CASE REPORT

A RARE PULMONARY TUMOUR - INFLAMMATORY PULMONARY MYOFIBROBLASTOMA

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ABSTRACT

The inflammatory myofibroblastic tumor is also known as inflammatory pseudotumor is a rare benign lesion, though becomes invasive and recur after excision. It accounts for 0.7% of lung tumors. We report a case of a young female 18 years of age who presented to our hospital with complaints of cough, chest pain, shortness of breath, intermittent fever, and undocumented weight loss for 1 year. She had a previous history of lung infection. Chest auscultation revealed reduced air entry on the right side. Laboratory tests showed an increased erythrocyte sedimentation rate of 130. A chest radiograph was done which showed homogenous opacity invading the right hemithorax. Contrast tomography of the thorax was performed which reported a large heterogeneously enhancing tumor of 13 x 10 x 12 cm with central necrosis occupying the right hemithorax practically collapsing the right hemithorax partially with left hemithorax without any alterations. No internal calcification was appreciated. It was causing compression on the trachea and esophagus and complete obliteration of the right main bronchus. The tumor showed distinct fat planes with surrounding structures. Mild right-sided pleural effusion was seen. CT-guided biopsy was done. Microscopic examination revealed linear cores of a tumor comprising proliferating spindle cells and scattered epithelioid cells with eosinophilic nucleoli admixed with abundant lymphocytes and plasma cells. Immunohistochemical analysis showed positive staining for ALK-1, smooth muscle actin (SMA), desmin, and pan-CK. In contrast tumor, cells were non-reactive to S-100. Based on this data, a diagnosis of inflammatory myofibroblastoma was retained. The purpose of this report was to highlight the rare disease with its CT features. Despite being a benign lesion its potential for local invasion and recurrence requires surgical resection.

Keywords: inflammatory myofibroblastoma, inflammatory pseudotumour, contrast computed tomography, histopathological assessment.

Introduction

An inflammatory myofibroblastic tumor (IMT) is regarded as a seldom type of intermediate tumor that most frequently occurs in adults of young age group.[1] They have comprised of myofibroblastic spindle cells along with an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils. The underlying etiology may be a genetic mutation or it can occur after infectious or autoimmune diseases.[2] Inflammatory myofibroblastic tumors (IMTs) can involve almost every organ. IMTs usually follow a benign course that may become locally invasive and malignant, chance for local recurrence, or may present with distant metastasis. Patients have a wide range of manifestation. Patient may be asymptomatic or they can have a cough, hemoptysis, dyspnea, pleuritic pain, constitutional symptoms, or pneumonia. Due to such

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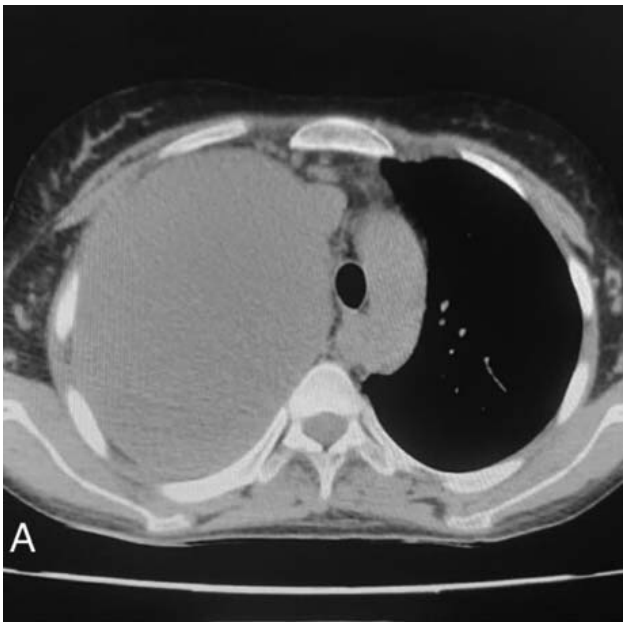
discrepancy in tumorous progression and nonspecific clinical findings, the clinical and radiologic diagnosis of lung IMTs is very ambiguous and this requires treatment to limit aggressive behavior of these tumors.[3]

Case Presentation

We report a case of a young female 18 years of age who presented in our hospital emergency department with complaints of cough, chest pain, shortness of breath, intermittent fever, and undocumented weight loss for 1 year. She had a previous history of lung infection and took ATT for 6 months for suspected TB. She had no history of any co morbid illness. Her presenting vitals were BP 110/80mm Hg, respiratory rate of 25/min, temperature 99 F, BMI 18 Kg/m square. She has pallor,however there was no evidence of clubbing, jaundice or koionychia. Chest auscultation revealed reduced air entry on the right side and dull percussion note on right side. Rest of her systemic examination was unremarkable. A chest radiograph was done which showed homogenous opacity invading the right hemithorax with contralateral tracheal and medistinal shift. Silhouetting of right hemidiaphragm was appreciated. Right CP angle was obscured. Left lung was unremarkable. Bones appeared normal. Ultrasound chest was done which showed mixed echogenicity lesion in right lung. No pleural effusion was appreciated. Contrast tomography of the thorax with IV contrast was performed using 2 mm reconstructed images from scan performed on multislice CT which were reviewed on workstation using different window widths and level settings, reported a large heterogeneously enhancing tumor of 13 x 10 x 12 cm with central necrosis occupying the right hemithorax practically collapsing the right hemithorax partially with left hemithorax without any alterations. No internal calcification was appreciated. It was causing compression on the trachea and esophagus and complete obliteration of the right main bronchus. The tumor showed distinct fat planes with surrounding structures. Mild right-sided pleural effusion was seen. No adjacent visceral or vascular encasment was seen.Left lung and visualized bones were unremarkable. Patient got admitted in medicine ward and was medicated accordingly. CT-guided biopsy was done. Microscopic examination revealed linear cores of a tumor comprising proliferating spindle cells and scattered epithelioid cells with eosinophilic nucleoli admixed with abundant lymphocytes and plasma cells. Immunohistochemical analysis showed positive staining forALK-1, smooth muscle actin (SMA), desmin, and pan-CK. In contrast tumor, cells were non-reactive to S-100. Based on this data, a diagnosis of inflammatory myofibroblastoma was retained. The purpose of this report was to highlight the rare disease with its CT features. Despite being a benign lesion its potential for local invasion and recurrence requires surgical resection.

Discussion

IMT affect patients of all age group, but the mean age of start of disease is 10 years old having female predilection.[1] They consist of myofibroblastic spindle cells admixed with plasma cells,varying degree of inflammatory infiltrates, lymphocytes, and occasional eosinophils. Genetic mutation plays an important role in in its occurrence, however may occur secondary to infection or autoimmune diseases.[2] Lungs are most frequently involved site in pediatric population. They are extremely rare in adults, accounting for less than 1% of adult lung tumors.[3] IMT usually affects lung, with morbidity reported to be 0.04 to 1.2% but it may originate in diverse locations like omentum, retroperi-



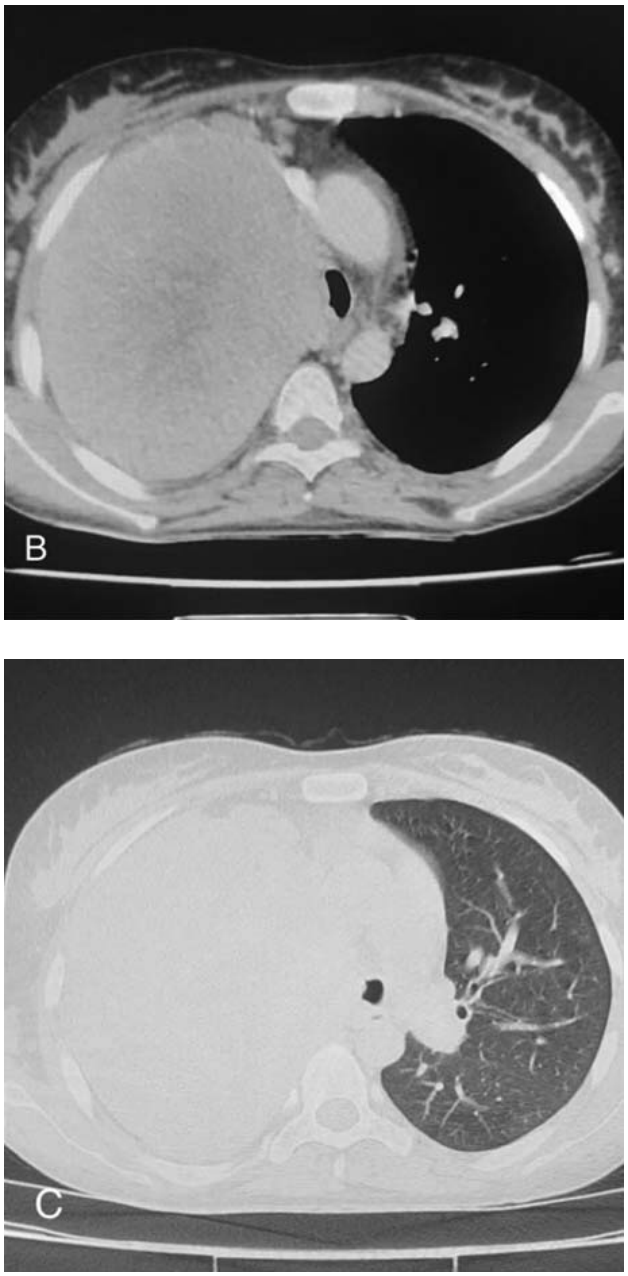
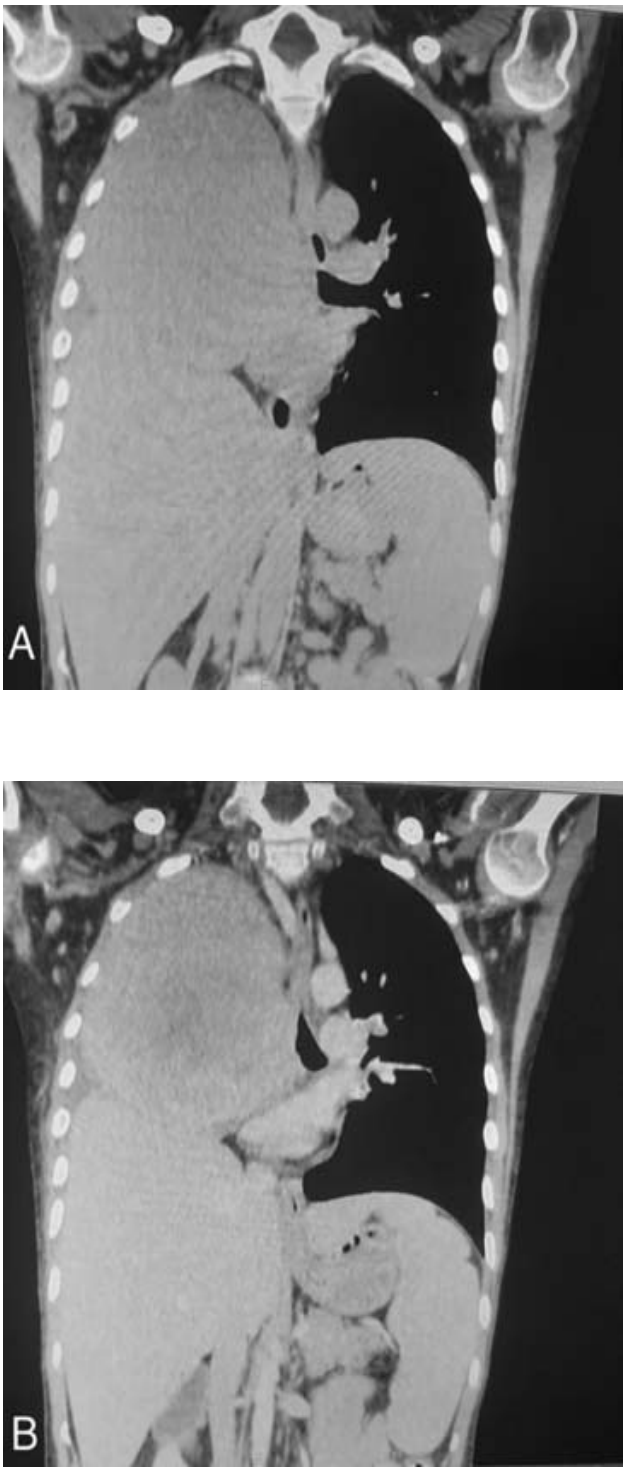


Figure 1: Chest CT axial sections with **(A)** mediastinal precontrast, **(B)** mediastinal post contrast and **(C)** parenchymal window showing right heterogeneously enhancing lung tumour.

toneum, pelvis, abdominal soft tissue, head and neck, gastrointestinal track, liver, spleen and larynx.[4] Lung biopsy is required for pathological analysis. About half of these tumors express anaplastic lymphoma kinase (ALK) gene rearrangement (2p23).[5] On CT scan imaging they appear as solid homogeneous or heterogeneously enhancing lesions that are characterized as single, well-circumscribes, lesion and having punctate calcifications more frequently in younger age group.[6] The mainstay of treatment is complete surgical excision. NSAIDS, steroids and chemotherapy have also been used as adjunct therapies.[7]



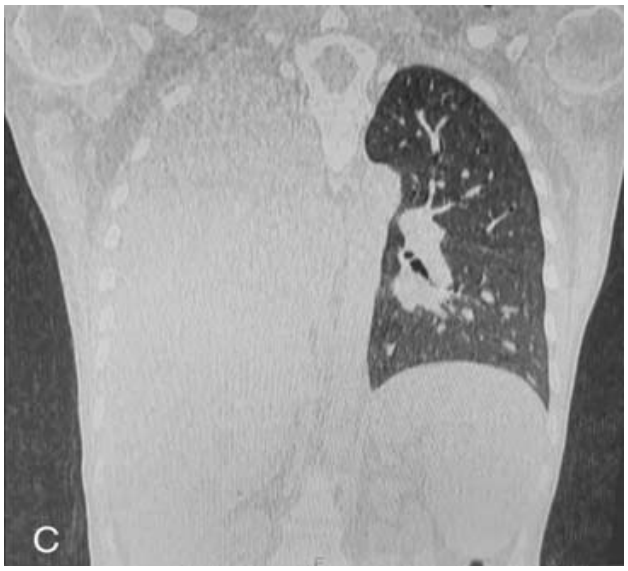
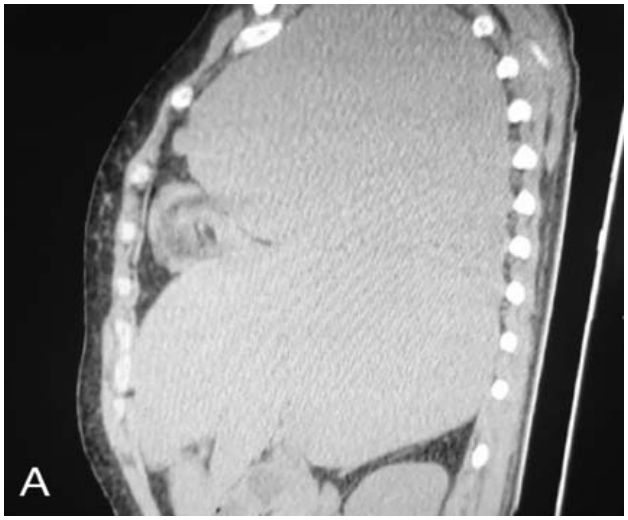
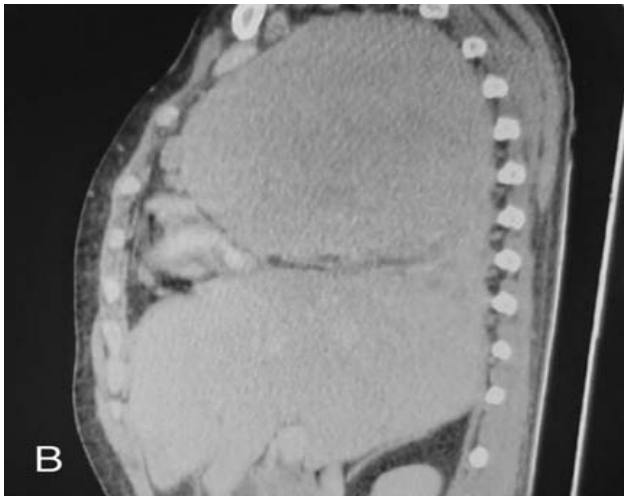


Figure 2: Chest CT coronal sections with **(A)** mediastinal precontrast, **(B)** mediastinal post contrast and **(C)** parenchymal window





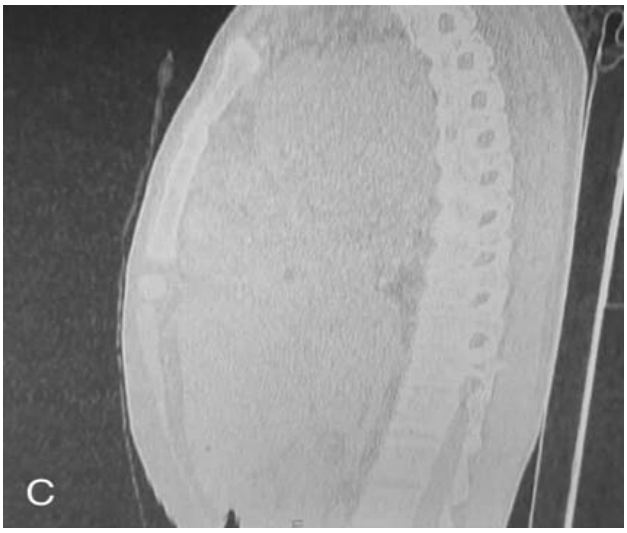


Figure 3: Chest CT sagittal sections with **(A)** mediastinal precontrast, **(B)** mediastinal post contrast and **(C)** parenchymal window

Conclusion

Because of the wide spectrum of clinical manifestation of IMTs, it remains a matter of debate as to whether IMTs represent an inflammatory response to malignancy or whether IMTs are a primary inflammatory process. Distinguishing untypical symptom, surgical resection, pathological evaluation and close follow-up will reach the successful diagnosis and treatment.

Conflict of Interest: None

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