Inflammatory Pseudo Tumour – A Rare Case of Recurrence

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ABSTRACT

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Inflammatory pseudotumour is considered to be a rare tumour occurring in childhood. It constitutes less than 1% of all lung tumours.[1] An endobronchial location of the tumour is a rare entity and its recurrence is still rarer. We here present the case of an endobronchial tumour presenting as acute respiratory distress which recurred after su Professor, Department of Pediatrics, SAT Hospital, Medical College Thiruvananthapuram- 695011, Kerala.

Keywords: Inflammatory pseudotumour, Endobronchial tumour

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CASE REPORT

A 9 year old male child presented with complaints of fever and cough of 3 weeks and dyspnoea of one day duration. On examination child was sick, tachypneic with intercostal retraction and subcostal retraction. Chest examination showed bilateral vesicular breath sounds, with decrease intensity on right side along with bilateral crackles. Chest x ray showed bilateral pneumonic patches. The child was started on crystalline Penicillin and Gentamicin for bronchopneumonia. The child's condition deteriorated further on the second day. Trachea was shifted to right and apex beat couldn't be localized on left side. Repeat chest x-ray showed gross mediastinal shift to right. We considered the possibility of foreign body or endobronchial Tuberculosis. But Mantoux test was negative and three subsequent gastric aspirate samples for acid fast bacilli was negative. Cold Agglutinin test was negative. Other investigations were within normal limits. Since the child continued to deteriorate CT scan thorax was taken which showed a smooth vascular growth just beyond the origin of right bronchus occluding more than 90% of the lumen. Bronchial adenoma or an endobronchial malignancy was considered. Endobronchial biopsy was taken, which was inconclusive. Hence an open thoracotomy was done and mass sent for histopathology. The final diagnosis of the rare condition pseudo inflammatory tumour was made.

The child had relief of complaints following the surgery. But symptoms of cough and dyspnea reappeared after 3 months of initial episode. A repeat excision biopsy revealed the same diagnosis. Presently child is under follow up and is symptomatically better.

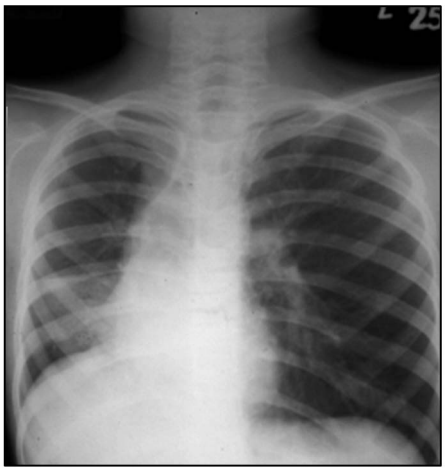

Figure 1. **Repeat Chest Xray showing gross mediastinal shift to the right.**

DISCUSSION

The tumour was first described by Pack and Baker in 1953.[3] The tumour is most commonly located in the peripheries of lungs and is often asymptomatic. Other common sites of involvement include liver, intestines, omentum, spleen, ovaries and stomach. Endobronchial location of the tumour has also been described.[4] 3%cases may be bilateral.[5]

The actual etiology of the condition is not known. An inflammatory reaction to infections, surgery or trauma has been proposed. The tumour has been reported

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to occur after Mycobacterium Tuberculosis infection in immuno compromised individuals.[6] Mycoplasma pneumonia infection and chromosomal abnormalities has also been suggested.[3]

This tumour is often asymptomatic. When manifested, clinical features are often non specific or may pertain to the organ involved. Common symptoms include fatigue, malaise, fever, chest pain, dyspnea and abdominal pain Endobronchial tumours can present with cough, dyspnea or with collapse lungs.

Investigation may show leucocytosis, raised ESR and thrombocytosis. Chest x ray may show collapse lungs distal to the site of obstruction .The tumor is of intermediate signal on T1 weighted images and high signal in T2 weighted images on MRI.[7] Definite diagnosis is by histological evaluation of the biopsy specimen which shows the characteristic picture. The tumour is likely to produce confusion in diagnosis. A fibroblastic predominance may be confused with sarcoma and inflammatory cell predominance may masquerade Hodgkin's disease or even plasmacytoma.[8] Spindle cells may be stained with vimentin.[5] Calcification is only seldom present in this tumour. However when present they are classed under a different entity called calcifying fibrous pseudotumour.[10]

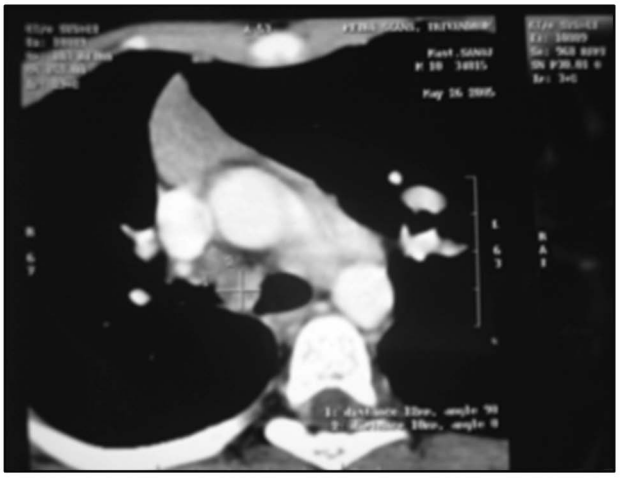

Figure 2. **MRI shows intermediate signal in T1 weighted images and high signals on T2 weighted images.**

The tumour is well circumscribed, non encapsulated, usually solitary firm parenchymal nodule covered by an intact pleura.[6] The mass characteristically contains inflammatory cells, fibrous stroma and spindle cells. There is wide variation in the histological picture which includes vascular proliferation, fibrosis, hyalinization, myxoid change, fat accumulation with the formation xanthoma cells, hemosiderin deposition, proliferation of alveolar cells in addition to inflammatory cells like plasma cells and lymphocytes.[6] Due to its wide variability the same tumour has been referred to as inflammatory myoblastic tumour, plasma cell granuloma or fibroxanthoma.[6] Three types have been described. This includes organizing pneumonia, fibrous histiocytoma and lymphoplasmocytic variety. The latter is the least common variant.[9]

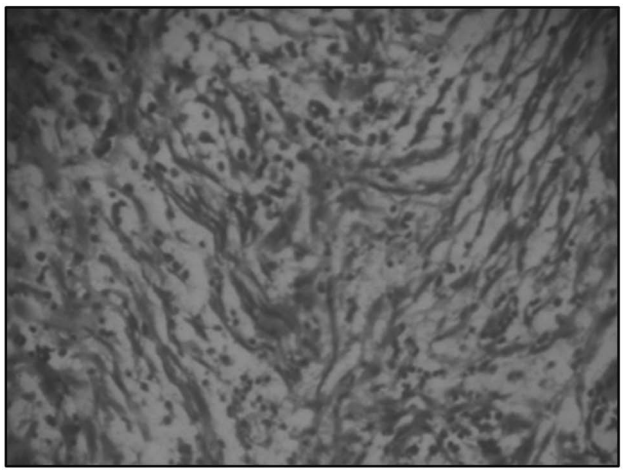

Figure 3. **Biopsy shows fibroblastic predominance.**

Treatment is essentially surgical excision.[6] In rare instances the tumour may behave like a malignancy with local invasion and metastasis. Local infiltration to pleura, spine, mediastinum and distant metastasis has been reported. Even death has resulted from the tumour. Radiotherapy and chemotherapy has been tried but with doubtful benefit. Steroids have doubtful value in management. However the size of the tumour has been shown to decrease with steroids.[9]

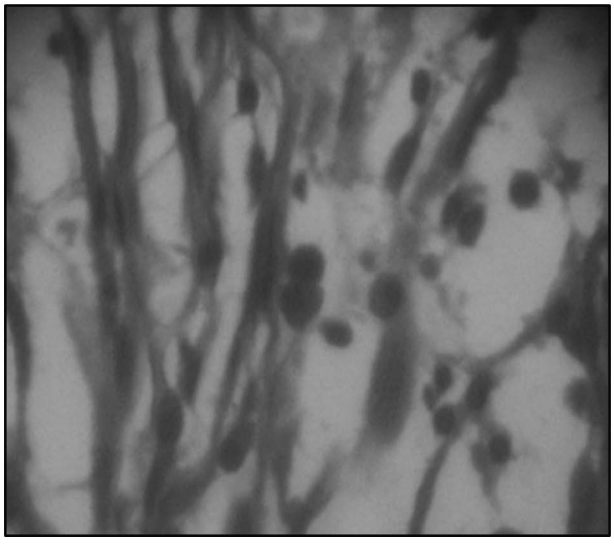

Figure 4. **Presence of inflammatory cells, spindle cells and fibrous stroma.**

The prognosis of the tumour is generally good. Poor prognostic factors include metastasis, necrosis more than 15% of surface area examined, local recurrence, bizarre giant cells, mitotic figures more than50/hpf,

advanced stage, high cellularity and poor circumscription.[5]

The case is being reported in view of its rarity and diagnostic dilemma that was associated with it. Recurrence of inflammatory pseudotumour is a rarer phenomenon and hence reported herewith

END NOTE

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