Case report

Cervicothoracic Lymphangioma in an Adult: A Rare Entity

Abstract

Background

Lymphangiomas are uncommon benign lymphatic malformations most commonly seen in the head and neck region. Cervical lymphangiomas with intrathoracic extension are very rare clinical entity and comprise only 1% of the cases.

Case Report

We report here a rare case of a 25 year old female, with complaints of a huge soft, cystic fluctuant and transilluminant swelling involving the right side of the neck and chest. On imaging, it revealed a multilocular thoracocervical mass, which was isointense on T1 and hyperintense on T2, occupying right anterosuperior, middle and posterior mediastinum compressing the upper lobe of the right lung and extending supraclavicularly. Patient was managed surgically via multidisciplinary approach. Histopathological examination confirmed the diagnosis of lymphangioma

Discussion and Conclusion

Imaging and histopathological examination play a key role in diagnosis of lymphangiomas. Such extensive lymphangiomas require complete surgical excision and should be performed by experienced surgeons as this is essential to prevent further recurrences.

Keywords: Lymphangioms, Thoracocervical, Histopathology, Magnetic Resonance Imaging

Introduction

Cystic lymphangiomas are rare benign dysembryoplasias of the lympho-ganglionic system, responsible for a tumor syndrome by angiolymphatic proliferation. They can occur everywhere in the body, but cervicofacial localizations are the most common. They represent between 2.6 and 5% of benign congenital cervical masses, and their clinical revelation is generally very early, in the first two years of life [7,8]. Lymphangiomas are rare congenital malformations of the lymphatic system that can appear at any age or site. In majority they are diagnosed at birth and 95% of them are found in the neck, the head, or the axilla. Although benign, and commonly asymptomatic, they can sometimes cause pressure and life threatening complications especially massive lesions involving the neck and mediastinum in newborn infants or cause a diagnostic confusion as a result of sudden enlargement following haemorrhage or infection [9,10].

Case Presentation

A 25 year old female patient presented to our otolaryngology department with complaints of painless swelling over right side of the neck, which was gradually progressive in size since 3 months. Patient had no complaints of change in voice, dyspnoea or dysphagia. She gave history of similar swelling in the past for which she underwent surgery 7 years back and histopathological examination (HPE) of the excised specimen report revealed Cystic hygroma. Following surgery she was asymptomatic for 6 years. There was no history of similar complaints at birth or in any of the family members.

Clinical examination of the neck, revealed a swelling of size 12x6x4 cm involving lower third of the right side of the neck and extending into the right supraclavicular area. A previous surgical scar was noted over the anterior aspect of upper chest. No discharging

sinuses were visualised over the swelling. There was no local rise of temperature or tenderness on palpation of the swelling. The swelling was soft, fluctuant, compressible and brilliantly transilluminant. On further evaluation, ultrasonography of the neck revealed a 12x8.9x8.6cm anechoic cystic lesion in the right side of neck in the subcutaneous plane with multiple septations with avascularity causing compression of the right internal jugular vein. Chest radiograph anteroposterior view revealed a large homogeneous opacity in the anterior mediastinum compressing the upper lobe of the right lung (Fig.1A). Magnetic resonance imaging (MRI) of the neck and chest revealed a thoracocervical mass of 27x12x14 cm occupying right anterosuperior, middle and posterior mediastinum compressing the upper lobe of the right lung and extending into right lower neck subclavicularly. It was compressing the lower part of the right internal jugular vein. Mass was multilocular and septate with thick walls. T1 weighted images were isointense whereas T2 weighted images showed hyperintensity (Fig.1). All the routine haematological investigations and 2D ECHO were under normal limits.

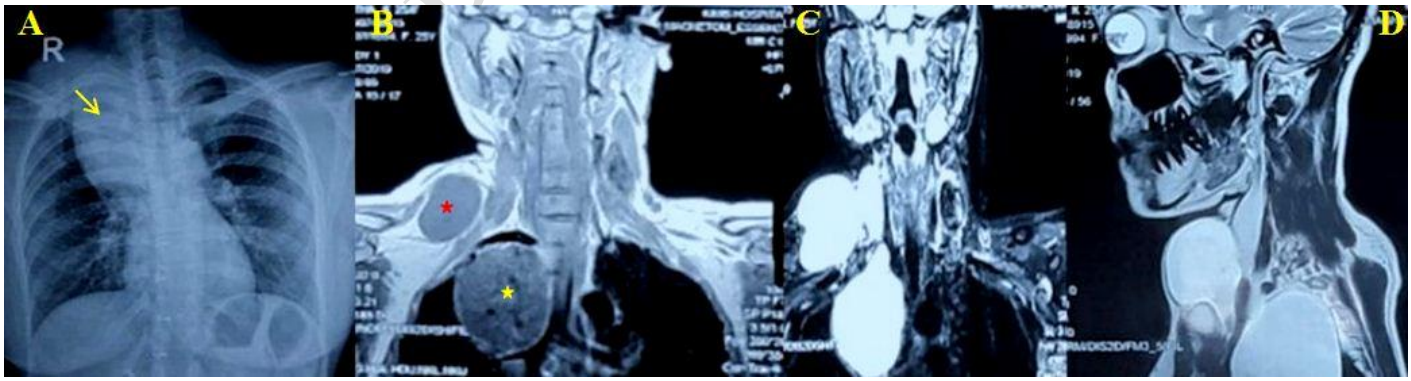
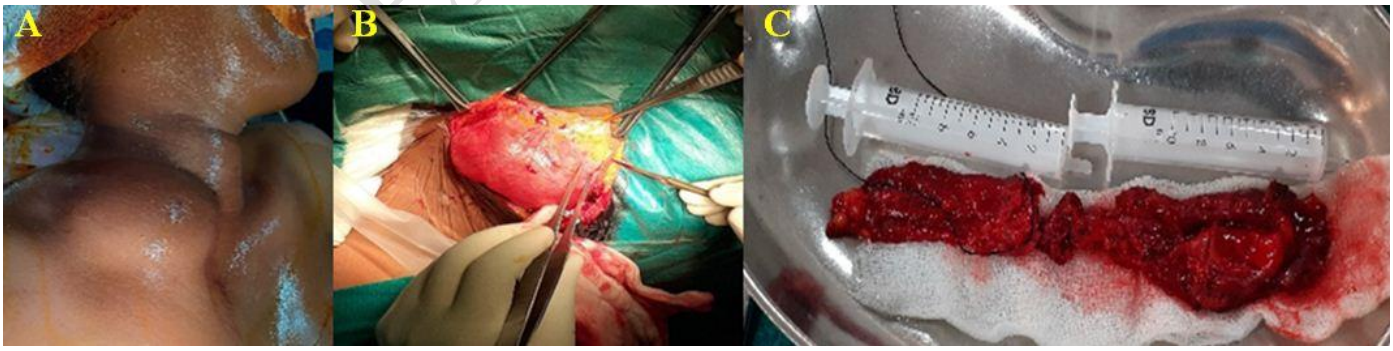


Fig. 1. (A) Chest radiograph AP view showing a large homogeneous opacity (yellow arrow) in the anterior mediastinum compressing the upper lobe of the right lung. (B,C and D)

Magnetic resonance imaging (MRI) of the neck and chest showing a thoracocervical mass (red asterix-cervical part, yellow asterix-thoracic part) which is isointense on T1 and hyperintense on T2 weighted images. (B) T1 weighted coronal section isointense on T1 weighted images and (C) T2 weighted coronal section and (D) T2 weighted sagittal sections.

After obtaining consent from the patient, under general anaesthesia surgical excision was performed by a team of Otolaryngologists and Cardiothoracic surgeons. A large dumbbell shaped cystic swelling was seen occupying the right hemithorax which was seen extending subplatysmally and substernally with a connection behind the clavicle, without causing any compression of the trachea and oesophagus. The cyst was completely excised via neck exploration and right anterior thoracotomy approach with sparing the internal jugular vein and the phrenic nerve (Fig. 2). The excised cyst was sent for HPE. Haemostasis was attained. Postoperative period was uneventful. HPE revealed collapsed cystic spaces with supporting fibromuscular connective tissue, lymphoid aggregates with focal giant cell reaction which was suggestive of Lymphangioma. Patient was followed up for 2 years, which showed no



recurrence.

Fig. 2. (A) Clinical image of large cystic swelling involving the lower neck and upper chest on the right side. (B and C) Intraoperative image of the cervicothoracic cyst.

Discussion

Lymphangiomas are degenerative lesions arising from the lymphatics, and can be classified as: simple lymphangiomas, cavernous lymphangiomas and cystic hygromas.[1] Cystic hygromas are composed of cysts and sinuses, varying in size from a few millimetres to several centimetres in diameter. They usually present as a cystic masses containing eosinophilic acellular lymph fluid.[1].

Although congenital in most of the cases which show usual presentation at birth, sometimes they seem to manifest for the first time in young adults which was the presentation in our case. They can appear anywhere in the head and neck. On palpation they feel cystic and transilluminate. They may remain static or involute, but in some cases they gradually increase in size and occasionally, especially after internal haemorrhage or infection, and can grow rapidly causing compression on trachea leading to life threatening airway obstruction. 90% of the lymphangiomas develop on the body surface, 75% are seen in the neck, mostly invoving the posterior triangle and 20% in the axilla.[2,3] 1% of the cystic lymphangiomas have mediastinal localization.[4] The common features helpful in the diagnosis of lymphangiomas are uni or multilocular predominantly cystic masses with clear borders, septations with lobular contours, contiguous extension along the neurovascular planes of mediastinum without mass effect.[5] These findings of lymphangiomas were also seen in our case, with rare presentation in an adult. On Imaging, lymphangiomas are hyperintense on T2-weighted MRI images, and are isointense on T1-weighted.[6] Surgical excision is considered as the treatment of choice.

Conclusion

The diagnosis of lymphangioma is usually made on clinical grounds, but CT and MRI scanning are complimentary in helping to accurately determine the size, exact anatomical location, and relationship to important structures as well as aiding in surgical planning. Surgical excision remains the treatment of choice but it is challenging due to close association with vital structures and is therefore best undertaken by a multidisciplinary approach to prevent recurrence.

Consent

Informed written consent was obtained from the patient.

"All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal."

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