LYMPHANGIOMA OF THE MAXILLARY SINUS- A RARE CASE REPORT

N. Sangeetha[1] Ramkumar[2] N. Malathi[1] H. Thamizchelvan[1] Sharada .T.Rajan[1]

[1]Department of Oral & Maxillofacial Pathology, Faculty of Dental Sciences, Sri Ramachandra University, Chennai 16, Tamilnadu, India [2] Department of Oral & Maxillofacial surgery, Faculty of Dental Sciences, Sri Ramachandra University, Chennai 16, Tamilnadu, India.

Corresponding Author: N. Sangeetha, Department of Oral and Maxillofacial Pathology, Faculty of Dental Sciences, Sri Ramachandra University, No 1, Ramachandra Nagar , Porur- Chennai 600116, Tamilnadu, India. Email: sangeetha_narashiman@yahoo.co.in, Ph: 919444157222

Abstract

Lymphangiomas are benign hamartomas of the lymphatic vessels. They are congenital malformations that are usually present at birth or diagnosed as early as two years of age. Though 75% of lymphangiomas occur in head and neck region, its occurrence in the maxillary antrum is a very rare entity. Very few cases have been reported in the literature so far. We report a case of lymphangioma of the maxillary sinus in a 29 year old male patient which was discovered as an accidental finding during open reduction of a maxillary complex fracture. Complete surgical excision was done with no reports of recurrence.

Keywords: Lymphangiomas; Hamartoma; Maxillary sinus

Introduction:

Lymphangiomas are developmental malformations that arise from the proliferation of the sequestered lymphatic tissues during embryogenesis. Though it was first reported by Redenbacher in 1828, an accurate description was given by Virchow in 1854[1,2]. Watson and McCarthy have classified lymphangiomas into 5 types: Simple, Cavernous, cellular/ hypertrophic, diffuse systemic and cystic hygromas[3]. Macroscopically the lesions are also classified into macrocystic (cystic space $\geq$ 2 cms), microcystic (<2cms) and mixed (macrocystic component $\geq$ 50% of the total lesion)[4]. Clinically lymphangiomas are often asymptomatic, painless and slow growing. Superficial lesions appear as translucent cluster of vesicles with a pebbly surface whereas deep seated lesions manifest as subcutaneous nodules. These lesions are known to be associated with a wide range of syndromes that include Turner's syndrome, Noonan's syndrome, Trisomies, Cardiac anomalies, Fetal hydrops, Fetal alcohol syndrome, Familial pterygium colli and Maffucci's syndrome. Treatment of lymphangiomas varies from simple surgical excision to cryotherapy, sclerotherapy, laser photocoagulation, chemotherapy and radiotherapy [2]. In the past decade sclerotherapy with Picibanil (OK-432- A mixture of group A streptococcus pyogenes incubated with benzylpenicillin) had produced promising results in the treatment of lymphangiomas [5].

Case Report

A 29 year old male patient reported to the outpatient department of oral and maxillofacial surgery with injury to the face following an assault (Fig 1). He complained of pain and swelling over the maxillary and mandibular region. His past medical history was insignificant except for constant nasal obstruction and chronic rhinitis. Computed tomography of the face revealed Pan facial trauma with multiple fractures in the maxillozygomatic complex and mandible (Fig

2). During open reduction of the maxillary fracture, a mucosal polyp was found attached to the anterior wall of the left maxillary sinus as an accidental finding. The tissue mass was completely excised and sent for histopathological examination with a provisional diagnosis of an Antral polyp. The excised soft tissue appeared translucent measuring 1.1X1.5X 0.5cms in dimension. A clear fluid oozed out from the tissue while grossing it into two halves (Fig 3).

The histopathological sections of the lesion revealed a pseudostratified ciliated columnar epithelium with juxta epithelial mixed inflammatory infiltrate (Fig4). The underlying lamina propria showed large dilated lymphatic spaces lined by endothelial cells and filled with pale eosinophilic fluid resembling lymph (Fig 5). These findings were at par with cavernous lymphangioma. Further the sections were stained with D2-40 antibody to confirm the diagnosis. All the lymphatic endothelial cells around the cavernous spaces showed strong positivity whereas the blood capillaries stained negative with D2- 40 (Fig 6). With respect to the above stated findings a diagnosis of cavernous lymphangiomas was made. The patient was followed up for 6 months and no recurrence was documented.

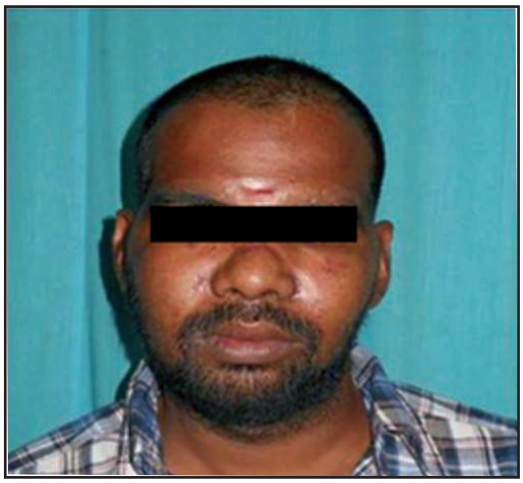

Fig 1: Patient with bilateral swelling of the face.

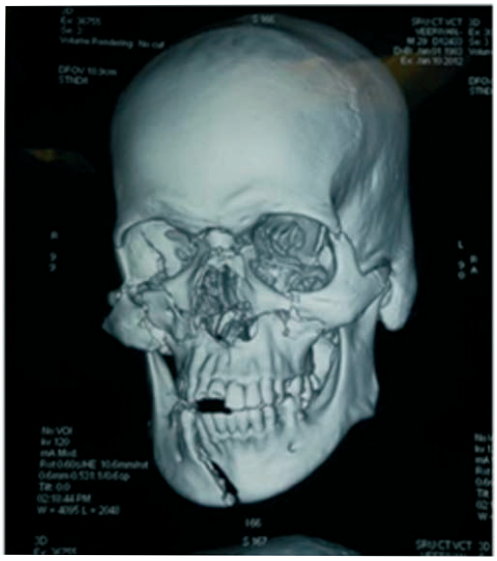

Fig 2: CT of the patient showing multiple fractures of the maxillofacial complex.

Fig 3: Photograph of the soft tissue specimen. Inset shows two halves of the grossed specimen.

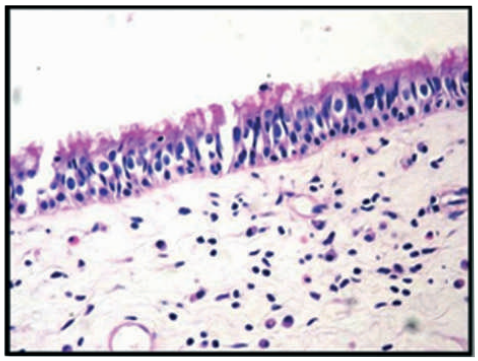

Fig 4: Histopathological section of the specimen showing psuedostratified ciliated columnar epithelium with juxtaepithelial inflammatory infiltrate. (H&E 40X)

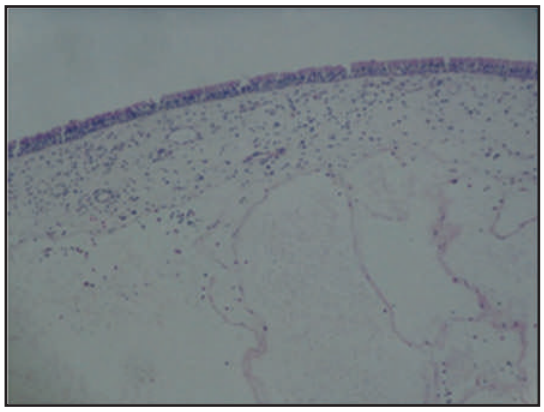

Fig 5: H&E (20X) section depicting the connective tissue with dilated lymphatic vessels lined by endothelial cells & filled with lymph.

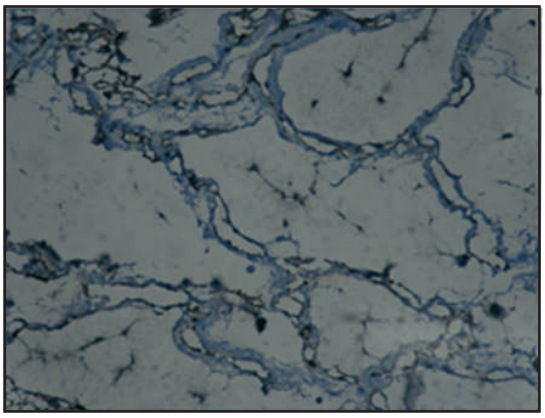

Fig 6: Photomicrograph of D2-40 positive lymphatic endothelial cells lining the cavernous spaces

Discussion

Lymphangiomas are vascular malformations that usually occur in neonates and children below 2 years with no sex predilection. According to Grasso et al the origin of these lesions might fall under one of the following hypothesis.

1. Blockage or arrest of the normal growth of the primitive lymphatic channels during embryogenesis.
2. Proliferation of primitive lymphatic sac that does not reach the venous system.
3. Laying down of lymphatic tissues in wrong sites during embryogenesis.

The incidence of lymphangiomas ranges from 1.2 to 2.8 cases per 1000 live births and account for about 30% of all the lesions that arise from the lymphatic vessels [6]. Head and neck has a marked predilection for the lesion, with the posterior triangle of the neck, submandibular and parotid regions being most commonly affected. In the oral cavity tongue is the usually affected site followed by the palate, buccal mucosa, gingiva and the lips [7]. Pouria M et al have reported a case of bilaterally symmetrical lymphangiomas of the gingiva in a 32 year old patient [8].

In our case the lesion was present in the anterior wall of the maxillary sinus which is a very rare location (A Pubmed search resulted with only three reported cases). Though lymphangiomas do not show any sex predilection all the reported cases including our case were males. Unusually the lesion was attached to the sinus wall by a short stalk. Price .C has reported one such case of a large pendulculated lymphangioma of the maxillary sinus [9]. These lesions within the sinus cavity usually present with nasal obstruction or mimic chronic sinusitis. An accurate location, extent and relationship of these intrabony lesions can be evaluated with an ultrasonography and MRI. Based on the histopathology, lymphangiomas are categorized into lymphangioma simplex (small lymphatic vessels with thin walled endothelium), cavernous lymphangioma (dilated lymphatic vessels with surrounding adventitia) and cystic lymphangioma (huge lymphatic vessels with surrounding fibrovascular tissues and smooth muscle)[2].

Vascular and lymphatic channels share similar histological features. With the advent of specific immunohistochemical markers differentiation of lymphatic vessels from blood vessels has become less promising. D2-40(Monoclonal antibody to Podoplanin) – A specific lymphatic endothelial marker tags the lymphatic vessels sparing the blood capillaries. Other specific lymphatic markers include LYVE-1(lymphatic vessel endothelial HA receptor-1), VEGR3 (vascular endothelial growth factor receptor 3) and Prox1[7].

Genetic studies have showed mutations in chromosome 13, 18, 21, VEGF-C and its

receptors in patients diagnosed with lymphangiomas [2].

Lymphangiomas can be treated with a wide range of treatment modalities with respect to their extent and location. Surgical excision was the treatment of choice in our case as the lesion was small and pedunculated. Lack of recurrence in our case might also be attributed to the pedunculated nature of the lesion

Conclusion

Lymphangiomas of the maxillary sinus is a very rare entity. Lesions with a stalk might mimic an antral polyp. Recurrence of up to 40% has been reported in cases with incomplete removal. A combination of treatments is needed for large growing lesions of the tongue; however a simple surgical excision is the golden standard for small growths. Surgeons and pathologists should be aware of the occurrence of these lesions in such rare anatomical locations for prompt diagnosis and early treatment.

References

1. *Dos Santos G T, Dos Santos J M, Pinheiro Soares L G, Silva Santos N R, Dos Santos J N, Barbosa Pinheiro A L. Surgical Treatment of Oral Lymphangiomas with CO2 Laser: Report of Two Uncommon Cases. Braz Dent J 2010;21(4): 365-369.*
2. *M Kheur S M, Routray S, IngaleY, Desai R S. Lymphangioma of Tongue: A Rare entity Indian Journal of dental advancements 2011;3(3): 635-637.*
3. *Hudson J.W. Submucoal lymphangiomas of the maxillary sinsus: Journal of oral and Maxillofacial surgery 2003;61:390-392.*
4. *Rao A P, Reddy N V, Krishnakumar R, Nirma R M, Muhammed S, Louis C J. Oral lymphangioma presenting as multiple papular lesions of the lip: Hong Kong Dent J 2011;8:107-10.*
5. *Gigue`re C M, Bauman N M, Sato Y,. Burke DK, Greinwald J H, Pransky S, et al. Treatment of Lymphangiomas with OK-432 (Picibanil) Sclerotherapy: Arch Otolaryngol Head Neck Surg 2002;128:1137-1144.*
6. *Grasso D L, Pelizzo G, Zocconi E, Schleef J. Lymphangiomas of the head and neck in children: ACTA Otorhinolarynologica Italica 2008;28:17-20.*
7. *Vasconcelos M G, Santos B C, Peixoto Lemos L C, Ribeiro B F, Paraíso Iglesias D P, Vasconcelo R G. Oral lymphangioma: case report. RSBO 2011;8(3):352-6*
8. *Motahhary P, Sarrafpour B, Abdirad A. Bilateral symmetrical lymphangiomas of the gingiva: case report. Diagnostic Pathology 2006; 1:9.*
9. *Price C. Cystic lymphangioma in the maxillary sinus: a case report. British journal of oral surgery 1976;14:41-46.*

Source of Support - Nil

Conflict of Interest - None declared

How to cite this article:

Sangeetha N, Ramkumar, Malathi N, Thamizchelvan H, Rajan T Sharada; Lymphangioma Of The Maxillary Sinus- A Rare Case Report:*Oral Max Path J*, 4(2), July-Dec 2013: 372-375