



Inflammatory Myofibroblastic Tumor of the Lung : Rare Case Report

KEYWORDS

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Introduction

The inflammatory myofibroblastic tumor is composed of myofibroblastic spindle cells accompanied by an inflammatory plasma cells, lymphocytes and eosinophils. The variety of terms used to describe this entity include inflammatory pseudotumor, cellular inflammatory pseudotumor, plasma cell granuloma, inflammatory myofibrohistiocytic tumor and more recently, inflammatory fibrosarcoma. Inflammatory myofibroblastic tumors (IMTs) are relatively rare type of a distinct histologic appearance and benign clinical course.[4] The inflammatory myofibroblastic tumor or inflammatory pseudotumor usually occurs in children and young adults.[1,2] Inflammatory myofibroblastic tumors in children are commonly found in lung, abdomen, retroperitoneum and extremities.[3] Most commonly it occurs in lungs and presents as benign tumor. Extrapulmonary tumors are aggressive in nature.[5] [6]

Most patients present with respiratory symptoms in pulmonary tumors.[7]

Case Report

A 11 year old girl with history of previous respiratory infections presented with complaints of dyspnea and cough. With initial treatment, symptoms were not subsided. On advising the patient HRCT thorax with contrast, a large well circumscribed lobulated heterogeneously enhancing lesion was seen in the peripheral aspect of right lobe which measured about 4.6 x 4 x 5.1 cm (AP X TR X CC). The lesion showed enhancing irregular solid component and few irregular non-enhancing necrotic components with irregular linear calcifications in the central aspect. Rest of the lung parenchyma and tracheo-bronchial tree appeared normal.

The pleural spaces and major mediastinal blood vessels were appeared normal on CT scan.

Patient was also subjected for contrast enhanced 18F – FDG whole body PET-CT scan. It showed increased FDG uptake in the large well defined soft tissue mass in the lower lobe of the right lung measuring 4.6 x 4.4 x 5.6 cm, SUV max 10.20 showing calcifications within. No active disease seen elsewhere in the body. Then patient was advised CT guided Right lung mass biopsy. Histopathology of right lung lesion biopsy showed sheets of spindle cells with plasmacytic rich infiltrate. These features are compatible with Inflammatory Myofibroblastic tumor. On immunohistochemistry the tumor cells were positive for ALK-1 and SMA with occasional plasma cells IgG4 expression.

After thorough preoperative investigations patient was

posted for right sided thoracotomy.

Right sided thoracotomy performed under general anesthesia with single lumen endotracheal tube. Right lower lobectomy was done. Final histopathology report was consistent with Inflammatory Myofibroblastic tumor. Patient recovered well after surgery.

Discussion

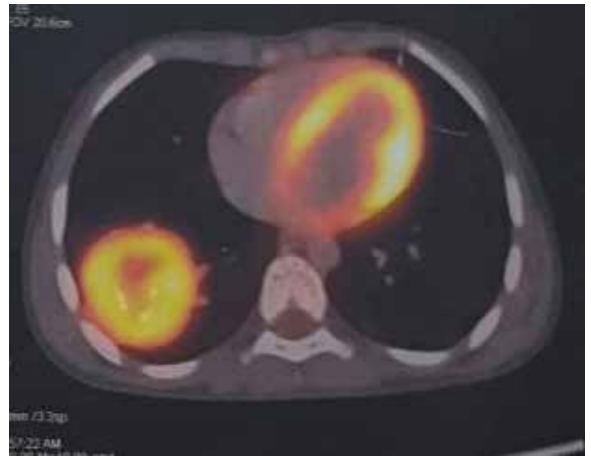
Inflammatory myofibroblastic tumor (IMF) can be defined as a localized mass consisting of a fibrous stroma and chronic inflammation, infiltrated with a predominance of plasma cells or histiocytes and an absence of anaplasia. Lung inflammatory myofibroblastic tumor is a rare cause of solitary lung nodule (0.7 % of lung tumors). The principal site of inflammatory pseudotumor is the lung, but it can also occur elsewhere in various organs. Although it presents mostly as benign tumor, they may be locally very aggressive. Recurrent and multifocal forms have been described. Since they are likely to mimic malignant neoplasms, accurate histopathologic diagnosis is necessary in order to decide appropriate surgical excision and avoid aggressive treatments. [8][11] Tumor recurrence is unusual following complete surgical resection. [9] The etiology and cellular origin of IMF tumor still remains unclear.[11] Some researchers believe that this neoplasm represents an immunologic response to an infectious agent or noninfectious agent, whereas others believe that IMF tumors are true neoplasms. [2] The clinical presentation of IMF tumors mostly depends on the site at which the tumors originate. IMF tumor of the lung is often asymptomatic but it can present with cough, shortness of breath, chest pain, dysphagia, or hemoptysis. Laboratory evidence of inflammatory response has also been described in association with IMF tumor of the lung.[2]

Conclusion

Inflammatory myofibroblastic tumor is rare but clinically important and pathologically distinct lesion of respiratory tract in children. Despite local invasiveness, local recurrences, and occasional reports of metastatic spread, IMF tumor can usually be treated successfully with conservative surgery. Because of the characteristic clinical presentation, the diagnosis of IMF tumor should be decided and similarities between IMF and malignant tumor make the differentiation difficult, and, hence, histologic examination is necessary preoperatively. So complete surgical excision in resectable disease shall be considered as primary treatment with favorable outcome.



PET CT image 1



PET CT image 3



PET CT image 2



PET CT image 4

REFERENCE

- 1] Makimoto Y, Nabeshima K, Iwasaki H ; Inflammatory myofibroblastic tumor of the posterior mediastinum: an older adult case with anaplastic lymphoma kinase abnormalities determined using immunohistochemistry and fluorescence in situ hybridization, *Virchows Archiv*; April 2005, Volume 446, Issue 4, pp 451-455 2] Souid et al,] IMF in Children; *CANCER* September 15, 1993, 72, 6 ,2042-2048 3] Wang K,ma L, Is radical surgery necessary to head and neck inflammatory myofibroblastic tumor (IMT) in children?; *Child's Nervous System* March 2009, 25, 3, 285-291 4] Venizelos I, Papatimos T, Pediatric inflammatory myofibroblastic tumor of the trachea: A case report and review of the literature, *Pediatric Pulmonology*, 43, 8, , August 2008, 831-835 5] Satomi T et al, A successfully treated inflammatory myofibroblastic tumor of the mandible with long-term follow-up and review of the literature, *Medical Molecular Morphology*, September 2010, 43, 3, 185-191 6] Dhoub A et al, Inflammatory myofibroblastic tumor of the lung: a rare cause of atelectasis in children , *Pediatric Radiology*, March 2013, 43, 3, 381-384 7] Jonathan I M et al, Inflammatory myofibroblastic tumour of the lung manifesting as progressive atelectasis, *Pediatric Radiology*, October 1999, 29, 11, 816-819 8] Zennaro H et al, Inflammatory myofibroblastic tumor of the lung (inflammatory pseudotumor): uncommon cause of solitary pulmonary nodule, *European Radiology* , July 1999, 9, 6, 1205-1207 9] kovach SJ et al, Inflammatory myofibroblastic tumors, *Journal of Surgical Oncology*, October 2006, 94, 5, 385-391 10] Monzon CM, Gilchrist GS, Burgert EO, O'Connell EJ, Telander RL, Hoffman AD, et al. Plasma cell granuloma of the lung in children. *Pediatrics* 1982; 70:268-74. 11] Yamrubboon W et al, Inflammatory Myofibroblastic Tumor of Abdomen: Computerized Tomographic (CT) and Pathological Findings, *J Med Assoc Thai* 2008; 91 (9): 1487-93