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Solitary Angiokeratoma of the Buccal Mucosa. Report of a Case

SUMMARY

Background*: Angiokeratoma is an asymptomatic, hyperkeratotic, capillary disorder of the skin present as solitary or multiple, keratotic papules or plaques, which may also be related to Fabry disease. Oral involvement may be observed in cases of widespread muco-cutaneous angiokeratomas, whereas solitary buccal angiokeratoma without systemic/ cutaneous involvement is extremely rare.*

Case Report*: A 45-year-old woman was referred with a 3-month, painless, bluish lesion, located on left buccal mucosa. The medical record of the patient was free of any systemic disease or medication. After a careful clinical oral, mucosal as well as skin examination, an excisional biopsy was taken. A routine haematoxylin-eosin staining and additional immunohistochemistry were performed. Differential diagnosis included haemangioma, haematoma or lesions of melanocytic origin. Clinical examination showed a solid, lobulated bluish lesion, located on left buccal mucosa without other skin or mucosal involvement. The microscopic findings revealed dilated vascular spaces covered by normal endothelium without atypia, extending into the epithelium, indicating the diagnosis of angiokeratoma.*

Conclusions*: Despite its rare occurrence, solitary angiokeratoma of oral mucosa should be included in the differential diagnosis of black-bluish lesions. Further investigation for other similar lesions throughout skin or mucosa is needed to avoid complications as haemorrhage.*

Keywords: Solitary Angiokeratoma; Oral Mucosa

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CASE REPORT (CR)

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Introduction

Angiokeratoma (AK) is a rare capillary vascular disorder, characterized clinically by asymptomatic, solitary or multiple, keratotic papules or plaques, and histologically by benign vascular ectasia of the papillary dermis[1]. Angiokeratomas (AKs) have been described as either a generalized systemic form, presenting as multiple asymptomatic papules on the skin, associated with metabolic diseases, or a solitary cutaneous form. On the basis of their clinical appearance, AKs have been grouped traditionally into localized (purely cutaneous) or widespread types. Though all types differ clinically, they share similar histological features[2].

Mucosal involvement, including the oral cavity, has been reported both in systemic forms and as a component of localized ones. Oral mucosal angiokeratomas (OAKs) are most commonly seen as a component of diffused corporal angiokeratoma in Fabry syndrome. They may also be associated, uncommonly, with AKs of the scrotum and/or gastrointestinal mucosa (jejunum)[1]. However, AKs found exclusively in oral mucosa have been rarely reported as multiple or solitary, mainly located at tongue[2-11].

The aim of this report is to describe a rare case of solitary buccal AK based on clinical and immunohistochemical findings and additionally, we provide a brief review of the literature for OAKs.

Case Report

A 45-year-old woman was referred with a painless, black-bluish (non-homogeneous), solid, lobulated mass (0.5x1cm) located at left buccal mucosa (Fig. 1). According to the patient, the lesion appeared 3 months previously, and was not related to local trauma. It did not change considerably in size and colour, so far. Her medical history was free of any systemic metabolic or other disease. The initial clinical diagnosis included vascular malformations, such as haemangioma, traumatic haematoma or lesion originated by melanocytes. A careful examination of skin and other mucosal linings was performed and the oral lesion was totally excised under local anesthesia afterwards. Serial sections stained with haematoxylin-eosin revealed dilated vascular spaces covered by normal endothelium without atypia, which extended into the overlying epithelium (Fig. 2). In some of the vascular structures, small thrombi were seen. An additional immunohistochemical staining with CD34 (DakoCytomation, monoclonal mouse anti-human, clone QBEnd), by using the automated Envision/HRP immunohistochemical technique (DakoCytomation A/S, Glostrup, Denmark) was performed for better observation of the endothelium of vascular formations (Fig. 3).

The combined clinical and microscopic findings set the final diagnosis of a solitary oral angiokeratoma. In the last follow-up consultation, 1 year after surgical excision, the patient remained asymptomatic, with no evidence of recurrence.

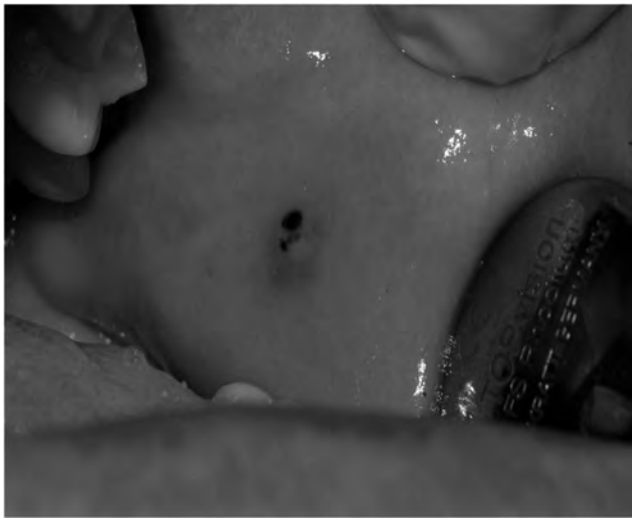

Figure 1. Clinical appearance of the buccal mucosa lesion

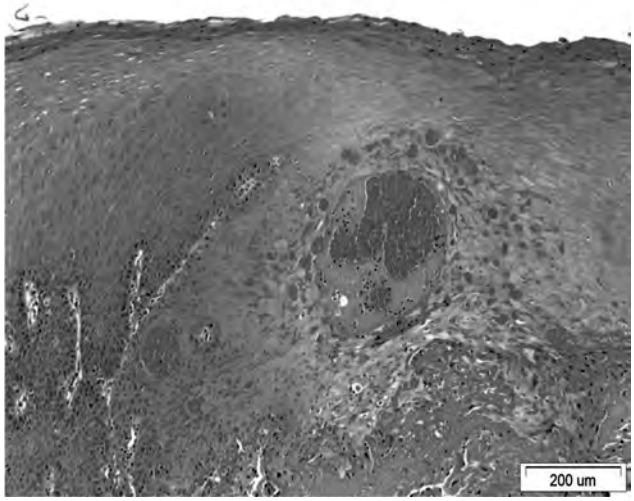

Figure 2. Dilated vascular spaces covered by normal endothelium without atypia, which extend into the overlying epithelium. The presence of small thrombi was observed in the vascular spaces (H&E x 200).

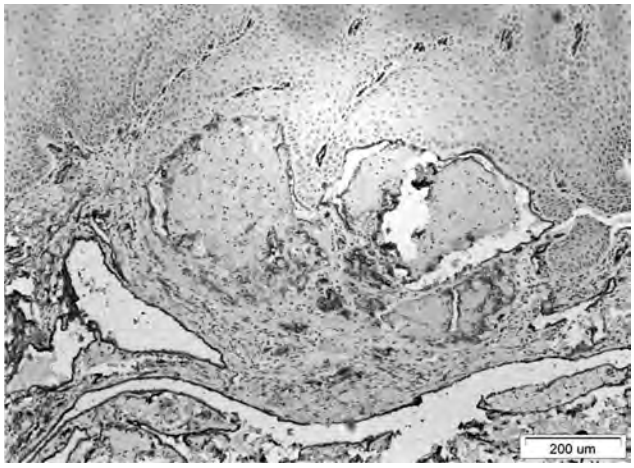

Figure 3. Endothelium of the vascular formations expressed positive immunostaining for CD34 (Original magnification x 200)

Discussion

The current taxonomy of AKs includes the following types: (1) the generalized systemic type, or diffused corporal angiokeratoma of Fabry; (2) the bilateral form occurring in the dorsal of fingers and toes angiokeratoma of Mibelli; (3) the localized scrotal form angiokeratoma of Fordyce; (4) the usually solitary papular angiokeratoma; and (5) the multiple papular and plaque-like circumscribed angiokeratoma[6].

AKs have been rarely described in the oral cavity, mainly at the tongue, sometimes in the context of diffused corporal angiokeratoma (Fabry's disease) or fucosidosis, where in mucosal involvement occur as part of a more generalized systemic disorder[2], and the presence of swollen and vacuolated (lipid-containing) endothelial cells, in addition to typical AK histopathology, is characteristically seen. Also, OAK is associated to AK of the scrotum, in the Fordyce type, or related to AKs of scrotum plus jejunum[10].

Clinically, OAKs present as single/multiple, erythematous, shiny papules, which may be studded with keratotic tops. They are firm on palpation, non-tendered, show telangiectatic vessels on diascopy, and may bleed, occasionally[1]. They may be misdiagnosed as hematomas, hemangiomas, lymphangiomas, focal epithelial hyperplasia, or nevi. Due to the low frequency of such a form of presentation, the clinical diagnosis is not always straightforward and the differential diagnosis must be established, not only with other vascular lesions, but also with melanocytic lesions of the oral cavity[10].

Although the first case of AK was reported more than a century ago, isolated OAKs appear to be a rare and relatively new subset, described for the first time in 1997 by Leung and Jordan[2]. To date, only 10 case reports of OAK, mostly as solitary lesions, have appeared in the literature. However, it has been hypothesized that the lesion is probably more frequent than reported, due to misdiagnosis[6].

The tongue, with a predilection for a dorsal surface, appears to be the most common site of solitary OAK in the oral cavity. In most patients, the lesions develop in early childhood and, as in other types of AK, females are affected more frequently[6-11]. Interestingly, multiple OAKs have been described exclusively in males[4,5].

Despite similar microscopic features in all types of AK with other vascular malformations, biopsy and diagnosis based on histology is necessary for the exclusion of melanoma or other melanocytic lesions[1]. The microscopic features of hyperkeratosis, acanthosis, and papillomatosis of the epithelium that encloses partially or completely, dilated vascular spaces covered by normal-appearing endothelium and containing erythrocytes and thrombi from the papillary part of the corium are characteristic for the diagnosis of OAK[7]. The immunohistochemical staining with CD 34 may be useful in order to distinguish the presence of vascular proliferation into the epithelium. OAKs are histologically similar to their cutaneous counterparts; the only difference described was the additional presence of parakeratosis[5]. Another main concern is the differential diagnosis of AK from circumscribed lymphangioma, which is a true tumor of the lymphatic capillaries[11].

Pathogenesis of AK is still uncertain. It is thought that the primary event is vascular ectasia within the papillary dermis just beneath the basement membrane. The epidermal pathological changes seem to be a secondary reaction. It has been speculated that the increased proliferative capacity on the surface of vascular malformations and the close proximity of the vascular spaces to the epidermis in AK could explain the reactive epidermal growth[7].

A recent classification has been proposed to denote solitary from multiple lesions, respectively, and this precise distinction between solitary and multiple lesions may help in management planning[1]. The treatment of OAK is the complete surgical excision, not only for microscopic diagnosis but also to avoid bleeding[6,7,10]. Alternatively, solitary AK may be treated by diathermy or cryotherapy, whereas multiple lesions will require laser excision[9]. The excisional biopsy with detailed histologic examination is important to confirm the diagnosis, as demonstrated in our case.

Conclusion

Despite its rare occurrence, solitary angiokeratoma of oral mucosa should be included in the differential diagnosis of black-bluish lesions like hematomas, hemangiomas and of melanocytic lesions including melanoma or nevi. Also, multiple AKs in adulthood may be related to Fabry syndrome. Nevertheless, further investigation for other similar lesions throughout skin or mucosa is needed after the diagnosis of AK, to avoid complications such as haemorrhage.

References

1. *Ranjan N, Mahajan VK*. Oral angiokeratomas: proposed clinical classification. *Int J Dermatol*, 2009; 48:778-781.
2. *Leung CS, Jordan RC*. Solitary angiokeratoma of the oral cavity. *Oral Surg Oral Pathol Oral Radiol Endod*, 1997; 84:51-53.
3. *Kumar MV, Thappa DM, Shanmugam S, Ratnakar C*. Angiokeratoma circumscriptum of the oral cavity. *Acta DermVenereol*, 1998; 78:472.
4. *Bhargava P, Bhargava S, Mathur D, Agarwal US, Bhargava R*. Angiokeratoma of tongue. *Indian J Dermatol Venereol Leprol*, 2001; 67:270.
5. *Vijaikumar M, Thappa DM, Karthikeyan K, Jayanthi S*. Angiokeratoma circumscriptum of the tongue. *Pediatr Dermatol*, 2003; 20:180-182.
6. *Varshney S*. Angiokeratoma circumscriptum of the tongue. *Int J Dermatol*. 2005; 44:886-888.

7. *Siponen M, Penna T, Apala-Sarkkinen M, Palatsi R, Salo T*. Solitary angiokeratoma of the tongue. *J Oral Pathol Med*, 2006; 35:252-253.
8. *Yildirim M, Kilinc N, Oktay MF, Topcu I*. A case of solitary angiokeratoma circumscriptum of the tongue. *Kulak Burun Bogaz Ihtis Derg*, 2007; 17:333-335.
9. *Ergun S, Mete O, Yesil S, Tanyeri H*. Solitary angiokeratoma of the tongue treated with diode laser. *Lasers Med Sci*, 2009; 24:123-125.
10. *Sion-Vardy N, Manor E, Puterman M, Bodner L*. Solitary angiokeratoma of the tongue. *Med Oral Patol Oral Cir Bucal*, 2008; 13:E12-14.
11. *Fernandez-Acenero MJ, Rey Biel J, Renedo G*. Solitary angiokeratoma of the tongue in adults. *Rom J Morphol Embryol*, 2010; 51:771-773.

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