Angiokeratoma of the Tongue: A Case Report and Review Literature

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Abstract

Angiokeratomas are vascular abnormalities that usually appear as multiple or solitary cutaneous plaques. Several clinical variants have been designated, with the same principal histopathological lesion. Mucosal affections, including the oral cavity, are infrequently found either as a component of the systemic variety, called angiokeratoma corporis diffusum, or are associated with cutaneous lesions in more locations. Isolated oral involvement seems to be rather infrequent. Here, we report a case of this rare entity affecting a 6-year-old boy in the dorsum of the tongue.

Keywords: Angiokeratoma, corporis diffusum, solitary cutaneous plaques, vascular abnormalities

Introduction

Angiokeratoma refers to a set of cutaneous vascular lesions that may be associated with some metabolic diseases. On histopathological examination, angiokeratoma is well-defined by dilated vessels in the dermis, only apparently included within acanthotic and hyperkeratotic epidermis. These vascular spaces are surrounded by rete ridges and can have thrombi inside them. Ulcerative lesions may be found on its surface. Clinically, the lesion is irregular, dark brown in color, and easily bleeds.[1]

Several clinical forms have been defined depending on the multiplicity and locality of the lesions.[2] The localized forms can be further classified in (a) solitary papular angiokeratomas; (b) scrotal or vulval angiokeratomas; (c) multiple congenital angiokeratomas; and (d) bilateral angiokeratomas in the dorsum of hands and feet (Mibelli type).

Angiokeratoma of the oral mucosa is rare and is usually associated with Fabry's disease, a condition where multiple angiokeratomas are found in the skin and oral mucosa.[3] This lesion also can be secondary to trauma, resulting in epithelial proliferation and hyperkeratosis[1]

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Case Report

A 6-year-old boy [Figure 1] presented with multiple violaceous raised lesions over the dorsal aspect of the anterior two-thirds of tongue present for a duration of 2 years. It started as a single raised lesion on the tip of the tongue which gradually progress in size and coalesced to form a plaque involved the whole anterior two-thirds of the dorsal surface of the tongue. Some of the papules were also present over the lateral surface of the tongue. The mother stated that strangely he ingest strongly spicy foods, no other type of trauma was reported. There was a history of occasional spitting of blood which occurred spontaneously, as well as on trauma. There were no any similar lesions elsewhere on the body.

On examination, there were multiple, grouped, violaceous shiny papular lesions, some of which were pink, bluish, and purple in color, present over the dorsal aspect of the anterior two-thirds of the tongue as well as the lateral surface. They were mobile, spongy, and did not bleed on

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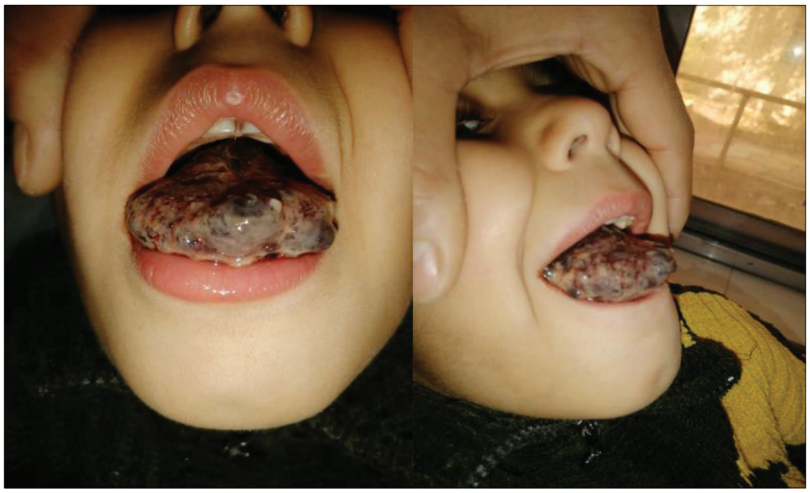

Figure 1: Angiokeratoma of the tongue

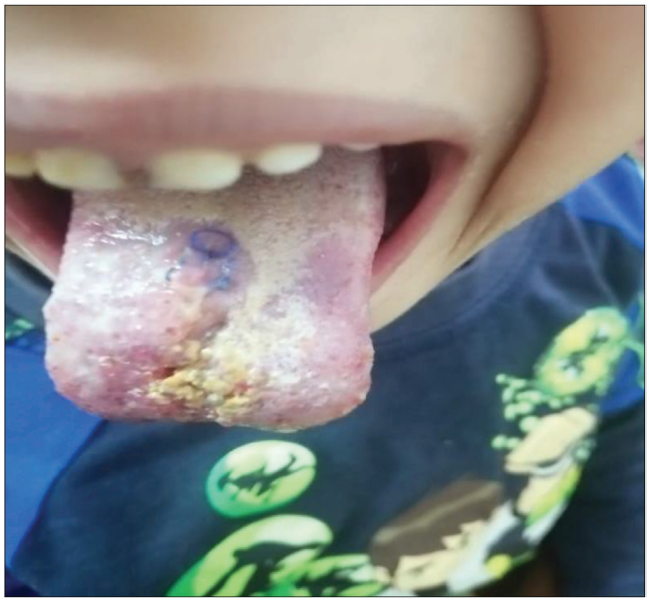

Figure 2: Angiokeratoma after incisional biopsy

touching. On examination, no other significant cutaneous or systemic abnormality was detected. All hematological and biochemical investigations were normal.

After full preoperative preparation, incisional biopsy was taken under general anesthesia with no significant peroperative bleeding. A biopsy specimen of a representative tongue lesion showed vascular lesion which has dilated thin wall blood vessels enclosed within hyperplastic squamous epithelial mucosa. On the basis of clinical examination and histopathological findings, a diagnosis of angiokeratoma of the tongue was made. Fortunately, after taking the biopsy, the lesion was improved [Figure 2]

Discussion

Mibelli[4] reported the first case of Angiokeratoma more than a century ago in 1889. Solitary angiokeratoma was also described in 1967.[5] These lesions are commonly found on the hips, thighs, buttocks, umbilicus, lower abdomen, scrotum, glans penis, and rarely oral mucosa.

The clinical variants of the disease are as follows:[6]

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 effect on vessel walls. Clinical features present between 10 and 15 years, minute red macules, slowly increase in size and become elevated and warty.
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 are 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 angiokeratoma corporis diffusum and is very uncommon in other types of angiokeratoma.

To the best of our knowledge, this case is the first reported case of angiokeratoma on the tongue without metabolic disease in Iraq. Angiokeratoma is a dark irregular lesion that bleeds occasionally. Therefore, it can be mistaken clinically for melanocytic nevus, malignant melanoma, verruca vulgaris, hemangioma, capillary aneurysms,

spitz nevus, or focal epithelial hyperplasia.[7,8] Biopsy with histologic examination is important to confirm the diagnosis. The pathogenesis of angiokeratoma is still uncertain. Vascular ectasia within the papillary dermis appears to be a primary event, whereas epidermal changes are a secondary reaction. Increased proliferative capacity on surface of vascular malformations and close proximity of vascular spaces to epidermis in angiokeratoma could explain the reactive epidermal growth.[7]

Treatment of oral angiokeratoma is surgical excision. Prognosis of Angiokeratoma is good, although recurrences have been noted in few cases.[9]

The pathogenesis of angiokeratoma is ambiguous. Localized angiokeratoma may be seen in a setting of acute or chronic trauma, chilblains, high venous pressure, or nevoid or vascular malformation. Several theories have been proposed in literature which includes the occurrence of lesions secondary to a local trauma, subcutaneous hematomas, tissue hypoxia,[10] as a result of high venous pressure or as a part of vascular malformation. The primary event is vascular ectasia within the papillary dermis. Epidermal changes seem to be a secondary reaction. It has been speculated that the increased cell proliferation on the surface of vascular malformations and the close location of the vascular spaces with the epidermis in angiokeratoma could explain the reactive epidermal growth. In hyperkeratotic forms of angiokeratoma circumscriptum, metalloproteinase may be seen just beneath the stratum corneum. Oral involvement may be seen in a setting of angiokeratoma corporis diffusum. Anderson-Fabry disease is an X-linked lysosomal disorder marked by a deficiency of α-galactosidase deficiency. Apart from oral involvement, it may be associated with cutaneous angiokeratomas distributed symmetrically in a bathing suit pattern. Fucosidosis is other lipid storage disorders in which oral angiokeratomas are seen.[11] The treatment options for angiokeratoma depend on the site and size of the lesions. Simple excision and closure is usually considered in case of few lesions. Cryotherapy, electrocautery, and radio-frequency are the modalities currently preferred for multiple lesions.[12] Kar and Gupta[13] described the CO_2 laser and pulse dye laser in angiokeratoma of tongue and found 75% improvement with it.

Conclusion

Angiokeratomas are vascular conditions characterized by vascular ectatic changes and overlying epidermal hyperkeratosis. Tongue angiokeratomas are uncommon and few cases worldwide were reported till now. We have presented a case of angiokeratoma of tongue without any systemic disorders occurring in an otherwise healthy 6-year-old child.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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