

## Solitary angiokeratoma of the tongue.

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### Summary

Angiokeratoma refers to a rare group of lesions that have a characteristic dermal vascular dilatation, often associated with a number of metabolic disorders. Different forms of angiokeratoma have been described, which share the same histologic features but differ clinically. We report a case of an oral angiokeratoma in a 6 year old boy. This is to our knowledge the fifth case of oral angiokeratoma and the third pediatric case ever described.

### Key words

Angiokeratoma, tongue.

**A**ngiokeratoma refers to a group of cutaneous vascular lesions that may be associated with certain metabolic disorders. On histological examination, angiokeratoma is defined by dilated vessels in the dermis, only seemingly included within acanthotic and hyperkeratotic epidermis. These vascular spaces are surrounded by rete ridges and can have thrombi within them. There may be ulceration of the surface. Clinically, the lesion is irregular, dark brown in color, and easily bleeds (9). Angiokeratoma of the oral mucosa is rare, and it is more commonly associated with Fabry's disease, a condition where multiple angiokeratomas are found in the skin and oral mucosa (8). This lesion also can be secondary to trauma, resulting in epithelial proliferation and hyperkeratosis (9).

In this report, our patient had no signs of systemic disease and no local predisposing trauma for the development of the angiokeratoma was reported.

### Case Report

We report a case of a six-year-old boy with no significant past medical history seen in the dermatology clinic presenting with a recurrent lesion on the left side of the tongue. The mass appeared 3 years prior. The biopsy, done in another institution, showed histologic features of angiokeratoma. The patient was treated with laser surgery (pulsed dye laser) on four different occasions, but due to the location of the tumor, it could not be completely removed. Three months after the last surgical procedure, the tumor started to regrow and bled into the oral cavity. He was then admitted to the hospital for a complete excision of the lesion. At the time of surgery, the lesion was noted to be a large, anterior-lateral tongue tumor beginning anterior to the papilla of the tongue base. The lesion extended over the oral tongue but did not cross the midline. Once again, due to technical difficulties, the tumor could not be completely removed. The specimen

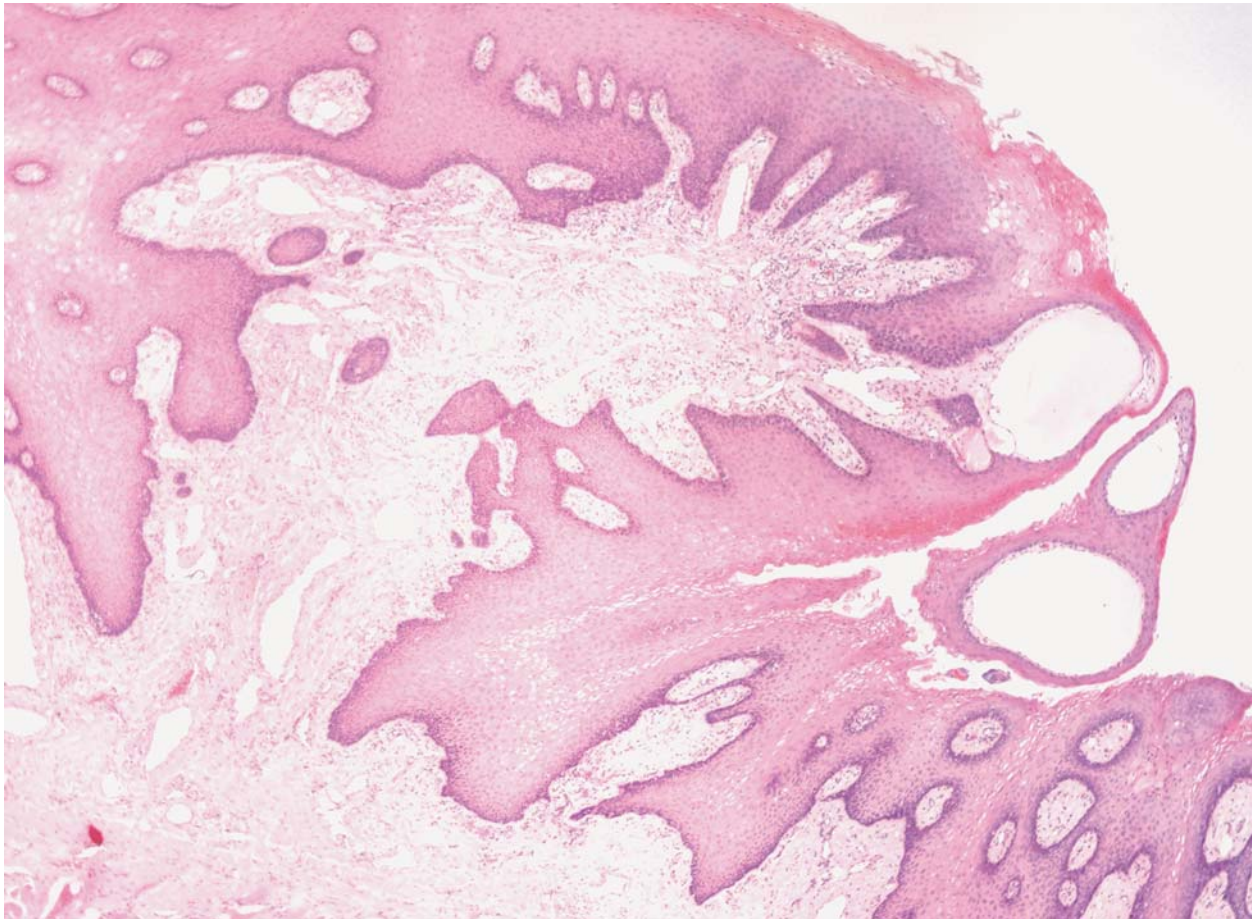


Fig. 1: Photomicrograph showing acanthotic epidermis and cystic spaces containing red blood cells (hematoxylin and eosin stain, 400x).

received measured 4.0 x 3.0 x 1.0 cm in overall dimension. The pale tan papillae of the tongue were visible grossly, as well as the thick muscular layer underneath. Microscopic sections demonstrated numerous dilated spaces in the squamous mucosa and submucosa filled with either blood or lymphatic fluid. Due to the location of the lesion, there was no associated hyperkeratosis but there was pseudoepitheliomatous hyperplasia and parakeratosis (Fig. 1).

### Discussion

Solitary angiokeratoma was first described in 1967 (4). These lesions are commonly found on the hips, thighs, buttocks, umbilicus, lower

abdomen, scrotum, glans penis, and rarely oral mucosa.

There are five categories of angiokeratoma as follows:

- 1) angiokeratoma corpora diffusum of Fabry (12, 14), which is the generalized systemic type and is secondary to metabolic diseases;
- 2) angiokeratoma of Mibelli, found on the dorsum of the fingers and toes, bilaterally. It is an autosomal dominant disease;
- 3) angiokeratoma of Fordyce, which is the localized scrotal form;
- 4) angiokeratoma circumscriptum, consisting of multiple papular lesions. It is common in female patients and during childhood;
- 5) solitary papular angiokeratoma, which has been associated to trauma.

**Table 1: Case reports of oral angiokeratoma.**

<i>Author</i>	<i>Year</i>	<i>Sex</i>	<i>Age (yrs)</i>	<i>Location</i>
Leung (11)	1997	Male	82	Oral cavity
Kumar (12)	1998	Male	16	Oral cavity
Karthikeyan (13)	2000	Male	30	Oral cavity
Vijaikumar (14)	2003	Male	12	Tongue
Farooq	2005	Male	6	Tongue

Although solitary cutaneous forms have already been reported in the literature (5, 10) localized lesions in the oral cavity are uncommon and only four previous cases were reported (Table 1). Oral mucosal involvement is a component of angiokeratoma corporis diffusum, however in this case no other associated lesions were identified. The pathogenesis of localized lesions is unknown and has been linked to arterovenous fistula (2) and to lymphangioma circumscriptum after repeated local injuries (7).

Treatment options usually include complete surgical excision, cryotherapy and laser ablation.

As far as laser treatment, numerous publications underlined the efficiency of this technique in the treatment of cutaneous angiokeratomas (1, 3, 11).

In this case, the lack of clinical improvement implies that laser may not be the ideal treatment in case of oral angiokeratoma, in which case a primary complete surgical excision seems to be a more appropriate treatment choice.

In conclusion, to our knowledge, we are reporting the fifth case of an isolated oral angiokeratoma and the third case in a pediatric population.

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