

Cystic lymphangioma of parotid in adult - A rare entity

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ABSTRACT

Cystic lymphangioma of parotid in an adult is a rare occurrence. Very few cases of cystic lymphangioma of parotid have been reported in the literature. Here, we report a case of parotid cystic lymphangioma in a 23-year-old female. Ultrasonography showed a well-defined multicystic lesion with thick internal septations in the parotid region. Fine needle aspiration showed mature lymphocytes only. The diagnosis of cystic lymphangioma of parotid was confined to histopathology. Surgery is the mainstay of treatment; however, medical management in the form of sclerotherapy may be required in infiltrative lesions or when surgical management is not possible. Although rare, cystic parotid lymphangioma should be kept as a differential in cystic lesions of the parotid gland in children as well as adults.

Key words: *Cystic lymphangioma, Parotid gland, Parotidectomy*

Lymphangiomas probably are the benign congenital malformation of the lymphatic channels. They commonly occur before the age of 2 years, and the occurrence of lymphangioma among adults is rare. Both the sexes are affected equally. They can develop anywhere in the body, but the cervicofacial region is the commonly affected [1]. Parotid cystic lymphangioma in an adult is rare owing to its location in adult age group. Here, we report a case of parotid cystic lymphangioma in adult female, which was treated with surgical excision.

CASE REPORT

A 23-year-old female presented to ENT outpatient department with a complaint of swelling on the right parotid region for past 1 month. Swelling was gradually increasing in size and not associated with any other complaints such as fever and pain. The clinical examination showed a 3 cm×2 cm, painless, non-tender, and non-pulsatile mobile parotid mass. The mass was not fixed to deeper structures or skin and no associated symptoms were found with the parotid mass. Systemic examination was unremarkable.

Ultrasonography (USG) showed a well-defined lesion measuring 2.7 cm×2.1 cm×1.4 cm with thick internal septations in the right angle of mandible (Fig. 1). No definite mural nodularity or internal vascularity was seen. The lesion clearly abuts the right parotid gland. The adjacent parotid gland was normal in echotexture. A possibility of the infected brachial cyst was given on USG. The fine needle aspiration cytology (FNAC) of the parotid mass yielded a straw-colored fluid. The cytological smears prepared from aspirated fluid showed mature lymphocytes only (Fig. 2). No squamous cells were seen. Cytological diagnosis of

benign cyst of parotid was signed out. The right parotid swelling was excised and sent for histopathological examination (HPE). A soft tissue piece with variegated appearance was received. HPE showed large caliber lymphovascular spaces with smooth muscles in the wall (Fig. 3). Aggregation of a mature lymphoid cell was noted on the wall. Benign salivary acini and reactive nodal tissue were also noted. HPE diagnosis of vasoformative lesion favoring cystic lymphangioma was signed out.

DISCUSSION

The origin of lymphangioma has been controversial. Theories include lymphangiomas as true neoplasms, hamartomas, or congenital dysplasias of the lymphatics. Sidle *et al.* conducted a study in 2005 using immunohistochemistry expression for angiogenic inducer vascular endothelial growth factor (VEGF) and angiogenic inhibitor pigment epithelium-derived factor (PEDF) [2]. They concluded that lymphangiomas exhibit tumor-like pathogenesis owing to the high expression of angiogenic inducers compared with the low expression of inhibitors. Recurrence may be influenced by this imbalance of angiogenic mediators. Further, research with antiangiogenic therapy using agents such as PEDF analogs or anti-VEGF receptor antibodies is indicated [1].

However, most of the physicians favor the theory that dysplastic lymphatic tissue is sequestered in a target tissue space or organ during fetal development. During the 8th week of gestation, six lymphatic sacs develop - two jugular, two iliac, one root of mesentery, and one abdominal aorta. Later, communications are established between various lymphatic sacs and venous

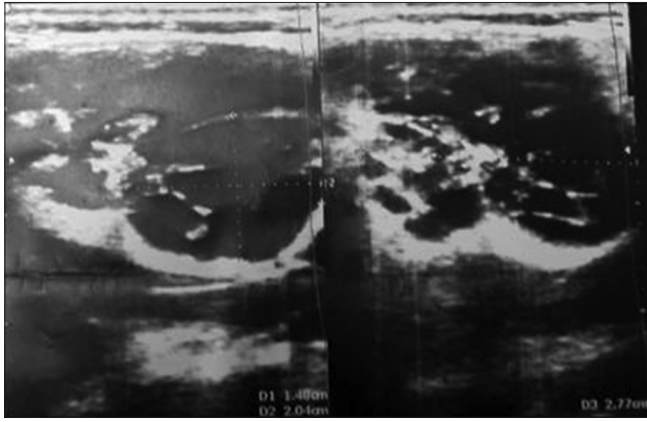


Figure 1: Ultrasonography showing well-defined lesion measuring 2.7 cm×2.1 cm×1.4 cm with thick internal septations in the right angle of mandible

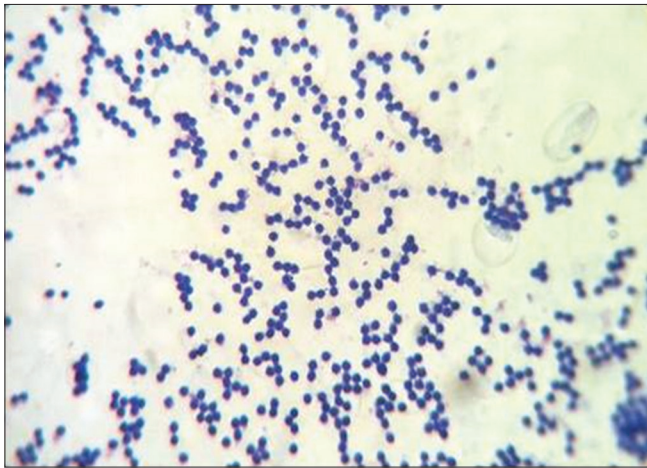


Figure 2: Cytological smears prepared from fluid aspirated from parotid swelling showing mature lymphocytes (Giemsa, ×40)

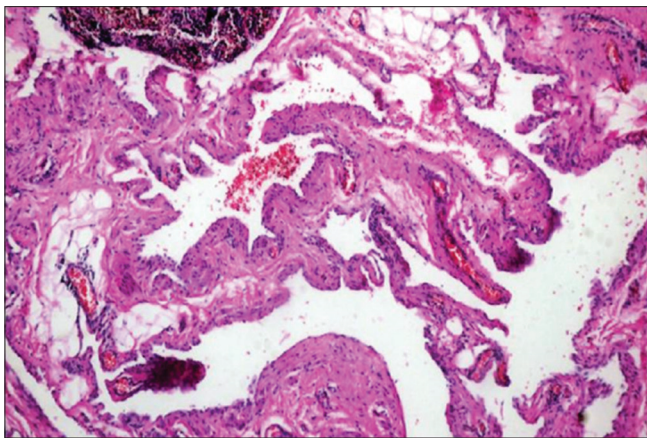


Figure 3: Histopathological examination showing large caliber lymphovascular spaces with smooth muscles in wall (H and E, ×40)

system [1]. Majority of the lymphangiomas arise from the parts of lymphatic sacs which fail to establish connections. Acquired lymphangiomas usually develop after surgery, trauma, infection, or chronic infection [1].

On the basis of microscopic characteristics, three types of lymphangiomas are described - capillary, cavernous, and cystic [3]. Capillary lymphangioma is composed of small,

capillary-sized lymphatic vessels. Cavernous is composed of dilated lymphatic channels. Cystic lymphangioma is 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 [3]. Microcystic lymphangiomas are composed of cysts, each of which measures <math><2\text{ cm}^3</math> while in macrocystic lymphangiomas, cysts measure >math>2\text{ cm}^3</math> in volume. Lymphangiomas of the mixed type contain both microcystic and macrocystic components.

Lymphangiomas being a congenital lesion are rare in adults and primary involvement of parotid gland is still an uncommon occurrence. Parotid masses and swellings are due to infection, inflammation, and neoplasm or congenital. The differential diagnosis of cystic parotid mass is Warthin's tumor, benign lymphoepithelial lesions, branchial cleft cysts, chronic sialadenitis, and cystic pleomorphic adenoma and cystic low-grade mucoepidermoid carcinoma [4].

On USG, parotid lymphangioma appears as hypoechoic, multicystic, multiseptate lesion surrounded by smooth thin or irregular walls. The differential for cystic lesions in parotid is Warthin tumor, sialocele, first branchial cleft cyst, lymphoepithelial cyst, necrotic lymph node(s), and infected lymph node(s) [5]. Computed tomography shows parotid lymphangioma as thin-walled, multicystic, homogenous mass with smooth septa belonging to the parotid gland. Magnetic resonance imaging is the imaging modality of choice for parotid lymphangioma showing hyperintensity T2-weighted images with well-defined margins. FNAC aspirates yellow colored fluid with lymphocytes and occasional histiocytes. Definitive diagnosis is made on post-operative histopathology.

Parotid lymphangioma evolves in one of the three ways - (a) spontaneous regression [6], (b) slow progression, and (c) rapid enlargement due to traumatic hemorrhage or infection [7]. Complication reported with parotid lymphangioma is rupture, infection, nerve compression secondary to hemorrhage within the cyst, and osteolysis of adjacent bone (Gorham-Stout syndrome). Malignant transformation has never been reported in parotid lymphangiomas [1].

Primary management of parotid lymphangioma 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 lesion is infiltrative. Conservative management with observation only is rarely followed. Decompression of cyst with aspiration is temporary and recurrence is ruled after this.

CONCLUSION

Parotid gland is an uncommon site for lymphangioma. Parotid lymphangioma should be kept as one of the differentials in cystic parotid lesions. Pre-operative diagnosis can be made with imaging modalities and FNAC. Surgery is curative in most of the cases; however, it may not be possible to obtain complete surgical

resection so partial excision or medical therapy (sclerotherapy) may be required.

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