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Neonatal Mediastinal Teratoma: A Cause of Neonatal Respiratory Distress

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Mediastinal teratoma is a rare cause of neonatal respiratory distress and frequently requires urgent surgical management. Arising from any structure in the mediastinum, they produce life-threatening respiratory and vascular insufficiency and present unique surgical challenges for the pediatric surgeon.

We report a one-month-old child who presented to the emergency department of Children's Hospital Number 2 (Bệnh Viện Nhi Dồng 2), Ho Chi Minh City, Vietnam with severe respiratory distress. Following stabilization and appropriate imaging, he underwent emergency thoracotomy and excision of the large hemithoracic mass. Histology confirmed a mature benign teratoma.

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Mediastinal mass is the most common intrathoracic lesion in the pediatric population[1]. They include a wide spectrum of cystic, solid, vascular and neurogenic origin. Comprised of three compartments, mediastinal masses could be divided anatomically into anterior, middle and posterior. Germ cell tumors arise from stem cells that failed to migrate to the gonads and are located in the anterior mediastinum. They commonly contain all three embryological layers: endoderm, mesoderm and ectoderm. Teratomas account for 50%-70% of such tumors[2].

Histologically, teratomas are broadly divided into mature well-differentiated tumors, immature poorly-differentiated tumors and those with malignant transformation[3]. Prenatal diagnosis of a mediastinal mass is often challenging, and its presentation is directly related to the size of the teratoma and its local effects on intrathoracic structures.

The aim of this presentation is to report a case of neonatal mediastinal teratoma causing neonatal respiratory distress.

THE CASE

A nearly one-month-old male child presented as an emergency to Children's Hospital Number 2, (Bệnh Viện Nhi Dồng 2), Ho Chi Minh City, Vietnam with severe respiratory distress.

The patient was immediately intubated and ventilated. A chest x-ray showed a marked displacement of the mediastinum to the right with a white out of the left hemithorax with obvious calcification, see figure 1. The compromised cardiovascular function had improved following intubation, but subsequent CT scan showed persistent and significant mediastinal displacement, see figure 2.

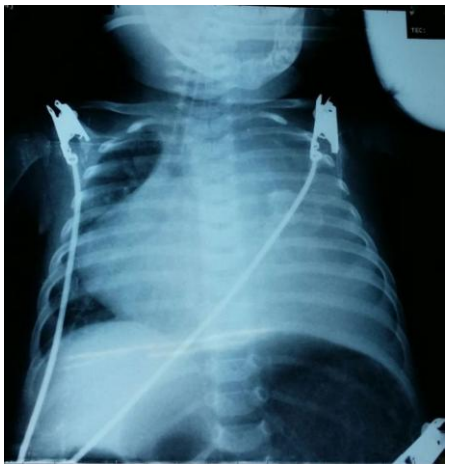

Figure 1: Chest X-ray on Admission

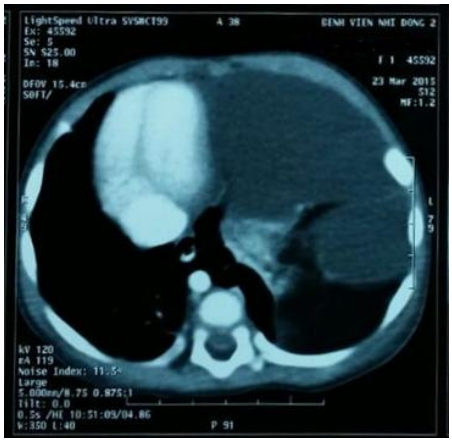

Figure 2: CT Chest Preoperatively

CT confirmed a large hemithoracic tumor with solid and cystic components and heavy calcification. The underlying left lower lobe was markedly collapsed. Hematological investigations were normal apart from markedly elevated alpha feto protein (αFP).

A left thoracotomy was performed; a large solid/cystic tumor was filling the entire left chest was excised in toto. It was necessary to aspirate the cystic component of the tumor to allow dissection and complete excision of the tumor from the left lung hilum, see figure 3.

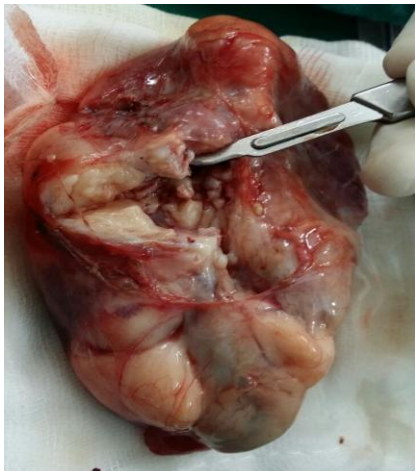

Figure 3: The Excised Tumor

The chest was closed with the placement of an underwater seal chest drain, see figure 4. The remainder of the postoperative course was uneventful, and he was discharged after five days. Histopathology confirmed a benign solid and mature cystic teratoma.

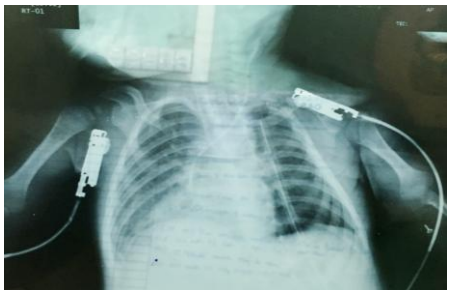

Figure 4: Postoperative Chest x-ray

DISCUSSION

Germ cell tumors are the third most common mediastinal masses in neonates after neurogenic tumors and foregut cysts. They are defined as benign or malignant teratomas[4]. Accounting for 3%-12% of mediastinal tumors, they present with local symptoms of compression or obstruction of a major vessel or airway, during which immediate intervention is required[5]. The size and location of the mediastinal mass plays a major role in the presentation, as well as tumor rupture, infection and cardiovascular compromise, these manifestations are more evident in children owing to limited compensatory reserve.

The majority of benign teratomas arise in the anterior mediastinum especially in neonates. Prenatal diagnosis is difficult due to the small size of the mass and more than often it is diagnosed in the acute setting when the child presents with respiratory compromise. Routine antenatal ultrasound (post 23 weeks) should always be attempted to exclude the presence or absence of mediastinal masses despite their rarity[6]. This allows antenatal surveillance as many apparent lesions disappear, but important complications such as cardiovascular compromise (hydrops fetalis) can be detected early. Awareness of an existing mediastinal lesion facilitates surveillance and elective intervention. This patient presented as an emergency with no antenatal history.

Early surgical excision is the most appropriate treatment using either a median sternotomy or a thoracotomy incision, but a sternotomy can also be converted to a thoracotomy if needed as in a trap door incision[7]. In our case, the fifth intercostals space thoracotomy incision provided adequate access to the tumor and feeding vessels. It is important that the lesion be completely removed to minimize the risk of malignant transformation in later life[8].

Preoperatively, the surgeon must be prepared to face technical challenges in tumor excision. These challenges occur when the teratoma arises from any mediastinal structure such as major vessels of the heart and the pericardium. In a major series of late presentation of 17 mediastinal teratomas in adults (age 15-57 years) invasion of the pleura and pericardium was commonly encountered. In four patients, the malignant transformation had occurred, and all died[8]. In our patient, the teratoma was not adherent or invading the chest wall or pleura; however the challenge lay in identifying its blood supply which arose from the hilar vessels. Aspiration of the cystic component of the teratoma allowed for good visualization and access to the hilar vessels. This technique allowed safe and swift excision of the tumor.

CONCLUSION

Pediatric mediastinal teratomas are commonly asymptomatic unless they grow to a sizable mass and cause compressive symptoms. The size of the tumor correlates to the time of presentation; however, some cases are found incidentally. The majority of mediastinal teratomas are benign, but surgical excision remains the management cornerstone. Tumor excision provides immediate relief from compression on intrathoracic structures but equally important prevents malignant transformation in a teratoma. Imaging is a crucial part of diagnosing and managing mediastinal masses. Chest x-rays and CTs provide details on size, location, nature and invasion of the tumor, which are key elements in planning an effective tumor excision.

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