Parotid Lymphangioma: A Rare Diagnostic Puzzle in Head and Neck Pathology

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ABSTRACT

Introduction

Lymphangioma is a rare, benign lymphatic malformation, most commonly seen in children and infrequently involving the parotid gland in adults. Its presentation often mimics other parotid masses, posing a diagnostic challenge. Diagnosis is typically based on clinical examination, imaging, and confirmed by histopathology.

Case Report

A patient presented with a congenital swelling on the left side of the neck. Clinical examination revealed a soft, lobulated, mobile, non-tender mass measuring approximately 5×5 cm in the infra-auricular region extending to the mandibular area. Fine-needle aspiration cytology (FNAC) suggested a benign cystic lesion. Surgical excision was performed via a modified Blair incision under general anesthesia. The mass was sent for histopathological examination, which confirmed lymphangioma.The postoperative period was uneventful, and the patient was discharged in stable condition.

Discussion

Though rare, parotid lymphangioma should be considered in the differential diagnosis of parotid region masses in adults. A combination of clinical assessment, imaging, and histopathological evaluation is essential for accurate diagnosis and appropriate management.

Keywords

Lymphangioma; Parotid Neoplasms; Salivary Gland Neoplasms; Head and Neck Neoplasms

Lymphangiomas are benign, congenital malformations of the lymphatic system, typically occurring in early childhood.[1] The incidence among children is 1.2–2.8 per 100,000.[2]

These lesions are rare in adults, particularly when located in the parotid gland, which is an uncommon site for lymphangiomas. Parotid lymphangiomas often present as cystic masses that can cause facial swelling or asymmetry.

Lymphangiomas are categorized into three types: capillary, cavernous, and cystic. The latter has the potential to infiltrate the surrounding tissue leading to a problematic surgical removal.[3]

Case Report

A 17 year-old male patient presented to our OPD with a history of swelling in left parotid region since birth. The swelling was insidious in onset, non- progressive and painless. There was no dysphagia or respiratory distress. Patient did not have any comorbidities.

Examination – On inspection a soft, bosselated mass seen in left parotid region ~ 5cm × 5 cm in size (Fig. 1).

On palpation, mass was multilobulated, soft, non-tender & mobile with well-defined borders. Skin over the mass was mobile .

Origin of the mass could not be ascertained on history and clinical examinations.

USG neck showed cystic lesion with fine homogenous echoes in left upper cervical region (Lymphangioma / low flow venous malformation).

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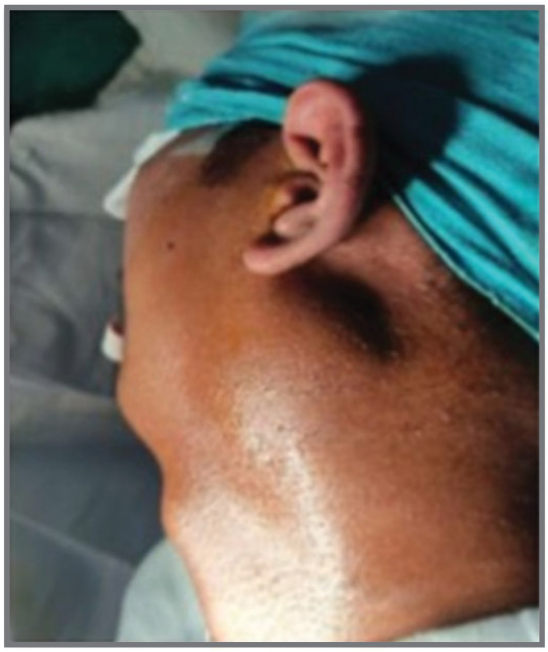

Fig. 1. Pre operative image showing a soft, multilobulated swelling in parotid region

MRI face and neck sections revealed a thin walled lobulated multiloculated cystic lesion involving posterior inferior part of superficial and deep lobes of left parotid gland showing slight intra parotid extension inferiorly in adjacent superficial inter facial planes-likely benign (lymphoepithelial cyst/lymphangioma) (Fig. 2.1, 2.2).

FNAC was suggestive of benign cystic lesion.

A provisional diagnosis of Lymphangioma of left parotid gland was made and surgical excision planned. We further planned a surgical procedure for excision of the lymphangioma.

An informed consent was obtained and the patient was taken under general anaesthesia. Under all aseptic precautions, the usual painting and draping was carried out and incision line was marked. A modified Blair's incision was taken in preauricular and cervical skin crease, superficial cervicofacial flap was raised, anterior border of sternocleidomastoid identified followed by exposure of posterior belly of digastric. The cyst was enveloped around internal carotid artery and jugular vein .Cyst was excised by finger and blunt dissection after removal from surrounding structures. (Figs. 3.1, 3.2).

Gross section revealed grayish white pale yellow soft tissue piece measuring 10*4*2 cm. Histopathology revealed large dilated cystic areas lined with flattened epithelium with intervening fibro collagenous tissue displaying numerous cluster of lymphoid tissue along with lymphoid aggregates. Parotid serous acini are seen at places adjacent to these dilated lymphoid spaces. (Fig. 4.1, 4.2).

Thus, a definitive diagnosis of Lymphangioma of left parotid gland was made. Intra-operative and post-operative periods were uneventful and no recurrence was seen (Fig 5).

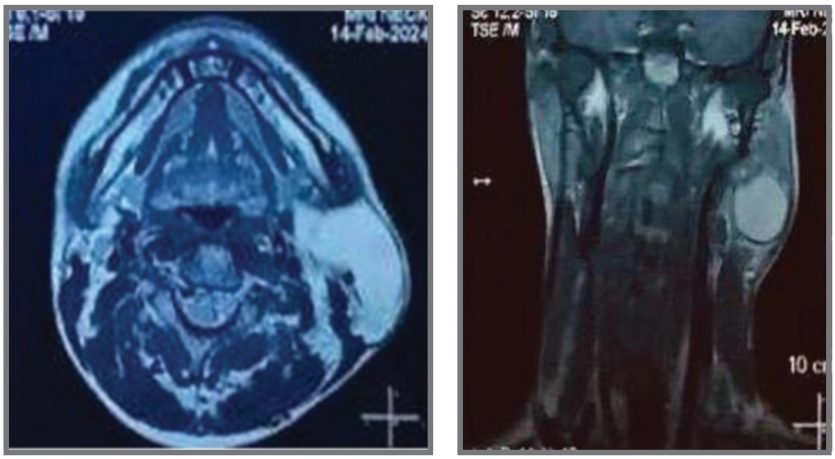

Fig. 2.1, 2.2. MRI section ,axial and coronal view showing a lobulated swelling in parotid region involving superficial and deep lobe with slight inferior extention to adjacent superficial inter facial plane

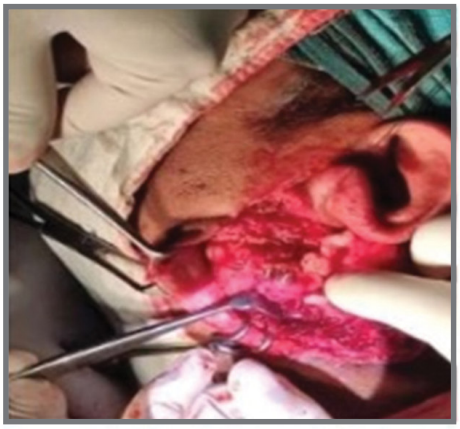
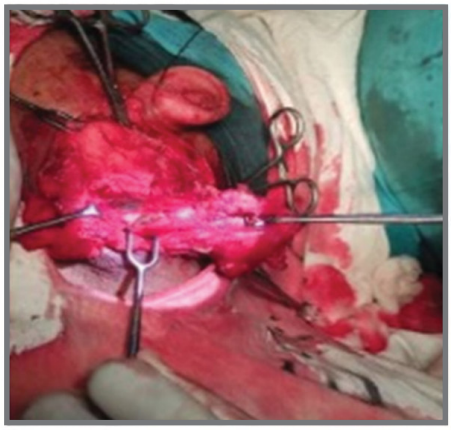

Fig. 3.1, 3.2. Intraoperative image showing excision of swelling

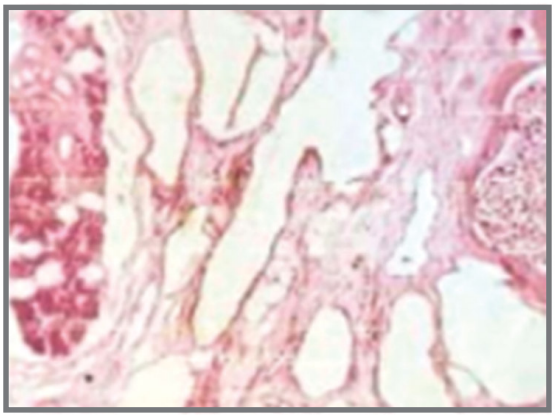

Fig. 4.1. Histopathology image showing large caliber lymph vascular spaces

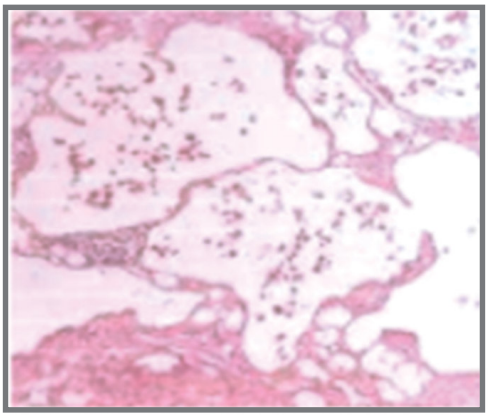

Fig. 4.2. Histopathology showing cluster of lymphoid aggregates

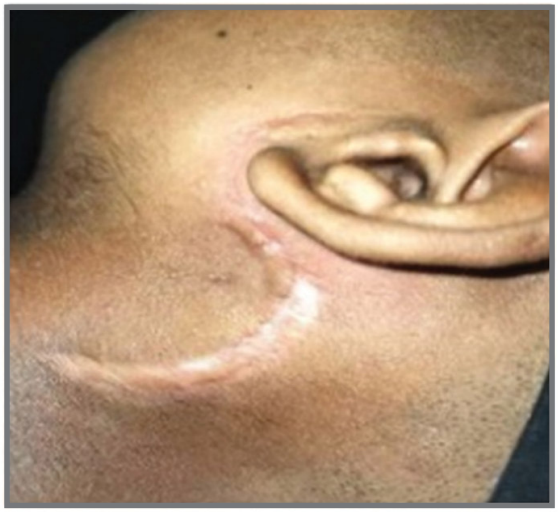

Fig. 5. Post operative scar at 1 year of follow up

Discussion

The origin of lymphangiomas remains controversial. Theories include lymphangiomas as true neoplasms, hamartomas, or congenital dysplasias of the lymphatics. Sidle et al. conducted a study in 2005 using immunohistochemistry to assess the expression of the angiogenic inducer vascular endothelial growth factor (VEGF) and the angiogenic inhibitor pigment epithelium-derived factor (PEDF).[4] They conclude that lymphangiomas exhibit tumor-like pathogenesis due to the high expression of angiogenic inducers compared with the low expression of inhibitors. However, most physicians favor the theory that dysplastic lymphatic tissue has been sequestered in a target tissue space or organ during fetal development. The majority of lymphangiomas arise from

parts of lymphatic sacs that fail to establish connections. Acquired lymphangiomas usually develop after surgery, trauma, infection, or chronic infection.

On the basis of microscopic characteristics, three types of lymphangiomas are described: capillary, cavernous, and cystic. Capillary lymphangioma consists of small, capillary-sized lymphatic vessels. Cavernous lymphangioma comprises dilated lymphatic channels. Cystic lymphangioma is a large, macrocystic lymphangioma filled with straw-colored, protein-rich fluid. Cystic types have the potential for extensive infiltration of surrounding tissues and lead to surgical difficulties. Cystic lymphangiomas are also classified into microcystic, macrocystic, and mixed subtypes, according to the size of their cysts.[5]

Lymphangiomas, being congenital lesions, are rare in adults, and primary involvement of the parotid gland remains an uncommon occurrence. The differential diagnosis of a cystic parotid mass includes Warthin's tumor, benign lymphoepithelial lesions, branchial cleft cysts, chronic sialadenitis, cystic pleomorphic adenoma, and cystic low-grade mucoepidermoid carcinoma.[6]

On ultrasonography (USG), parotid lymphangioma appears as fine homogeneous echoes in the left upper cervical region. The differential for cystic lesions in the parotid includes Warthin's tumor, sialocele, first branchial cleft cyst, lymphoepithelial cyst, necrotic lymph nodes, and infected lymph nodes.

MRI of the face and neck reveals a thin-walled lobulated multiloculated cystic lesion involving the posterior inferior part of the superficial and deep lobes of the left parotid gland, showing slight intra-parotid extension inferiorly into adjacent superficial interfascial planes—likely benign (lymphoepithelial cyst/lymphangioma). Fine needle aspiration cytology (FNAC) aspirates are suggestive of a benign cystic lesion.

The definitive diagnosis is made on postoperative histopathology, which reveals large dilated cystic areas lined with flattened epithelium with intervening fibro-collagenous tissue displaying numerous clusters of lymphoid tissue along with lymphoid aggregates. Parotid serous acini are seen at places adjacent to these dilated lymphoid spaces.

Primary management is surgical, which includes enucleation, superficial, or total parotidectomy. Medical therapy in the form of sclerotherapy can be given in cases where surgery is difficult or the lesion is infiltrative. Conservative management with observation is rarely followed. Decompression of the cyst with aspiration is temporary. However, in our case, we plan a surgical procedure for excision of the lymphangioma via a modified Blair's incision.

Conclusion

Lymphangiomas of the parotid gland in adults are rare, and their presentation can pose diagnostic challenges due to their similarity with other cystic lesions. In this case, a 17-year-old male presented with a congenital, non-progressive swelling in the parotid region, which was ultimately diagnosed through imaging as a cystic lymphangioma. Surgical excision remains the mainstay of treatment, with complete resection offering both diagnostic confirmation and therapeutic relief. Early recognition and proper imaging are crucial in guiding management and achieving favorable outcomes, particularly in rare anatomical locations such as the parotid gland.

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