

Cervical Cystic Lymphangioma: A Case Report

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Abstract

Cystic lymphangiomas are birth defects affecting the lymphatic system. These are rare benign dysembryoplastic lesions predominate in the region of the head and neck, more particularly in the posterior cervical triangle. They usually occur in childhood and are exceptional in adults. The diagnosis is evoked by the clinic (lateral-cervical swelling) and imaging (ultrasound and tomodensitometry), then confirmed by histology after surgery which forms the basis of treatment. We report a case of cervicothoracic cystic lymphangioma in a patient of 45-year-old with a review of the literature.

Keywords: Cystic lymphangioma; Neck; Thorax

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Introduction

Cervico-thoracic cystic lymphangiomas are a rare entity. They are suspected in front of a later cervical mass. The interest of medical imaging: CT scan and cervicothoracic MRI, plays an important role in their diagnosis, but only the pathological examination confirms it. Surgery remains the most suitable treatment. We report a case of cervicothoracic cystic lymphangioma with a review of the literature.

Case Report

This is a 45-year-old patient with no notable pathological history, admitted to the otorhinolaryngology and cervicofacial surgery department for management of a right cervicothoracic mass, evolving for nine months, gradually increasing in volume without any sign of cervical compression (dysphonia, dysphagia, or dyspnea). The clinical examination finds a healthy-looking skin next to the mass which measures 6 cm long axis, the latter does not rise on swallowing, of consistent painless and non-hookable at its lower pole.

Cervical ultrasound complemented by venous Doppler ultrasound revealed a left paramedian hypoechoic fluid mass measuring 70 mm long in close contact with the vascular axis of the neck which remains permeable. This mass extended into the thorax. The cervicothoracic CT scan showed a left cervical mass with a liquid component measuring approximately 7cm in height, compressing the vascular structures alongside in particular the jugular vein which is pushed back inwards, the laryngotracheal axis remains intact and not deviated (**Figure 1**).

The patient underwent a cervicotomy, with a cervical incision by

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Paul André, which allowed the complete excision of this mass. The post-operative consequences were simple and the pathological report was in favor of cystic lymphangioma. Subsequent checks did not show any recurrence after a 3-year follow-up (**Figure 2**).

Discussion

Lymphangiomas are rare benign tumors. Three types of lymphangiomas can be distinguished: capillary lymphangiomas comprising small vessels with a narrow lumen, cavernous lymphangiomas with dilated, anfractuous, and intercommunicating lumen, and cystic lymphangiomas or cystic hygroma with large confluent cavities filled with light yellow fluid [1].

Two pathogenic theories are mentioned in the literature [2]: the mechanical theory explaining the occurrence of these cysts following lymphatic obstruction or contusion, but this theory is seldom confirmed by clinical history, and the most widely accepted congenital theory today. The lymphangioma would come from sequestration [3] of the embryonic lymphatic sac

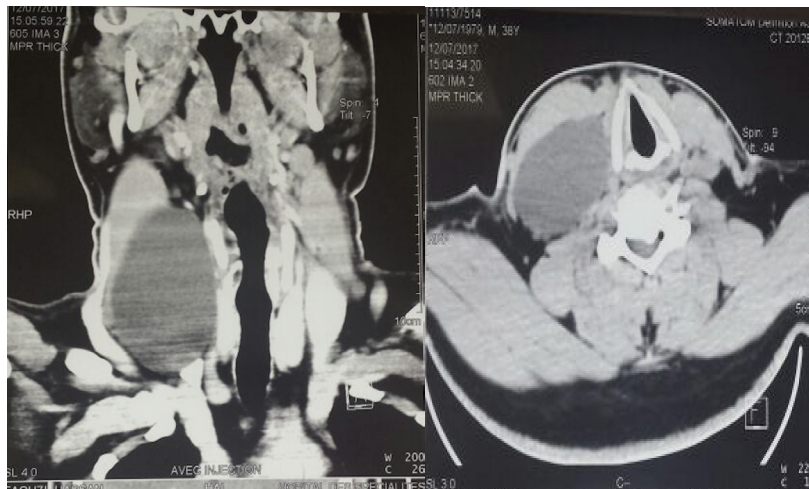


Figure 1 CT scan images in coronal (A) and axial (B) section with injection of contrast product showing a cyst-like mass with fluid content pushing back the vascular axis of the neck.

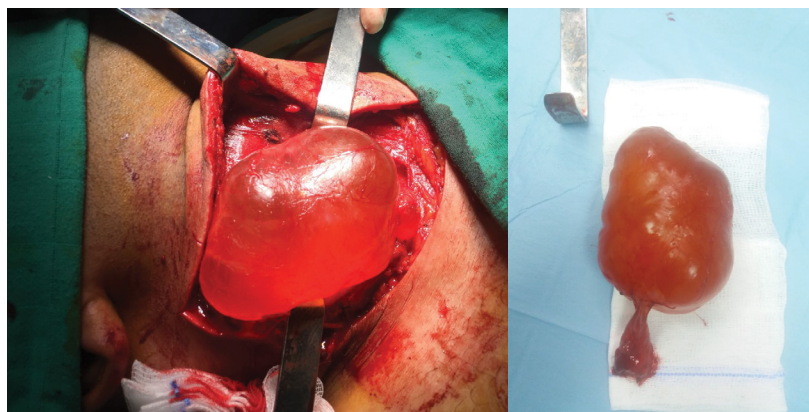


Figure 2 (A) Peroperative image of the intervention shows the extent and appearance of cyst (B) Image of the cystic lymphangioma piece.

which would gradually fill with lymphatic fluid. The failure to establish anastomosis between normal and pathological vessels, and the accumulation of lymphatic fluid, would be responsible for the genesis of this lesion [4].

Cystic lymphangiomas are most often isolated, exceptionally diffuse carrying out lymphangiomatosis [3]. Apart from the brain, lymphangiomas can be found all over the body, they can localize in the abdomen, oral cavity, mediastinum, axillary and inguinal region but lymphangiomas mostly affect the cervical region in about 75% of cases [1,3]. The cervical localization is found more in childhood: 90% before the age of 20 years but can be discovered at any age of life due to the latency of evolution [2,4]. Other localizations were also cited: the retro-peritoneal, splenic, colonic [4], muscular [5], and even at the level of the spermatic cord localization. The cervicothoracic localization of our observation would advance the hypothesis according to which it would result from the migration of lymphatic elements initially sequestered at the cervical level and which would have followed in their downward displacement of other migratory elements

such as the thymus, bronchial, the heart or the pericardium [1,5]. This suggests that cervical cystic lymphangiomas have a mediastinal extension.

The clinical symptoms depend on the size of the tumor and the topography of the cystic formation. Apart from the palpable cervical mass, cystic lymphangiomas have no clinical specificity. Thus, the circumstance of discovery of cervicothoracic cystic lymphangiomas is sometimes revealing symptomatology such as the cervical mass as was the case for our patient; sometimes borrowing symptomatology, but in 50% of cases, they are discovered by chance during a chest x-ray.

The standard radiograph shows an opacity of the anterior or posterior mediastinal site, the appearance of which is not specific. Ultrasound shows a hypoechoic or anechoic appearance, sometimes with sediment or fine internal echoes and posterior reinforcement of echoes [2,6]. The CT scan shows a tumor of low fluid density (10-36 HU) but the septa are sometimes not revealed until after injection of the contrast product [2,4]. Magnetic resonance imaging would seem to be useful for exploring this

tumor but would prove to be less efficient than computed tomography in the event of complications [4].

Only histology can provide a definite diagnosis [1,2]. The treatment is essentially surgically allowing the complete excision of the tumor which is essential for a complete cure. Other therapeutic means have been tried without success, such as radiotherapy, drainage by mediastinoscopy, and chemical sclerosis by intravenous cyclophosphamide [2,6], they are specially reserved for unresectable tumors because of their size, their location or because of the general condition of the patient [6]. Video thoracoscopy, drainage by mediastinoscopy, or by scanno puncture or guided echo do not ensure the excision of the wall of the cyst source of recurrence. The approach depends on the location and the cystic extensions. Posterolateral thoracotomy is recommended by many authors for pure mediastinal localization [2,4,6]. The cervical and mediastinal location may lead to the choice of a suprasternal cervicotomy sometimes associated with a median sternotomy depending on the endothoracic extension,

visceral adhesions to the large vessels, to the nervous structures, or the trachea. In our case we performed a cervicotomy with an incision by Paul Andre which allowed us to remove the tumor in its entirety, the relative ease of the intervention is based on the fact that the cystic formation was well-circumscribed of moderate volume with a plan of obvious cleavage without inflammatory flare and adhesions to the vascular structures allowing the total excision of the tumor. The postoperative results are generally excellent.

Conclusion

Despite the rarity of cervicothoracic cystic lymphangiomas, they can be detected from an isolated low later-cervical mass with images of cystic appearance on the cervicothoracic scanner which also studies the intrathoracic extension and its relationships with vascular structures. Complete surgical excision, often by cervicotomy, is the basis of treatment, and the short and long-term post-operative effects are often excellent.

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