CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 76 (2020) 174–177

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

A rare case of cystic hygroma in neck and extending into thoracic cavity☆

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ARTICLE INFO

Article history:
Received 10 July 2020
Received in revised form
13 September 2020
Accepted 13 September 2020
Available online 3 October 2020

Keywords:
Cystic hygroma
Maxillofacial team working with cardio thoracic team
20 cm cystic lesion

ABSTRACT

INTRODUCTION: Cystic hygroma is a benign swelling of the neck among the pediatric population. It is a fluid-filled sac resulting from blockage in the lymphatic system and is commonly located in the cervical region and axilla.
CASE PRESENTATION: We present a rare case of a large cystic hygroma in the neck and the thorax in a 12 year's old male. The lesion was diagnosed with the help of MRI which showed a cystic lesion extending from the carotid sheath to the mediastinum. And confirmed by fine-needle aspiration cytology (FNAC). The lesion was treated by surgical excision along with a cardiothoracic team.
DISCUSSION: We will explore in our discussion the diagnostic modalities for such lesions and the different treatments available for Cystic Hygromas.
CONCLUSION: We concluded from this case that accurate diagnosis and careful planning with multidisciplinary team approach and choosing the right treatment as indicated has a great impact on the results for such difficult cases.

1. Introduction

Cystic Hygromas, also called lymphatic malformation, are anomalies of the lymphatic system characterized by single or multiple cysts within the soft tissue [1]. In most cases of cystic hygromas 70–80% involving the neck and lower part of the face, other sites are axilla, superior mediastinum, retroperitoneum, mesentery pelvis, and lower limbs [2]. Cystic hygromas contain either a clear or cloudy fluid. Cystic hygroma is one of the easiest and earliest anomalies that can be detected by prenatal sonographic examination in the second trimester. Approximately 50–60% of these malformations appear before one year of life and 80–90% before the end of the second year of life [3]. Occasionally, Cystic hygroma is inherited as an autosomal recessive. Yet, in most cases the cause is idiopathic [4]. Cystic hygromas involving the neck are thought to arise secondary to failure of the jugular lymph sacs to join the lymphatic system resulting in tiny sac-like structures sprouting from the existing cystic space. Lymph like fluid is secreted into these endothelial lined cystic spaces. Thus, local dilation and enlargement of the cystic spaces will be the result. All work has been reported in line with the SCARE criteria [5].

2. Case presentation

A 12-year-old boy presented to the department of pediatrics. He was then referred to the care of both cardiothoracic and maxillofacial surgery teams. The patient had a painless swelling on the left side of his neck. The swelling started 4 months ago when it was noticed by the child's parents, but it was too small. He was complaining of dyspnea during normal day exercises. Upon examination, the swelling was present over the region extending from carotid sheath to mediastinum on the left side passing behind the sternum (Fig. 1). The swelling was soft on palpation, with poorly defined margins, nontender and when subjected to light test was brilliantly translucent. Fine needle aspiration cytology [FNAC] revealed that the mass contained clear yellow fluid in which no tumor cells were present and was diagnosed as cystic hygroma. Magnetic resonance imaging (MRI) of the lesion showed a well-defined cystic lesion in the left upper anterior triangle of the neck deep to the sterno cleido mastoid muscle involving the carotid sheath and extending into mediastinum on the left side passing

☆ No patient or author details are included in the figures.
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https://doi.org/10.1016/j.ijscr.2020.09.090

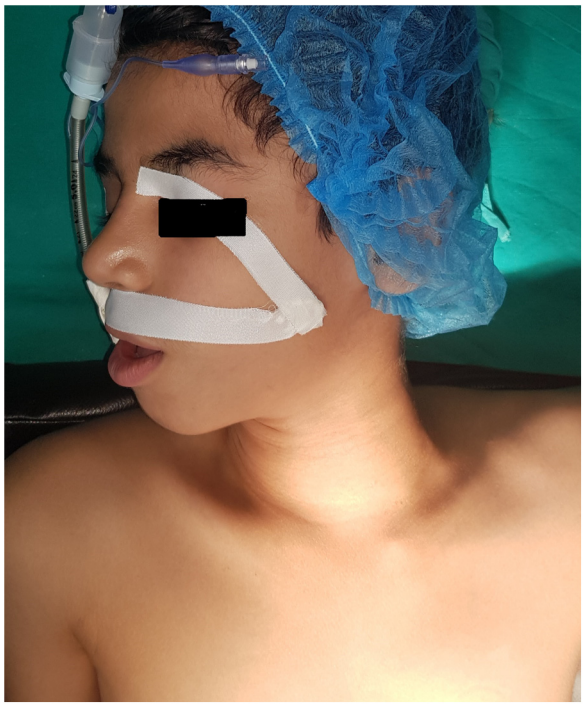

Fig. 1. Pre operative view of the patient on the OR table.

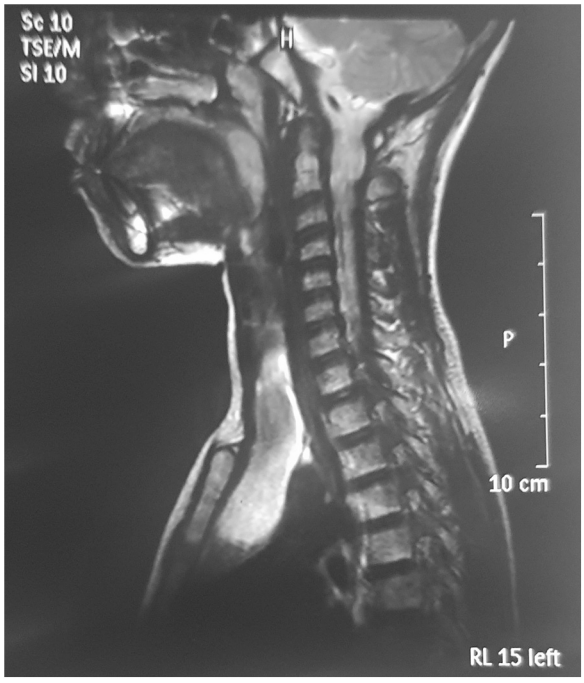

Fig. 2. MRI showing the extension of the lesion from the neck into the thoracic cavity.

behind the sternum (Fig. 2). When the lesion was found to be inseparable from the left side of the sternum total surgical excision of the lesion was planned together with the cardiothoracic team and open thoracotomy was done and the lesion was dissected from the mediastinum and the pleura up to the suprasternal notch (Fig. 3). The lesion was adherent to the carotid sheath and was dissected from it through a midline neck incision by the maxillofacial surgery team. Following excision, the cyst was measured to be 20 cm in length (Fig. 4). 6 months follow up with no recurrence evidence.

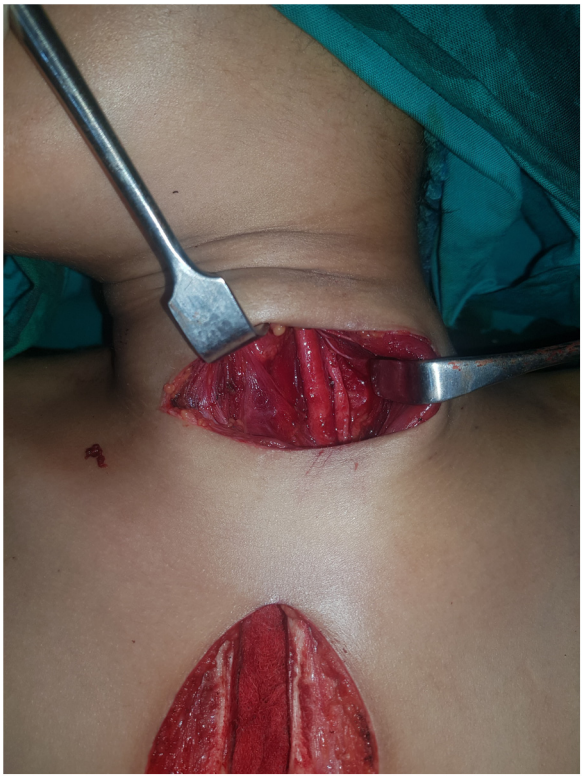

Fig. 3. Internal carotid artery and internal jugular vein after dissecting the lesion from the carotid sheath.

3. Discussion

Lymphangiomas are congenital lymphatic malformations that comprise 5.6% of all benign lesions of infancy and childhood. They can affect any part of the body, their presentation depends on the surroundings, but a strong predilection for the neck and axilla has been noted. Lymphangiomas are detected before the age of two years. Histologically, there are three types of lymphangiomas, capillary, cavernous and cystic. These lesions are composed of dilated lymphatic channels with one or two endothelial layers, with or without an adventitial layer. These dilated lymphatics can vary in size, depending upon the location and surrounding tissues and is the basis for classification [6]. Capillary lymphangioma [or simple lymphangioma] is composed of small, capillary-sized endothelium-lined lymphatics, whereas cavernous lymphangioma is made up of larger lymphatic channels with adventitial coats. Cystic Hygromas are multilocular masses, consist of large microscopic lymphatic spaces. Cystic hygromas are deeply seated in areas of areola or loose connective tissues. They appear early in life as large soft-tissue masses on the axilla, neck or groin. They are soft, vary in size and shape, and tend to grow extensively if not surgically excised [6]. They are multilocular cysts filled with clear or yellow lymph fluid. Cystic hygromas may be associated with Turner syndrome, Noonan syndrome, trisomies, fetal alcohol syndrome, chromosomal aneuploidy, cardiac anomalies and fetal hydrops [7]. Cystic hygroma can be diagnosed based on radiographic appearance on MRI, biopsy (FNAC), and clinical symptoms [8]. FNAC of cystic hygroma is watery clear in most cases and milky to hemorrhagic in few cases. Histologically the fluid contains cholesterol crystals, lymphoid cells mainly small lymphocytes in variable number and endothelial cells [9]. Clinically the mass is soft, non-tender and ill-defined. Symptoms may develop when the cyst enlarges to where it compresses surrounding tissue or organs. It may show obstructive

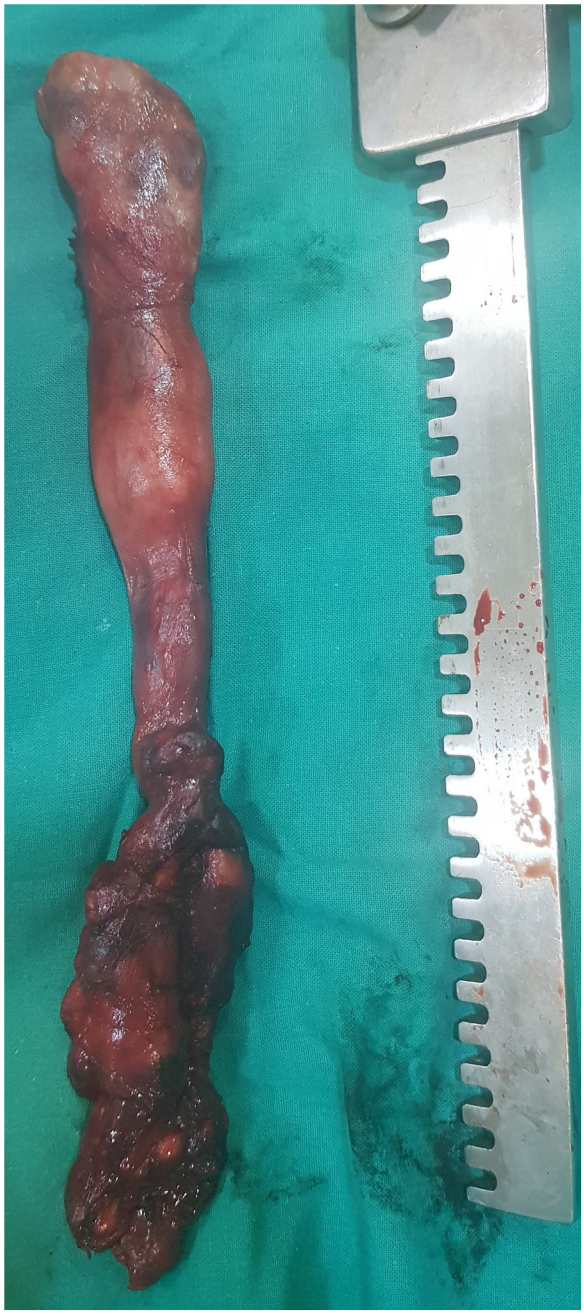

Fig. 4. The excised hygroma measuring about 20 cm in length.

symptoms such as dysphagia, dysphonia, and airway obstruction [10].

Prenatally, the diagnosis of cystic hygroma can be made with high reliability through an ultrasound examination. Ultrasound characteristic of the presence of a large cystic mass occupying the posterolateral aspect of the fetal neck. Large, septated, multilocular hygromas seem to have a worse prognosis than the non septated ones. The differential diagnosis includes occipital encephalocele, cervical meningocele, and cystic teratoma of the neck. Two types of cystic hygromas have been described. The first type which is the most common type consists of a localized lymphatic defect without associated fetal hydrops or other abnormalities, the prognosis is good with a surgical repair that can be performed at any time during the neonatal period. The second type of cystic hygroma is diagnosed in early pregnancy by sonography, this type is associated with chromosomal abnormalities in approximately 60% of cases. In this type, fetal death usually occurs shortly after diagnosis.

For treatment, many asymptomatic patients may require nothing except watch and see, and only require intervention when there is symptoms or serious disfigurement. There are either prenatal or postnatal treatment options. There are a few reports concerning intrauterine treatment of cystic hygroma without chromosomal abnormalities. Experimental OK-430 injection and sclerotherapy using special agents like bleomycin and doxycyclin have been reported and is believed to shrink the growth although it may require several injections [11]. Moreover, postnatal treatment of choice is complete excision with an extremely low mortality rate, but recurrence, infection wound seroma and nerve damage occur in 30% of cases. In addition, the recurrent rate varies, and it depends on the complexity of the lesion and the completeness of the excision. Difficulties in surgical management is due to the proximity of cystic hygromas in Head and Neck area to vital structures such as parotid gland, facial nerve, carotid sheath and this makes complete surgical excision is considered a challenge.

4. Conclusion

Observation is recommended for small, non-expanding lesions. This case of cystic hygroma was diagnosed based on FANC and its extension diagnosed carefully based on MRI radiographic pictures. The lesion was huge and extended from the carotid sheath to the mediastinum. Therefore, a Careful surgical plan was made to avoid injury to adjacent structures. A team of experienced surgeons from varying fields including cardiothoracic surgery, maxillofacial surgery, and anesthesia worked together to ensure a successful surgical outcome.

Declaration of Competing Interest

We wish to confirm that there are no known conflicts of interest associated with this publication and there has been no significant financial support for this work that could have influenced its outcome.

Funding

No funding was received for this work.

Ethical approval

In accordance to declaration of Helsinki.

Patient guardians wrote informed consent for surgical intervention and for publication.

Consent

"Written informed consent was obtained from the patient guardians for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request".

Author's contribution

Dr Mohamed Touny:

Writing the paper.

First Surgery Assistant.

Professor Mohamed El Sayed: Consultant in charge of the surgery.

Primary surgeon.

Postoperative follow up of the patient.

Dr Nesreen Ibrahim:
Data collection.
Analysis of data.
Patient follow up.
Dr Zainab Al-Azzawi:
Patient follow up.
Data collection.
Revision of manuscript.
Professor Ibrahim Kasb
Cardio thoracic consultant in charge of the case.

Registration of research studies

N/A.

Guarantor

Dr Mohamed Touny.
Professor Mohamed El Sayed.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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