Case Report

Venolymphatic vascular malformation of the parotid gland extending into the parapharyngeal space: A rare presentation

Yasmeen Khatib, Madhura Dande, Richa D Patel, Vinod Gite[1]

Departments of Pathology and [1]ENT, Dr. R. N. Cooper Hospital, Mumbai, Maharashtra, India

Abstract

Vascular malformations (VMs) are structural malformations of vascular development causing soft tissue abnormality with functional and esthetic impairment. They are named by their predominant vessel type as arterial, venous, lymphatic or mixed types. VMs of the parotid gland are extremely rare and constitute a distinct entity of parotid pathology that requires specific diagnostic tools and management. Till 2013, only fifty cases of VMs of the parotid have been described in the literature. We present a case of a venolymphatic malformation of the parotid gland extending into the parapharyngeal space in a 21-year-old male who presented with a swelling on the left side of the face extending into the neck. Diagnosis was suggested by ultrasonography and computed tomography scan and was confirmed by magnetic resonance imaging examination. Complete surgical excision of the lesion was done with a favorable outcome. Diagnosis was confirmed based on histopathology and immunohistochemical studies.

Key Words: Parotid, vascular anomaly, venolymphatic malformation

Address for correspondence:
Dr. Richa D Patel, Department of Pathology, Dr. R. N. Cooper Hospital, Mumbai - 400 056, Maharashtra, India. E-mail: dr.richa. 13@gmail.com
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INTRODUCTION

Venolymphatic malformations (VLMs) are tumor-like lesions combining dysplastic lymphatic and venous vessel structures.[1] The International Society for the Study of Vascular Anomalies has divided vascular lesions into hemangiomas and vascular malformations (VMs).[2] Hemangiomas are characterized by rapid growth in neonatal life, endothelial proliferation and spontaneous slow regression while VMs are present at birth have normal rate of endothelial turnover and continue to grow throughout the patients' lifetime.[3] They are also classified as slow-flow (capillary, venous, lymphatic) and fast-flow (arterial, arteriovenous [AV]) lesions.[3] These two types of lesions have different clinical behavior and require different diagnostic and therapeutic strategies. The present case of venolymphatic malformation of the parotid gland extending into the parapharyngeal space was asymptomatic and presented for cosmetic purpose. Magnetic resonance imaging (MRI) confirmed the diagnosis and described the exact location and extent of the lesion which was successfully treated by surgery.

CASE REPORT

A 21-year-old male patient came with a complaint of swelling over the left side of the face extending into the neck since birth. The swelling was initially small and gradually increased in size for the last 4–5 years, reaching up to the present size

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of 8 cm × 5 cm. The swelling was not associated with pain or any other symptom. There was a history of operative procedure done at the age of 2 years, the details of which were not available. There was no other positive family history or medical history. On clinical examination, the swelling extended from the parotid region into the neck [Figure 1a]. It was soft, nonpulsatile, nontender, nonfluctuant, compressible with smooth borders. There was no evidence of thrill or bruit over the swelling. There was no associated lymphadenopathy.

Ultrasonography (USG) with color Doppler and computed tomography (CT) was suggestive of a low-flow venous malformation occupying the lower pole of the superficial lobe of parotid. The part of retromandibular vein was seen traversing through the mass. A tiny phlebolith was also seen.

Multiplanar, multisequence MRI of neck was performed which demonstrated a predominantly T2-hyperintense, T1-isointense mass in the left parotid region with extension into the deep ipsilateral parapharyngeal space [Figure 1b-d]. No invasion of the adjacent tissue was seen. On post contrast examination, there was minimal overall contrast enhancement. There was a large draining vein at the lateral aspect of the lesion [Figure 1c]. There was a hypointense T2-focal area in the lesion corresponding to the focal calcification on CT. Considering the involvement of multiple compartments, abnormal prominent draining vein and focal calcification, a diagnosis of venolymphatic malformation was made.

In view of the above investigations, the patient was taken up for surgery and superficial parotidectomy was performed along with the excision of the lesion from the parapharyngeal space [Figure 2a]. On gross examination, a partially encapsulated mass was received measuring 5.5 cm × 4 cm × 3 cm. Externally, it was congested. The cut surface was spongy, reddish brown and grossly, many vascular channels of varying sizes were seen [Figure 2b].

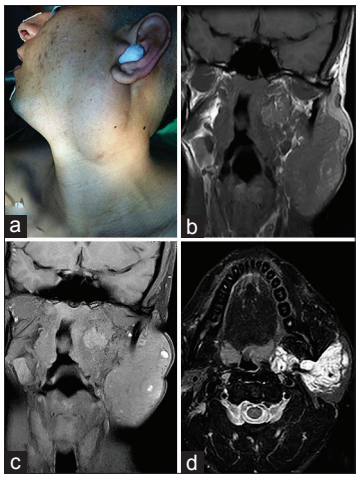

Figure 1: (a) Clinical photograph showing swelling over preauricular region extending into the neck, (b) pre-contrast magnetic resonance imaging showing the lesion, (c) post-contrast magnetic resonance imaging showing mild post-contrast enhancement of the lesion with a large draining vein at the lateral aspect, (d) T2-weighted magnetic resonance imaging showing the predominantly T2-bright lesion in the superior part of the parotid

Microscopically, the lesion was composed of numerous vascular channels of varying sizes, many of which were dilated [Figure 3a-d]. Some of the vessels were lined by single flat layer of endothelium suggestive of venules and veins. Other small to medium sized vascular channels were lined by flattened endothelial cells. These had patchy smooth muscles in the vessel wall and were filled with eosinophilic proteinaceous material and were suggestive of lymphatic channels. The adjacent tissue showed fatty stroma and presence of lymphocytes, lymphoid follicles and smooth muscle [Figure 3b and d]. The lesion was seen infiltrating into salivary gland tissue. On immunohistochemistry (IHC), the vascular endothelial cells expressed CD31 [Figure 4a] while the lymphatics were highlighted by D2-40 [Figure 4b]. Based on the histology and the IHC findings, a diagnosis of venolymphatic malformation was made.

Postoperatively, the patient was followed up and did not have any complaints pertaining to the swelling.

DISCUSSION

VLMs are slow-flow lesions composed of both venous and lymphatic elements. They were previously called lymphangiohemangiomas or hemangiolymphangiomas.[3] Nearly 50% VMs occur in the head and neck region with lip, oral cavity, tongue, masseter, temporalis muscle and

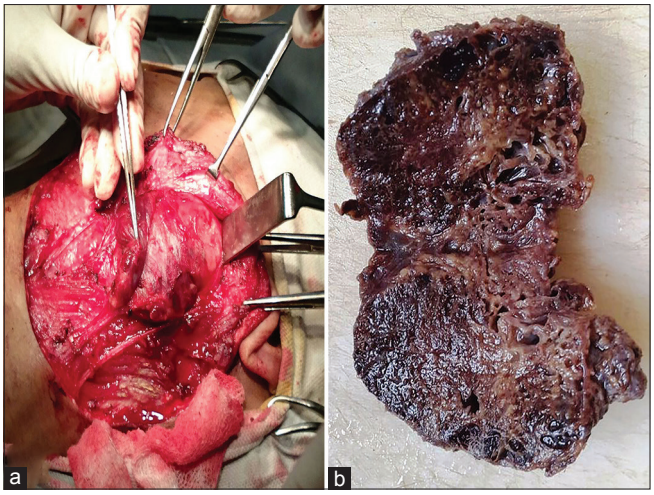

Figure 2: (a) Photograph showing intraoperative removal of the mass from parotid, (b) gross photograph showing spongy cystic reddish brown mass

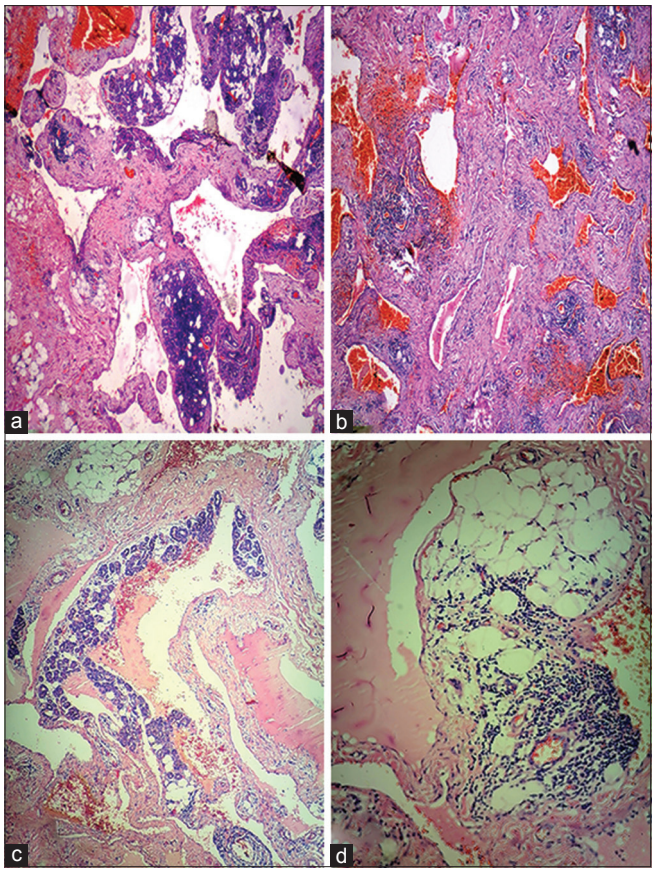

Figure 3: (a) Photomicrograph showing salivary gland along with vascular channels (H&E stain, ×40), (b) photomicrograph showing venous and lymphatic channels with lymphoid follicles within the stroma (H&E stain, ×40), (c) photomicrograph showing endothelial lined lymphatic and venous channels. Stroma shows adipose and lymphoid tissue (H&E stain, ×40), (d) photomicrograph showing presence of lymph inside the lymphatic channels along with acini of parotid gland (H&E stain, ×100)

airway mucosa being the common sites of involvement.[4] They have also been reported rarely in the mediastinum, lower extremity, intestine and heart.[5] They are present at birth and enlarge as the patient grows and never involute. These lesions are the result of an embryonic abnormality of the vascular system. VMs are caused by a disturbance in the late stages of angiogenesis (truncal stage) and result in the persistence of AV anastomosis present during embryonic life. Venous malformations are mediated by germline or somatic mutations in the TEK (chromosome 9p) gene which encodes the endothelial cell tyrosine kinase receptor TIE2. The increased phosphorylation of TIE2 leads to the uncoupling between endothelial cells and normal recruitment of smooth muscle cells.[6] Their presence has also been linked to the errors in the receptor tyrosine kinase TIE2 gene. Hormonal influences, infection, trauma or surgery caused by birth control pills, puberty and pregnancy may cause VM to enlarge and become more symptomatic. Parotid gland is a very rare site of presentation.[7,8]

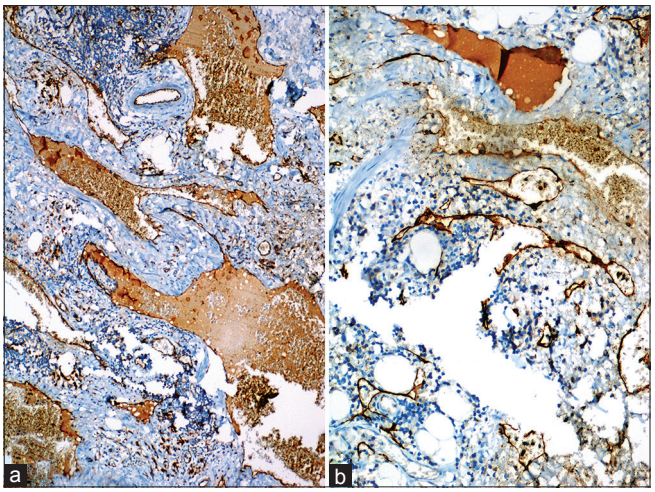

Figure 4: (a) CD31 positivity in venous and lymphatic channels (IHC stain, ×40), (b) D2-40 (podoplanin) positivity in lymphatic channels (IHC stain, ×40)

Achache M found 10 cases of VMs of parotid in 614 parotidectomy procedures.[9] Beahrs *et al*.[10] have reported only 0.5% and Byars *et al*.[11] have reported 0.6% in their study of a large no of parotidectomy specimens. There was a clear female preponderance with female:male ratio of 9:1. Diagnosis was made usually in the fourth decade of life though age ranged from 19 to 54 years. Two cases showed extension into the parapharyngeal space as the present case. Clinical presentation was of a mass present since birth or childhood with a slow growth more after puberty as in the present case. These VMs were seen as slow-growing soft mobile masses with no facial nerve palsy. Most patients develop few or no symptoms. Symptoms when present were related to either long-standing nature of the tumor (dyspnea, dysphagia with raised hemivelum) or to micro-thrombotic events (pain causing phleboliths). Nearly 90% cases were treated with surgery. As they are rare, VLM of parotid are not usually considered in the differential diagnosis and are mistaken for common cystic parotid lesions, pleomorphic adenoma and Warthin's tumor. Fine-needle aspiration cytology shows hemorrhagic aspirate and is noncontributory.

USG and color Doppler imaging has proven to be helpful in the initial diagnosis and shows a slow flow lesion, presence of phleboliths and vein traversing through the lesion.[3] Although CT scan with contrast can be done, MRI has proven to be the mainstay in the diagnosis and to correctly delineate the full extent of the lesion which is needed to plan the treatment. A specific aspect of venous malformation is the presence of phleboliths with a hyperintense T2-weighted signal on MRI.[9] Diagnosis is confirmed on histopathological examination which shows the presence of dilated ectatic venules lined by flat endothelial

lining and lymphatic vessels showing patchy smooth muscle in the wall. Vessels may show congestion, thrombosis or calcifications (phleboliths).[9] Lymphatic vessels may show proteinaceous material. Stroma shows the presence of adipose tissue, lymphoid follicles and smooth muscle as was seen in the present case. IHC shows positivity to vascular markers such as CD31 and CD34 while lymphatics are specifically stained by D2-40 or podoplanin.[1,5]

Treatment of VMs depends on the size, location, symptoms and proximity to the vital structures. Moreover, as VM have poorly defined borders and tendency to infiltrate normal tissues, they require calculated treatment decisions in the effort to preserve the surrounding architecture. Treatment of VLM of parotid is usually surgical resection with sparing of the facial nerve. Total resection is required to prevent recurrence. Other treatment modalities for VM include sclerosing agents such as ethanol, sodium tetradecyl sulfate and bleomycin which are injected under imaging guidance or even intraoperatively. Laser therapy using neodymium: Yttrium-aluminum-garnet laser is also used in some cases.[12]

CONCLUSION

VLMs of the parotid gland are rare lesions which are present from birth but show a spurt in growth during puberty or due to hormonal changes. They are usually asymptomatic. MRI is necessary to confirm the diagnosis, know the extent of lesion and to plan the treatment. Histopathology shows the presence of both veins and lymphatics which can be confirmed by IHC studies. The treatment of choice is surgical removal in majority of the cases.

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Conflicts of interest

There are no conflicts of interest.

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