Immature Mediastinal Teratoma - A Cause of Recurrent Respiratory Distress

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Abstract:

The word Teratoma is derived from Greek words "terato" meaning monster and "onkoma" meaning swelling. A teratoma is a germ cell tumour derived from pluripotent cells and is made up of elements of different types of tissues from one or more of the three germ cell layers. The worldwide incidence of teratoma is about 1 in 4000 live births (Shetty et al, 2000). Frequencies of the most common sites are sacrococcygeal (40%), ovary (25%), testicle (12%), brain (5%) and others including neck and mediastinum (18%).

Thoracic masses arising from the mediastinum are a common diagnostic difficulty in the paediatric age group. These masses represent a wide variety of histological morphologies and require many different forms of surgical & non-surgical treatment.

The choice of appropriate operative procedure depends on patient’s age, tumour size, location, expansion, co-existent morbidity and cardio-respiratory reserve, Surgeon’s experience gives a favourable outcome and long term prognosis (Zisis et al, 2005).

Children especially those with benign tumours have very high survival rate after surgical resection (Grosfeld et al, 1994). Herein, we report a case of an infant who was diagnosed and treated for benign mediastinal teratoma with emphasis on its clinical presentation.
Case Report:

A 5 month old male baby weighing 5.5 kg was admitted in pediatric emergency with complaints of low grade fever, cough and breathlessness for last 8 days. He had such repeated episodes in the past, each of them lasting for 4-5 days since the age of 1 month. He was repeatedly treated by general practitioner as a case of pneumonia and given antibiotics along with steroids during each episode. On admission, physical examination revealed that infant was afebrile, with heart rate of 140/mm. and respiratory rate of 70/min. His blood pressure was 60/40 mm Hg in right upper arm. Oxygen saturation was 77% without oxygen on admission which increased to 89% with 3 litre / minute of oxygen. Systemic examination revealed marked intercostal and substernal, retraction: percussion note was impaired in infraclavicular and mammary region in left side with diminished air entry on left side and normal air entry on right side of chest. Rest of systemic examination was within normal limits.

After clinical assessment, provisional diagnosis of pneumonia with hydropneumothorax. Oxygen supplementation, intravenous fluid, intravenous antibiotics and bronchodilator were administered. His complete blood count, electrolytes, liver and renal function tests were within normal limits. X-ray chest(PA view) showed well defined homogenous rounded opacity on left side involving mid and upper zone causing mediastinal shift towards right side.

A tentative diagnosis of mediastinal mass of unknown origin was made. The transverse section of contrast enhanced CT scan of thorax showed a large well defined, heterogeneous, hypodense mass measuring 7x6 cm with few areas of calcification. The mass was causing compression over left main bronchus and displacing the mediastinal structures. (Fig. II)

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Baby was operated on day 5 admission. Through left lateral thoracotomy a mass measuring 8x6x4 cm arising from upper part of posterior mediastinum was removed in toto and the pedicle was arising directly from the aorta which was carefully separated and ligated. (Fig. III)

The mass was pushing the heart towards right side and left upper and middle lobes of lung were compressed. (Fig. IV)

The post operative period was uneventful. X–ray chest on second post–operative day showed complete expansion of the left lung. He was discharged in stable condition on 12th post–operative day. (Fig. V)

Gross examination of the resected specimen measured 8x6x4 cm and weighed 200 gms. It had a glistening capsule. On cutting the specimen a gritty sensation was felt. Cut surface had variegated appearance showing solid mucin filled cystic areas, haemorrhagic areas, cheesy pultaceous material.
Microscopically, multiple sections showed various mature tissues of three germ layers arranged haphazardly like intestinal mucosa, pseudo-stratified ciliated columnar epithelium, stratified squamous epithelium with adenexal structures like sebaceous glands, sweat glands, hair roots and keratin filled cystic areas. It also showed fatty tissue, cartilaginous tissue and neural tissue. These histopathological findings were consistent with "Immature teratoma. At the 8 months follow up child is healthy without any recurrence.

Discussion:

The mediastinum is second most common site for teratomas in the paediatric population (Friedmann et al, 2003). Primary mediastinal teratomas account for approximately 8-20% of mediastinal neoplasms (Lancaster et al, 1997). They are rare tumours in childhood accounting for only 7% of all germ cell neoplasm (Lakhoo et al, 1993). Approximately 8% of mediastinal tumors are benign teratoma; 82% of these are in the anterior, 4% in the posterior and 14% in the middle mediastinum (Lewis et al, 1983).

Most of the symptoms due to mediastinal teratoma result from compression of adjacent structures (Savras et al, 2005). Our patient presented chiefly with respiratory problem consistent with other reports in the literature (Tansel et al, 2006). The most important factors affecting the management of a patient with a mediastinal mass are the nature of the disease, age, presenting symptoms & the location of the mass.

So, the differential diagnosis of posterior mediastinal mass includes lymphoma, thymoma, bronchogenic cyst, neurogenic cyst and esophageal duplication. Calcification (occasionally teeth) noted in a tumor as seen on chest X-ray & CT Scan are consistent with the diagnosis of teratoma until proven otherwise. In addition, thymic, bronchogenic, or pericardial cyst can also be differentiated sonographically by the presence of a thin wall in contrast to a teratoma which is always a thick walled cystic lesion (Wu et al, 2002). Microscopically, mesodermal, ectodermal and endodermal elements are seen in varying proportion. Pulmonary teratomas are mostly composed of mature, cystic, somatic tissue, although malignant elements may occur (Saini et al, 2006).

The surgical approach to these tumours has changed significantly over time. In the past, death was usually related either to the mass effect of the tumor or the complication of general anesthesia. With refinement in surgical technique & anesthetic management, the mortality rate has significantly decreased following surgical intervention. Mature mediastinal teratomas are benign, do not infiltrate adjacent organs, and can be resected completely with good results (Chen et al, 2007). After complete resection of mediastinal cysts & benign tumors the prognosis is generally excellent. In a review of 153 children with non-testicular mature teratomas, the 6 year relapse–free survival for completely resected teratoma was 96% as compared to 55% for incompletely resected teratoma (Goblet et al, 1998).
Conclusion:

In early infancy, mature mediastinal teratoma can cause life-threatening respiratory distress and therefore, should be considered in differential diagnosis of recurrent respiratory distress.

References:


