptum has been reported to coexist with angiokeratoma of Fordyce (found on the scrotum) and “caviar spots” (angikeratomas of the tongue). Oral mucosal involvement is most commonly a component of angiokeratoma corporis diffusum.[9] It is usually associated with the syndromes such as Klippel-Trenaunay-Weber syndrome, Cobb syndrome, and other mixed vascular malformations.[4] Angiokeratoma though appears in the oral cavity, mostly associated with systemic disease and represents as multiple papules in buccal mucosa and tongue, but in our case, the patient had no systemic disease and represented with a single lesion in the tongue which caused dysphagia for the patient. However, they
differ clinically in appearance and morphology but share a similar histological feature. Thrombosis of these vessels are common and is responsible for the clinical mimicry of melanoma. In 1967 Imperial and Helwig introduced the term “ verrucous hemangioma” and distinguished it from angiokeratoma and its variants. Verrucous hemangioma is a congenital, localized, vascular malformation, histologically characterized by dilated capillaries and large cavernous, endothelial-lined, blood-filled spaces extending well into the reticular dermis and subcutaneous fat and with hyperkeratosis. On histopathological examination, our case of solitary angiokeratoma revealed all characteristic histological features of angiokeratoma circumscriptum which are dilated dermal papillary capillaries drained by dilated venules. The overlying epidermis shows a variable degree of acanthosis, papillomatosis, and compact hyperkeratosis. Angiokeratoma circumscriptum are asymptomatic, benign, vascular malformations that require no treatment. Nevertheless, surgical treatment is often rendered for improving function, cosmesis and a better quality of life. Other ablative therapies such as cryotherapy, the laser may also be effectively used to treat angiokeratomas, but our case reported with a complaint of functional impairment and had dysphagia and hence was treated by excisional biopsy under general anesthesia. A follow-up of 2 years revealed no recurrence of the lesion and patient is leading a normal life. To the best of our knowledge, only ten cases of isolated angiokeratoma in the oral cavity have been reported in the literature review by Kang et al. in 2014. Majority of the cases were found to be on the tongue. Only two cases were found to be in buccal mucosa. Until date, only two reported cases of solitary angiokeratoma of the tongue by Siponen et al. (2006) in a 54 year female and Erkal et al. (2013) in a 67-year-old female. Our case supposed to be the third reported solitary Angiokeratoma of the tongue and the first youngest age group (11 years) male patient.

Conclusion

Angiokeratoma of the tongue is a rare cutaneous vascular lesion mostly associated with the systemic disorder and syndromes. We are reporting the third solitary case of this rare variant of Angiokeratoma of the tongue; out of which our patient is the youngest aged 11 years old boy who did not show any other systemic disorders and cutaneous lesions, and being successfully treated without morbidity.

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 initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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