

原文題目(出處)：	Solitary Angiokeratoma of Oral Mucosa: A Rare Presentation. Case Rep Dent 2013, Article ID 812323
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內文：

Introduction

1. Angiokeratoma is an acquired vascular lesion which is characterized histologically as one or more dilated blood vessels lying directly subepidermally and showing an epidermal proliferative reaction.
2. They can be divided into localized and systemic types. Mucosal involvement, including the oral cavity, has been described both as localized and systemic types, as a component of Fabry's disease, or as a component of fucosidosis.
3. To classify isolated oral mucosal angiokeratomas, other classification systems have been proposed by Ranjan and Mahajan .
4. solitary angiokeratomas of the oral mucosa seem to be a rather infrequent occurrence, and very few cases have been reported in the literature. Since 1997 till date, only 16 cases involving oral cavity have been reported.

Case Report



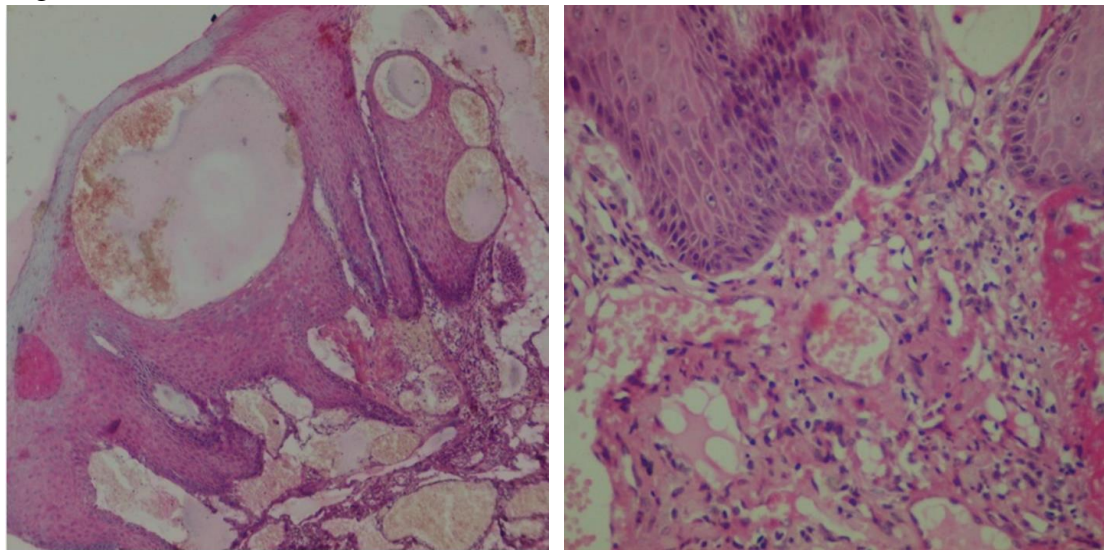
1. A 38-year-old male patient reported to the Department of Oral Pathology with chief complaint of growth on tip of tongue since last 10 years. He noticed small painless growth, and then the growth steadily increased in size involving right side of the tip of the tongue.
2. Patient also experienced bleeding associated with trauma during mastication.
3. No abnormality was revealed in his medical, personal histories and general examination. The patient had a habit of tobacco chewing for about 10–15 years.
4. On clinical examination, it was observed that well-circumscribed sessile growth is present on the dorsal surface of tip of the tongue, approximately 1 × 1 cm in dimension, ovoid in shape. Growth was dark brownish in color with a granular surface texture.
5. On palpation, growth was non tender and rough.
6. After routine hematological investigations, under local anesthesia, the lesion was completely excised and taken for histopathological investigation. The gross specimen is irregular in shape approximately 1 × 1 × 0.5 cm in size, brownish in color, and soft

in consistency with rough surface.

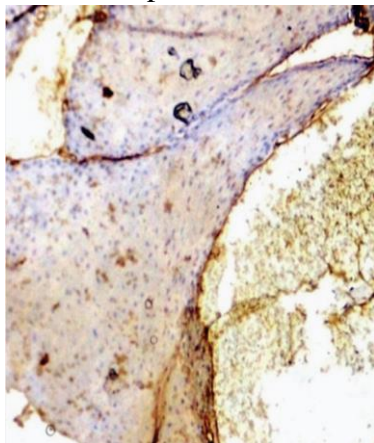


7. Histopathologically, parakeratotic stratified squamous epithelium of varying thickness with long slender rete ridges and in some areas large bulbous rete ridges is evident.

8. Papillary connective tissue shows numerous large dilated blood filled spaces and lined by endothelial cells. Chronic inflammatory cell infiltration around blood vessels and rete ridges is also present. All these features were suggestive of a diagnosis of angiokeratoma (下圖)



9. For the confirmation of proliferation of blood vessels, CD34 marker was used. The lesion was positive for CD34 (下圖)



10. After diagnosis, the patient underwent further examinations, and no lesions were found elsewhere in his skin or mucous membranes. The case was considered a solitary angiokeratoma affecting the tongue.

11. In the last follow up after six months, the patient was disease-free and asymptomatic.

Discussion

1. Solitary angiokeratoma was first described in 1967 by Imperial and Helwig.

2. These lesions are commonly found on the hips, thighs, buttocks, umbilicus, lower abdomen, scrotum, glans penis, and rarely oral mucosa. Thorough search, we found only 16 case reports of solitary angiokeratomas affecting oral cavity.
3. Solitary angiokeratomas have been described in the oral cavity, mainly the tongue. Also, one case was also reported on the tonsillar pillar.
4. Pathogenesis of the lesion includes relation to trauma, high venous pressure, or vascular malformation.
5. The increase in proliferation of the epithelium is because of the close proximity of the vascular spaces. In case of angiokeratoma, the blood vessels are in close proximity to epithelium, and hence their close proximity to epithelium suggests the secondary proliferation of epithelium.
6. Review of all the past cases suggests that it is more common in female, but the present case patient was male. The most common site of involvement in the oral cavity is the tongue, the anterior dorsal surface. In present case the site of involvement was also the tongue.
7. The only clinical problems these lesions can cause are bleeding, discomfort or cosmetic changes. Most cases were asymptomatic.
8. Therapy has usually been surgical excision in most of the published cases. A recent report has employed diode laser in a 16-year-old woman.
9. Usually, no recurrences have been described, but few recent cases suggest the recurrence. In the present case, after surgical excision, no recurrence is found after 6-month follow up.
10. Oral mucosal involvement is a component of angiokeratoma corporis diffusum. If further lesions elsewhere are present, then the possible association with systemic diseases could be expected in wide spread cases.
13. Fabry's disease and fucosidosis can be suspected on histopathological grounds by the presence of swollen endothelial cells with a vacuolated cytoplasm in addition to the histology of angiokeratoma.
14. The present case did not show swollen endothelial cells. Also, no other associated lesions were identified. Hence, the present case can be categorized as an isolated solitary angiokeratoma of oral cavity affecting tongue
15. Solitary angiokeratoma of the tongue in adults has proposed a clinical classification for oral angiokeratomas.
 - (1) Type 1: primary (purely mucocutaneous and not associated with systemic disorders)
 - >Type 1A, isolated angiokeratomas of the oral cavity
 - Type 1As solitary
 - Type 1Am multiple
 - >Type 1B, mucocutaneous angiokeratomas, that is, oral angiokeratomas associated with cutaneous angiokeratomas
 - Type 1Bs solitary
 - Type 1Bm multiple
 - >Type 1C, angiokeratomas occurring simultaneously in oral cavity ,skin and gastrointestinal mucosa
 - Type 1Cs solitary
 - Type 1Cm multiple
 - (2) Type 2: secondary (as a component of a generalized systemic disorder)
 - >Type 2A, as a component of Fabry's disease
 - Type 2As solitary
 - Type 2Am multiple

- >Type 2B, as a component of fucosidosis
 Type 2Bs solitary
 Type 2Bm multiple
16. Considering the same classification, the present case can be categorized as Type 1As, that is,isolated solitary angiokeratoma.
17. Immunohistochemical staining is implied. In the previous literature,antigens used were CD31, CD34, and LYVE-1 (lymphatic vessel endothelial hyaluronan)
 (1) CD31 and CD34 were found positive and LYVE-1 (lymphatic vessel endothelial hyaluronan) was negative.
 (2) In the present case, antigen used was CD34. CD34 antigen that was used is considered as a reliable marker for the proliferating blood vessels.
18. The differential diagnosis of angiokeratoma is important because of its similarity to some other lesions. Other vascular lesions like hemangioma, and lymphangioma can be ruled out with the help of histopathological investigation.
 (1) In case of hemangioma, small capillary lined by single layer of endothelial cells supported by connective tissue stroma is seen.
 Also, endothelial cell proliferation is also noted. These blood vessels are completely lain within the connective tissue, while in case of angiokeratoma, blood vessels are supported by epithelium and lie very close to the epithelium.
 (2) In case of lymphangiomas, multiple intertwining lymph vessels lie very close to the epithelium and are seen also in papillary connective tissue. Presence of blood-filled spaces and endothelial lining also helped to differentiate angiokeratoma from lymphangiomas.
19. Angiokeratoma can be clinically confused with the aggressive lesions like malignant melanoma, especially in case of angiokeratoma when the vessels are thrombosed.In malignant melanoma presence of atypical melanocytes. These cells show prominent nuclei often with prominent nucleoli. Such appearances are not seen in case of angiokeratoma.

Conclusions

1. Oral angiokeratomas of the oral cavity are rare tumors.
2. Although they can appear as isolated lesions, their presence should prompt further investigations to rule out systemic disease.

題號	題目
1	Which following descriptions is not true for angiokeratoma? (A) an acquired vascular lesion (B) Histologically,one or more dilated blood vessels lying directly subepidermally (C) Since 1997 till date, only 16 cases involving oral cavity have been reported. (D) Treatment consists of radiation therapy
答案(D)	出處：此篇 journal, Oral and Maxillofacial Pathology, 2nd ed P.484
題號	題目
2	What is the most appropriate treatment for angiokeratoma? (A) Surgical Excision (B) Local enuleation (C) Decompression (D) No treatment
答案(D)	出處：此篇 journal, Oral and Maxillofacial Pathology, 2nd ed P.484