CASE REPORT

Large cystic lymphangioma of the parotid gland in the adult

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Abstract Cystic lymphangioma (CL) of the parotid gland is an uncommon benign congenital tumor and very few cases have been reported in adults. We report and discuss the case of a large CL of the parotid gland in a 66-year-old woman. On computed tomography (CT) the tumor appeared as a large, well defined, unilocular cyst of the parotid region. The lesion was surgically removed and the diagnosis was confirmed on postoperative histology.

CL of the parotid gland in adults presents difficult challenges to the surgeon for both differential diagnosis of parotid masses and treatment. Surgery remains the treatment of choice despite its difficulty due to the infiltrating character of the lesion. Sclerotherapy is an alternative in macrocystic type when surgical management is not possible. Despite being benign tumors, CL are attended by a high rate of morbidity.

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1. Introduction

Lymphangiomas are a benign congenital malformation of the lymphatic system. They occur anywhere in the body, but most often in the cervicofacial region, especially in the posterior triangle of the neck. Most of lymphangiomas appear before the age of 2; however their occurrence in an adult remains less common. Both sexes are affected equally.

Very few cases of CL of the parotid gland in the adult have been reported. We describe the case of a woman with a large CL of the parotid region treated by a total surgical resection.

2. Case presentation

A 66-year-old female was admitted to the department of surgery for a large formation of the right cheek region. This mass appeared 8 years ago. Past medical history included arterial hypertension and a surgery for cataract 3 years ago.

On admission the patient looked healthy. Physical examination found in the right parotid area a large, painless, non-tender, non pulsatile mass. Its diameter measured 15 cm (Fig. 1). The mass was not fixed to the deeper structures or to the skin. The skin over the mass, oral cavity and pharyngeal areas were normal. There was no trismus, motor or sensory deficits, lymphadenopathy, regional veins congestion or compressing signs.

Ultrasonography (US) revealed a cystic swelling of the right parotid gland. Moreover it found a thyroid nodule in the left lobe of the gland. Cervicofacial CT showed a large, well
limited, unilocular cystic formation in the parotid area. It measured $150 \times 130$ mm with punctiform calcifications in the periphery and moderately enhanced by contrast in favor of a cystic lymphangioma of the parotid gland (Figs. 2 and 3).

The patient was admitted to the operating room. General anesthesia and orotracheal intubation were performed. The operative approach was a preauricular incision that curved back under the ear lobule and descended along the sternocleidomastoid muscle. The cyst had arisen from the superficial lobe of the parotid gland. A surgical enucleation of the lesion was performed. The external carotid artery was ligated and removed with the mass because it was involved with the lymphangioma (Fig. 4). The operation ended by inserting a sucking drain.

Postoperatively paralysis of the buccal branch of the facial nerve was noticed and collection of fluid at the resection site occurred which was evacuated by syringe aspiration. The patient was discharged from hospital after five days.

Pathology report confirmed the nature of the parotid mass as a cystic lymphangioma.

3. Discussion

The origin of lymphangiomas is not clearly understood. Most authors favor the theory that dysplastic lymphatic tissue is sequestered in a target tissue during the fetal development. In fact, 6 lymphatic sacs (2 jugulars, 2 iliac, 1 root of mesentery and 1 abdominal aorta) develop in the 8th week of gestation. Later, communications are established between lymphatics of various regions and with the venous system. The majority of lymphangiomas may arise from parts of lymph sacs that fail to establish these connections. The disconnected lymphatic structures may dilate and evolve to a cystic formation. Lymphangiomas may also be acquired after surgery or trauma, infection, or chronic inflammation.

Lymphangiomas are divided into three categories: capillary, cavernous, and cystic type. Cystic types have the potential for extensive infiltration of surrounding tissues and
lead to surgical difficulties. Therapeutically, they are classified into macrocystic, microcystic or mixed lymphangiomas.

The lesion appears as a soft, compressible, nontender, transilluminant mass of the parotid region. It varies in size from a few millimeters to more than several centimeters as in the present case. The overlying skin is usually normal; however it can be thin, atrophic or bluish. In adults this lesion rarely causes compressing signs.

On ultrasonography, the cystic lymphangioma appears as a hypoechoic, multicystic, multiseptate lesion surrounded by smooth thin or irregular walls. There is no blood flow on Doppler US. On CT, it appears as a thin walled, multicystic, homogeneous mass with smooth septa belonging to the parotid gland. The enhancement occurs in only 50% of cases. CT also evaluates the extent to deep and adjacent structures. Magnetic resonance imaging (MRI) is the imaging method of choice for the diagnosis of lymphangiomas. It shows hyperintensity on T2 weighed images, hypointensity on T1 weighed images, and multiple cysts with well-demarcated margins on T2. Fine-needle aspiration sucks up a yellow or serosanguineous fluid with mature lymphocytes and histiocytes. Definitive diagnosis is usually based on postoperative histology.

Parotid masses can be also due to infectious, neoplastic, vascular or congenital origins such as acute or chronic parotitis, hemangioma, dermoid cyst, lymphoepithelial cyst, adenoma, intraparotid facial nerve neurofibroma or schwannoma, cancers and lymphomas.

Malignant transformation has never been reported. The mass evolves in three ways: (1) Kumar et al. described a case of spontaneous regression of a cystic lymphangioma of the parotid region. (2) A slow progression. (3) A rapid enlargement (spontaneous or traumatic hemorrhage into the cyst or after infection). Infection within the cyst is usually caused by staphylococcus or streptococcus species. Nerve paralysis by compression could be secondary to the hemorrhage within the cyst. Lymphangioma can infiltrate and cause osteolysis of adjacent bones (Gorham Stout syndrome). An ulceration or rupture of the cyst has been reported.

Some surgeons have recommended conservative management with observation only in asymptomatic cysts. Spontaneous regression is possible but rare. Aspiration of the cyst is rarely indicated and it is used as a temporary measure for emergency decompression. Recurrence of the lesion is the rule after this procedure.

Surgical total resection is the treatment of choice, but it is not always possible because of the infiltrating nature of the lesion. Enucleation, superficial or total parotidectomy are the three possibilities for the ablation of the lymphangioma. In this case, we preferred to enucleate this benign lesion to avoid the operative difficulties. In spite of that, we were obliged to sacrifice the external carotid artery to obtain complete resection of the cyst. When the dissection is difficult, the risk of the injury of neighboring structures is high and the excision of the cyst may be incomplete. Ameh reported 24% postoperative morbidity.

The injury of the facial nerve, particularly the marginal mandibular branch is the most injured nerve. Injury of blood vessels, collection fluid at the resection site, lymphatic discharge, wound infection, parotid fistula and recurrence are the other complications of the surgical treatment. Meticulous dissection, wide field exposure, the discovery of extratemporal part of the facial nerve, perfect hemostasis, the use of bipolar cautery and nerve simulator are helpful to obtain best outcomes.

Percutaneous sclerotherapy can be considered the treatment of choice when surgery is difficult. Once within the cyst, the sclerosing agent provokes a local inflammation that leads to the retraction of the lesion. Adverse reactions are minor like a low-grade fever, vomiting, a local inflammatory reaction and skin discoloration. A variety of sclerosing agents have been used with fluctuating results: bleomycin, OK-432, triamcinolone, alcohol, and fibrin sealant. OK-432 is a lyophilized incubation mixture of group A Streptococcus pyogenes of human origin and appears to be a relatively safe and effective treatment. Macrocytic lesions respond better to OK-432.

Radiotherapy has now been abandoned due to the risk of malignant transformation, while the rate of recurrence is high after laser excision. Radiofrequency is beginning to be used for the ablation of the lymphangioma.

4. Conclusion

Cystic lymphangioma of the parotid gland in adults is a rare entity and poses the problem of differential diagnosis of parotid masses. Its infiltrating nature to the adjacent structures may present important challenges to the surgeon. The pretherapeutic assessment (CT, MRI) and a meticulous procedure are mandatory for best outcomes. Although surgery is the treatment of choice, it is not always possible to obtain complete surgical resection, so partial excision of the lesion or medical therapy (sclerotherapy) is the other alternative.

Conflict of interest

None declared.

References