Isolated mediastinal cystic lymphangioma in children: About two cases

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\textbf{Summary}
Lymphangiomas are a heterogeneous group of benign vascular malformations of the lymphatic system composed of cystically dilated lymphatics. Most cystic lymphangiomas are found in the cervical region. Isolated mediastinal cystic lymphangiomas are very uncommon. Herein we present two pediatric cases of isolated mediastinal cystic lymphangioma. In the first case, a seven-year-old girl presented with chest pain and dyspnea revealing a giant anterior mediastinal cystic lymphangioma extending inferiorly into the upper part of the abdomen. In the second case, a four-week-old boy presented with an acute respiratory distress revealing a voluminous posterior mediastinal cyst lymphangioma. In the view of literature, these cystic lymphangiomas are accepted to be atypical because of their locations and large size.

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\textbf{Introduction}

Cystic lymphangioma, also known as cystic hygroma is a congenital malformation of the lymphatics resulting in blockage of lymphatic flow.\textsuperscript{1} As they develop in the body surface, cervical region (75%) and axillary region (20%), the majority of them are detected before the age of 2 years. Of cervical lymphangiomas only 2–3\% may be associated with an intrathoracic extension.\textsuperscript{2} An isolated mediastinal lymphangioma without a cervical component is an uncommon occurrence which account for less than 1\%.\textsuperscript{3}

Herein we present two pediatric cases of isolated mediastinal cystic lymphangioma. In the first case, a seven-year-old girl presented with chest pain and dyspnea revealing a giant anterior mediastinal cystic lymphangioma extending inferiorly into the upper part of the abdomen. In the second case, a four-week-old boy presented with an acute respiratory distress revealing a voluminous posterior mediastinal cyst lymphangioma.
Case reports

Case 1

A 7-year-old girl, with no significant previous medical history, presented to the pediatric department with dyspnea and left chest pain. Physically, she was febrile, mildly tachypneic. Pulmonary examination revealed a matity at the left side on percussion and a diminished breath sounds at auscultation. A chest radiogram showed a large homogeneous opacity in the left lower lung field (Fig. 1).

Investigations revealed the following: hemoglobin: 11.3 g/dl, white cells: 13,500, C Reactive protein: 105 mg/l. Serology of hydatid cyst was negative. Ultra sound showed a multicystic lesion in the lower third of the left hemithorax associated with a pleural effusion.

Computed tomography (CT) of the chest showed a voluminous 10 × 15 × 24 cm multilocular cystic lesion in the anterior mediastinum. This lesion extended inferiorly into the upper part of the abdomen through a diaphragmatic defect until the interhepato-diaphragmatic space (Fig. 2).

The radiologic appearance suggested a diagnosis of lymphangioma of the mediastinum. Laparoscopy was performed. A very large cystic lesion was found. It compresses the segment II of the liver. There was discharge of hemorrhagic fluid at fine-needle aspiration of the cyst. The dissection of the mediastinal mass over the diaphragmatic defect was performed without thoracoscopic procedure. Than the diaphragmatic defect was sutured. Complete excision of the cystic lymphangioma was performed.

Histopathology confirmed it to be a lymphangioma. The post-operative course was uneventful, and she was discharged on the seventh post-operative day. At discharge, chest radiographic findings were normal. The patient is asymptomatic three years after surgical excision with no recurrence.

Figure 1 Chest X-ray showing a large left-sided mediastinal mass.

Case 2

A 4-week-old boy weighing 3.85 kg with no significant previous medical history, was admitted to the Pediatric Department for dyspnea, fever, cough, in context of epidemic of viral bronchiolitis.

Physical finding were: tachypnea (a respiratory rate of 70 breaths/mn), tachycardia, intercostal and suprasternal retraction. Rhonchi and crackles rales were revealed at auscultation.

A chest radiogram showed a bilateral pulmonary hyperinflation associated at a homogeneous opacity in the upper part of the right lung considered as an atelectasis.

Respiratory symptoms persist, and 48 h later, the neonate developed an acute respiratory distress. Blood gas results were the following: pH: 7.15; P\textsubscript{CO\textsubscript{2}}: 56 mmHg and P\textsubscript{O\textsubscript{2}}: 96 mmHg. The baby was immediately managed by tracheal intubation and controlled mechanical ventilation. After 17 days of ventilation, he was extubated, but the acute respiratory distress had recidived and the neonate was reventilated.

A chest radiogram showed a bilateral pulmonary hyperinflation associated at a homogeneous opacity in the upper part of the right lung (Fig. 3). The diagnosis of cystic lymphangioma was suspected.

Under general anesthesia, a left postero-lateral thoracotomy was performed. Intraoperatively, there was a large multicystic mass of the posterior mediastinum, this mass displace and compresses the left lung, encompasses the aorta and the esophagus and exerted to the diaphragm, the mass extend to the right side of mediastinum. To facilitate dissection, the mass was decompressed with a trocar with removal of a clear fluid. Complete excision of a macrocystic lymphangioma were performed. A thoracostomy tube drain was left in situ.

Figure 2 CT scan of the chest showing a voluminous multilocular cystic lesion in the anterior mediastinum.
symptoms. In the second case an acute respiratory distress compression, and hemoptysis are found.6

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chest pain, acute respiratory distress, and respiratory presenting symptoms are respiratory: dyspnea, cough,

thed mediastinal cystic lymphangioma, the most common location is exceptional.2,5,7 In our first case, a giant cystic

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lymphangioma was located in the anterior mediastinum which extended into the abdomen. And in the second case,
giant cystic lymphangioma of the posterior mediastinum is reported.

These mediastinal cystic lesions are mostly asymptomatic, and the diagnosis usually is entertained only after mediastinal widening is noted in chest radiogram.3

However, in children under two years of age with isolated mediastinal cystic lymphangioma, the most common presenting symptoms are respiratory: dyspnea, cough, chest pain, acute respiratory distress, and respiratory failure. Rarely symptoms like recurrent respiratory symp-
toms, dysphagia, stridor, vocal cord paralysis, venous compression, and hemothysis are found.6–13

In the first case, chest pain and dyspnea were the main symptoms. In the second case an acute respiratory distress was revealing a giant mediastinal lymphangioma in a neonate.

A post-operative chest radiograph finding were normal. Histopathology confirmed the diagnosis of cystic lymphangioma. The post-operative course was uneventful. Seven-years follow-up showed no evidence of recurrence.

Discussion

Lymphangioma are a heterogeneous group of benign vascular malformations of the lymphatic system, composed of cystically dilated lymphatics.4 Most cystic lymphangioma are found in the cervical region and only 1–2% extend into the mediastinum.1

Isolated mediastinal cystic lymphangiomas are very uncommon, they represent less than 1% of all cystic lymphangioma.2 Cystic lymphangioma make up 5–6% of mediastinal masses in children.5

Most of mediastinal cystic lymphangiomas are not diagnosed during childhood until growing to be very large, and three quarters of these lesions are diagnosed in adults.2,6

The majority of mediastinal cystic hgyromas are located in the superior and anterior compartments. Posterior location is exceptional.2,5,7 In our first case, a giant cystic lymphangioma was located in the anterior mediastinum which extended into the abdomen. And in the second case, a giant cystic lymphangioma of the posterior mediastinum is reported.

Pathologically, lymphangiomas are classified into three types, i.e unilocular, cavernous and intermediate types.14 In order to correlate pathological and imaging findings, Charruaa et al.,17 have studied CT an MR imaging features of mediastinal lymphangioma in adults. They conclude that the most common unilocular type is a non-enhancing-walled mass on CT. A less frequent cavernous type can be suggested based on a multiseptated and lobulated mass on CT and/or MR examination.17

Spontaneous regression of mediastinal cystic lymphangioma has not been reported. Usually a progressive increase in size occurs and, in most cases, it would not be prudent to recommend observation of a cystic lymphangioma of the mediastinum.3

Surgical excision is the treatment of choice.6,7,18 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.11 Since lymphangioma is a benign tumor, but infiltrating the adjacent vital structures must be delineated and safeguarded.11

A good prognosis could be achieved if the tumor is completely resected.6,11,19 Thoracoscopic treatment of mediastinal cystic lymphangioma has been reported.20 But compressive cysts with lung distension and mediastinal shift remain a contraindication for these procedure.20 Unlike cervical lymphangiomas, treatment of mediastinal lymphangioma with intralesional OKT-432 is a limited option.
since a reactionary increase in the size of the cysts can compromise respiration.21

In conclusion, an isolated lymphangioma of the mediastinum in the pediatric population is exceedingly uncommon.

Here we reported two atypical cases because of age at diagnosis in the second case (a neonate with acute respiratory distress), their location (a posterior mediastinal cyst lymphangioma) and their large size exceeding 10 cm of diameter in the second case, and a giant thoracoabdominal cyst lymphangioma in the first case. In both cases, complete resections were performed without any complications and without any recurrences.

Conflict of interest statement

None of the authors have a conflict of interest to declare in relation to this work.

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