The Journal of Medical Research

Case Report

JMR 2017; 3(2): 45-47
March- April
ISSN: 2395-7565

www.medicinearticle.com
Received: 11-03-2017
Accepted: 30-04-2017

Isolated mediastinal cystic lymphangioma- A rare case report

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Abstract

Cystic lymphangioma (cystic hygroma) is a congenital malformation of the lymphatics resulting in blockage of lymphatics. Cystic hygroma are mainly found in cervical region (75%) and axillary region (20%) though they are also found in some other locations. An isolated mediastinal lymphangioma without a cervical component is an uncommon occurrence which account for less than 1%. We report a 7 year old girl who presented with sudden onset severe breathlessness. Contrast enhanced computed tomography (CECT) thorax done on emergency basis revealed large mass of mixed heterogeneity with areas of necrosis occupying entire left hemithorax with complete collapse of left lung, provisional diagnosis being a mediastinal tumour. The patient underwent emergency surgical resection with piece meal total excision of the tumour other than a small strip densely adhered to the subclavian vein. She started having increase in size of tumour within a month hence was electively taken up for re exploration and complete resection of tumour was performed. Biopsy showed cystic lymphangioma with xanthogranulomatous inflammatory lesion. Patient stable at 3 months follow up.

Keywords: Mediastinal tumour, Respiratory distress. Emergency surgery, Rare diagnosis.

INTRODUCTION

Cystic lymphangioma (cystic hygroma) is a congenital malformation of the lymphatics resulting in blockage of lymphatic flow. They are mainly found in cervical region (75%) and axillary region (20%) Majority of them are detected before the age of 2 years as they are superficial [1]. They can also be found in the retroperitoneum, pelvis, and chest wall [2]. Of cervical lymphangiomas only 2-3% may be associated with an intrathoracic extension. An isolated mediastinal lymphangioma without a cervical component accounts for less than 1% [1,2] Cystic lymphangioma constitute5-6% of mediastinal masses in children [1].

Cystic lymphangiomas are sometimes acquired, which are principally seen in middle-aged adults, and caused by a process of chronic lymphatic obstruction secondary to surgery, chronic infection, or radiation. Malignant transformation of cystic lymphangiomas has not been described [2].

CASE REPORT

A 6 year old girl was referred to our department in emergency with sudden onset severe breathlessness. On examination there were absent breath sounds in all zones of left lung. Routine blood investigations were normal. Xray Chest showed large space occupying lesion in left hemithorax with mediastinal shift to the right side. CECT thorax revealed large mediastinal mass with areas of necrosis within occupying the entire left hemithorax with complete collapse of left lung. There was large mediastinal shift to the right side with associated pneumonia of the right lung. (Fig 1)

On aspiration there was hemorrhagic fluid so in order to relieve the dyspnoea and stabilise the patient till emergency surgery an intercostal drain was inserted and around 200 ml of hemorrhagic fluid drained. The serum alpha fetoprotein (AFP), ß-HCG and LDH (samples taken preoperatively) were normal. In view of severe breathlessness, patient taken up for emergency surgery. A left posterolateral thoracotomy was done through 5th intercostal space. Intraoperatively there was a large well encapsulated tumour occupying almost the entire anterior mediastinum on the left side with complete collapse of left lung. The tumour showed loculations with areas of necrosis and hemorrhage in the lower part. (Fig 2).

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The upper margin of the tumour was densely adhered to the subclavian vein. Entire tumour was removed piece meal leaving behind some tumour densely adherent to the subclavian vein. As patient was hemodynamically not stable, further resection was not done at that time. 2 drains were inserted. Patient had uneventful recovery in postoperative period and was discharged on postoperative day (POD) 8. Post op Chest Radiograph showed adequate expansion of left lung. Biopsy proved it to be cystic lymphangioma with xanthogranulomatous inflammatory lesion. A follow up CT scan showed an increase in size of the tumour after a month (Fig 3). Hence she was electively taken up for resurgery and completion resection of tumour with careful separation from subclavian vessels was done (Fig 4). There was complete expansion of lung intraoperatively (Fig 5). Patient was stable postoperatively and was discharged on POD 7. Post op CT scan shows well expanded lung with no recurrence (Fig 6). She is stable with no recurrence at 3 months follow up.

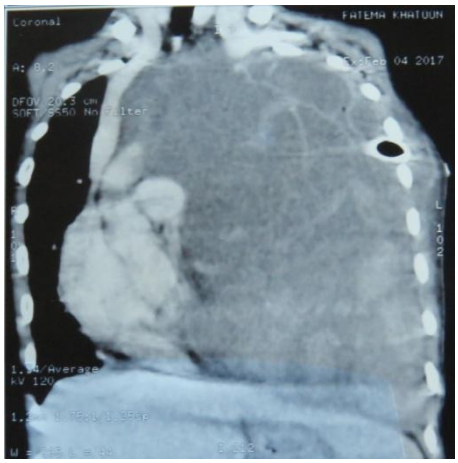

Figure 1: Mediastinal Tumour in Left hemithorax with shift of mediastinum

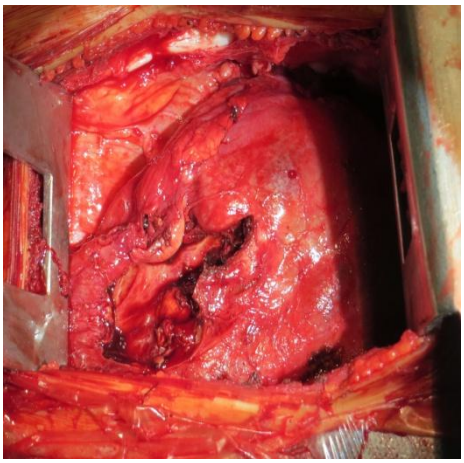

Figure 2: Large encapsulated Tumour with loculations

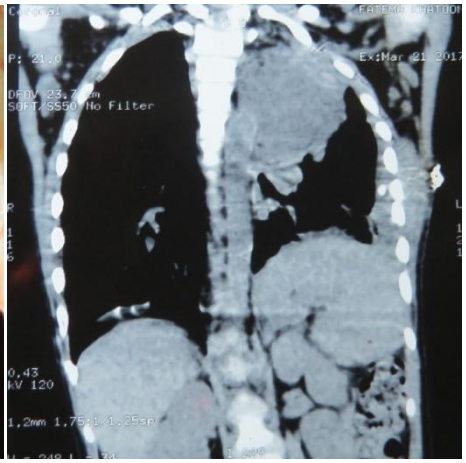

Figure 3: Postop early recurrence of tumour with refilling of cyst

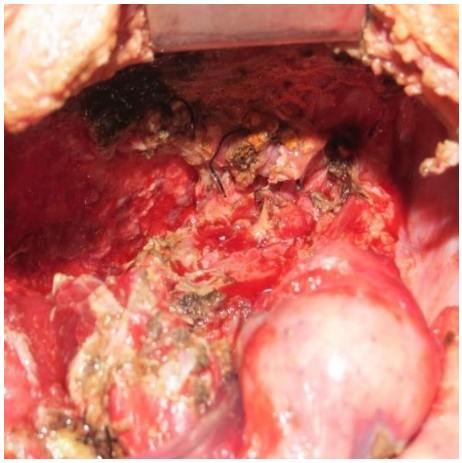

Figure 4: Completely resected tumour in second surgery

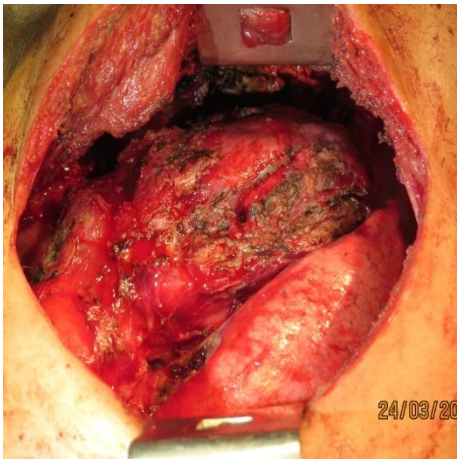

Figure 5: Fully expanded lung at end of 2nd surgery

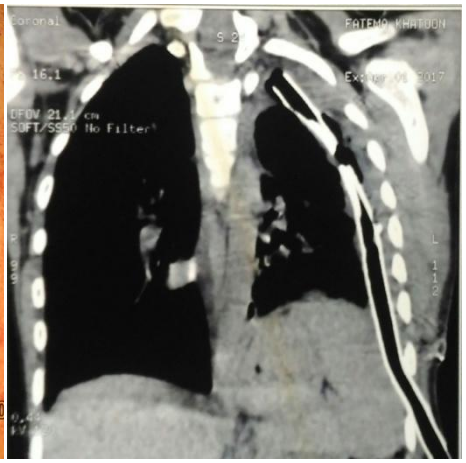

Figure 6: No tumour recurrence with expanded lung

DISCUSSION

Cystic lymphangioma (hygromas) are benign developmental anomalies of vasculolymphatic origin [3] resulting in blockage of lymphatic flow. They develop most commonly on the body surface, cervical region (75%) and axillary region (20%) being the commonest [1]. Hence majority of them are detected before the age of 2 years [1,3]. They can also be found in the retroperitoneum, pelvis, and chest wall [2].

Of cervical lymphangiomas only 2-3% may be associated with an intrathoracic extension. An isolated mediastinal lymphangioma without a cervical component is uncommon accounting for less than 1% [1-3]. Cystic lymphangioma make up 5-6% of mediastinal masses in children [1].

The majority of mediastinal cystic hygromas are located in the superior and anterior compartments. Posterior location is exceptional [1]. They are usually slow growing and are usually asymptomatic until they reach dimensions large enough to cause compression of adjacent structures causing respiratory distress, feeding difficulties, or vascular compromise or be a target for secondary infection [1-3]. Their rapid growth can also cause hemorrhage [2]. These symptoms are more common in children under 2 years [1].

In many cases patients may be asymptomatic and it may be an incidental finding in imaging studies [1,2].

Our patient presented in emergency with respiratory distress and had to be taken up for emergency surgery after partially relieving dyspnoea by drainage of hemorrhagic fluid relieving some compression.

Detection of a cystic lesion on a chest X-ray or a computed tomography scan of the chest are primary diagnostic indicators. Computed tomography of the chest demonstrates well-capsulated, smoothly marginated and cystic lesion[1] with involvement or deviation of natural structures, absence of calcification, and various high attenuation areas within the lesion [2,4]. Other cystic masses that must be considered in

the differential diagnosis include bronchogenic cyst, cystic thymoma, cystic teratoma, and malignant lymphoma [1,4].

Definitive diagnosis is made through histopathological examination [2]. CT scan in our patient showed large mass of mixed heterogeneity with areas of necrosis occupying entire left hemithorax with complete collapse of left lung and mediastinal shift to the right side.

Surgical excision is the treatment of choice for cystic lymphangioma [1,2,4]. A surgical approach via a thoracotomy is easy and provides excellent exposure for a meticulous dissection of all the cysts which is essential to prevent recurrence[1]. Some other types of adjuvant treatment, such as radiotherapy or injection of sclerosing agents (OK-432 or bleomycin), have been proposed. However they are still controversial [2,4] as a a reactionary increase in the size of the cysts can compromise respiration [1].

With complete tumor resection, the prognosis is good, although cases of local recurrence have been described, especially after partial resection [2].

In our patient, we were able to resect the entire tumour in piecemeal fashion other than a small part which was densely adherent to the subclavian vein in view of the general condition of the patient and nature of emergency surgery. A second surgery at 1 month for an increase in size of the residual tumour involved complete resection of the tumour. Child is asymptomatic and no recurrence at 3 month follow up.

The pathologic diagnosis of lymphangioma is made on the basis of the presence of dilated cystic spaces lined with endothelium, often containing lymphocytes [5]. Lymphangiomas are classified pathologically into three types, i.e. unilocular, cavernous and intermediate types [1].

Histopathological examination of specimen of our case showed cystic hygroma with xanthogranulomatous inflammatory lesion.

Postoperative complications can include infection, chylothorax, and fistula formation, as well as injury to the phrenic nerve, vagus nerve, lungs, or major vessels [2].

CONCLUSION

This is a case report of a rare case of an isolated mediastinal cystic lymphangioma with acute presentation with severe breathlessness probably due to hemorrhage within the lesion in a 7 year old child with timely surgery as life saving followed by a second exploration for complete curative resection of tumour.

Conflicts of Interest: No conflicts.

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