Mature Mediastinal Teratoma- A Rare Cause of Recurrent Respiratory Distress

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Abstract:

Teratomas are interesting because of their obscure origin, bizarre microscopic appearance and sometimes unpredictable behaviour. Although they occur infrequently in children, clinician should be aware of their clinical features, natural history, pathology and principles of treatment. In an attempt to document some of the features , we report a case of an infant with large mediastinal teratoma presenting with severe and recurrent respiratory distress. Patient was operated successfully with no recurrence in 1 year follow up.

Key Words: Infant, Mature Teratoma, Mediastinal mass, Respiratory Distress.

Introduction:

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 made up of elements of different types of tissue from one or more of the three germ cell layers. The world wide 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 mediastinum are a common diagnostic dilemma in 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, cardiorespiratory 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 it's clinical presentation.

Case report:

A 4 month old male baby weighing 5.43 kg was admitted in peadiatric emergency with complaints of low grade fever, cough and breathlessness for last 8 days. He had recurrent suchepisodes in the past, each

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of them lasting for 4-5 days since the age of 10 days. 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, cyanosed, having cushingoid facies with heart rate of 140/min. and respiratory rate of 70/min. His blood pressure was 60/40mm 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 subcostal retraction; percussion note was impaired in infraclavicular and mammary region in right side with diminished air entry on right side and normal air entry on left side of chest. Rest of systemic examination was within normal limits. After clinical assessment, provisional diagnosis of pneumonia was made. Oxygen supple-mentation, intravenous fluid, intravenous antibiotics and bronchodilator were administered. His complete blood count electrolytes liver and renal function test were within normal limits. Arterial blood gas analysis showed PCO₂ of 26 mm Hg, PO₂ as 98 mm Hg and pH 7.4 suggestive of hyperventilation. X -ray chest (PA view) showed well defined homogenous rounded opacity on right side involving mid and upper zone causing mediastinal shift towards left upto the lateral chest wall (Fig.IA). Lateral view of chest revealed that the mass was extending anteriorly upto retrosternal space and posteriorly upto prevertebral space (Fig.1B). A tentative diagnosis of neoplastic 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 9x7cm with multiple areas of calcification. The mass was causing compression over right main bronchus and displacing the mediastinal structures (Fig. II).

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a) Postero-anterior View

Fig. IA & IB: Radiographies showing turnour mass originating from mediastium



Fig. II: CT Scan showing hypodense mass with multiple areas of calcification.

Baby was operated on day 3 by pediatric surgeon. Through right lateral thoracotomy a large mass measuring 10x8x6 cm arising from upper part of anterior mediastinum was removed in toto and the pedicle was ligated. The mass was pushing the heart towards left side and right upper and middle lobes of lung were compressed. The child was ventilated electively for 12 hours.

The post-operative period was uneventful. Xray chest on first post-operative day showed complete expansion of the right lung (Fig.III). He was discharged in stable condition on 12th post-operative day.

Gross examination of the resected specimen measured 10x8x6 cm and weighed 500 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 and bunch of hairs (Fig. IV). Microscopically, multiple sections showed various mature tissues of three germ layers arranged haphazardly like intestinal mucosa, pseudostratified ciliated columnar epithelium, stratified



Fig.III: X-ray chest PA veiw on day 1 of Post Operative period.



Fig. IV: Photograph showing resected mass of teratoma.

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 (Fig.V). These histopathological findings were consistent with "mature teratoma. At the 1 year follow up child is still healthy without any recurrence.



Fig. V: Microphotograph showing three germ layer.

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 (Savas 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.

Almost all masses in the anterior mediastinum consist of lymph nodes, thymic masses, germ cell tumors or thyroid. So, the differential diagnosis include lymphoma, thymoma and bronchogenic cyst. Calcification (occasionally teeth) noted in a tumor as seen on chest X-ray & CT scan are consistent with the diagnosis of teratoma until proven other wise . 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 tumour 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 infilterate 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 nontesticular 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.

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