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ANGIOKERTOMA OF TONGUE: A RARE ENTITY

Oral Pathology

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ABSTRACT

Angiokeratoma is non neoplastic ectasia of superficial dermal vessels.[1] It is characterized by large dilated blood vessels in the superficial dermis and hyperkeratosis mainly involving lower extremities and trunk.[2] It is usually associated with systemic disease. However, we are reporting this case of solitary angiokeratoma of the tongue which is very rare, only seven cases are reported till date.

KEYWORDS

Angiokeratoma, oral mucosa, tongue

INTRODUCTION:

Angiokeratoma is benign cutaneous vascular malformation mostly occurring in lower extermities and trunk.[2,3] Mibelli in1989 first reported the case of angiokeratoma in the fingers and toes.[4,5] The etiology of this is unknown, but may be congenital and acquired. They may present as multiple or single, itchy and painful swelling. These lesions are present in 70% of males and 30% of females.

Oral mucosal involvement is seen as part of angiokeratoma corporis diffusum.[6] It is usually associated with the syndromes such as Klippel-Trenaunay-Weber syndrome, Cobb syndrome, and other mixed vascular malformations.[5] Angiokeratoma though appears in the oral cavity, mostly associated with systemic disease and represents as multiple papules in buccal mucosa and tongue, but in our case, the patient is seven years old with a solitary lesion on the tongue and had no systemic disease.

CASE REPORT:

A seven year old girl presented in department of ENT with chief complaints of swelling on dorsum of tongue which is gradually increase in size since 4 years. It is not associated with pain, discharge and bleeding. There was no previous history of local trauma. There was no history of similar lesions elsewhere on the body. On intraoral examination a single dark-red to blue–black, 1x1 cm swelling with well defined margins, which did not blanch on pressure, was seen on dorsal surface of the tongue with no plaque formation. The lesion was non-tender and did not bleed on manipulation. The rest of the cutaneous and systemic examination was normal. We received grey white soft tissue bit measuring 0.8x07x04 cm. Microscopically there was marked ectasia of papillary dermal vessels surrounded by overlying epithelium which showed acanthosis, hyperkeratosis and papillomatosis. Rete ridges were elongated completely enclosing the vascular channels, collarette seen at the edge of lesion. These vessels are lined by flattened epithelium without atypia. On the basis of histopathological picture a diagnosis of angiokeratoma of tongue was made, as shown in figure 1, 2 and 3.

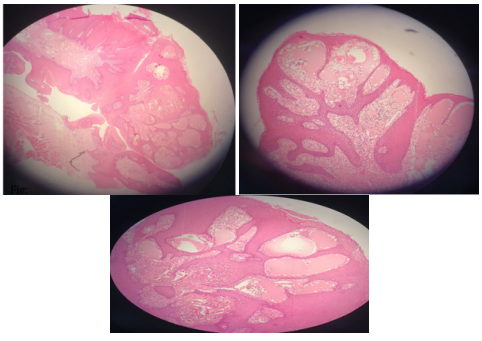

Figure 1, 2 And 3

DISCUSSION:

Angiokeratoma is non neoplastic ectasia of superficial dermal vessels. It has five variants but all variants have same histopatholgical features. They may be solitary and multiple. 1. Angiokeratoma corporis diffusum: In this lesions are usually seen from umbilicus to genitalia. Age of onset is 20-30 years and males more severly affected. They are associated with Anderson-Fabry disease in which multiple angiokeratomas appear in late childhood. Rarely, they are also seen in X linked recessive chromosomal lipid disorder due to deficiency of lysosomal alpha galactosidase. [1,6] 2.Angiokeratoma of Mibelli: It is rare genodermatosis with an autosomal dominant trait. Lesion are found over bony pominenace on dorsa of fingers, toes, knees and elbows bilaterally. More often seen in children and adolescents with a predilection for females.[4] 3.Angiokeratoma of Fordyce: Lesions may be single or multiple vascular papules present in scrotum, vulva, penis, upper thigh and lower abdomen. Mostly seen in middle aged and elderly patients.[1] 4.Angiokeratoma circumscriptum: It is least common variant, present as hyperkeratotic, papular nodule that coalesce to form plaque, always unilateral on arm, trunk or legs.[1] 5.Solitary and multiple angiokeratomas: They are present clinically as warty papule predominantly on lower extremities of healthy person and may be associated with trauma.[1]

Isolated oral cavity involvement is very rare with only 18 cases reported in the world literature.[7] First case of solitary isolated oral cavity involvement was reported by Leung et al[8] in 1997 on buccal mucosa of an 82-year-old male. Isolated solitary as well as multiple angiokeratomas of tongue without plaque formation is very rare. The first case of isolated angiokeratoma over tongue was reported by Vijai Kumar et al.[9] Solitary angiokeratomas seems rather infrequent, with only seven cases reported till date. [2,8,10,11,12,13,14] Here, we are reporting a eighth case of solitary angiokeratoma in a 7-year-old girl.

CONCLUSION

Angiokeratoma of the tongue is a rare cutaneous vascular lesion mostly associated with the systemic disorder and syndromes. We are reporting the eighth solitary case of this rare variant of angiokeratoma of the tongue; out of which our patient is the youngest who did not show any other systemic disorders and cutaneous lesions, and being successfully treated.

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Conflicts of interest
There are no conflicts of interest.

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