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Case Report

Dentistry Section

Isolated Angiokeratoma of Oral Cavity: A Rare Case Report

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ABSTRACT

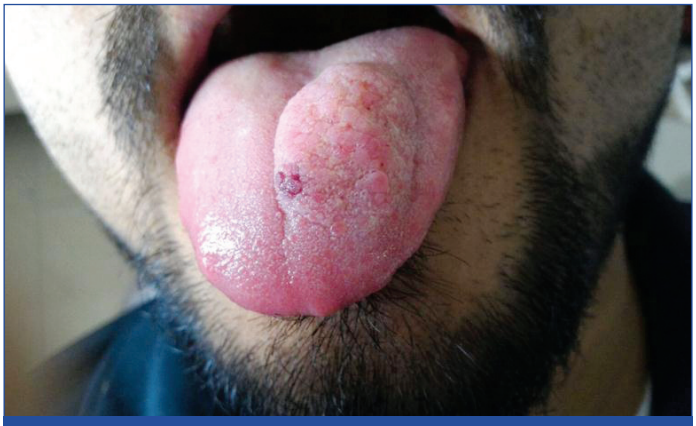
Angiokeratomas are rare benign mucocutaneous lesions that result from distension of subepidermal vessels, along with hyperkeratotic epidermis. Mucosal angiokeratomas are more commonly seen as a constituent of systemic variant called angiokeratoma corpus diffusum or may co-exist with similar lesion in the other sites of the body as seen in Fordyce's or congenital forms. Solitary lesion in oral cavity is very infrequently seen and till date very few cases have been reported in literature. The biologic significance of angiokeratomas may vary greatly, which may range from lesions that have very little clinical repercussion to widespread eruptions that are a manifestation of potentially fatal, systemic, metabolic diseases therefore it is important that these lesions should be identified and evaluated to rule out underlying pathologic conditions. Hereby, authors reported a case of a 24-year-old male who presented with a unilateral swelling on dorsal tongue which gradually increased in size over 1.5 years with history of occasional bleeding and pain. It is common to see involvement of oral cavity in systemic variety but very rare in localised solitary form, therefore while considering the differential diagnosis of the swellings of tongue this entity should be taken into account.

Keywords: Blood vessels, Histopathology, Oral, Solitary, Tongue, Tumour

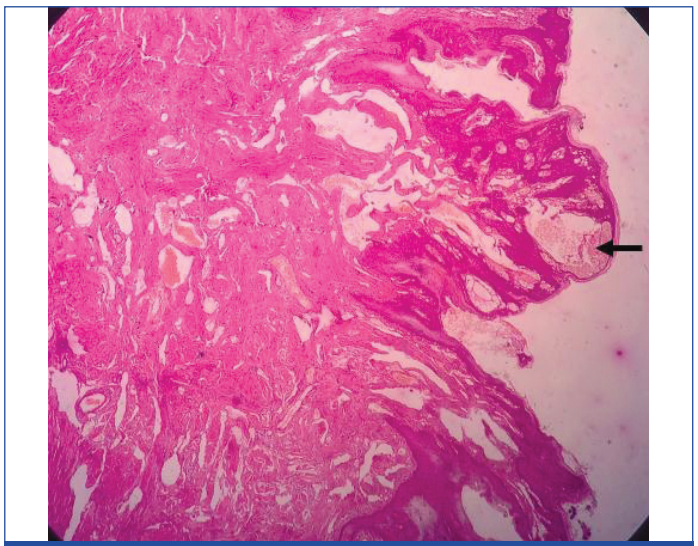
CASE REPORT

A 24-year-old male patient attended the dental clinic complaining of a raised growth on tongue from last 1.5 years. The swelling was initially small and a gradual increase in size was observed. The patient also complained of occasional pain and bleeding occurring following trauma due to mastication that subsided on its own. The medical history was non significant with no history of symptoms suggestive of any metabolic derangement in patient of his family.

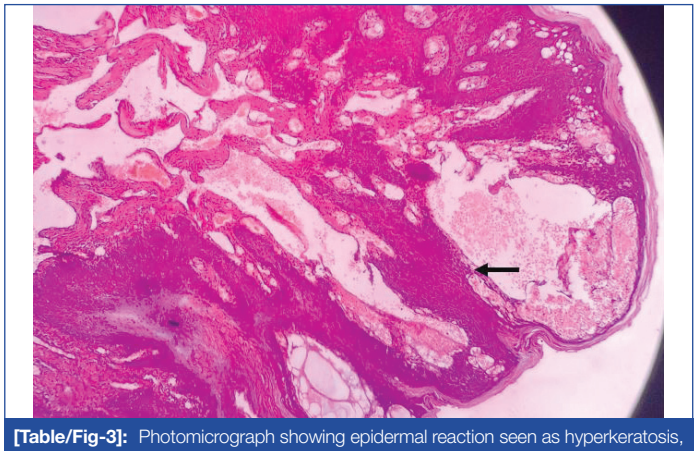
On intraoral examination there was a solitary, well-defined, non tender, sessile nodular growth on the left side of the dorsal surface of tongue measuring approximately 2.5×2 cm [Table/Fig-1]. The growth had a granular surface texture with dark bluish-red papule at one area which did not blanch on pressure. There were no other changes in the oral cavity and no similar lesion was noted elsewhere on the skin. Patient's Complete Blood Count (CBC), Liver Function Tests (LFT), Kidney Function Tests (KFT) and urine analysis were within normal range. The provisional diagnosis of benign mesenchymal lesion was given and the lesion was excised under local anaesthesia and the specimen were sent for histopathological examination. Microscopy revealed dilated thin walled cystic spaces lined by endothelial cells in the papillary dermis. The overlying epithelium was characterised by hyperkeratosis, acanthosis and elongation of the rete ridges encircling the dilated vascular spaces. The surrounding connective tissue showed mild chronic inflammatory cell infiltration [Table/Fig-2,3]. Based on the classical histopathologic features and absence of any other lesion anywhere in the body, diagnosis of solitary angiokeratoma was given. During two years follow-up there were no relapse and patient remained asymptomatic.



[Table/Fig-1]: Oral angiokeratoma over the dorsum of the tongue.



[Table/Fig-2]: Photomicrograph showing dilated blood vessels in papillary dermis (H&E,100X).



[Table/Fig-3]: Photomicrograph showing epidermal reaction seen as hyperkeratosis, elongated rete ridges (H&E, 200X).

DISCUSSION

Angiokeratomas are rare benign acquired vascular lesions which are characterised by the existence of distended dermal vessels with hyperplasia of epidermis.The pathophysiology of the lesion includes injury or irritation to the wall of capillaries in papillary dermis, venous hypertension or arteriovenous malformation [1]. They can be divided into localised and systemic types depending on the multiplicity and location of the lesions [2].

Four different clinical subtypes of angiokeratomas have been recognised these are: solitary, angiokeratoma corporis diffusum, Mibelli and Fordyce [3]. [Table/Fig-4] depicts the detail comparison between the various clinical variants. All the varieties of angiokeratomas however have similar histologic features [4].

Clinical variants	Solitary angiokeratoma	Angiokeratoma corporis diffusum (Fabry disease)	Mibelli angiokeratoma	Fordyce
Aetiology	Caused by chronic irritation, trauma or injury to the wall of a blood vessel in papillary dermis.	Represents a cutaneous manifestation of a group of hereditary enzymatic disorders; results from deficiency of the lysosomal enzyme hydrolase alpha-galactosidase A	Mibelli angiokeratoma is a condition that is inherited in an autosomal dominant fashion.	Usually associated with varicocoele, inguinal hernia and thrombophlebitis. The lesions may develop after surgical injuries to the genital veins.
Age and sex	Affect mainly young adults	Usually appear shortly before puberty; X-linked disease, exclusively affect males; females may be asymptomatic carriers.	Usually appear in childhood or adolescence and they are more common in females.	Affects elderly people, however, there are examples of congenital cases.
Site	Most common site of occurrence is the lower limbs however any anatomic site can be affected including the oral cavity.	Fabry disease has bathing-trunk distribution; affecting the lower part ofabdomen, genitalia, buttocks, and thighs. Oral mucosal involvement is common.	Commonly seen affecting the dorsum of the fingers, toes and interdigital spaces.	Most common in the scrotum and vulva.
Clinical presentation	Small, warty, black, well-circumscribed papules. Sometimes solitary angiokeratomas develop thrombosis and recanalisation with the development of secondary intravascular papillary clinically endothelial hyperplasia. Due to their colour, these lesions may be clinically confused with malignant melanoma.	Dark red punctate papules with size less than 1 mm in diameter in mild cases, and female carriers an Asymptomatic superficial corneal dystrophy also known as cornea verticillate, is frequently seen and helps in diagnosis. Other symptoms include dry skin, anhidrosis, hyperthermic crises, capillary changes in the nail matrix leading to acroparaesthesiae. In rare cases patients may also report with existing Klippel-Trenaunay-Weber syndrome.	Several dark papules with a slightly hyperkeratotic surface, and may be associated with acrocyanosis and chilblains.	Characterised by the occurrence of multiple papules with diameter 2-4 mm, dark purple in color.

[Table/Fig-4]: Comparison between the various clinical variants of Angiokeratoma [3].

Oral mucosal angiokeratomas are most commonly seen as a component of angiokeratoma corporis diffusum and is very uncommon in other types. In 1967 Imperial R and Helwig EB were the first to describe solitary angiokeratosis as a distinct entity [4]. Later Schiller PI and Itin PH found that Isolated angiokeratoma represented almost 70-83% of all angiokeratomas and was the most common type [5]. Isolated lesions were commonly found on the lower leg area, abdomen, scrotal wall, shaft of penis and rarely oral mucosa [6]. First case of isolated oral angiokeratomas without any metabolic disease was reported in literature by Leung CS and Jordan RC in 1977 [7]. Since then, very few cases have appeared in literature. The dorsal surface of the tongue is the most common site, other sites include ventral surface [8,9], lateral tongue [10], tip of the tongue [1] buccal mucosa [7] and tonsillar pillar [11].

The pathogenesis of isolated angiokeratomas is unclear but external trauma is believed to play a role [5]. The lesions are usually asymptomatic however it may be associated with episodes of bleeding and pain related to irritation or trauma. Clinically isolated angiokeratomas have varied presentations as papular, nodular, plaques, wart like growth and their colour may vary from pink, dark red, bluish black to brown which may be mistaken for number of lesions such as melanoma, wart, naevi, seborrheic keratosis, verruca vulgaris, haemangioma, lymphangioma, and occasionally malignancy [12-14]. The final diagnosis in the present case was made based on microscopic examination. Histologically oral angiokeratoma and those arising on the skin shows similarity by exhibiting acanthosis and hyperplasia of overlying stratified epithelium. However, while cutaneous lesions show hyperorthokeratosis, the oral lesion shows hyperparakeratosis [15]. The papillary dermis shows the presence of large dilated blood vessels lined by endothelial cells that appear normal. These blood vessels are filled with erythrocytes and thrombi may be present [4]. The elongated rete ridges surround many of these blood vessels. These features are also seen in other oral lesions like lymphangiomas and haemangiomas and hence differential diagnosis is important. Lymphangiomas exhibit lymph vessels in the papillary connective tissue in close proximity to epithelium [16,17]. However, in angiokeratomas the blood filled spaces are lined by endothelial cells and this helps to differentiate from lymphangiomas. Haemangiomas are usually associated with proliferation of small blood vessels in the deep connective tissue stroma. These blood vessels are lined by single layer of endothelial cells and proliferation of endothelial cells may also be seen [18]. In contrast blood vessels lie underneath a papillomatous epithelium in angiokeratoma [19].

The present case was seen in young male who first noticed a raised lesion on tongue 1.5 years back that gradually increased in size and occasionally bled. The lesion was solitary without any evidence of similar lesion elsewhere and exhibits the classical histopathological features like hyperplastic epithelium, rete ridges obliterating the dilated dermal spaces containing erythrocytes and hence the final diagnosis of solitary angiokeratosis was made.

Generally, solitary angiokeratomas do not require further evaluation by imaging studies, however when multiple lesions are noted and some other underlying disease is suspected then imaging studies like Computed Tomography (CT) Angiogram (plain and contrast), Magnetic Resonance Imaging (MRI) Angiography, doppler studies can be done to see hypertrophic changes of arterial and venous channels [20]. Desmoscopy is helpful tool for accurate diagnosis for solitary angiokeratomas by allowing differentiating from other cutaneous tumours such as malignant melanomas and pigmented basal cell carcinomas [21].

The treatment for solitary angiokeratoma includes surgical excision, diathermy, electrocoagulation, cryotherapy, multiple lesions however require laser ablation [22].

CONCLUSION(S)

Authors present a case of solitary angiokeratoma on tongue arising in a young patient without any underlying pathology. These cases are very rare and require thorough evaluation to rule out any underlying systemic condition. Thus the knowledge of this entity is required for proper identification and histopathological diagnosis

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