Pulmonary Blastoma in a Child with Recurrence: A Case Report and Literature Review

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Abstract
An 8 year old child treated case of pleuropulmonary blastoma a very rare primary pulmonary malignancy in children presented with fever, respiratory distress and bulging chest wall. He had typical clinical and radiological features and ultimately succumbed to illness.

Introduction

Pulmonary blastoma, termed as 'embryoma' is a rare primary tumour even rarer in children. It has a uniformly poor prognosis and a typical clinical course and radiologic features.

Case Report

A then 5 year old child had presented 3 yrs ago with fever, cough with expectoration and was managed conservatively, the symptoms deteriorated and a CT was requested which showed a large right sided pneumothorax with complete collapse of the ipsilateral lung and contralateral shift of mediastinum to the left. Right pleural based soft tissue mass lesion was also noted and further investigation was advised.

An ICD was inserted for symptomatic relief and a follow up CT showed complete resolution of pneumothorax with partial loculated pleural effusion and collapse consolidation of the right lower lobe. Fluid was aspirated and sent for bacteriological investigation, patient put on empirical AKT assuming the lesion to be an empyema.

A follow up radiograph and CT showed complete opacification of the right hemithorax with a large heterogeneously enhancing pleural/extra pleural mass lesion involving the entire hemithorax and a suspicion of Askin’s tumour was raised. However biopsy was carried out which showed pleuro pulmonary blastoma with rhabdomyoblastic differentiation.

On subsequent surgery an organized partially necrotic mass filling the right pleural cavity was removed. Persistence of symptoms led to a repeat CT which revealed a large residual lesion.

The patient was started on chemotherapy (3 mt), chemoradiation (3 mt). A follow up CT showed significant resolution, residual nodular lesion with calcification, adjacent fibrosis and cystic change and mild pleural effusion.

A further surgical or medical management was debated, patient was put on maintenance chemotherapy (9 mts). Post chemo completion CT showed a residual loculated collection with peripheral calcification. Patient was followed up with monthly radiographs which showed no detectable change. A PET scan was advised but the patient was lost to follow up.

After 2 years the patient now presented with fever, respiratory distress and bulging anterior chest wall. Radiograph revealed an increase in the size of radioopacity. An ultrasound examination showed large lesion with heterogenous echogenicity with calcific foci displacing the liver inferiorly with surrounding pleural effusion. A CT scan showed typical 12 x 12.7 x 7.5 cm large hypodense lesion with whorls of hyperdensity showing heterogeneous post contrast enhancement with peripheral calcification, pushing the liver inferiorly, causing collapse of the underlying lung, pleural and pericardial involvement and loss of fat planes with vessels. However no bony destruction was noted nor was any distant metastasis (Fig. 6).

The patient was being managed conservatively with further investigations but there was sudden rapid deterioration and patient succumbed to cardiorespiratory failure. No autopsy was performed.
Fig. 1: Topogram and CT image showing pneumothorax

Fig. 2: CT images showing hypodense collection with an ICD

Fig. 3: CT topogram showing complete opacification of hemithorax due to a pleural based mass lesion
Fig. 4: Post op CT showing large residual mass lesion

Fig. 5: Post chemo completion CT showing near complete resolution of mass lesion, with residual lesion showing peripheral calcification, follow up radiograph showing no change in residual mass
Discussion

Pulmonary blastoma was termed as 'embryoma of lung' by Barnard in 1952 based on histologic similarity to foetal lung. In 1961 Spencer coined the term pulmonary blastoma postulating that the tumour originated from primitive pluripotential mesoderm. Pulmonary blastoma having a peak incidence in 3rd and 4th decade is not very rare in paediatric age group. Most of the paediatric patients are symptomatic at presentation, the symptoms being fever, tachypnoea, respiratory distress, haemoptysis.

Imaging features

Most commonly lung neoplasms in children are metastatic disease, like metastasis from Wilm's tumour, osteosarcoma, Ewing's sarcoma and rhabdomyosarcoma. Primary neoplasms of the lung are rare. These include papilloma, hamartoma, carcinoids which are

Fig. 6 : Radiograph and CT showing large recurrent mass lesion with typical radiological features
benign. Primary malignant neoplasms include various sarcomas and pulmonary blastomas. Of these pulmonary blastomas and sarcomas are peripheral in location.\textsuperscript{2,3}

Pulmonary blastomas at presentation on radiographs may show pneumothorax, small pulmonary nodule to large opaque hemithorax. Larger lesions are more common in children than adults. Punctate calcification readily visible on CT may be noted on radiographs at presentation but usually develop following chemotherapy.

Ultrasound is superior to radiographs as it shows heterogeneous echogenicity mass distinctly from the pleural effusion.

CT scan typically shows low attenuating lesion with projections and whorls of high attenuation solid tissue. Such lesions may be confused with empyema but are considered characteristic of pulmonary blastoma. Association with congenital cysts of the lung is reported, but such lesions may be a component of tumour itself. Pneumothorax arises when these cysts rupture.\textsuperscript{5,6}

**Pathology**

Colby et al have divided pulmonary blastoma into three subtypes up\textsuperscript{1}

1. WDFA (PET) epithelial variety is rare in children with average age in 5\textsuperscript{th} decade. Prognosis is better than the classic variety.

2. Classic/Biphasic pulmonary blastoma: They have tubular (epithelial) components and immature mesenchymal components. Grossly it is a nodular tumour in the periphery of the lung. It is considered to be a specific form of carcinosarcoma. They may have a p53 mutation and have a poorer prognosis.

3. Pleuro pulmonary blastoma is seen in children younger than 10 yrs and is a solid cystic sarcoma. Cysts are lined by metaplastic epithelium producing a pseudobiphasic pattern.

The tumour shows features of other mesenchymal derivatives.

Pulmonary blastomas are classified as “Carcinomas with pleomorphic sarcomatoid or sarcomatous elements” according to WHO classification.\textsuperscript{7}

**Management and Prognosis**

Surgery is the preferred treatment. Combination chemotherapy and radiotherapy can be given. Prognosis is poor regardless of age if size at presentation is greater than 5 cm. Most patients die within 2 yrs of presentation despite therapy. Metastatic disease though uncommon at presentation is frequently found at autopsy.\textsuperscript{5,6}

**References**

7. Sternberg’s Diagnostic Surgical Pathology 3\textsuperscript{rd} edn.