



A Rare Case of Inflammatory Myofibroblastic Tumour in a young Female Presenting with Pancoast Tumour of Left Lung

Authors

Soumitra Das^{1*}, Abhisek Lath², Anadi Roy Chowdhury³

Department of Pathology, R.G. Kar Medical College & Hospital

*Corresponding Author

Soumitra Das

Abstract

Inflammatory Myofibroblastic Tumour is a controversial lesion as it was previously presumed to be an inflammatory lesion. However due to its infiltrative growth pattern, its neoplastic potential is a matter of interest. In this case report we studied a female in her third decade of life who presented with symptoms of superior vena caval obstruction and was radiologically diagnosed as Pancoast Tumour. In pathological examination it showed evidence of the histological features of Inflammatory Myofibroblastic Tumour.

Keywords: *Inflammatory Myofibroblastic Tumour, Pancoast Tumour, Left Lung.*

Introduction

Inflammatory Myofibroblastic tumour (IMT), also known as, inflammatory pseudo tumour and plasma cell granuloma, is a rare disease process. It was previously presumed to be an inflammatory response to a previous injury with unknown etiology. However, due to the notable capability of infiltration and destruction of adjacent structures as well as their ability to recur, as noted in few cases, some of its characteristics were distinctly concluded to be that of a neoplastic process.¹ In recent times, specific cytogenetic characteristics were also identified and established, indicating a neoplastic nature of the lesion.² However, owing to the diverse clinical and radiological findings, a diagnosis of Inflammatory Myofibroblastic tumour is difficult without surgery and consequent histopathological findings, leading to such diagnoses being more of an incidental finding, especially in pulmonary lesions.

Prominent established genetic change associated with this neoplastic lesion is the translocation in the

ALK tyrosine kinase receptor region at chromosome 2p23. About 50 % of IMT s in lungs have this translocation.³⁻⁶ In this case report, we have discussed the histopathological findings of a case of pulmonary Inflammatory Myofibroblastic tumour, that was presumed to be a case of Pan coast tumour. The histopathological findings were correlated with clinical and radiological observations and confirmation was done immunohistochemically.

Case Report

A 27 year old female presented with a history of positional cough for 8 months along with swelling in the left supraclavicular region for 4 months. Her ECG showed sinus rhythm and complete blood counts were normal, except for a haemoglobin value of 9.4 gm/dl. All her biochemical tests were normal, except for raised serum TSH value of 9.72 microIU/ml.

Her chest X-ray revealed an opacity in the upper lobe of the left lung. The cytology from a fine needle aspiration of the left supraclavicular swelling

stated the evidence of reactive fibroblastic proliferation along with inflammatory cells in a background of RBCs, suggesting the possibility of a Benign Fibroblastic lesion.

She also underwent a whole body multi slice (dual) spiral CT scan which reported a mass like area of consolidation measuring 7.1 cm x 6.7 cm x 4.4 cm in the apex and adjacent anterior segment of left lung. Mild surrounding ground glass opacities were seen in the adjacent lung parenchyma. The lesion was noted to extend into the adjacent superior basal segment of lower lobe of the same side. The lesion was also noted to abut into the chest wall and slightly infiltrate into the sternocostal region as well as into the adjacent mediastinum. Minimal bilateral pleural effusion was also noted. The overall features suggested a lymphoma or bronchogenic carcinoma with less likelihood of an infective etiology.



Figure 1: Chest X-ray revealed an opacity in the upper lobe of the left lung

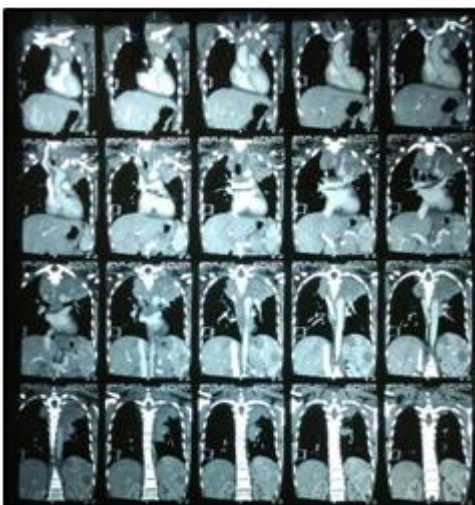


Figure 2: CT scan showed an area of consolidation measuring 7.1 cm x 6.7 cm x 4.4 cm in the apex and adjacent anterior segment of left lung.

In due course of time, the patient developed swelling of face along with shortness of breath and progressive cough. A digital X-ray of the barium swallow esophagus was done which revealed a soft tissue mass in left upper chest and right paratracheal region, causing mild narrowing of esophagus. She then underwent a contrast enhanced CT scan of the thorax which alike prior investigations, also revealed a pleural based consolidating mass in the upper lobe of left lung field, measuring 7cm x 4.5 cm, but the mass was noted to involve the mediastinum and chest wall and causing erosion of left 2nd and 3rd ribs. In the mediastinum, it reported presence of confluent homogenous mediastinal nodes extending to left supraclavicular nodes causing obstruction of the superior vena cava due to vascular encasement. Minimal bilateral pleural effusion was also noted. Overall features ascertained a possibility of Lymphoma.

A CT guided FNAC from the left lung and mediastinal lesion revealed smears showing numerous foamy macrophages, few in clusters and few discretely along with mild inflammatory infiltrate, suggesting the possibility of an inflammatory lesion. The trucut biopsy from the same lesion showed linear cores of fibrocollagenous tissue with areas rich in ganglion cells and nerve fibres with surrounding areas of lymphoid aggregates and was inconclusive of any diagnosis.

The patient was then admitted for surgery with a provisional diagnosis of Pancoast tumour causing superior vena cava obstruction symptoms. Under general anaesthesia, thoracotomy was done and a left thoracic mass was noted, involving left upper lobe and upper part of lower lobe of left lung. It was densely adherent to the apex of the chest wall, left pulmonary artery, left bronchus, arch of aorta and descending thoracic aorta. Approximately 150 ml of pleural effusion fluid was drained out. Open biopsy was taken from the mass.

Pathological Findings

The resected tumour was received in the department of Pathology of the institute as multiple fragmented bits of tissue altogether measuring 5.5cm x 4.5 cm x

4cm. Microscopic examination revealed fragmented bits of tissue comprising predominantly of spindle cells arranged in short fascicles and with focal storiform pattern. Admixed within the spindle cells were inflammatory cells, including lymphocytic, plasma cells and histiocytic infiltration. The individual spindle cells had elongated oval nuclei, with inconspicuous nucleoli, surrounded by eosinophilic cytoplasm with indistinct cytoplasmic borders. No distinct cytological atypia or mitotic activity was noted in the sections examined.

Immunohistochemical analysis showed positive staining for Smooth Muscle Actin (SMA). In contrast, the sections were not reactive to ALK. Hence diagnosis of Inflammatory Myofibroblastic Tumour was finally reached at.

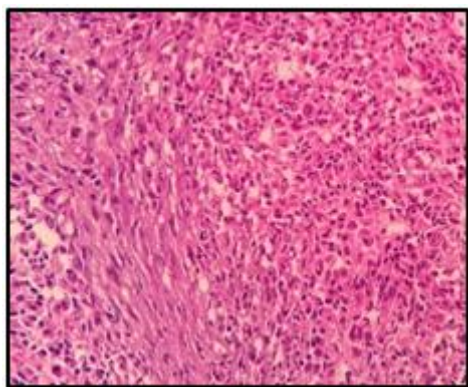


Figure 3: Histomorphological features of Inflammatory Myofibroblastic tumour (H&E, 400x)

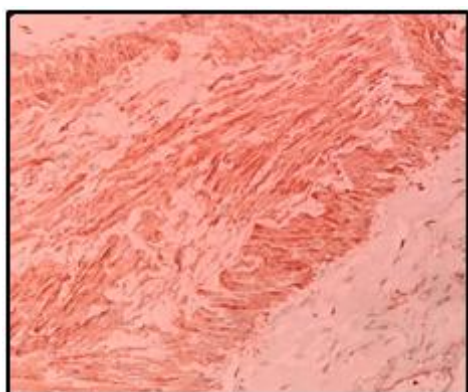


Figure 4: Photomicrograph showing SMA positivity in the tumour (H&E, 400x)

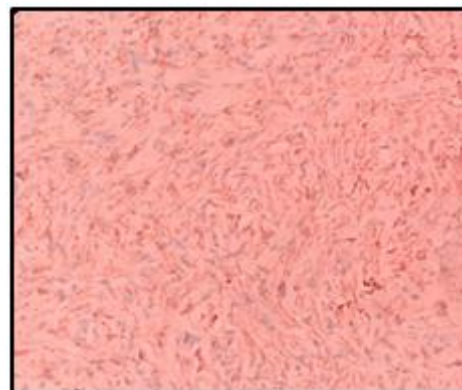


Figure 5: Photomicrograph showing ALK negativity in the tumour (H&E, 400x)

Discussion

IMTs, including pulmonary lesions are more common in children and young adults. Among lung lesions, children account for about 25% to 40% of IMTs in all age groups.⁷ Usually, such lesions present as a mass with or without accompanying localized or generalized clinical features, like cough, chest pain, fever, haemoptysis.⁸⁻¹⁰ Systemic manifestations are usually mediated by Interleukin-1, interleukin-6, and tumour necrosis factor, secreted by macrophages, fibroblasts, and other constituent cells of an IMT. In the case described above, the patient initially had progressive cough, but later due to vascular obstruction by the tumour, she developed facial edema too.

Pulmonary IMTs are the most common IMTs, of which lung is the most common site. Other pulmonary sites include trachea and bronchus. The extrapulmonary sites include mesentery, liver, spleen, mediastinum, central nervous system, soft tissues.¹¹⁻²⁵ In rare cases, multiple sites may be involved. In cases of lung involvement, there is a predilection for the right side⁸, though in this case the lesion was located in left lung. The radiographic features according to a study was found to be a solitary peripheral mass in 85 % cases and extra parenchymal involvement was found in 18 % cases.²⁶ Some cases have intrapulmonary mass with contiguous extension to local structures like pericardium, oesophagus, parietal pleura, mediastinum, left atrium, diaphragm and regional blood vessels. In this case, with due course of time, the lesion was noted to infiltrate into the

sternocostal region as well as the chest wall and later also to the confluent mediastinal and left supraclavicular nodes causing obstruction of the superior vena cava.

On gross examination, the lesion is usually a firm, sharply circumscribed but non encapsulated mass, size varying between 4-6 cm in diameter. The cut surface is usually whorled to homogenous, and grayish white to focally yellowish white in colour. Calcifications may be present in 25-30 % cases either grossly or microscopically.

Microscopically, two different patterns have been described. Fibrohistiocytic spindle cell type is characterised by fascicles of spindle cells with focal storiform pattern, spindle cells showing elongated nuclei surrounded by a rim of amphophilic to eosinophilic cytoplasm, inconspicuous nucleoli, generally devoid of mitotic atypia. Spindle cells are admixed with mixed inflammatory cells, predominantly foamy macrophages. The plasma cell type is characterized by mainly plasma cells with lesser predominance of fibroblastic spindle cells. The case described here revealed histopathological features more in favour of the former type.

For immuno histochemical detection, spindle cells show positivity for SMA and vimentin. About one-third of the cases have been demonstrated to have ALK (Anaplastic lymphoma Kinase) rearrangement, detected by FISH or IHC using ALK 1 antibody. Despite such recent methods of diagnoses, ALK 1 positivity have been observed mainly in children and its absence does not exclude the diagnosis.

Conclusion

This case presented with considerable diagnostic dilemma both due to its growth pattern as well as microscopic features. However, evidence of such findings in few cases in the past alongwith immunohistochemical features acted as a major aid in drawing the conclusion.

References

1. Suster S, Moran CA. Biopsy Interpretation of the Lung. Philadelphia: Wolters Kluwer; 2013:358-60.
2. Su LD, Atayde-Perez A, Sheldon S, et al. Inflammatory myofibroblastic tumour: cytogenetic evidence supporting clonal origin. *Mod Pathol*. 1998;11:364-368.
3. Yousem S, et al. Inflammatory myofibroblastic tumour. In: Travis W, et al., eds. Classification of tumours. Lyon: IARC Press, 2004:105-106.
4. Chun YS, et al. Pediatric inflammatory myofibroblastic tumour: anaplastic lymphoma kinase (ALK) expression and prognosis. *Pediatric Blood Cancer* 2005;45(6):796-801.
5. Debelenko LV, et al. Identification of CARS-ALK fusion in primary and metastatic lesions of an inflammatory myofibroblastic tumour. *Lab Invest* 2003; 83(9):1255-1265.
6. Dehner L. Inflammatory myofibroblastic tumour: the continued definition of one type of so-called inflammatory pseudotumour. *Am J Surg Pathol* 2004;28:1652-1654.
7. Cerfolio RJ, et al. Inflammatory pseudotumours of the lung. *Ann Thorac Surg* 1999;67(4):933-936.
8. Kim JH, et al. Pulmonary inflammatory pseudotumour—a report of 28 cases. *Korean J Intern Med* 2002;17(4):252-258.
9. Sakurai H, et al. Inflammatory myofibroblastic tumour of the lung. *Eur J Cardiothorac Surg* 2004;25(2):155-159.
10. Narla LD, et al. Inflammatory pseudotumour. *Radiographics* 2003;23(3):719-729.
11. Rodrigues M, et al. Inflammatory myofibroblastic tumour of the larynx in a 2-year-old male. *ORL J Otorhinolaryngology Relat Spec* 2005;67(2):101-105.
12. Seki S, et al. A clinicopathological study of inflammatory pseudotumours of the liver

- with special reference to vessels. *Hepatogastroenterology* 2004;51(58):1140–1143.
13. Pungpapong S, Geiger XJ, Raimondo M. Inflammatory myofibroblastic tumour presenting as a pancreatic mass: a case report and review of the literature. *JOP* 2004;5(5):360–367.
 14. Oz Puyan F, et al. Inflammatory pseudotumour of the spleen with EBV positivity: report of a case. *Eur J Haematol* 2004;72(4):285–291.
 15. Lo OS, et al. Inflammatory pseudotumour of the liver in association with a gastrointestinal stromal tumour: a case report. *World J Gastroenterol* 2004;10(12):1841–1843.
 16. Fang JC, Dym H. Myofibroblastic tumour of the oral cavity. A rare clinical entity. *N Y State Dent J* 2004;70(3):28–30.
 17. Dehner LP. Inflammatory myofibroblastic tumour: the continued definition of one type of so-called inflammatory pseudotumour. *Am J Surg Pathol* 2004;28(12):1652–1654.
 18. Dasgupta D, et al. Liver transplantation for a hilar inflammatory myofibroblastic tumour. *Pediatr Transplant* 2004;8(5): 517–521.
 19. Browne M, et al. Inflammatory myofibroblastic tumour (inflammatory pseudotumour) of the neck infiltrating the trachea. *J Pediatr Surg* 2004;39(10):e1–e4.
 20. Yoshida T, et al. Inflammatory pseudotumour of the liver: report of a case diagnosed by needle biopsy. *Hepatol Res* 2003;27(1):83–86.
 21. Ueda M, et al. A case of inflammatory pseudotumour of the liver hilum successfully treated with aggressive hepatectomy. *J Pediatr Surg* 2003;38(11):E9–E11.
 22. Thompson RJ, Barrett AM, Dilley P. Congenital multifocal inflammatory pseudotumour: a case report. *J Pediatr Surg* 2003;38(10):E17–E19.
 23. Sobesky R, et al. Inflammatory pseudotumour of the common bile duct. *Endoscopy* 2003;35(8):698–700.
 24. Koea JB, et al. Inflammatory pseudotumour of the liver: demographics, diagnosis, and the case for nonoperative management. *J Am Coll Surg* 2003;196(2):226–235.
 25. Kapusta LR, et al. Inflammatory myofibroblastic tumours of the kidney: a clinicopathologic and immunohistochemical study of 12 cases. *Am J Surg Pathol* 2003;27(5):658–666.
 26. Agrons GA, et al. Pulmonary inflammatory pseudotumour: radiologic features. *Radiology* 1998;206(2):511–518.