Clinically obscure venous malformation in the sub mandibular triangle: A rare presentation

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ABSTRACT

Vascular lesions have been classified as hemangiomas or vascular malformations depending on the presence of cellular proliferation. These lesions have been known to cause significant morbidity and even mortality in both children and adults. Confusing nomenclature, relative rarity of these lesions, inappropriate diagnosis, lack of precise imaging studies and deficiency of multidisciplinary approach are some of the factors which result in ineffective treatment of such patients. Here, we report a rare case of venous vascular malformation presenting as a mass in the sub mandibular region with no clinical stigmata which was diagnosed with the help of various imaging techniques and subsequently treated by surgery.

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INTRODUCTION

Hemangiomas and vascular malformations are relatively common tumors of the face and neck region which represent one of the largest groups among soft tissue tumors and typically present with clinical stigmata as skin discoloration, dilated subcutaneous vessels and compressibility suggestive of the diagnosis. Vascular lesions were earlier categorized on the basis of the size of the channels and the type of fluid in the lesions, according to which blood-containing lesions were called hemangiomas and lymph-containing lesions were referred to as lymphangiomas or cystic hygromas. Hemangiomas were further differentiated on the basis of channel size into capillary hemangiomas, strawberry hemangiomas and cavernous hemangiomas.¹ This classification system resulted in great confusion and in 1982 was replaced by a biologic classification system by Mulliken and Glowacki² which separates endothelial malformations into two large groups, hemangiomas and vascular malformations, on the basis of their natural history, cellular turnover, and histology. Hemangiomas are benign,

localized tumors of the blood vessels where as vascular malformation result from anomalous development of vascular plexuses.³ Hemangiomas are vascular neoplasms with endothelial hyperplasia, these tend to be small or absent at birth and often go unnoticed. Soon after birth they undergo proliferation and rapid growth which may last for several months. This is followed by a stationary period and then a period of involution. Conversely, vascular malformations form secondary to an error of embryonic development with normal endothelial turnover; these are always present at birth and grow commensurately with the child, although the lesion may expand secondary to trauma, infection, hormonal changes, or embolic or surgical intervention. They do not involute and remain present throughout the patient's life. Vascular malformations are subcategorized on the basis of their histological makeup as lymphatic, capillary, venous, arteriovenous, and mixed malformations and according to hemodynamic features as slow flow (either capillary, venous, lymphatic, or combined forms) or "fast-flow" (arteriovenous fistulas and arteriovenous malformation).⁴ Malformations with arterial

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components are considered high-flow lesions and those without arterial components are considered low-flow lesions.¹

Here, we present a rare case of an obscure venous vascular malformation in the sub mandibular triangle, a low-flow lesion arising from the facial vein which without involving the gland was present in the fascial planes extending from the buccal to the lingual side of the mandible.

CASE REPORT

A 25-year-old male presented with a swelling in the left sub mandibular region, he gave a history of a small swelling being present since childhood. Recently, the swelling had increased in size and was causing discomfort in neck movements. There was no change reported in the size of the swelling with meals or on straining and there was no history of pain. He was otherwise asymptomatic.

Clinical examination revealed a large solitary, compressible swelling, palpable both on buccal and the lingual side of the mandible with the overlying skin normal in color, texture, with a temperature similar to the surrounding skin, also the lesion was non pulsatile. No other mass or lymph node was palpable in the rest of the neck. Intraorally, a swelling was visible both on the lingual and buccal side of the left mandibular arch with bluish discoloration of the overlying mucosa. Saliva flowed freely from the ipsilateral Wharton's duct and no calculi were palpable. Initial clinical diagnoses included a reactive nodal mass, sub mandibular gland neoplasm and branchial cleft cyst.

Fine needle aspiration revealed frank blood. Computed Tomography (CT) angiography and Magnetic resonance imaging (MRI) was advised. CT of neck region showed a lobulated predominantly hypodense lesion in left side of neck involving the superficial soft tissue and extending along, both the buccal and lingual aspect of mandible. No definite bony involvement was seen. CT angiography revealed the feeders to be facial vessels.

MRI demonstrated a heterogeneous, well-defined mass, $7 \times 8 \times 5$ cm in size in sub mandibular triangle of neck on the left side. T1-weighted MRI images showed a large, lobulated, predominantly hyperintense space occupying lesion. The lesion was seen extending toward the lingual side curving around the mandible, compressing the left sub mandibular gland and extending anteriorly along the lingual margin of mandible toward the midline. The lesion was abutting and compressing the mylohyoid muscle and mildly displacing the carotid vessels posteriorly. The underlying bone showed no erosion. On the buccal side, the lesion was seen extending anteriorly along the body of mandible, abutting the masseter muscle with loss of fat planes and no definite evidence of muscle invasion. No signal void was seen ruling out the possibility of high-flow lesion (Fig. 1A and B).

A diagnosis of venous vascular malformation of the fascial planes in the sub mandibular region was made on the basis of history, clinical and radiological findings. Surgical excision of the lesion was planned as the lesion was non infiltrative, well defined and confined to a single fascial plane.

At surgery, a reddish blue colored lesion was seen lying deep to the superficial fascia and platysma above the sub

Fig. 1 (A) T1 W magnetic resonance image (sagittal section) showing a lobulated mass in sub mandibular region extending along the buccal and lingual side of the mandible. (B) T1 W MRI axial image showing the posterior extension of lesion to the parapharyngeal space.

mandibular gland. The facial vein, which was feeding the tumor, was ligated and the specimen was carefully removed in toto (Fig. 2). Post-operative period was uneventful. The drain was removed on the 2nd post-operative day and the patient was discharged on the 7th post-operative day after suture removal. Histopathological examination of the specimen revealed lobules of blood filled cavernous spaces lined by flattened endothelium. Intervening fibroadipose tissue was seen scattered with chronic inflammatory cells confirming the diagnosis.

The patient is completely asymptomatic after 1-year-post-operative follow-up.

DISCUSSION

The sub mandibular triangle normally contains the sub mandibular gland, lymph nodes, and fat. The differential diagnosis of sub mandibular triangle masses generally includes inflammatory lesions of the gland or lymph node; neoplasms as pleomorphic adenoma, lymphangioma, lipoma, intramuscular hemangioma and cystic masses as branchial cleft cyst or malignancies. The distinction between these diseases is apparent by history, physical examination and imaging techniques.⁵

While characterizing vascular malformations, the most important feature to be differentiated is whether the lesion is a high- or low-flow vascular malformation. Malformations with arterial components are considered high-flow lesions and those without arterial components are considered low-flow lesions, which can be distinguished by MRI and angiography as was done in our case. Venous vascular malformations are low-flow lesions which have

previously been referred to as non involuting cavernous hemangiomas in both clinical and radiologic literature. They are thought to be caused by an error in vascular morphogenesis, secondary to hereditary or sporadic mutations, altered gene expression, or environmental factors.⁶ These are dysplasias of small and large venous channels associated with a variable amount of hamartomatous stroma. The venous channels connect with adjacent veins.¹ Venous vascular malformations though common in the facial region are rarely found in the sub mandibular region. They may present as solitary or multiple lesions, localized or infiltrating various tissue planes, an isolated skin varicosity or as a spongy blue mass. They usually present with typical clinical stigmata that include skin discoloration, dilated subcutaneous vessels, pulsatility, and compressibility which aids in diagnosis. The location and extent of lesion may produce additional clinical features as pain, discomfort and stiffness, bleeding, distortion of speech or dentition, or obstruction of the airway. On palpation they are easily compressible and deflate with elevation, differentiating them from hemangiomas which on palpation will reveal a fleshy tumor that does not empty completely when compressed. Venous malformations typically involve the skin of the face, limbs, or trunk but also are found in the internal viscera, skeletal muscles and bones.⁷

In our case, none of the usual clinical features were present, concealing the diagnosis; only after Fine needle aspiration and imaging was the lesion diagnosed representing the obscure nature of some of the vascular malformations.

Many techniques like ultrasonography, MRI, CT and angiography assist in defining the extent of lesion. Imaging techniques are more often than not required for correct diagnosis and treatment planning as they are most useful in characterizing the lesion and discovering the anatomic extent of disease. Vessel density as depicted on Doppler sonography has been used in differentiating other types of masses from vascular malformations.¹ MRI is more expensive but highly sensitive and specific, providing more information than other techniques. MRI portrays the extent of involvement within tissue planes and demonstrates flow characteristics.⁸ Deep hemangiomas and venous malformations show usually hypointense to intermediate signal on T1-weighted images, also multiple rounded signal intensity voids representing phleboliths may be noted. Phleboliths are rare but characteristic finding. These lesions can sometime be hyperintense on T1 images, as was seen in our case. On T2 images, these lesions are usually hyperintense. Features of high-flow lesions (AVMs) include serpiginous signal voids, absence of a dominant mass and intraosseous extension with decreased marrow signal on T1-weighted images which were not seen in our case ruling out the possibility of a high-flow lesion.⁹

Fig. 2 Excision of lesion with ligated vessel.

Angiography is commonly used nowadays and is useful in super selective embolization, for sclerotherapy of venous malformations and extripation of vascular malformations.

Low-flow vascular malformations are generally difficult to treat. Surgical resection, medical therapy, and transarterial embolization have all had limited success. The treatment of choice for venous vascular malformations is percutaneous sclerotherapy with or without surgical excision¹⁰ as spontaneous regression does not occur. Either complete sclerosis or excision is important, as it has been shown that incomplete treatment leads to symptom recurrence. In our case, the lesion was confined to a single fascial plane resulting in complete surgical excision.

Other treatment methods for venous malformations apart from sclerotherapy and surgery include pressure, cautery, embolization, cryotherapy, laser surgery, vascular ligation, and corticosteroids.

Indication for medical or surgical treatment of a venous malformation is usually pain which is secondary to thrombosis of the malformation. Other indications include symptomatic but stable tumors in older children, sudden rapid acceleration of tumor growth, gross functional impairment, local skin necrosis, thrombocytopenia, and cosmetic deformity. Choice of therapy required again varies with the type, location and extent of lesion. Involvement of adjacent neurovascular structures complicates the treatment. Local recurrence may occur due to incomplete surgical resection and is more common with diffuse malformations. Surgery is often contraindicated if risks associated with the resection outweigh the presumed improvement in appearance or function that may be derived from surgery.

The reported case due to the obscure nature, location and absence of typical clinical features of lesion represents the diverse and rare forms of vascular malformations which may present at any clinical setup, here we would also emphasize that accurate knowledge of differentiating clinical features and characteristic imaging findings along with multidisciplinary approach is required to successfully manage such cases.

CONFLICTS OF INTEREST

All authors have none to declare.

REFERENCES

1. Donnelly LF, Adams DM, Bisset GS III. Vascular malformations and hemangiomas. *AJR Am J Roentgenol*. 2000;174: 597–608.
2. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg*. 1982;69: 412–422.
3. Shafer WG, Hyne MK, Levy HM. *A Textbook of Oral Pathology*. 4th ed. W.B.Saunders Company; 1983.
4. Finn MC, Glowacki J, Mulliken JB. Congenital vascular lesions: clinical applications of a new classification. *J Pediatr Surg*. 1983;18(6):894–900.
5. Martin JF, Roy AH, Roland JT. Clinically unsuspected venous malformations limited to the submandibular triangle: CT findings. *Am J Neuroradiol*. 1995;16:491–494.
6. Legiehn GM, Heran MK. Venous malformations: classification, development, diagnosis, and interventional radiologic management. *Radiol Clin North Am*. 2008;46:545–597.
7. Hein KD, Mulliken JB, Kozakewich HP, et al. Venous malformations of skeletal muscle. *Plast Reconstr Surg*. 2002;110(7): 1625–1635.
8. Fordham LA, Chung CJ, Donnelly LF. Congenital vascular and lymphatic malformations of the head and neck. *Radiol Clin North Am*. 1999.
9. Baker LL, Dillon WP, Hieshima GB, Dowd CF, Frieden IJ. Hemangiomas and vascular malformations of the head and neck: MR characterization. *AJNR Am J Neuroradiol*. 1993;14:307–314.
10. Berenguer B, Burrows PE, Zurakowski D, Mulliken JB. Sclerotherapy of craniofacial venous malformations: complications and results. *Plast Reconstr Surg*. 1999;104(1):1–11. discussion 12–5.