CASE REPORT

Case report of a chest wall cystic hygroma in a teenager

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SUMMARY

Cystic hygroma or cystic lymphangioma is a congenital malformation of lymphatic origin. Their occurrence on the chest wall is very rare, and they progressively grow with age infiltrating into the local tissues, around muscle fibers and nerves, making them difficult and hazardous to remove. There are various treatment modalities of such lesion. Based on the literature surgical excision is the preferred treatment of choice in cystic hygroma because it gives a better cure rate compared to other modalities. We report a case successful excision of anterolateral chest wall cystic hygroma in a teenager in Hospital Serdang.

KEY WORDS:
Cystic hygroma; Chest wall lesion; Surgical excision; Sclerotherapy

INTRODUCTION

Cystic hygroma is a benign congenital malformation of the lymphatic system that is also known as a macrocystic lymphatic malformation and was first described in 1828 by Redenbacker. Eighty percent of cystic hygromas occur in the neck, usually in the posterior cervical triangle. Other sites include the axilla, superior mediastinum, mesentery, retroperitoneal region, pelvis and lower limbs. [1]

Their occurrence on the chest wall is very rare, and they progressively grow with age infiltrating into the local tissues, around muscle fibers and nerves, making them difficult and hazardous to remove. We report a case of anterolateral chest wall mass in a teenager and the approach in our center.

CASE REPORT

A 15years old male presented to the cardiothoracic clinic with progressively enlarging right anterior chest wall mass for a month duration. The patient denied a history of a similar lesion in the past. On examination, the swelling measured 10cm x 12cm in size and extended from lateral 2/3rd of the clavicle to 4th rib inferiorly and from right mid-axillary line to midclavicular line on the same side. CT scan defined a well-defined cystic lesion in the right anterolateral chest wall superficial to the rib and unable to get a clear plane between the mass and subclavian vessels. Subsequently magnetic resonance imaging revealed a large septated cystic lesion in the right axilla, extending down to the right lateral chest wall. The lesion appeared encasing the subclavian vessels, and the subclavian vessels appear compressed by this lesion (figure 1). The case was further discussed with our vascular team and planned for surgery. The patient underwent surgical exploration of the mass via an axillary apical incision along the lateral aspect of the third rib. The mass was encapsulated with a thin wall and adherent to surrounding musculature. The lower border of the lesion was identified and carefully released from latissmus dorsi and serratus anterior muscles. Dissection carried towards the apex and meticulously divided the upper border that was encased in a thin layer around clavicopectoral fascia. Contents of the cyst leaked intra-operatively towards the final phase of the dissection, but complete removal of the wall of the septated cyst was achieved (figure 2). Long thoracic nerve and thoracodorsal nerve encountered during the dissection were seen and preserved.

Post-operatively the patient recovered well and had mild winging of the scapula on the right side. There was no swelling or any other neurological deficits. He was discharged well on day three post op. The histopathology report was consistent with cystic hygroma. During the subsequent follow-up visits, the patient was well with no neurological deficit or recurrent swelling.

DISCUSSION

Cystic hygroma is a congenital malformation of the lymphatic system. Lymphangiomas are usually classified as a capillary, cavernous or cystic lymphangiomas. They may also be classified more conveniently, by the size of the cyst, as microcystic, macrocystic and mixed lymphangiomas. Microcystic lymphangioma consists of cysts measuring less than 2 cm in size, whereas the size of cysts in the case of macrocystic lymphangioma is more than 2 cm. The mixed lymphangioma is characterized by cysts of variable sizes, i.e. some cysts are more than 2 cm in size, and others are less than 2 cm.

There are some proposed mechanisms to explain the pathophysiology of cystic hygroma. Embryologically, these lesions are believed to originate from sequestration of lymphatic tissue from lymphatic sacs, during the development of lymphatico-venous sacs. These sequestered tissues fail to communicate with the remainder of the lymphatic or venous system. Later on, dilatation of the sequestered lymphatic tissues ensues, resulting in the cystic morphology of this lesions.

Cystic hygroma of the chest wall is a very rare anatomical condition. A review of the literature showed only 15 reported

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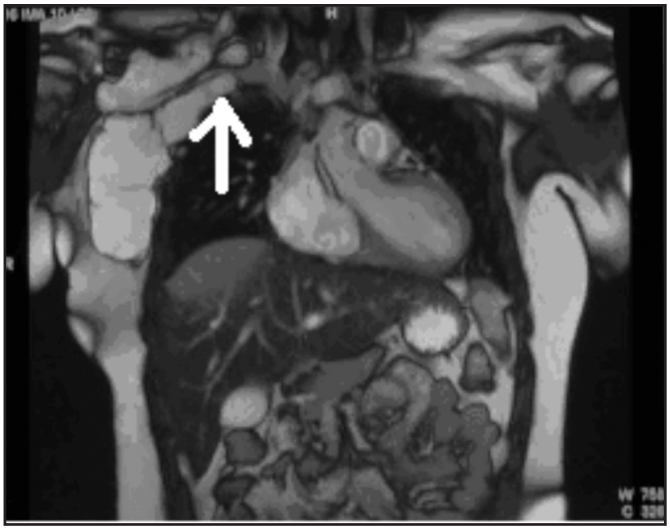

Fig. 1: MRI image, There is encasement of the subclavian vessels and the subclavian vessels appear compressed by this lesion.

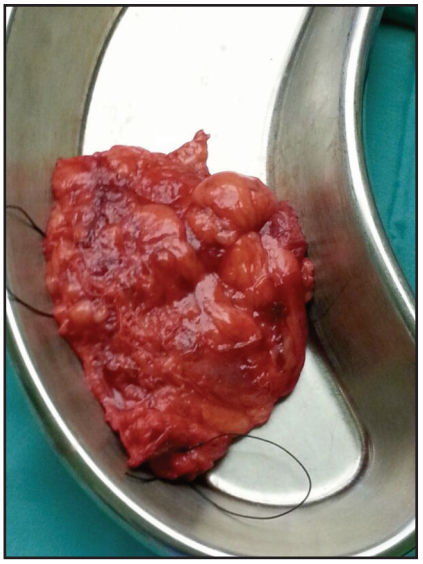

Fig. 2: Post resection specimen.

cases in the world literature. The effect of these lesions depends on their position and relationship to surrounding structures, although the most common teenage presentation is of a painless lump in an otherwise asymptomatic patient. However, in this case, the lesion was reported as encasing subclavian vessels and delaying excision might result in an unfavorable outcome.

Aspiration of the cystic hygroma can be performed as a temporary measure to reduce the size of the cystic hygroma, and thereby, reducing its pressure effects on the respiratory and feeding passages.

Many recent case reports and case series have increasingly documented remarkable results for management of cystic hygroma with sclerosant agents. Although sclerotherapy is now well established in the treatment of neonatal cystic hygromas, there have been relatively few cases reported of its use in teenagers and adult patients. Caution has been urged with the use of agents such as OK-432 that induce a local immune response that often results in a temporary rapid increase in the size of the cystic hygroma.[2] Besides OK- 432 few other sclerosant agents such as bleomycin, pure ethanol, bleomycin, sodium tetradecyl sulfate, and doxycycline have been tried but showed unfavorable outcomes with malicious adverse effects.

In this case, it was thought that the ideal treatment would be complete surgical excision as multiloculated cystic hygroma may not respond to sclerotherapy. The success of surgery has been found to correlate with histology, encapsulation, complete excision and anatomical location. The issue in surgery that commonly encountered in cystic hygroma is the failure to remove the sac completely without rupture. It's a recognized problem as these tumors usually have a thin, fragile wall. Intra-operative rupture of the lesion complicates complete removal as it obscures the limits of the structure. However, Riechelmann reported very low levels of recurrence (1/9 patients) following subtotal excision when small plaques of tumor wall were known to be left in situ.[3]

The possible complications of surgery are damage to nerves, axillary vessels and incomplete excision in the case of infiltration to the surrounding structures. The post-operative complications observed after surgical excision of cystic hygroma are wound infection, hemorrhage, hypertrophied scar and lymphatic discharge from the wound. In about 20 % of cases, there is recurrence even after apparent complete excision of the lesion.[4]

Based on the literature surgical excision is still the preferred treatment of choice in cystic hygroma because it gives a cure rate of 81% compared to other treatment modalities such as aspiration, partial excision, and sclerotherapy.[5]

We opted for surgery in this case with an aim to achieve a better cure rate and to avoid further complication of encasement of axillary vessels if delayed.

CONCLUSION

Cystic hygroma of the chest wall is rare condition and management plan should be carried out following a multidisciplinary discussion. Surgical excision always has the better outcome according to literature, but a careful patient selection is vital to avoid post-operative complications.

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