CASE REPORT

An unusual presentation of intramasseteric vascular malformation

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Abstract

Vascular malformations (VMs) are congenital anomalies of the capillary, venous, lymphatic, and arterial system. They may be distinguished from vasoproliferative tumors (hemangiomas) which present with increased endothelial cell turnover, whereas VMs are structural anomalies of vessels without endothelial cell proliferation. VMs, in general, occur in superficial areas such as lips, tongue, and buccal mucosa, and an intramuscular location is quite rare. An intra-masseteric location may be mistaken for a parotid swelling and may also pose problems in terms of proximity to facial nerve. One such rare case of intra-masseteric VM occurring in a 23-year-old female with an unusual clinical presentation is being presented in this case report. The patient complained of a swelling associated with pain on her right cheek since 9 months. The swelling develops only when her head and torso is bent to 90° or more for at least 5-10 min and gradually disappears on attaining an upright posture (turkey wattle sign). Color Doppler ultrasonographic findings were suggestive of a VM. The lesion was excised under general anesthesia and was sent for histopathologic evaluation which was suggestive of an intramuscular hemangioma/malformation. The patient did not develop any post-operative complications and had recovered well after the surgery.

Keywords: Hemangioma, magnetic resonance imaging, masseter, turkey wattle sign, vascular malformation

Introduction

The head and neck region has a complex vascular anatomy and hence may be prone to develop vascular anomalies. The term hemangioma has been used for several decades to describe the vast variety of vascular anomalies despite varied clinical behavior and differences on histopathology and imaging. In 1982, Mulliken and Glowacki proposed a classification for vascular anomalies based on pathologic features as follows (1) vasoproliferative or vascular neoplasms (hemangiomas) and (2) vascular malformations (VMs). This classification was adopted by The International Society for the Study of Vascular Anomalies, in 1996, and has been regularly reviewed every 2 years and is widely accepted. The major distinction between the two categories is with regard to endothelial cell turnover, vasoproliferative neoplasms have increased endothelial cell turnover, whereas VMs do not have increased endothelial cell turnover. Hemangiomas comprise infantile and congenital hemangiomas. Infantile hemangiomas present between 2 weeks and 2 months of life, whereas congenital hemangiomas are fully formed at birth. Infantile hemangiomas rapidly proliferate during the 1st year of life, after which, they enter into the involution phase and by the age of 8-9 years, they completely regress. Congenital hemangiomas are further classified into non-involuting congenital hemangiomas, which may partially involute but complete resolution is not seen and rapidly involuting congenital hemangiomas, which regress completely within 2 years. VMs are usually present at birth and grow proportionately with the child, showing no signs of spontaneous resolution. VMs can be categorized as low-flow lesions (combinations of capillary, venous, and lymphatic components) and fast- or high-flow malformations (combination of arteries with other vascular structures). It has been reported that although most VMs are noticed at birth, some appear in early childhood while some do not appear until adolescence or early adulthood. VMs tend to develop in superficial areas in head and neck region such as lips, tongue, and buccal mucosa. The occurrence of VMs in skeletal muscles is rare, approximately 1% of VMs are known to occur in skeletal muscles. In general, muscles of the lower limb, upper limb, and trunk are generally involved and muscles in the head and neck muscles are rarely affected. Only 4.9% of all intramuscular malformations occur in the masseter. The following describes a rare case of intramuscular VM with an unusual presentation.
Case Report

A 23-year-old female presented with a complaint of a swelling associated with pain on her right cheek since 9 months. History revealed that the swelling developed only when her head and torso was bent to 90° or more for at least 5-10 min. This has been described as “Turkey wattle sign,” i.e., the appearance of a swelling in a dependent position. The appearance of the swelling was associated with pain of moderate intensity radiating to the forehead. There was gradual disappearance (approximately 10 min) of the swelling and pain on attaining an upright posture. There was no history of prior trauma, tooth pain, or allergy. The medical history was non-contributory. On examination, as there was no evidence of swelling on the cheek in the upright position, the patient was asked to bend her torso for at least 5 min. In this posture, the swelling was not very obvious on inspection, however, a solitary ovoid swelling measuring 2 cm × 1.5 cm was palpable in the right masseteric area anterior to the parotid gland [Figure 1a and b]. There was no abnormality of the skin over the swelling with no obvious discoloration. There was no local rise in temperature. The swelling was tender, firm and pulsations were not evident. There was no enhancement of the swelling on clenching. On attaining an erect posture, the swelling gradually disappeared. There was no reduction in mouth opening. Intraoral examination revealed no odontogenic cause for the swelling. The orifice of the right Stensen’s duct did not show any abnormality, and the salivary flow was normal. Considering the age of occurrence of the lesion and the typical appearance of the swelling in a dependent position, a provisional diagnosis of vascular malformation was given. A differential diagnosis of venolymphatic, pure venous, pure lymphatic and arteriovenous malformation was given. The panoramic radiography did not reveal any calcification in the region of the right masseter. Fine needle aspiration cytology revealed a fibrovascular core with adipocytes and scanty cellularity. On gray-scale ultrasonography (US), a compressible, non-homogeneous lesion with irregular borders was evident within the masseter muscle. The poor acoustic enhancement was evident. The lesion consisted of both cystic (anechoic areas) and solid areas (isoechoic areas). Color Doppler ultrasound showed reduced vascularity (low internal flow) and monophasic flow [Figure 2]. Magnetic resonance imaging (MRI) revealed a bulky right masseter muscle. A well-defined lesion measuring 26 mm × 12 mm × 24 mm (anteroposterior × transverse × craniocaudal) was noted in the outer muscle bulk. The lesion is predominantly heterointense (hypo and isointense) in T1 weighted images [Figure 3a] and hyperintense in T2 weighted images [Figure 3b]. Fluid-fluid levels were seen in T2 weighted fat suppressed images along the anterior aspect of the lesion. A focal area of diffusion restriction was also seen in the outer aspect of lesion in T2 fat-suppressed images (flow voids). The US and MRI were suggestive of a venous/lymphatic malformation. A complete surgical excision was planned under general anesthesia. The masseteric area was exposed through a curvilinear incision in front of the ear and extending onto the neck [Figure 4]. A soft swelling 3 cm × 2.5 cm × 1 cm was exposed adherent to the masseter muscle. The lesion was excised with some of the underlying muscle fibers. Hematoxylin and eosin stained sections revealed large endothelium-lined vascular channels with engorged red blood cells and adipocytes.
cells. These channels were seen in between the muscle fibers along with neural bundles. Few lymphatic channels were also evident. Both extravasated red blood cells and lymph were also seen [Figure 5a-c].

**Discussion**

Although the exact pathogenesis of VM is unknown, a few studies suggest a simultaneous increase in proteolytic enzymes such as high molecular weight matrix metalloproteinases along with tissue remodeling.[5] The formation of lymphatic malformations can be correlated to the overexpression of receptors such as vascular endothelial growth factor R3 and Prox-1 that are responsible for the development of lymphatic vascular channels.[5] In capillary malformation, there is an altered cell growth, differentiation, and proliferation of endothelial cells which is caused by delayed Ras/mitogen-activated protein kinases activation after receptor tyrosine kinases activation.[6] Mutation of angiopoietin receptor gene Tie2/TEK in venous malformations has been recently discovered.[6] Tie2 signaling pathway is crucial for angiogenesis and vascular maturation.[6] Furthermore, studies on patients with venous malformation show an upregulation of tissue growth factor beta and basic fibroblast growth factor.[2] Mutation of RASA1 gene has been noted in subjects with arteriovenous malformations.[2] Some studies also demonstrate an elevation in plasma levels of angiopoietin-2 and Tie-2 receptor in subjects with active arteriovenous malformation.[5]

VMs are common in whites than in dark skinned individuals.[7] In a case series, the age range of VMs was found to vary between 15 and 65 years with a mean age of 38 years.[8] A few reports suggest that VMs have a predilection to occur in males,[7] whereas other reports suggest that both genders are equally affected. Involvement of skeletal muscles by VMs is rare.[8] In head and neck region, the masseter, sternomastoid and trapezius muscles are involved in the descending order of frequency.[9] Intramuscular malformations often have a delayed presentation due to lack of obvious skin involvement or deformity until they reach a certain size, subjected to trauma/infection or if the involved muscle is in a state of contraction.[10]

Purely venous malformations may be evident at birth however they may be noted in adults. Phlebothrombosis is common, leading to distention, firmness, and tenderness of affected soft tissues.[11] Lymphatic malformations are often noticed at birth and most of them are obvious by 2 years of age.[12] Both the lesions are easily compressible.[2,3,7,14-16] Purely venous and purely lymphatic malformations are rare, and they generally occur in combination. Venous malformation has the tendency to expand and contract with the change of patient’s posture as it is a dependent lesion.[13] Arteriovenous malformations are present at birth. They may not become noticeable until later in childhood or adulthood. The overlying skin feels warmer to touch and a palpable thrill or bruit is often felt.[14]

In the present case, there was no discoloration of overlying skin and no local rise in temperature. The lesion was compressible and the lesion appeared in a dependent position. Hence, a venolymphatic malformation was considered.

Radiologic investigations of vascular lesions include plain radiography, sialography, US, computerized tomography scanning, and MRI. Plain radiographs may reveal opacities that may be mistaken for salivary calculi, although radiopaque parotid calculi are uncommon (20%) and usually small.[8] The presence of a well-defined opacity in the parotid region on the plain radiograph is usually a phlebolith, dystrophic calcification within a lymph node, or a pleomorphic adenoma.[8] Sialography may confirm the extraductal position of the calcification.[8] US is commonly used to the delineate the margins of the lesion when it is located more superficially.[16] Experienced radiologists can differentiate malformations (specifically venous, arteriovenous and lymphatic anomalies) from hemangiomas based on US.
Infantile hemangiomas on gray scale US are well defined, solid and homogenous.[11] On color Doppler US, arterial and venous wave forms are evident, and hypervascularity is noted. Mature hemangiomas are similar, except that they are smaller (approximately 1 cm) and have a low flow. Venous malformations are compressible, solid echogenic masses on gray scale and on color Doppler, monophasic or no flow is evident.[2] Lymphatic malformations are multicystic on gray scale on color Doppler, no flow is evident.[3] In arteriovenous malformations, arterial and venous signals along with arterialization of venous structures and hypervascularity will be noted on color Doppler US.[2]

MRI provides images with good soft tissue definition of both normal anatomy and pathology, but it is also sensitive to blood flow within vessels, enabling the nature and extent of VMs to be defined more easily.[6] Most of the vascular lesions on MRI appear as iso to intermediate signal on T1 weighted images, hyperintense on T2 weighted images and flow voids may be seen. Flow voids represent an obstruction in the flow. Venous malformations show a heterogenous intermediate signal on T1.[2]

In the present case, the lesion comprised both solid and cystic areas. Hence, a malformation was considered, and a hemangioma was ruled out as hemangioma appears as a homogenous solid mass. Among the various types of malformations, arterial malformations were ruled out as there was no evidence of high flow signals. Pure venous and lymphatic malformations were ruled out as the lesion was not homogenous. Hence, a venolymphatic malformation was considered.

Histologically a hemangioma in its proliferating phase is composed of rapidly dividing endothelial cells forming syncytial masses with and without lumens. Electron microscope examination of proliferating hemangioma reveals multilamination of the basal lamina beneath the epithelium.[17] During the involutive phase, endothelial cell activity diminishes and the cellular parenchyma is replaced by fibrofatty tissue. At this stage, a surgical specimen may have a “cavernous” histologic appearance, which may be confused with a venous malformation.[17] In contrast, histologic evaluation of VMs shows no evidence of cellular proliferation, but rather a progressive dilatation of vessels of the abnormal mural structure. VMs are lined by flat, quiescent endothelium, lying on a thin single laminar basement membrane.[17] Further a study conducted in 2005 on 167 benign vascular lesions demonstrated the presence of the nerve bundles in VMs which were however not noticed in hemangiomas. It was concluded that the presence or absence may be used to differentiate between the two.[11]

Management of hemangiomas varies depending on the type and severity of complications. Corticosteroids are the mainstay of medical management in either topical, intralesional, or systemic forms.[11] If the lesion does not respond to corticosteroids, second-line of pharmacologic agents include vincristine or interferon alfa-2b.[11] Intralesional injection of corticosteroid should be considered for a small, well-localized cutaneous hemangiomas. Triamcinolone (25 mg/ml), at a dosage of 3-5 mg/kg, is injected slowly at low pressure with a 3 ml syringe and fine gauge needle.[11] For problematic, endangering, or life-threatening hemangiomas corticosteroids are orally administered. Prednisone or prednisolone is prescribed at a dosage of 2-3 mg/kg/day.[11] Propanolol has been recently introduced in the management of infantile hemangiomas.[2] For medical management of an individual with hemangiomas, imaging is not absolutely indicated however for non-pharmacological management imaging is necessary.[2]

Management of intramuscular VM should be individualized based on tumor location, flow characteristics, symptoms, accessibility, depth of invasion, patient age, and cosmetic considerations.[15] Sclerotherapy, laser therapy, or cryosurgery have been used in the management of low flow VMs which are compressible and accessible.[11] Surgery is reserved for non-compressible lesions.[7] Sclerotherapy is aimed at obliterating the lumen by causing fibrosis of patent vessels.[16] Sodium tetradecyl sulfate, ethanol, polidocanol, hypertonic saline, and sodium morrhuate are the commonly used sclerosing agents.[7] US can be used to prevent intra-arterial injection and to assess the extent of sclerosis.[7] Laser therapy in the form of neodymium-doped yttrium aluminium garnet, argon and carbon dioxide have been successfully used in the management of VMs.[7] Angiography is warranted for identifying the feeder artery and defining the complete extent of vessels before the management of high-flow malformations.[5] The first-line of therapy for arteriovenous malformations and arteriovenous fistulas is embolization.[7] This is followed by aggressive ablative therapy.[7] Embolization is a procedure in which the vessels are occluded by the introduction of a foreign body such as gelfoam, oxg medieval, silastic spheres, steel pellets, ivalon, microfibrillar collagen, and silicone rubber.[7]

Surgical approaches to masseter muscle VMs are pre-aureolar approaches using superficial skin flaps, resection in combination with superficial parotidectomy, or intraoral excision.[7]

Conclusion

The diagnosis of vascular lesions can be challenging. Unlike most other pathologies, histopathology does not yield a definitive diagnosis in case of vascular lesions. Hence, the clinical features, imaging, and histopathology should be considered in the diagnosis of these lesions.

References

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