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RECURRENCE OF SOLITARY ANGIOKERATOMA OF TONGUE

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ABSTRACT

Angiokeratoma is a rare vascular disorder, only first to be reported in 1997, mostly seen as one of the earliest clinical features of systemic muco-cutaneous disorders. The presence of oral angiokeratomas as solitary entity is very rare. Only 7 such presentations are reported in the English medical literature. We present a case of solitary angiokeratoma of the dorsum of the tongue in a 65 year old woman that showed recurrence 2 years after a surgical intervention.

INTRODUCTION

Angiokeratoma is a rare benign, capillary vascular disorder that may manifest as multiple raised plaques or papules. It may also present as one of the components of metabolic disorders. The term was derived from Greek Literature meaning blood vessels, horns and swellings respectively.[1] The rarity of occurrence of this pathology has made it difficult for medical specialities to hold much knowledge about the varied clinical features and the behavior of the lesion leading to the delay in diagnosis of this lesion. Solitary oral angiokeratomas are very seldom found. The first such reference of its existence was documented by Leung in 1997 and since then only 7 such cases exist in the current medical literature.[2] We present another such case of solitary angiokeratoma of dorsal tongue which had recurred after 2 years post incomplete excision. This paper also depicts that incomplete removal will result in recurrence of this benign lesion.

Case Report

A 65 year old lady reported to the clinic with an asymptomatic, pink, solitary growth on the dorsum of the tongue at the centre of the junction of anterior 2/3 and posterior 1/3 just anterior to the circumvallate papillae. The growth was elliptical in size, about 1.5 x 1cm in size, firm, non pulsatile, non compressible, non tender, and with sharp margins. There was absence of any induration on palpation of the swelling. It was also not easily bleeding on palpation. (Figure.1)

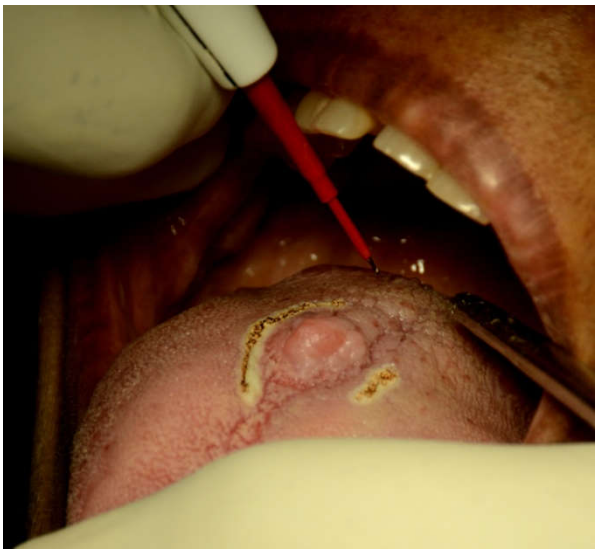

Figure 1 The lesion on the dorsum of tongue and wide local excision marking being performed with diathermy

Her past medical history, allergy history and personal history were otherwise unremarkable. The lady did not present with any history of trauma or infection to the concerned area also.

She had visited a dentist 2 years ago who took a biopsy from the same site. The excision was performed by him with clinical diagnosis of squamous cell carcinoma however the report of the biopsy was unrepresentative. At that time when the patient had reported to us and the site looked to be in healing phase.

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The case was pursued very carefully due to the original history delivered by the patient. The area of concern had remained asymptomatic for past 2 years when she had only noted a slow growth at the same region 6 months ago.

A wide local excision of the lesion with three dimensional clearance along with healthy tissue margins was marked and performed under local anesthesia and closure was performed with 3-0 vicryl. Post operative healing was uncomplicated. The patient has been under constant review for past 3 months and is without any recurrence.

Histopathologically there was presence of hyperkeratinization, acanthosis, and papillomatosis of the epidermis as well as presence of numerous dilated vessels in the dermis. (Figure.2) The clinicopathologic diagnosis was angiokeratoma of tongue and absence of any malignancy. The patient and her kin had been counseled upon the rarity of this lesion and its benign nature with lack of recurrence potential.

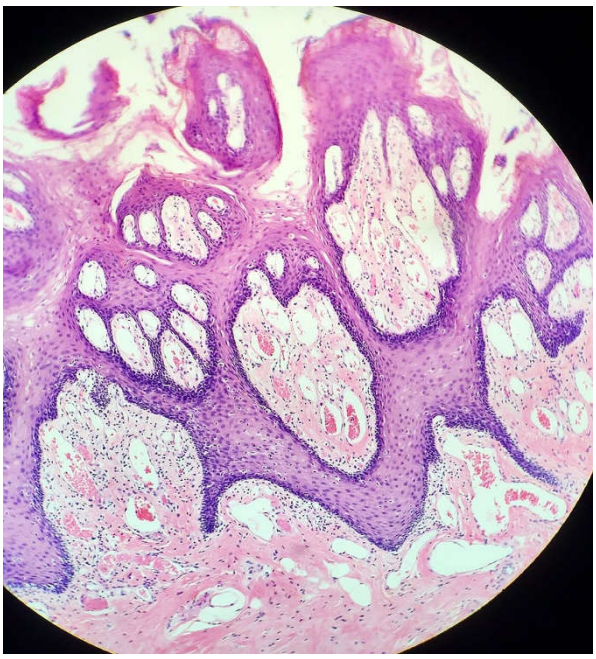

Figure 2 The histopathology showing keratinization, acanthosis and papillomatosis

DISCUSSION

The term angiokeratoma was first coined by Milbelli in 1889 in a 14 year old girl and in 1896 Fordyce reported the first case of atypical angiokeratoma of the scrotum. In 1898 Fabry and Anderson described similar cases on skin involvements and telangiectesias along with cardiac and renal involvements. Fabry first named the condition as purpura nodularis haemorhagica and later changed to Angiokeratoma Corporis Diffusum in 1915.[3] Leung was the first to have reported first solitary case of angiokeratoma in oral cavity in 1997.[2]

Angiokeratoma, rare muco-cutaneous disorder involving capillaries,[3] present as asymptomatic, hyperkeratotic, pink to purple, plaques or papules either solitary or multiple in number.[3,4] The lesion are generally classified into two broad categories depending on localized form or widespread form.[3,4] The localized form is further sub- divided as [3,4]

- Solitary, papular form formed on the legs
- A bilateral variety that involves the dorsa of fingers and toes : Mibelli type
- Solitary angiokeratoma of scrotum or vulva: Fordyce type
- Congenital angiokeratomas multiple in number, usually in the lower extrimities

The generalized form is Angiokeratoma Coporis Diffusum associated with metabolic disorders as Fabry's Disease, fucosidosis, amyloidosis, vasculitis.[3]

Fabry's disease is a rare X linked lysosomal disorder characterized by α galactosidase deficiency.[5] The manifestations range from gastrointestinal disturbances, pain in hands, feet and fever hypothesized to be due to capillary obstruction leading to nerve fibre damage and autonomic neuropathy with the earliest noticeable signs as blue to black papules or macules in the elbow, flanks, hands and knees. Enzyme Replacement Therapy with α- galactosidase A preparations and have shown to improve or reverse the progression of multi organ failure in Fabry's Disease.

Milbelli type generally occurs in young adults affected by chilblain and acrocyanosis that result in damage to the blood vessel walls of both hands and feet. It is presumed to have autosomal dominant trait with a female predilection.[6] They present as slow growing, raised, warty, red macules on the dorsum of knees, hands and feet bilaterally.

Fordyce type only occurs solitarily on scrotal skin of adults due to increased blood pressure in the scrotal veins or skin of vulva. [6]

Solitary and macular or popular variety, first described in 1967 by Imperial and Hertwig, occurs in young individuals on the lower extrimities. It has been thought to be due to congenital deficiency of elastic tissue in lower extremity veins.[6]

The first case of solitary angiokeratoma of tongue (ventral surface of tongue) was reported in 1998 by M.Vijay et al.[11] The second such case of was reported in 2009 by Ergun *et al* and treated with diode laser.[3] The first usage of immunohistochemistry in this lesion was by Maria J. Fernández *et al* in 2010 for a lesion at tip of tongue. It was proven that definitive diagnosis of angiokeratoma was if the lesion was positive for CD34 and CD31 but negative for LYVE-1, a lymphoma marker.[9] R.Sinha *et al* were the first to have present the same lesion in the posterior tongue in 2011.[12] Kar *et al* had reported 5th such case where the lesion involving ventral and lateral aspect of tongue, clinically mimicking lymphangioma only to be histopathologically diagnosed as angiokeratoma.[10] The last reported case of solitary angiokeratoma of tongue was on the right lateral border of an 11 year old in 2018.[13]

The etiopathology of the disease is still very unclear but has been hypothetised by Foucar and Nason as abnormal haemodynamics just below the soft tissue.[3] Sion- Vardi n *et al* further hypothetized the primary cause to be vascular event as ectasia within the papillary dermis just beneath the basement membrane.[8]

The dorsal surface of tongue reported as most common site for solitary angiokeratoma with female predilection whereas multiple oral angiokeratomas are most common in males.[7] Clinically, angiokeratoma may present as varied forms resembling melanocytic nevus, malignant melanoma, verruca vulgaris, capillary aneurysm. However it is mandatory to check the skin and mucous membranes to rule out systemic involvement.[5] The diagnosis of the lesion is of utmost

importance in the treatment of solitary angiokeratoma. Though articles have reported it having the clinical presentation of median rhomboid glossitits[3], ours presented at the same site but as a sessile growth pink in color with well defined margins and of size of 1.5 x 1cm. Other studies potray that clinical features are like that of circumscribed lymphangioma, hemangioma or lymphohemangioma.[7]

The treatment of angiokeratoma is generally wide excision of the lesion. Owing to benign nature of solitary oral angiokeratomas it is the only treatment modality and medical management have no role.[7] The 7 cases mentioning oral angiokeratoma of tongue have not reported of any further recurrences.[3,4] Our patient had undergone an incomplete excision before leading to its recurrence. This is the most distinguishable property of these benign lesions.

The other treatment options that have been reported are cryosurgery and diode laser excision.[3,7] Ergun *et al* have mentioned that use of diode lasers are beneficial to conventional surgery, or use of diathermy to control of intra operative bleeding. It is also superior to cryosurgery as it delivers less post operative pain.[3]

Histopathologic features of oral angiokeratoma are same as skin lesions with presence of acanthosis and papillomatosis of the squamous epithelium.[7, 9] The only difference is presence of hyperorthokeratinization in cutaneous lesions while oral lesions depict hyperparakeratosis.[10] There are presence of dilated vascular spaces surrounded by elongated rete-pegs of thrombi within the dilated spaces in the papillary dermis in both skin and oral lesions.

CONCLUSION

Solitary angiokeratoma of tongue is a rare entity in itself, only 7 known in English scientific literature. The diagnosis of this lesion is key to the treatment. This entity must be kept as the differential diagnosis of any purple, to bluish-black color, pappular or macular or any elevated lesion of the oral cavity. Moreover once such an entity is noted a thorough examination of entire oral mucosa and skin must be performed to rule out systemic involvement as well as tests for α galactosidase activity. The treatment of such lesion is wide and complete local excision, else would show a recurrence of the lesion in due course of time irrespective of its benign properties.

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Captions to Illustrations

Figure.1 : The lesion on the dorsum of tongue and wide local excision marking being performed with diathermy
Figure.2: The histopathology showing keratinization, acanthosis and papillomatosis

References

1. Mibelli,V.: Di una nuova forma di cheratosis, ``Angiocheratoma,'' G Ital Mal Vener 30:285-301,1889.
2. Leung CS, Jordan RCK, Solitary angiokeratoma of the oral cavity, Oral Surg Oral Med Oral Pathol Oral Radiol Endod, 1997, 84(1):51–53.
3. Sertan Ergun, Özgür Mete, Semen Yesil, Hakký Tanyeri, Solitary angiokeratoma of the tongue treated with diode laser, Lasers Med Sci (2009) 24:123–125
4. Roland Imperial, Angiokeratoma- A Clinicopathological study, Arch Derm, Feb 1967, Vol 95
5. A. Zampetti, C.H. Orteu, D. Antuzzi, M.R. Bongiorno,§ S. Manco,M. Gnarra,A. Morrone, G. Cardinali, D. Kovacs, N. Aspite, D. Linder, R. Parini, C. Feliciani* and the Interdisciplinary Study Group on Fabry Disease (ISGF), Angiokeratoma: decision-making aid for the diagnosis of Fabry disease, British Association of Dermatologists 2012 166, pp712–720
6. G.C Ravi, B.T. Nagaraj, Amitha, Multiple papular angiokeratoma of the tongue, Archives of Medicine,2010, Vol 3 No.2:1
7. Dimitrios Andreadis, Athanasios K. Poulopoulos, Anthi Asimaki, Eleni Albanidou-Farmaki, Anastasios K. Markopoulos, Solitary Angiokeratoma of the Buccal Mucosa- Report of a Case, *Balk J Dent Med*, 2014; 18:157-160
8. Sion-Vardy N, Manor E, Puterman M, Bodner L. Solitary angiokeratoma of the tongue. Med Oral Patol Oral Cir Bucal. 2008 Jan1;13(1):E12-4.
9. Maria J. Fernández-Aceñero, J. Rey Biel,G. Renedo, Solitary angiokeratoma of the tongue in adults, *Romanian Journal of Morphology and Embryology* 2010, 51(4):771–773
10. Kar HK, Gupta L. A case of angiokeratoma circumscriptum of the tongue: Response with carbon dioxide and pulsed dye laser. *J Cutan Aesthet Surg* 2011; 4:205-7.
11. M. Vijai Kumar, Devinder M. Thappa, Srikanth Shanmugam, Chaganti Ratnakar, Angiokeratoma Circumscriptum of the Oral Cavity, Acta Derm Venereol (Stockh) 1998; 78: 1
12. M Dutta, S Ghatak, G Biswas, R Sinha, Large, solitary angiokeratoma in the posterior third and base of the tongue: case report, *The Journal of Laryngology & Otology* (2011), 125, 1083-1086.
13. Kumar KS, Giri G, Pandyan DA, Subramanian A, Basu R. Solitary angiokeratoma of tongue: A case report and review of the literature. *Indian J Dent Res* 2018; 29:844-6.

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