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Case Report

Ear, Nose and Throat Section

A Rare Case Report of Venous Malformation of the Submandibular Gland-Masson's Haemangioma

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ABSTRACT

Masson's haemangioma was first described by Masson in 1923 as "haemangioendothelioma vegetant intravasculaire". It is common in skin and subcutis which appears as red blue nodule. It also occurs in fingers, trunk, head and neck, heart, larynx and hypopharynx. Masson's haemangioma is a rare venous malformation. Treatment is complete surgical excision. It is rarely known to recur. It is a locally occuring lesion with no reports of metastasis. Venous malformation can be distinguished by their characteristic imaging findings at doppler ultrasound vs Magnetic Resonance Imaging (MRI) and direct phlebography. A 30-year-old male presented with swelling in the left submandibular region for one month. On examination a cystic swelling was present in left submandibular region. Ultrasound Sonography test (USG) neck with doppler revealed multilocular cystic swelling with low level internal echoes in left submandibular region suggestive of low flow venolymphatic malformation. The mass was surgically excised and sent for histopathological examination and reported as masson's haemangioma. Masson's haemangioma is a rare venous malformation. Appropriate history, clinical examination and investigation leads to the correct diagnosis and treatment. Incomplete removal of the mass leads to recurrence. The patient was still on follow-up and no recurrence was noted.

Keywords: Cystic swelling, Intravenous atypical vascular proliferation, Salivary gland, Submandibular swelling, Venolymphatic malformation

CASE REPORT

A 30-year-old male presented to the Ear, Nose and Throat Out-Patient Department (ENT-OPD) with complaints of swelling on the left side of neck region (submandibular region) for the duration of one month. The swelling was insidious in onset, gradually progressing and attained the current size. No history of associated pain, trauma, fever, dental caries. Patient had no similar complaints and no history of any surgery in the past.

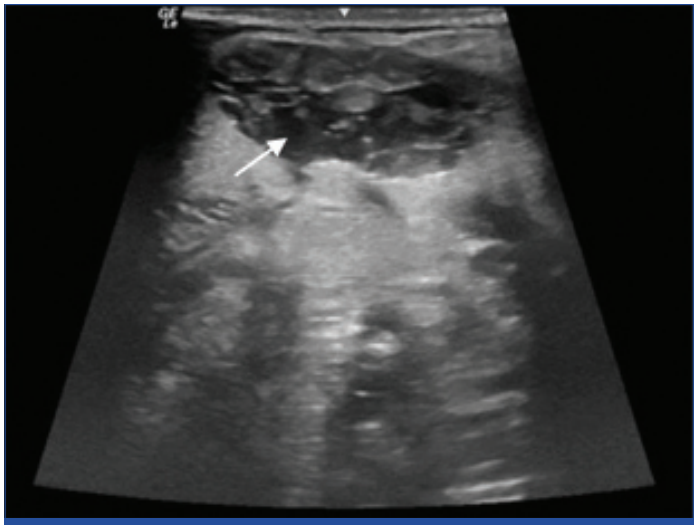
On general examination vitals were found to be normal. And on local examination of ear, nose and throat were also found to be normal. On Local examination of the neck a cystic swelling of size approximately measuring 3×2 cm present in the left submandibular region [Table/Fig-1] was evident. Swelling was ovoid in shape, cystic in consistency, compressible, no warmth, no tenderness, transillumination- negative. On palpation, swelling was movable not fixed to the superficial skin and no other palpable lymph nodes in the neck. Further investigations were done. The Ultrasound of neck (USG) showed a multilocular cystic lesion measuring 3.6×1.9 cm with low level internal echoes in left submandibular region [Table/Fig-2]. The lesion is related to submandibular gland medially, sternocleidomastoid muscle laterally and carotid space posteriorly. The USG of neck with doppler showed submandibular vein communicating with the lesion which was noted draining in to left internal jugular vein and reported as feature suggestive of low flow venolymphatic malformation [Table/Fig-3]. A provisional diagnosis of venous malformation of submandibular gland was made and treatment planned included surgical excision. The patients was still on follow-up and no recurrence was noted.

Operative procedure: Under general anaesthesia with orotracheal intubation patient was in supine position with head extension and head turned to right side. Under sterile aseptic precaution, skin incision marked using sterile maker. A horizontal incision placed two finger breadths below the left angle of mandible extending from midline to the anterior border of sternocleidomastoid muscle. Then flap elevated along subplatysmal plane and fascia covering the gland was dissected and elevated. Cystic swelling which was

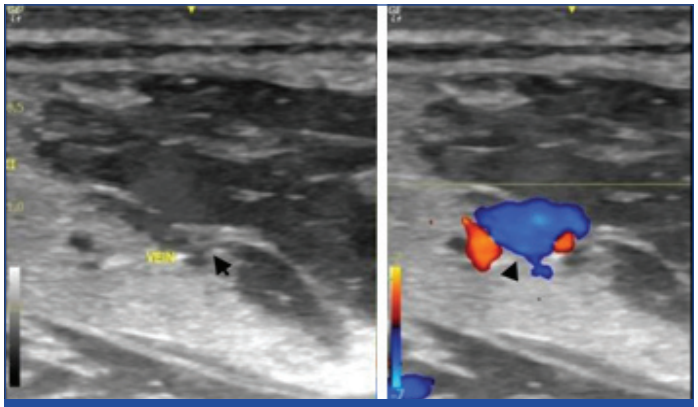
[Table/Fig-1]: Preoperative picture of the left submandibular gland.

irregular in shape was identified, attached to the submandibular gland tissue [Table/Fig-4]. Cystic swelling seen in close proximity with the left submandibular gland with the feeding vessel facial artery was ligated and cut. The marginal mandibular nerve was identified and preserved. Tendon of digastric, mylohyoid muscle and hypoglossal nerve were identified and preserved. The cystic swelling was delineated from the surrounding structure and sent for histopathological examination. Haemostasis obtained, suction drain placed and wound suturing was done. Sterile dressing applied and postoperative period was uneventful and healthy scar present.

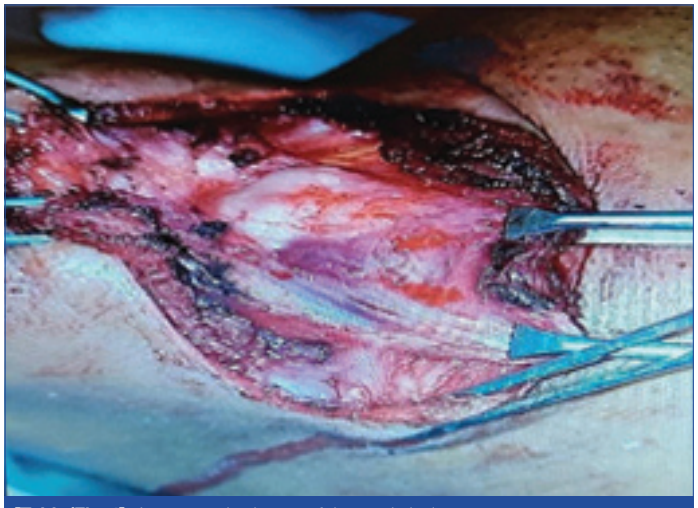
Histopathology: On histopathological examination it was diagnosed as organising thrombus with reactive papillary endothelial hyperplasia (masson's haemangioma). A single lymph node was identified which shows reactive changes. Histopathology sections showed endothelial proliferation within the vessel, multiple papillary projections, fibrin thrombus within, no evidence of necrosis [Table/Fig-5].



[Table/Fig-2]: Ultrasound image of left submandibular gland showing multilocular cystic lesion with low level internal echoes.



[Table/Fig-3]: Ultrasound with doppler image showing submandibular vein communicating with the lesion.

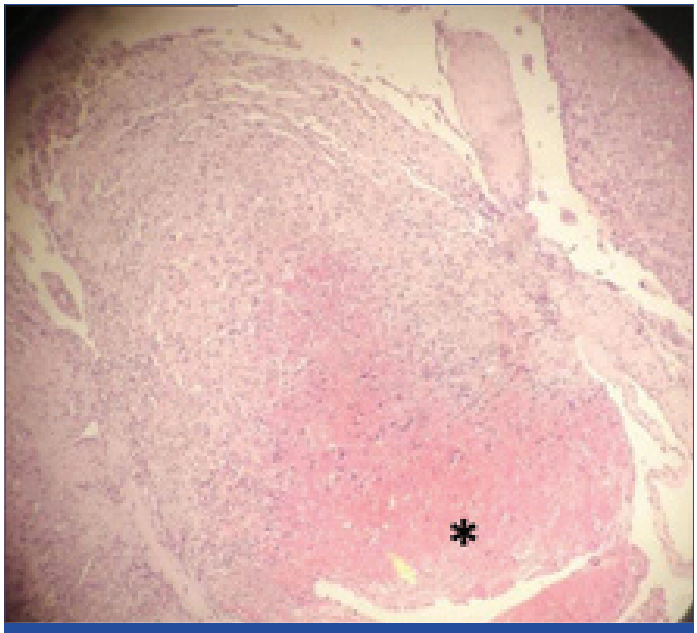


[Table/Fig-4]: Intraoperative image of the cystic lesion.

DISCUSSION

Masson's haemangioma is a peculiar rare benign intravascular lesion, rather than a neoplasm. It is an unusual form of thrombus organisation with endothelial cell proliferation. It has two forms-primary form is the pure form occurs in normal vessels and is found in extremities (fingers) and head and neck and secondary form is the mixed form occurs in pre-existing vascular lesions like haemangioma, arteriovenous malformation, pyogenic granuloma [1]. Sometimes masson's present as an extravascular form (uncommon type) seen in trunk.

Masson's haemangioma is considered as a reactive endothelial proliferation following traumatic vascular stasis [2]. Venous malformations are the most common vascular malformations. Vascular malformations are more commonly found in adults. Venous



[Table/Fig-5]: Histopathological picture of Masson's haemangioma showing fibrin thrombus (*) under low power magnification. (H&E magnification 10X).

malformation is an abnormal venous network with slow blood flow. The pathogenesis of this lesion is variable. It was said that endothelial proliferation leads to the thrombus formation. Another theory was proposed saying that the lesion developed in a pre-existing thrombus. It has various sites of occurence including the skull, thyroid, head and neck, breast, oral cavity, trunk and extremities [3]. Other names for Masson's haemangioma are Masson's intravascular haemangioendothelioma, Intravascular Papillary Endothelial Hyperplasia (IPEH) or reactive papillary endothelial hyperplasia, Masson's pseudoangiosarcoma, Masson's lesions, intravenous atypical vascular proliferation, intravascular angiomatosis [4]. Similar to our study, in study conducted by Shrestha KK et al., showed dilated blood vessels with multiple papillary projections with thrombus in some blood vessels lumen [5]. In Guledgud MV et al., study histopathological examination shows dilated vein lined by benign endothelial cells showing papillary processes with fibrin deposits and areas of haemorrhage were seen [6].

Increased levels of fibroblast growth factor may predispose to endothelial hyperplasia. Although benign, this lesion is clinically important because it presents as a mass lesion histologically mistaken for angiosarcoma and it tends to recur if incompletely resected [7,8]. Ki-67(MIB-1) is a large nuclear protein detected on immunohistochemistry. CD 105(endoglin) is a transmembrane protein highly expressed on human vascular endothelial cells in immunohistochemistry. Masson's haemangioma develops by exuberant recanalisation of a thrombus, secondary to trauma and may be superimposed on pyogenic granuloma or cavernous haemangioma. It rarely recurs after complete excision. Incidence of venous malformation is 1:5,000 to 1:10,000. It accounts 40% of most common benign vascular lesions. TIE-1(tyrosine kinase with immunoglobuline like and EGF like domains). Receptor mutation has been found in some patients with venous malformation syndrome (blue rubber bleb nevus syndrome). According to Hamburg classification by Prof. Stefan Belov, he classified vascular defect into arterial, venous, arterio- venous, lymphatic and combined vascular. Based on anatomical forms it is classified into two subcategory extra truncular and truncular [9]. Most common high flow malformation is arteriovenous malformation and arteriovenous fistulae. Low flow malformations are lymphatic malformation, venous malformation, glomuvenous malformation and non-shunting mixed lesion [10].

Histologically the following are required for definitive diagnosis of Masson's haemangioma:

- Endothelial proliferation within the vessel.

- Multiple papillary projections.
- Fibrin thrombus within.
- No evidence of necrosis.

These characteristic features help in differentiating Masson's haemangioma from angiosarcoma with which it is often confused and from other soft tissue tumours [11]. Clinically differential diagnosis of this lesion is venous malformation of submandibular gland, cystic haemangioma, submandibular lymphadenopathy, tumours of the submandibular gland (benign vs malignant), thrombosed vein. Radiographic differential diagnosis are lymphangioma, haematoma, kaposi sarcoma, haemangioendothelioma, pyogenic granuloma, angiosarcoma of salivary gland tumour, submandibular mucocele [12]. Complete surgical excision is the treatment of choice for Masson's haemangioma. It is a locally occuring lesion with no reports of metastasis and recurrence is rare. Venous malformation can be distinguished based on their characteristic imaging findings in Doppler ultrasound vs MRI and direct phlebography [13].

CONCLUSION(S)

Masson's haemangioma is a rare venous malformation of the submandibular gland. Appropriate history, clinical examination and investigation leads to the correct diagnosis and treatment. Complete surgical excision is the treatment of choice for Masson's haemangioma. Incomplete removal of the mass leads to recurrence.

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