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SURGICAL RESECTION OF A LOWER-NECK CYSTIC LYMPHANGIOMA: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Lymphangioma, also called cystic hygroma, is a benign congenital malformation of lymphatic vessels seen almost exclusively in children under two years of age. A presentation to the adult age is very rare. A 62 years-old patient was admitted by the consultation for a mass in the right supraclavicular region. A complete surgical excision was performed. Cystic lymphangioma is described like a low-flow vascular malformation, which appear when the lymph sacs are separated from the venous drainage system. Lymphangioma of the neck and mediastinum is usually asymptomatic. Generally, due to the growth the patient may chest pain, cough, dyspnea, dysphagia, or vascular compression syndromes if the lesion reaches dimensions large enough to cause compression of adjacent structures Surgical excision by cervicotomy is the basis of the treatment that can assure the best chance of surgical excision.

INTRODUCTION

Lymphangioma, also called cystic hygroma, is a benign congenital malformation of lymphatic vessels seen almost exclusively in children under two years of age (90%). Is extremely rare in adults often follow lymphatic obstruction secondary to extra-lymphatic pathology.[6]

These malformations are often found in the head and neck and are considered a sequestration of lymphatic tissue that has retained its growth potential. There are three type of lymphangioma: capillary, cavernous and cystic.[1,2] The cystic lymphangioma present a dilatation of capillary and sinusoidal lymphatic vessels (the latter with continued growth of the stromal component) which remain connected to the lymphatic network.[3]

The curative management of lymphangioma is surgical, by a complete resection, however, recurrence can be noted when it is incompletely resected.[8] Complications from non-surgical resection include hemorrhages and infections.[4]

CASE REPORT

A 62 years-old patient was admitted by the consultation for a mass in the right supraclavicular region. This mass is presented since a few of mouths. The patient does not have any particular medical history. At the clinical examination we found a well-defined cystic formation, painless, which measures 6 cm. The skin in regard of the cystic formation is normally colored. (Fig.1) The nasofibroscopic exam is normal. The cervical MRI describes found a cystic formation situated outside the vessels of the neck and an intimate contact with jugular carotidien package. (Fig.2) The cerviothoracic scan found the same characteristics as the MRI. (Fig.3) The hypothesis was a second embryo fist bunch versus a cystic lymphangioma.

Surgical treatment

We performed a right cervicotomy with the removal of the cystic mass under general anesthesia. We have practiced an incision of approximate 7 cm in the right cervical supraclaviacular region. Incision of the subcutaneous plan lifting platysma flap and finding the cystic formation well defined, filled with a liquid. (Fig.4, 5) An aspirative drain was left in place. The histopathologic exam shows a cystic lymphangioma which measured 7x5x3 cm. The immunohistochemical study showed a labeling of the endothelial cells by the antibodies anti-CD31, ERG and D2-40.

Postoperative course

The postoperative course was uneventful. The patient was discharged at the third postoperative day. Clinical exam and the cervicothoracic scan at 1 year after the intervention do not show any sign of recurrence of the cyst**.**

FIGURES

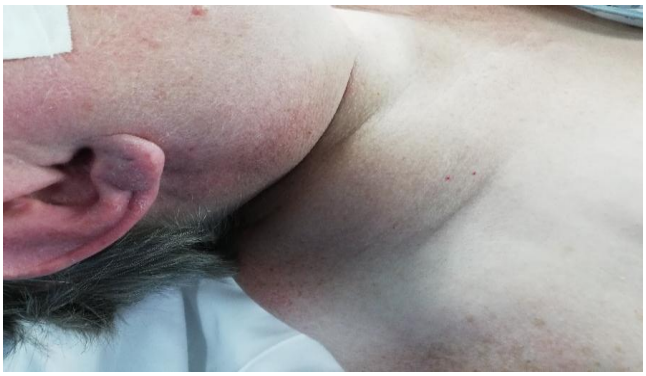

Fig.1: The clinical aspect of the right supraclavicular mass

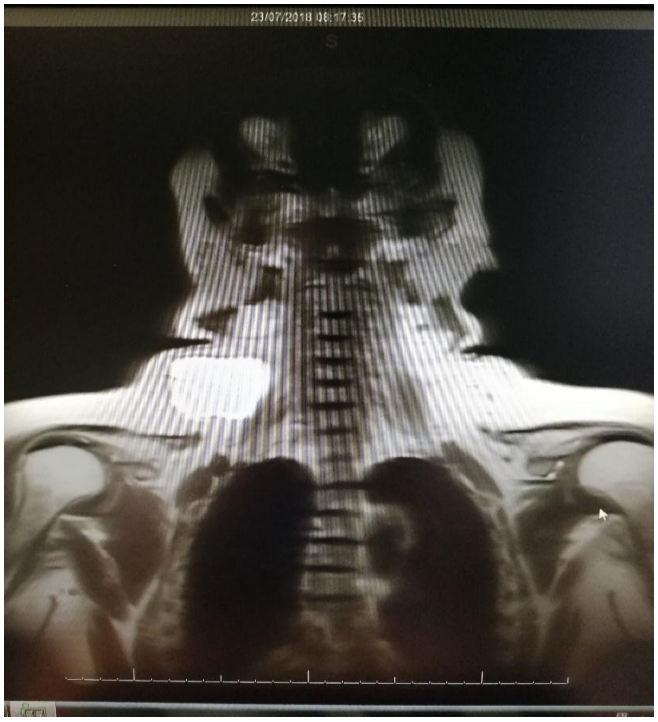

Fig.2: The MRI image of the cyst in T1 frequency.

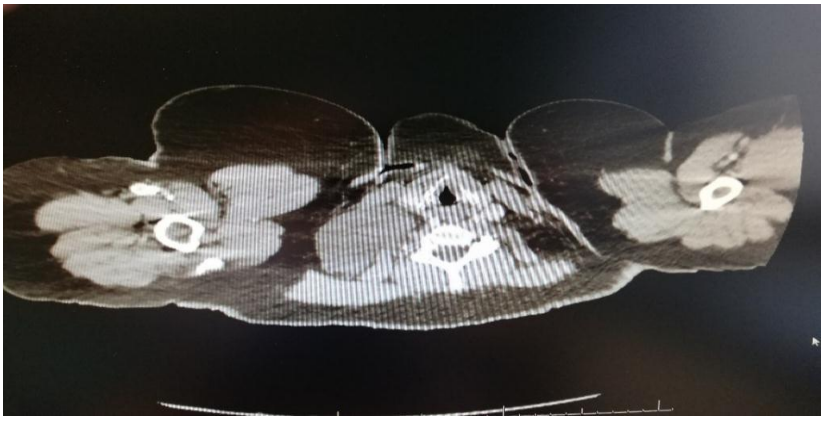

Fig.3: The CT image of the cyst in axial section.

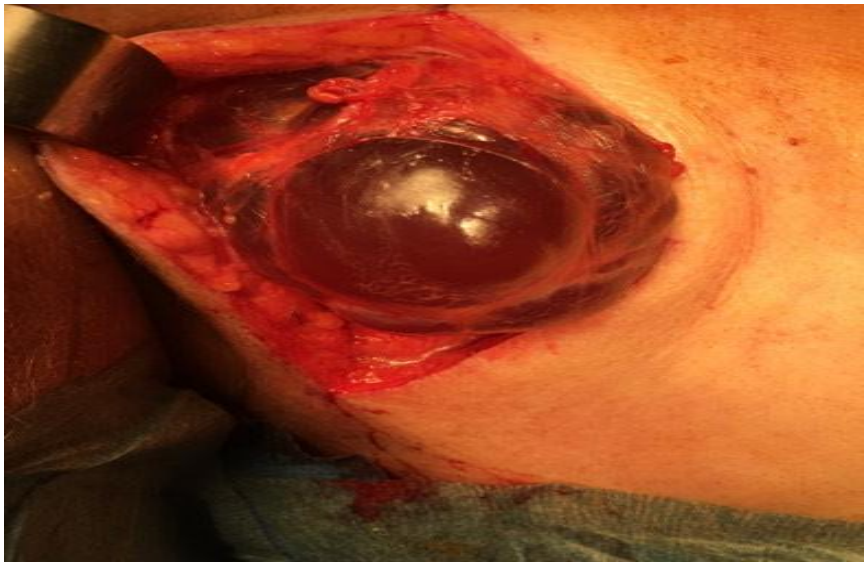

Fig.4. Intraoperative image with the cyst.

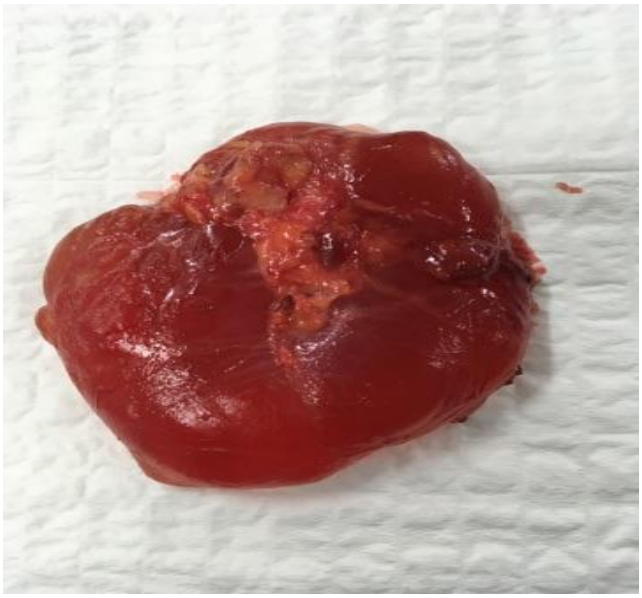

Fig 5: The intraoperatory piece that was sent in anatompathological exam.

DISCUSSION

The cystic lymphangioma is a rare condition, accounting for 5% of vascular abnormalities.[7,4] Cystic lymphangioma is described like a low-flow vascular malformation, which appear when the lymph sacs are separated from the venous drainage system. Two pathogenic theories are mentioned in the literature[9]: the mechanical theory explaining the occurrence of these cysts following an obstruction or a lymphatic contusion; but this theory is rarely confirmed by clinical history, and the most accepted congenital theory currently.

The neck region is the most common location (75%), especially in the posterior triangle and in the back cervical cavity. The cervical localization is more frequently found in childhood: 90% before the age of 2, but can be discovered at any age of life because of the latency of evolution.[12] It can be also found in axillary region (20%). Locations less frequently may be the retroperitoneum, mesentery, omentum, colon, pelvis, groin, bone, skin, scrotum and spleen.[10] Cervical cystic lymphangiomas have mediastinal extension according with the hypothesis of migration of lymphatic element initially sequestered at the cervical level which would have followed in the downward movement the elements migratory like tymus, the bronchial buds, the heart or pericardium.[14]

There are three types of lymphangiomas: capillary lymphangiomas including small vessels with narrow lumen, cavernous lymphangiomas with dilated, anfractuous and inter-communicating light, and cystic lymphangiomas or cystic hygroma with large confluent cavities filled with light yellow fluid.[11] The cystic lymphangiomas can be classified according to the size of their cysts in: microcystic lymphangioma which is composed of cysts less than two centimeters in volume; macrocystic lymphangioma with cysts greater than two centimeters and mixed lymphangioma whose cysts vary in size.[13]

Clinical findings

Lymphangioma of the neck and mediastinum is usually asymptomatic. The clinical presentation is polymorphic.[5] Generally, due to the growth the patient may chest pain, cough, dyspnea, dysphagia, or vascular compression syndromes if the lesion reaches dimensions large enough to cause compression of adjacent structures.[4,15] Apart from the palpable cervical mass, cystic lymphangiomas have no clinical specificity.[16]

There are fluctuating tumors of very variable volume, evolving by successive pushes and regressions and being able to adhere to the deep planes, even to expand to the mediastinum. The form can be soft, regular and well-limited swelling, or manifesting directly as a complication related to compression and repression of neighboring structures: respiratory signs, dysphagia, peripheral neurological disorders, superinfection with skin fistulation or intra-cystic hemorrhage.[17]

Differential diagnosis

Other cervicofacial lesions of cystic nature can be represented by : bronchial cyst, thymic cyst, cyst of the thyreo-glossal tract, abscess collected, hematoma, tumors necrotic, the teratoma cystic.[18]

Diagnosis

The cystic nature of the tumor is suspected on clinical examination and it must be confirmed by imaging data.[18]

The ultrasound reveals the cystic tumor in a form of a well-limited hypoechoic or anechoic mass, sometimes with sediment or fine internal echoes and posterior reinforcement of echoes; it is useful for the prenatal diagnosis of cystic lymphangioma.[9]

Standard X-ray shows anterior or posterior mediastinal opacity but is not specific.[16]

The CT scan shows a low-density tumor (10-36 HU) but the septa are sometimes revealed after the injection of the contrast medium. It shows a mass of hypodense content, well circumscribed without invasion of surrounding anatomical structures.[19]

MRI is complementary to and the CT scan and reveals the reports of lymphangioma with neighborhood structures, but would be less efficient than computed tomography in case of complications.[20] The cystic lymphangioma is of liquid signal: in hyposignal in T1 and hypersignal in T2. The partitions are hyposignal in T1 and T2. Gadolinium injection shows little or no parietal and septal enhancement.[25]

Only the histology can allow a diagnosis of certainty. Lymphangioma, histopathological, is described like lymphatic vessels which are lined by thin endothelial lining. The lymphatic space may appear empty or with proteinaceous material and occasional lymphocytes, macrophages and neutrophils,[21,22]

Treatment options

The treatment is essentially surgical, allowing complete excision of the tumor.[16,23] In 39% a recurrence rate was been seen because of its infiltrative nature.[24] The recurrence after excision occurred when the surgical resection was performed incompletely, reported by Ward et al.[30] Recurrence 3 month after excision was reported by Flanagan and Helwig.[31] A long-term follow-up is indicated. Other therapeutic alternatives are: drainage-aspiration, steroids,

injection of sclerosing agents via 100% ethanol or OKT-432, laser excision, radiofrequency ablation and cautery, radiotherapy used a long time ago is currently abandoned.[26,27] All these are associated with high recurrence rates and moreover subsequent surgery becomes technically difficult owing to sclerosed tissue.[29] Systemic chemotherapy and interferon-α is an alternative with limited success, reserved to the patients with extensive inoperable lesions.[28]

In our case, the therapeutic decision was to perform a complete excision of the cystic mass under general anesthesia. The lesion did not involve any important structures of the neck. The surgical excision was motivated by the size and also to avoid eventually complication later by the growth.

CONCLUSION

Cystic lymphangioma is a benign tumor usually known by its occurrence at a very early age. Is usually manifested by an isolated lower cervical mass. Surgical excision by cervicotomy is the basis of the treatment that can assure the best chance of surgical excision. A definitive diagnosis can be obtained from a final histopathology.

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