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Case Report

Radiology Section

Imaging in a Case of Extensive Paediatric Mediastinal Lymphoma Presenting as Haemothorax

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ABSTRACT

Lymphomas are the third most common paediatric malignancies and the most common paediatric mediastinal masses. On imaging they present as large, lobulated, homogenous, predominantly anterior mediastinal masses with possible extension into other mediastinal compartments and vascular encasements. Effusions, lung involvement and extensive disease are more common in paediatric patients; however haemothorax is an uncommon occurrence. In our case, multi-compartmental involvement, vascular encasement and haemothorax were the striking features.

Keywords: Chest imaging, Paediatric oncology, Thoracic mass

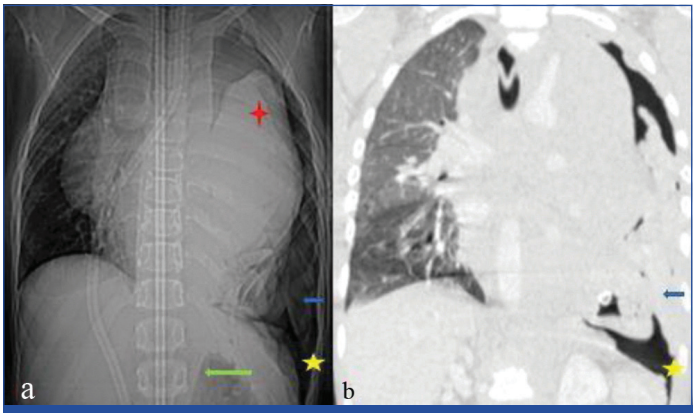
CASE REPORT

A 13-year-old male child presented with history of difficulty in breathing and chest pain on exertion since 15 days which was aggravated since the last two days. He had no complaints of cough, fever and weight loss, loss of appetite or past tuberculosis. An incidental history of past trivial trauma to chest was provided which occurred two weeks ago.

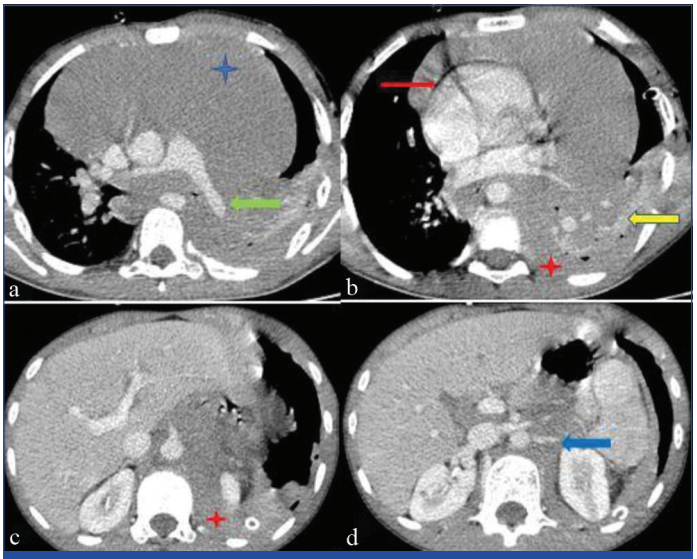
On preliminary chest X-ray done at a peripheral centre, a large left sided pleural collection was seen which was causing tracheal and mediastinal shift to the right. A chest tube was put which drained ~1000 mL of haemorrhagic fluid. Patient was referred to our centre for further management.

On examination, the patient had reduced breath sounds and palpable cervical nodes. Blood profile suggested a raised total leukocyte count with a normal differential count. Pleural fluid analysis suggested haemorrhagic fluid without any malignant cells or tubercular markers.

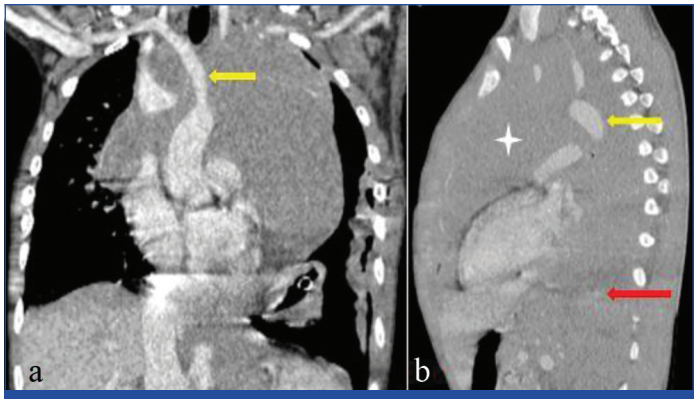
Contrast CT scan done to determine the cause of haemothorax which revealed a large left sided pneumothorax with minimal pleural collection and collapsed left lung [Table/Fig-1]. A large, well defined, multi-lobulated, homogeneous minimally enhancing iso-dense lesion was noted involving all compartments of the mediastinum, predominantly involving superior and anterior mediastinum, extending into middle as well as posterior mediastinum. The centre of the lesion was in the anterior mediastinum with a maximum width of 14.5 cm. The lesion was extending from thoracic inlet to the diaphragm encasing all major vessels (aorta with its branches, pulmonary vessels as well as bilateral internal jugular veins, superior vena cava as well as bilateral brachiocephalic veins), however, there was no evidence of luminal compression or thrombus in these vessels. It was occupying and widening the retrosternal space and involving the costal pleura along the anterior rib cage on the left side with focal bulge into the chest wall at the level of 5th intercostal space. Antero-inferiorly, lesion was involving the diaphragmatic pleura in the left hemi-thorax, abutting the left lobe of liver. Medially, the lesion was closely abutting and encircling the pericardium. Laterally, in the left haemothorax, lesion was compressing the adjacent left lung as well as left main bronchus, hence being the possible cause for left lung collapse. In the posterior mediastinum and abdomen, it was extending as paravertebral soft tissue from the level of arches of aorta upto the renal arteries; predominantly on the left side. Lesion was encasing the oesophagus, descending the thoracic aorta, the proximal coeliac trunk, left renal artery and left renal vein [Table/Fig-2,3].



[Table/Fig-1]: a: CT Tomogram shows left sided pneumothorax (Yellow Asterix) with collapsed lung (Blue Arrow). Widening of superior mediastinum seen with large soft tissue mass having lobulated contour causing shift of mediastinum to right (Red Asterix). Widened para-spinal soft tissue is seen (Green Arrow). b: Coronal Lung Window images confirm collapsed left lung with pneumothorax.



[Table/Fig-2]: Axial Post Contrast Mediastinal Window Images show: (a) a large multi-compartment mediastinal mass lesion primarily located in anterior mediastinum (Blue Asterix), which is encasing vessels (Green arrow). (b) Pericardial encasement (Red Arrow) and possible left lung infiltration seen (Yellow arrow). (c and d) Posteriorly, lesion is extending into the posterior mediastinum and retroperitoneum (Red Asterix) encasing the renal vessels (Blue Arrow).



[Table/Fig-3]: Post-contrast mediastinal window: (a) Coronal reconstruction shows superior and anterior mediastinal homogenously enhancing soft tissue lesion encasing brachiocephalic artery (Yellow Arrow). (b) Sagittal reconstruction shows the lesion is occupying the retrosternal space (White Asterix). It is encasing the pericardium and large vessels along with extension into the posterior mediastinum and retroperitoneum (Red Arrow).

Final radiological opinion was a diffuse, homogenous, extensive soft tissue density lesion, without any necrosis, calcification or cystic changes, involving multiple mediastinal compartments; along with pleural and retroperitoneal involvement. The lobulated morphology of the lesion, extension along anatomical planes and nature to encase vascular structures without causing compression, suggested Lymphoma as the probable diagnosis.

A CT guided biopsy of the mediastinal mass was done which showed proliferation of medium to large sized atypical lymphoid cells with high nucleo-cytoplasmic ratio, deeply basophilic nuclei, inconspicuous nucleoli and scant cytoplasm suggesting a diagnosis of malignant small round cell tumour favouring Non-Hodgkin's Lymphoma of Mediastinum. Bone marrow biopsy showed hyper-cellular marrow which was diffusely infiltrated by atypical lymphoid cells suggesting marrow infiltration by lymphoma cells. Immunohistochemistry of mediastinal biopsy showed expression of CD3, CD4, CD8 and TdT markers on tumour cells. The blast cells expressed CD10 and CD34. This suggested a diagnosis of Non-Hodgkin's Lymphoma of T-lymphoblastic type. Patient was started on Allupurinol and Cyclophosphamide therapy, but was unfortunately lost to further follow-up.

DISCUSSION

Lymphomas comprise of 13% of all paediatric malignancies and are the most common paediatric mediastinal masses [1]. The role of imaging in a suspected paediatric mediastinal mass is to determine the compartment of origin, extension and morphological appearance, presence of any airway or vascular compression and associated secondary features [2].

In an extensive lesion involving more than one compartment, determination of compartment of origin may be done by localising the centre of lesion by measuring the largest lesion diameter in axial imaging. Also, evaluation of the pattern of displacements of adjacent structures is helpful in determining the lesion origin [3,4]. In our case, though extending into all mediastinal compartments, lesion centre was located in the anterior mediastinum.

Mediastinal lymphomas are the 3rd most common paediatric malignancy and the most common cause of anterior mediastinal masses in children comprising of almost half of all paediatric mediastinal masses [3]. Most mediastinal lymphomas are secondary to systemic disease. Hodgkin's lymphoma is the more common form of mediastinal lymphoma seen in children less than 10 years while Non-Hodgkin's Lymphoma is seen in both 1st and 2nd decades. Clinical presentations may include dyspnoea, stridor and SVC syndrome [4,5].

Lymphomas of anterior mediastinum often appear as discrete or conglomerated large nodal lesions, showing diffuse homogenous appearance which may compress the surrounding trachea or vascular structures. Necrosis, haemorrhage and calcifications are rare. In anterior mediastinum, they occupy and obscure the retrosternal space. While involvement of both anterior and middle mediastinum is common in adults, both anterior and posterior nodal chains are equally involved in paediatric patients [4,5].

Presence of extensive disease and effusions, both pleural and pericardial, are more common in paediatric patients in comparison to adults [5,6]. Pulmonary nodules which may be cavitating, diffuse interstitial thickening and effusions are also more common in Non-Hodgkin's disease [5].

Mediastinal lymphomas have multiple varied presentations. Lee SJ et al., found extremely vascular presentation of Thoracic Lymphoma [7]. Rafeeq B et al., observed in a case that lymphoma presented as a cardiac tumour [8]. Hokka D et al., while describing a spontaneous rupture of mediastinal thymoma in an adult, argue that non-traumatic haemothorax in a lymphoid tumour is extremely rare and occurs only in case of vascular erosion [9]. In our case, it is hence unclear whether the trivial trauma reported two weeks back had any effect on the clinical presentation of haemothorax or not.

Imaging differentials when the mass is limited to anterior mediastinum include, thymoma which shows a continuation with thymus and does not displace the trachea [3,5]. Seminoma is another imaging differential for a smooth homogenous anterior mediastinal mass [4]. Other germ cell tumours have a more heterogeneous appearance with fat or calcific densities within [10]. However, multi-compartmental involvement with diffuse encasement of vascular structures is strongly indicative of lymphoma [3-5].

Haemothorax has been a known occurrence with cases of choriocarcinoma and uncommonly with diffuse large cell lymphomas in adults [11,12]. Though, pleural effusions are common in lymphoid pleural involvements in children, haemothorax is uncommon.

CONCLUSION

Paediatric mediastinal, though are common in occurrence but lymphomas have a large spectrum of clinical and radiological presentations. Multi-compartmental mediastinal involvement and vascular encasement only are often clinchers for diagnosis.

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