

Case Report

Intramuscular Vascular Malformation of Masseter Muscle—A Rare Entity

Dr. Mubeen Khan¹, Dr. Kondajji Ramchandra Vijayalakshmi², Dr. Bettadahalli Thimmaiah Kavya³

¹Professor & Head, ²Associate Professor, ³Post Graduate Student, Department Of Oral Medicine & Radiology, Govt. Dental College & Research Institute, Bangalore, Karnataka, India

***Corresponding author**

Dr. Bettadahalli Thimmaiah Kavya

Email: drkavyasmile@yahoo.com

Abstract: Facial vascular malformations can cause dental emergencies that result in fatal or life-threatening and disfiguring situations. A knowledge of vascular malformation, its clinical presentation and its complications can prevent iatrogenically related accidents and minimize potential spontaneous crisis for the patient in the dental clinic. A vascular malformation (VM) is a morphogenetic abnormality of blood and/or lymphatic vessels with normal ultra-structural characteristics and endothelial hyperplasia. It results from a developmental arrest after the endothelial stage of embryologic vascular development contrary to hemangiomas, which appear to be a failure of differentiation at the endothelial stage. VM has a normal endothelial cell growth cycle that affects the veins, the capillaries and lymphatics and they do not involute. Intramuscular vascular malformations are uncommon tumors in the head and neck, although masseter muscle is the most common site, accounting for approximately 5% of all intramuscular vascular malformations in the head and neck region. Because of the rarity of these tumors, their deep location and unfamiliar presentation, inaccurate preoperative diagnosis and inappropriate treatment planning are common problems. The present report describes intramuscular arteriovenous malformation of masseter are presented highlighting the typical clinical presentation with MRI which should alert the dental physician to the possibility of such a lesion.

Keywords: Masseter, vascular malformation, intramuscular, ultrasonography, MRI

INTRODUCTION

Vascular malformations (VM) are errors of vascular morphogenesis present at lower incidence accounting 7% of all benign tumors. About 1/3rd of VM occurs in head and neck region, although intramuscular vascular malformation is comparatively rare. According to Batsakis the incidence rate of intramuscular vascular malformation is 45% in lower limbs, 14% in the abdomen region and 14% in head and neck region [1]. In the head and neck regions, the favoured sites are the masseter, trapezius, and sternomastoid muscles. Clinicians who diagnose and treat oral conditions should be aware of the lesions and the impact they can have on routine procedures. Proper recognition and therapeutic intervention can help to avoid serious complications and potentially tragic outcomes.

CASE REPORT

A male patient aged 20 years reported to our department with a chief complaint of swelling on the right side of the face. The swelling had been present since 10years, which was gradually increasing in size. There was a history of enlargement of the swelling on the right side of lower jaw when the patient lies down

and reduces gradually in sitting posture. The patient's medical and family history were unremarkable.

Head and neck examination revealed, a diffuse swelling on the right lower one third of the face extending anteroposteriorly from the right corner of the mouth to right tragus of the ear. Superoinferiorly from the right ala tragus line to inferior border of the mandible. There was no apparent abnormality seen in the skin over the swelling. On palpation, the swelling was non tender, soft and not totally compressible and no pulsations were felt. Further the swelling appeared more prominent on contraction of masseter muscle. Intra oral examination revealed no significant findings in the mucous membrane and teeth, contributory for diagnosis.

Based on the history and clinical findings a provisional diagnosis of vascular lesion on the right masseter was established. Differential diagnosis of parotid gland tumor and lymphangioma was considered.

Patient was subjected for orthopantomograph, which did not reveal any significant finding. Fine

needle aspiration cytology revealed the presence of frank blood suggesting vascular etiology. Doppler ultrasound imaging showed a relatively well defined eco-complex lesion with internal hypochoic areas in the intramuscular compartment of right masseter muscle, demonstrating colour flow on compression of the lesion suggesting differential diagnosis of intramuscular haemangioma or arteriovenous malformation.

MRI with MR of neck angiography revealed a well-defined nonenhancing, lobulated, heterogenous lesion measuring about 3.3x1.7x4.4 cm which is showing isointensity on T1W1 and hyperintensity in T2/PDFSW1 with phleboliths, in the intramuscular plane of right masseter, suggestive of intramuscular vascular malformation of right masseter.



Fig-1: Diffuse swelling in the right lower one third of the face



Fig-2: Swelling evident on contraction of masseter muscle i.e, "Turkey Wattle" sign

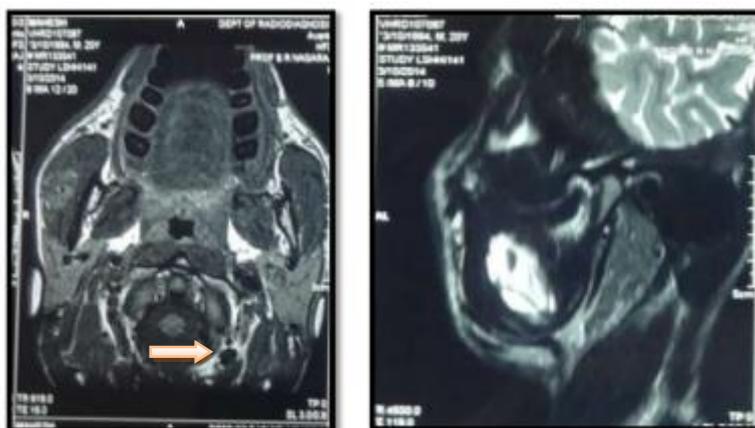


Fig-3: MRI with MR Angiography of neck showing the lesion



Fig-4: showing post-surgical scar in the angle of the mandible

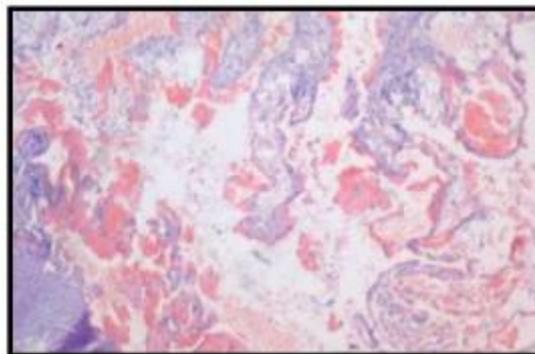


Fig-5: Photomicrograph (10x) of histologic section showing loose fibrocollagenous tissue with dilated vascular channels within muscular layer

MANAGEMENT

Patient was referred to a vascular surgeon and surgical excision was done using right submandibular approach under general anaesthesia. Histopathological examination of the specimen showed loose fibrocollagenous tissue and skeletal muscle fibre with dilated vascular channels within muscular layer suggestive of arterio-venous malformation of right masseter.

DISCUSSION

The intricate vascular anatomy of the head and neck probably predisposes the region to vascular anomalies. Vascular lesions are divided into two major groups - hemangiomas and vascular malformations - based on histologic and clinical presentation. Hemangiomas are identified by rapid endothelial cell proliferation in early infancy, followed by involution over time. Vascular malformations have a normal endothelial cell growth cycle that affects the veins, the capillaries, arteries or the lymphatics and they do not involute. Both hemangioma and vascular malformations are included in the broad scientific term “vasoformativetumors” [2].

A vascular malformation is a morphogenetic abnormality of blood and/or lymphatic vessels with normal ultrastructural characteristics and endothelial hyperplasia. It results from a developmental arrest after

the endothelial stage of embryologic vascular development - contrary to hemangiomas, which appear to be a failure of differentiation at the endothelial stage [2, 3].

Hemangiomas are characterized histologically by endothelial hyperplasia and increased numbers of mast cells during their proliferation phase. During the involution stage, they are characterized by fibrosis, fatty deposits, and multilaminated basement membrane formation beneath the endothelium. They are frequently documented at birth or within the first 10 days of life. Almost all show rapid clinical expansion during the first year of life and frequently involute slowly during the next 5 years [4].

Compared to haemangioma, Intramuscular vascular malformations are uncommon tumors in the head and neck region. Less than 1% of vasoformative tumors throughout the body occur in skeletal muscle; 15% of them arise in head and neck musculature. The masseter muscle is the most common site, which accounts for approximately 5% of all intramuscular vascular malformations in the head and neck region [5, 6].

Exact etiology is unknown. One hypothesis postulates that placental cells, such as the trophoblast may be the cell of origin for vascular malformations.

Therefore, vascular malformations may arise secondary to some event in utero. The relationship between vascular malformations and placental tissues is controversial and needs further investigation [7, 8].

The pathophysiology of vascular malformation represents an arrest in the development of the mesenchyme primordia in the undifferentiated capillary network stage. As differentiation progresses, primitive vessels penetrate deeper into the subcutaneous layer, the muscle, or the bone tissue. The final development stage involves the gradual replacement of the immature plexiform network by the mature vascular channels and gives rise to a vascular malformation [4, 8, 9].

In our case the swelling was small, hence the compressibility was not totally demonstrable and the swelling was prominent on contraction of masseter muscle (positive turkey wattle sign). However there was no noticeable discoloration of the skin, nor the bruits and pulsations were present.

Intramuscular vascular malformation usually present in the third decade of life with a male predominance as reported by Rai *et al* [10]. Palpation of the mass is often misleading because they are often located deep within a muscle and can vary in consistency from a diffuse, soft, compressible mass to one that is very firm. Discoloration of the overlying skin is unusual and the presence of pulsations, thrills, or bruits is rare. Enlargement of the swelling with valsalva or head dependency (turkey wattle sign) is also an unusual finding. Anatomically because of the deep intramuscular location of these tumours and lack of specific signs suggesting vascular etiology, accurate preoperative diagnosis has been reported in less than 8% of cases.

In our case there was no significant findings in OPG however Doppler ultrasonography and MRI showed features suggestive of vascular malformation. Fine needle aspiration cytology revealed frank blood in our cases. Doppler ultrasonography and magnetic resonance imaging with MR angiography is the best imaging method used in differential diagnosis and topographical characterization of vascular malformations and tumors of cervicofacial area.

As the swelling presents with no history of pain, tenderness and they are always slow growing can be mistaken for salivary gland tumors, benign masseter muscle hypertrophy, myositis ossificans, lymphangioma, angiosarcoma etc., however ultrasonography and MRI should always be advised to rule out vascular malformation and hemangiomas.

MANAGEMENT

Though there are different modalities of treatment, depending on tumor location, accessibility, depth of invasion, patient age and cosmetic

consideration, the management of intramuscular vascular malformation should be individualized. Various treatment modalities are medical therapy, embolotherapy, sclerotherapy, Laser, Cryosurgery, Surgical therapy. Complications can be of two types: 1. complications from the disease process includes hemorrhage, infection, and ulceration. 2. Complication because of treatment includes recurrence and restricted mouth opening, dysphagia etc. [10].

In our case only surgical approach was opted as the lesion was of considerable size and esthetically compromised. A follow up of 6 months did not show any complications or recurrence.

CONCLUSION

Oral physician should keep in mind the differential diagnosis of vascular malformation whenever there is a swelling in the angle of the mandible or parotid region. Fine needle aspiration cytology is recommended whenever there is soft swelling in these regions. Ultrasonography and MRI also enhances the diagnostic accuracy and facilitate optimal treatment planning.

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