CASE REPORT

Primary Pleural Inflammatory Pseudotumor in a Child

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Background: Inflammatory pseudo tumor, a rare non-neoplastic lesion, commonly presents as slow growing solid lesion in the lung, but many extra-pulmonary locations have been described. **Case characteristics**: A 4-year-old girl who presented with respiratory distress due to massive pleural effusion. Computed tomography revealed large hypodense non-enhancing lesion in the left hemi thorax. Surgical exploration revealed large semisolid pleural collection filled with gelatinous material with normal underlying lung. **Outcome**: Histopathology revealed spindle shaped cells with abundant myxoid stroma. Child recovered after surgery and was asymptomatic at 5 years follow-up. **Message**: Primary pleural inflammatory pseudotumor may be a rare cause of pleural effusion in a child.

Keywords: *Myxomatous tumor, Plasma cell granuloma, Pleural effusion.*

Inflammatory pseudotumors are rare non-malignant lesions; lung is a common site that can cause secondary pleural effusion. Primary involvement of pleura is extremely rare. We report primary pleural inflammatory pseudotumor in a child.

CASE REPORT

A 4-year-old girl was referred to our center for massive pleural effusion. She was treated for left sided empyema 5 months previously, with thoracotomy and decortication. In view of presence of myxomatous lesion in the pleural cavity, she was referred to us for further management. At the time of admission, she was febrile and tachypneic; trachea was shifted to right side with reduced air entry on left side. In view of previous operative details and recurrence of pleural effusion, a possibility of neoplastic lesion was considered. Pleural fluid analysis did not show any malignant cells.

Computed tomography of chest revealed large hypodense, non-enhancing lesion occupying the entire left hemithorax with collapse of the underlying lung with mediastinal shift to opposite side (***Fig.* 1**). During thoracotomy, left pleural cavity was found to be filled with extensive gelatinous, nodular, jelly-like lesion involving whole of the parietal pleura with partial involvement of visceral pleura, with normal underlying lung. Evacuation of the jelly like lesion with partial decortication was performed; post-operative period was uneventful. Histopathology revealed spindle shaped cells in an abundant myxoid stroma (***Web Fig.* 1)**. The cells exhibited mild atypia, wavy nuclei and abundant cytoplasm with occasional mitotic figures (***Web Fig.* 1**). Immuno-histochemistry revealed myxoid neoplasm positive for vimentin, CK, S100, desmin, myogenin, and negative for CD117 suggestive of inflammatory pseudotumor of the pleura. Histopathology of lung tissue was unremarkable (***Web Fig.* 1**). Patient was doing well at 5 years follow-up.

DISCUSSION

Inflammatory pseudotumor, also called plasma cell granuloma, is a rare benign tumor, accounting for 0.7% of all lung tumors [1,2]. It has a propensity to clinically

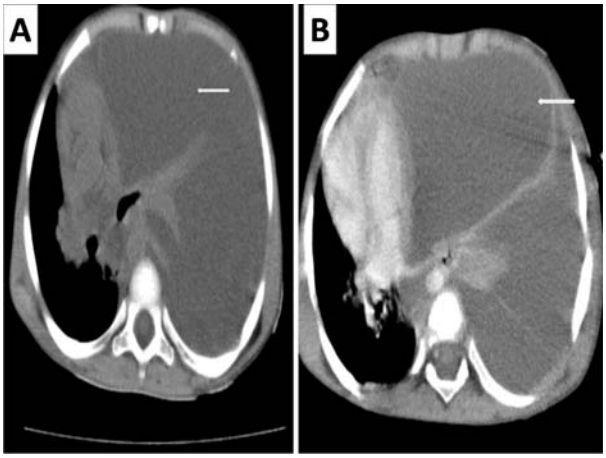

FIG.1 *Contrast-enhanced computed tomography of thorax (mediastinal window) showing large hypodense non enhancing mass occupying the entire left hemithorax, causing mediastinal shift to opposite side with underlying collapsed lung (arrow).*

and radiologically mimic a malignant disease. More than one-third of cases are closely related to recurrent respiratory infections caused by microorganisms such as Mycoplasma, Nocardia, Actinomycetes, Epstein Barr virus and human herpes virus [3]. These are most commonly seen in the lung and orbit, but have been reported from nearly every site in the body. Primary tumor arising from the pleura is rare, especially in children and in most cases pleura is involved secondarily [4].

Inflammatory pseudotumor has been reported in all ages, though less common in children, with slight predominance in females. Most patients present with nonspecific symptoms, and the tumor is discovered incidentally on a chest *X*-ray performed for other reasons [3]. In our patient, the initial presentation was like para-pneumonic effusion with fever, cough and breathlessness, and the patient underwent initial surgery for suspected empyema. Radiological findings were suggestive of massive pleural collection with collapse of underlying lung. Our patient had exclusive pleural involvement, without involving underlying lung parenchyma or mediastinal structures. Evacuation of gelatinous material with pluerectomy resulted in complete cure.

Diagnosis of inflammatory pseudotumor is mainly by histology that is characterized by myofibroblastic spindle cells mixed with a hyalinized stroma with inflammatory infiltrates. The presence of abundant myxoid stroma in our patient was unusual.

Surgical excision of the tumor remains the standard treatment as most tumors are amenable for complete removal. Reports of recurrence of tumor have been noted as late as 11 years after surgery, and this highlights the need for long-term follow-up [5]. Other modalities of treatment include chemotherapy with steroids, non-steroidal anti-inflammatory drugs, immunomodulation and radiotherapy [6].

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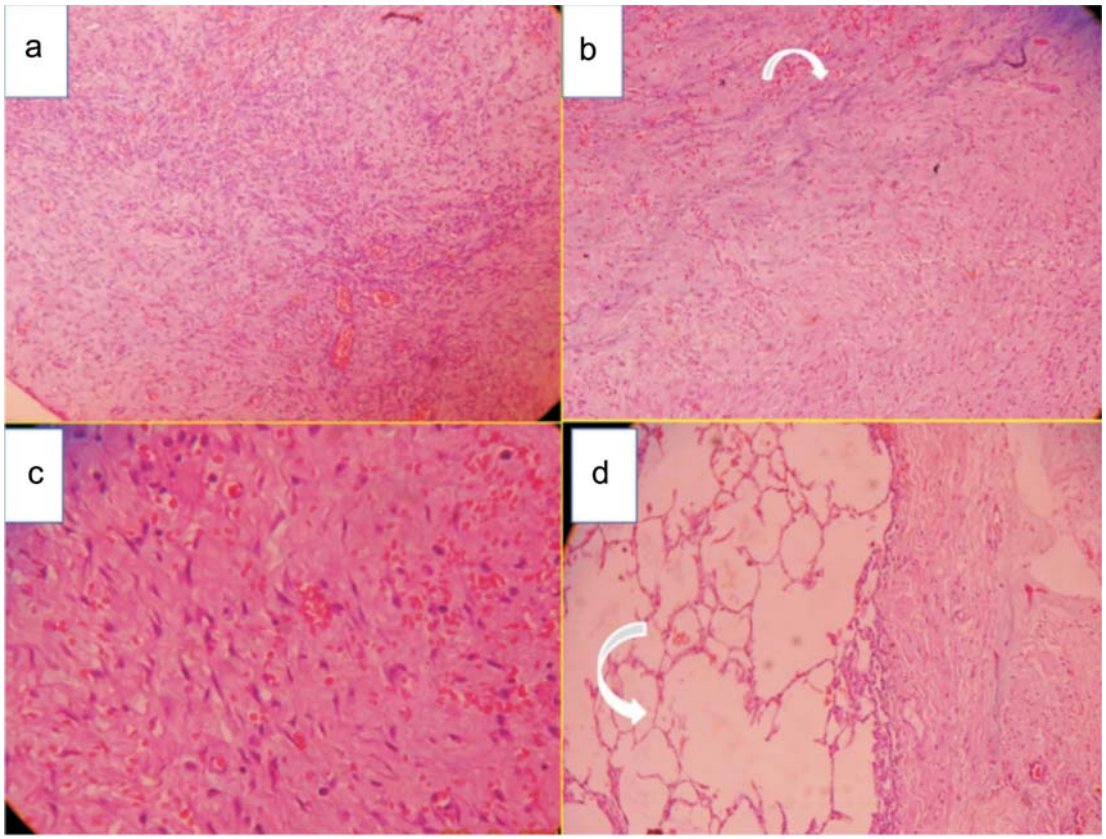

WEB FIG. 1 *Histopathological pictures (stained with hematoxyilin and eosin); (a) Low power view – showing inflammatory cells (plasma cells, lymphocytes, neutrophils, eosinophils) along with fibroblasts and blood vessels; (b) Low power view –curved arrow showing myxoid stroma in a background of inflammatory cells; (c) High power view showing proliferating benign looking spindle shaped cells with collagen; and (d) Low power view- showing lung tissue with adjacent pleura(Arrow showing normal underlying lung tissue).*