Hemangiolympangioma of the Tongue - an unusual clinical presentation creating a Diagnostic Quandary

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ABSTRACT
Hemangiolympangioma is rare vascular malformations consist of both Lymphatic and Blood vessels which are usually congenital and diagnosed early in life between birth and 2 years of age. We hereby highlight a case of an unusual presentation of hemangiolympangioma on the tongue of a 29-year-old male patient.

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INTRODUCTION
Hemangiolympangiomas (HLA) is a rare vascular malformation comprised of both endothelial and lymphatic components. International Society for the study of vascular anomalies 2018 classifies vascular anomalies into vascular tumours and vascular malformations(1). Hemangiolympangioma is a subtype of lymphangioma with a blood component. The common site of presentation occurs in the anterior and posterior cervical triangle of the neck, oral and maxillofacial region, duodenum, colon, bladder, testis, and vertebral column. Very few case reports have been reported the occurrence of hemangiolympangioma in the orofacial region. This case presented as a Diagnostic quandary among the clinicians and created a clinical dilemma because of its aberrant clinical presentation which is discrete from the literature.

Case Report
A 29-year-old patient reported to the department of oral medicine and radiology with the chief complaint of multiple growth & enlargement on the left side of his tongue since his 2 years of age. History of presenting illness revealed that growth was initially smaller in size and gradually progressed to attain the current size and reported no other similar growth elsewhere in the body.

The patient’s medical or dental history was non-informatory.

On examination, inspection reveals diffuse enlargement of the left side of the tongue, extending superoinferiorly from sulcus terminalis to ventral surface of the tongue and mediolaterally from the midline of the tongue to lateral border, along with multiple exophytic growth, ranging from 5-10 in number on the left lateral border which was nodular in shape with smooth surface texture and mucosa covering was normal in colour.

On palpation, exophytic growth was firm in consistency, non-tender, each measuring about 0.5*0.5 cm in size.

The Dorsum of the tongue shows a papular surface with high lingual frenum attachment evident on the ventral surface.
Tongue movements upward, lateral, protrusion and retraction was limited due to high lingual frenal attachment.

Fig.2. Dorsum, Lateral and Ventral surface of the tongue

Based on the history and clinical examination, a provisional diagnosis of lymphangioma of the tongue was made.

Differential diagnosis included neurofibromatosis, lymphoid aggregates, focal epithelial hyperplasia, mucosal neuromas were given.

Incisional biopsy of exophytic growth was done under local anaesthesia in proper aseptic conditions and tissue was sent to histopathological examination. Histopathology revealed parakeratinized stratified squamous epithelium with elongated slender rete processes. The underlying connective tissue stroma is fibro cellular composed of slender and stellate shaped fibroblast cells and dense collagen bundles. The presence of numerous thin-walled blood vessels of varying dimension is diffusely present from the papillary layer to the deep connective tissue. Walls of the blood vessel are lined by thin to plump endothelial cells. The lumen of the vessels are either empty or consist of RBC, suggestive of Hemangiolymphangioma.

Based on history, clinical examination and histopathological features, the final diagnosis was made as Hemangiolymphangioma of the tongue.
The patient is on continuous follow-up, and the lesion remained stable.

Fig.3. Post treatment

Discussion

International Society for the study of vascular anomalies classified vascular anomalies into vascular tumours and vascular malformations. The distinction between the two types is whether there is increased or decreased endothelial cell turnover, based on histopathology(2). Vascular tumours have increased endothelial cell turnover, while vascular malformations have structural abnormalities of the veins, arteries, capillaries and lymphatic vessels. Hemangiolympangiomas is a rare vascular malformation and comprised of both endothelial and lymphatic components. Vascular malformations were named per size of channels and type of fluid contained in the lesion(3).

The aetiology and pathogenesis of HLA are not elucidated in the literature. The coexistence of these two pathological entities may be related to abnormal development of the lymphatic structures during the embryonic period(4).

In this case, the clinical features made suspicion of various differential diagnosis because of its distinctive appearance. Various pieces of literature emphasize that HLA could be associated with a multitude of syndromes such as Rendu–Osler–Weber syndrome, Sturge–Weber–Dimitri syndrome, blue rubber bleb nevus syndrome, Parkes–Weber syndrome, Bannayan’s syndrome, Sturge–Weber–Krabbe syndrome, Klippel Trenaunay syndrome, Servelle–Martorell syndrome, Maffucci’s syndrome and von Hippel–Lindau syndrome which is negative in this case.

The diagnosis of hemangiolympangioma is confirmed using histopathology and can be managed by laser therapy, embolization, or surgical excision(5). The treatment option is dependent on the depth of vascular malformation and anatomical site and it has a good prognosis. Treatment elucidated in the literature is sclerotherapy, embolization and surgical excision. Since in this case, the entire left side of the tongue is involved, an incisional biopsy of the exophytic growth is excised and the patient is under continuous follow up to watch out for any recurrence.

Conclusion

In conclusion, hemangiolympangioma are rare entities encountered clinically and very few cases have been published in the literature. Hereby, we report the distinct clinical presentation of hemangiolympangioma of the tongue and the significance of its masquerading feature. The clinician must delineate its nature and course of treatment to the patient as it could be alarming for the patient.

References


