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Multiple papular angiokeratoma of the tongue

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Abstract

The term Angiokeratoma is applied to a number of quite distinct conditions that share a clinical presentation with asymptomatic hyperkeratotic vascular skin lesions and a histological combination of superficial dermal vascular ectasias and hyperkeratosis. It can be either localised or systemic form.

We are reporting a case of multiple papular angiokeratoma of the tongue, which is a very rare form of presentation of a large solitary angikeratoma, more so of the oral variety.

Case report

A 7 yr old boy presented with bluish- pink-coloured, small-raised lesions over the dorsal part of his tongue, which had been present since birth, which bled occasionally. There was no history of trauma, cold, injury, or similar lesions elsewhere in her body.

On clinical examination, an oval lesion of approximately 3.5cm x 1.5 cm. bluish- pink lesion was observed on the left side on the dorsum of the tongue. The lesion was bluish-pink in colour with a nodular appearance. Surface was firm and did not bleed on palpation. There were no other changes in the oral mucosa. On dermatologic examination, no similar lesions were found anywhere on the skin. The lesion was excised in-toto, under general anesthesia and sent for HPE. The histopathological diagnosis was "angiokeratoma of tongue".

A case of a solitary angiokeratoma of the tongue is being reported. We report the first ever case in the tongue, with a large, multiple papular angiokeratoma.

Key words: *multiple papular angiokeratoma, oral cavity, tongue.*

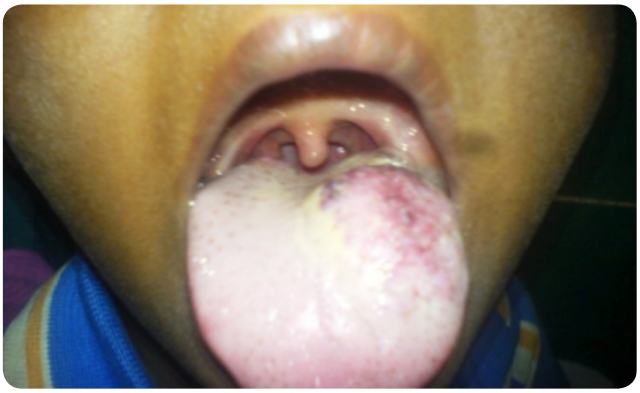

FIGURE 1. Nodular mass over the dorsum of tongue.

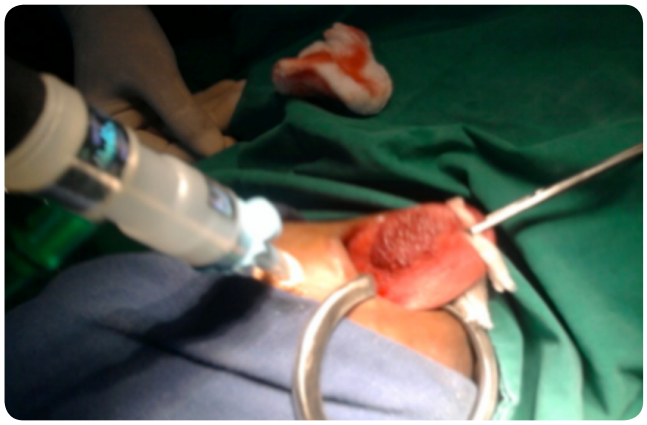

FIGURE 2. Multiple papular appearance of the mass.

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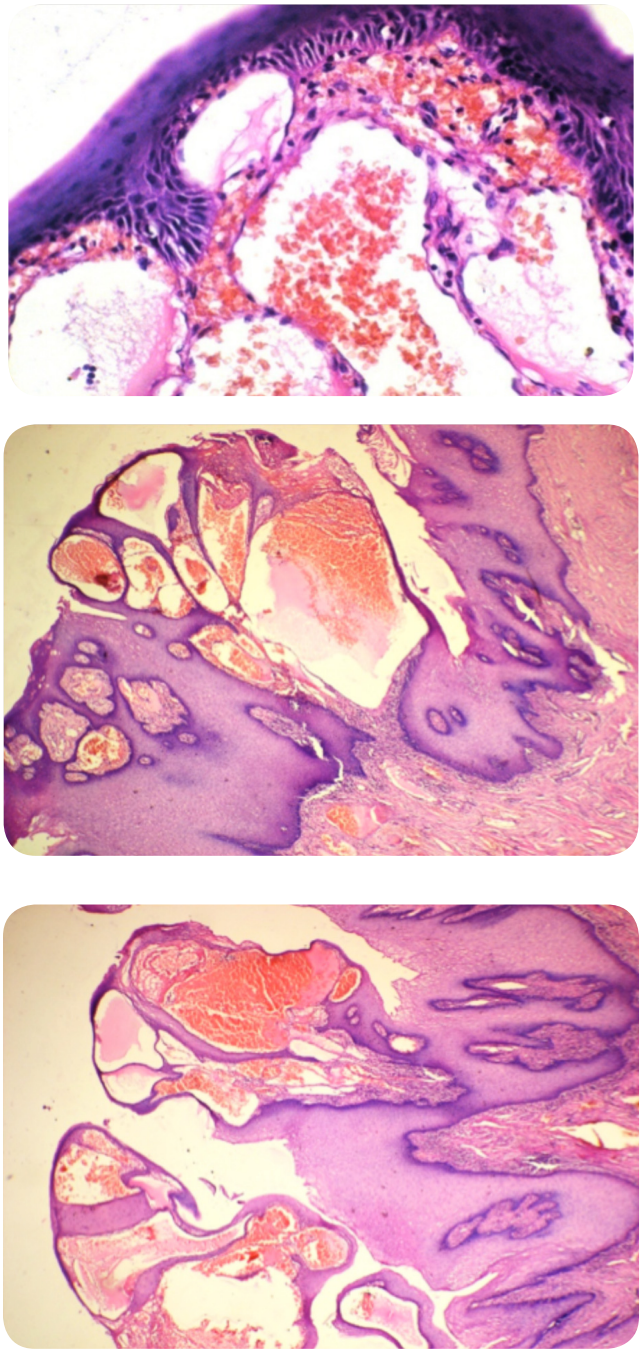

FIGURE 3, 4 & 5. Hyperkeratosis, acanthosis & papillomatosis of stratified squamous epithelium. Subepithelieal tissue & papillae show dilated vascular spaces enclosed within the papillae and filled with blood.

Discussion

Angiokeratomas are a group of telangiectasias with prominent epidermal response in the form of hyperkeratosis. The epidermal changes in all forms of angiokeratoma are secondary. The different entities causing the vessel ectasia lead to many clinical variants of angiokeratoma. Current classification distinguishes between a widespread form (angiokeratoma corporis diffusum), which is usually associated with an inborn error of metabolism, and localised forms, which include, solitary angiokeratoma, Fordyce's angiokeratoma, angiokeratoma circumscriptum naeviformae and angiokeratoma of Mibelli.

Clinicalvariants:

1. Mibelli type: The "Mibelli-type" occurs on the acral sites, mainly digits, of young people affected by repeated attacks of chilblains and acryocyanosis, which result in a deleterious effects on vessel walls. There is a definite family predisposition with an autosomal dominant trait and girls affected more.
   Clinical features -- presents between 10-15 yrs, minute red macules, slowly increase in size and become elevated and warty. Site- dorsal and lateral aspect of finger and toes, dorsum of hands and feet, knees and elbows.
2. Fordyce type: The "Fordyce-type" occurs on the scrotal skin of young and adults as a secondary effect to an increased blood pressure in scrotal veins. An equivalent form affecting adult females and occurring analogously on the skin of the vulva has been recorded.
3. *Solitary and multiple types;* The "solitary and multiple papular types" of young individuals affect the lower extremities and is considered a consequence of a congenital deficiency of elastic tissue in regional veins.
4. Angiokeratoma circumscriptum: This is a nevoid hamartomatous lesion arising early in life during infancy or childhood, sometimes in association with other congenital malformation of other sites.
5. Angiokeratoma corporis diffusum: It is a clinical variant of angiokeratoma that is typically associated with an enzyme deficiency in the metabolism of glycoprotein, most notably Fabry disease, resulting in many other systemic manifestations.

Oral angiokeratoma is very rare. It is most commonly found as a component of the generalized systemic disorder in Fabry's disease or fucosidosis, where multiple angiokeratomas can be found on the skin and the oral mucosa. Oral angiokeratoma affecting the tongue can also be in the congenital form, angiokeratoma circumscriptum, in the Fordyce type, where angiokeratoma of the scrotum is associated with angiokeratoma of the tongue or angiokeratomas occurring simultaneously in the jejunum, scrotum and oral cavity.

In the present case the lesion was solitary and there were no other lesions on the skin or in the oral cavity. Previously,there have been very few reports in the literature, of solitary angiokeratoms of the tongue, especially without metabolic disease. There are no reports of angiokeratoma,with multiple papular lesions and such a big size too. Oral angiokeratoma has histologic features similar to those arising on the skin. In both sites acanthosis and papillomatosis of the squamous epithelium can be seen). However, while the cutaneous lesions show hyperorthokeratosis, the oral lesions show more hyperparakeratosis.

Most of the vascular spaces were surrounded by elongated rete ridges with thrombi noted within the dilated vascular spaces. These features were also present in the oral lesions.

Angiokeratoma is a dark, irregular lesion which bleeds occasionally. Therfore it can be mistaken clinically for melanocytic nevus, malignant melanoma, verruca vulgaris, hemangioma, capillary aneurysm, Spitz nevus or focal epithelial hyperplasia. The excisional biopsy with meticulous histologic examination is important to confirm the diagnosis, as demonstrated in our case.

The pathogenesis of angiokeratoma 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 angiokeratoma could explain the reactive epidermal growth.

Treatment of oral angiokeratoma is surgical excision. It is required for histopathologic diagnosis and to cease episodes of bleeding or discomfort and to search for possible causative factors.

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