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CASE REPORT

Solitary angiokeratoma of the tongue

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Angiokeratoma is a rare, cutaneous vascular disorder that can occur in several clinically distinct conditions. It usually presents as multiple, red to blue or black, asymptomatic papules on the skin. Oral mucosal involvement is common in the systemic form, but very rare in the localized forms of angiokeratomas. We report the second case of a solitary papular angiokeratoma of the oral cavity.

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Case report

The patient was a 54-year-old woman with asthma and cardiac arrhythmia. For a few months, she had had a small papule on the ventral surface of the tongue which bled occasionally. Her physician had punctured the lesion with no discharge and the lesion persisted. She was then referred to the Oulu University Hospital for excision of the lesion.

On clinical examination in the Otorhinolaryngology Clinic, a tumor of approximately 3–4 mm in diameter was observed on the right anterior ventral surface of the tongue. The lesion was purple in color and had a slightly granulomatous appearance. There were no other changes in the oral mucosa or in the ears, nose or throat.

The lesion was excised and sent for histopathological examination. On microscopy, the specimen was lined by an acanthotic stratified squamous epithelium, which was non-keratinized in the sides and hyperparakeratinized in the middle part. Immediately beneath the hyperparakeratinized epithelium, a group of dilated cystic spaces lined by endothelial cells were seen (Fig. 1). The cystic spaces contained erythrocytes. The epithelial rete pegs were elongated on both sides of the vascular spaces forming a cup-shaped appearance. In the surrounding connective tissue, a chronic inflammatory infiltrate was observed. These histopathological features are consistent with angiokeratoma. With regard to the rarity of a solitary angiokeratoma in the oral mucosa, the patient was also examined by a dermatologist, who found no angiokeratomas anywhere on the skin.

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Comments

Angiokeratomas can be divided into localized and systemic forms. The localized forms include: (i) usually solitary papular angiokeratoma that typically occurs on the legs, (ii) localized angiokeratoma of the scrotum and vulva (Fordyce type), (iii) congenital form, angiokeratoma circumscriptum naeviforme that presents as multiple hyperkeratotic papular and plaque-like lesions, usually unilaterally on the lower leg, foot, thigh, buttock or occasionally elsewhere, and (iv) bilateral angiokeratomas that occur on the dorsa of the fingers and toes (Mibelli type). The generalized systemic form, angiokeratoma corporis diffusum, is usually associated with a metabolic disorder, the most common being Fabry's disease and fucosidosis. The pathogenesis and clinical presentation is variable, but the histological features are similar in all the forms (1, 2).

Histologically, angiokeratoma is characterized by hyperkeratosis, acanthosis and papillomatosis of the epidermis and dilated vascular spaces with or without organizing thrombi in the papillary dermis. The vascular spaces are partly or completely enclosed by elongated rete ridges, creating a basket-like or cup-shaped appearance. In Fabry's disease and fucosidosis, swollen and vacuolated (lipid-containing) endothelial cells are a characteristic finding (1–3).

The pathogenesis of angiokeratoma is still uncertain. 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 location of the vascular spaces with the epidermis in angiokeratoma, could explain the

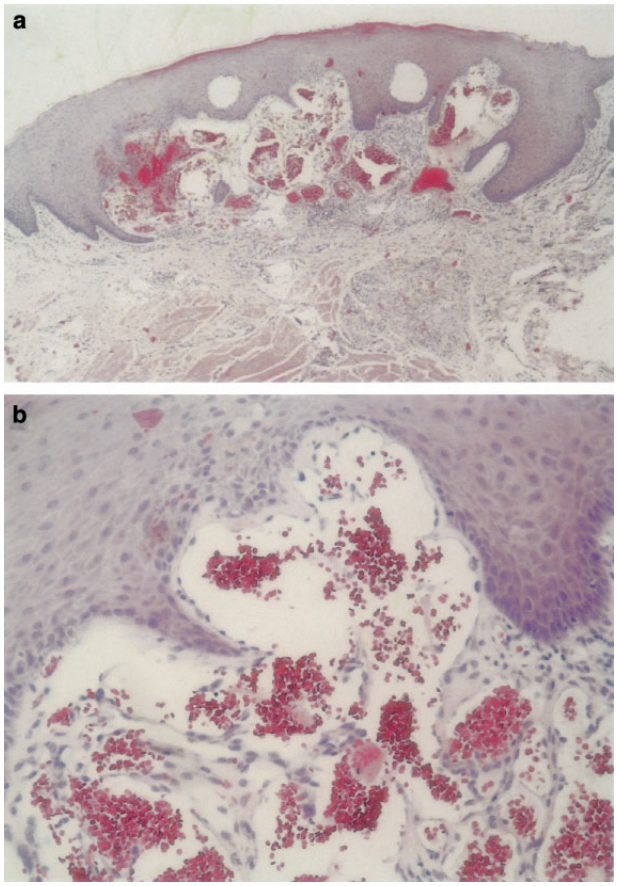

Figure 1 (a) Hyperparakeratosis and acanthosis of the epithelium with dilated vascular spaces in the subepithelial connective tissue. (b) The vascular spaces containing erythrocytes (hematoxylin and eosin stain: a, ×4; b, ×20).

reactive epidermal growth (1). An acute or chronic trauma, chilblains in the Mibelli type, high venous pressure or nevoid or vascular malformation are thought to be the causative factors in localized forms of angiokeratoma. The metabolic disturbance leading to weakness of the capillary wall and secondary ectasia is considered to be the primary cause for the development of angiokeratoma corporis diffusum (1).

Oral angiokeratomas are rare and are usually seen as part of a systemic condition such as Fabry's disease (4). To our knowledge, only four cases of a localized form of angiokeratoma occurring solely in the oral cavity have been reported previously: three cases compatible with angiokeratoma circumscriptum of tongue in 5-, 12- and 16-year-old boys (5–7), and one solitary angiokeratoma of the buccal mucosa in an 82-year-old male (3). In addition, two cases of angiokeratoma of the scrotum (Fordyce type) associated with angiokeratoma of the tongue (8, 9) as well as three cases of angiokeratomas occurring simultaneously in the jejunum, scrotum and oral cavity (10) have been published.

Angiokeratomas can be mistaken clinically for melanocytic nevus, melanoma, verruca vulgaris, hemangioma, capillary aneurysm or Spitz nevus (1). It is therefore important to confirm the diagnosis by histopathological examination. Treatment is generally not necessary, but may be required because of episodes of bleeding, discomfort or cosmetic reasons. It is also important to search for a causative factor although it is not possible to find one in all cases.

References

1. Schiller PI, Itin PH. Angiokeratomas: an update. *Dermatology* 1996; **193**: 275–82.
2. Braun-Falco O, Plewig G, Wolff HH, Burgdorf WHC. *Dermatology*, 2nd edn. Berlin, Heidelberg: Springer-Verlag, 2000; 1573–4.
3. Leung CS, Jordan RCK. Solitary angiokeratoma of the oral cavity. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1997; **84**: 51–3.
4. Baccaglini L, Schiffman R, Brennan MT, Lancaster HE Jr, Kulkarni AB, Brahim JS. Oral and craniofacial findings in Fabry's disease: a report of 13 patients. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2001; **92**: 415–9.
5. Kumar MV, Thappa DM, Shanmugam S, Ratnakar C. Angiokeratoma circumscriptum of the oral cavity. *Acta Derm Venereol* 1998; **78**: 472.
6. Vijaikumar M, Thappa DM, Karthikeyan K, Jayanthi S. Angiokeratoma circumscriptum of the tongue. *Pediatr Dermatol* 2003; **20**: 180–2.
7. Bhargava P, Bhargava S, Mathur D, Agarwal US, Bhargava R. Angiokeratoma of tongue. *Indian J Dermatol Venereol Leprol* 2001; **67**: 270.
8. Karthikeyan K, Sethuraman G, Thappa DM. Angiokeratoma of the oral cavity and scrotum. *J Dermatol* 2000; **27**: 131–2.
9. Jansen T, Bechara FG, Stucker M, Altmeyer P. Angiokeratoma of the scrotum (Fordyce type) associated with angiokeratoma of the oral cavity. *Acta Derm Venereol* 2002; **82**: 208–10.
10. Rappaport I, Shiffman MA. Multiple phlebectasia involving jejunum, oral cavity and scrotum. *JAMA* 1963; **185**: 437–44.