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Inflammatory Myofibroblastic Tumour of Lung, Masquerading As Tuberculosis – A Case Report

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Abstract: *Inflammatory myofibroblastic tumor is a histologically distinctive lesion that occurs primarily in the viscera and soft tissue of children and young adults. It is considered a tumor of borderline malignancy because of its tendency to recur locally (at least at certain sites) and its ability to metastasize rarely. It is composed of a variable mixture of collagen, inflammatory cells, and usually cytologically bland spindle cells showing myofibroblastic differentiation. There are many uncertainties about the pathogenesis of IMT. Currently, surgery is the mainstay of the treatment for IMTs. It has three histological subtypes. The differential diagnosis of this lesion depends on the clinicopathologic setting, including the patient's age, gender, tumor location, and number of lesions. Rarely inflammatory myofibroblastic tumors have a conspicuous population of large multinucleated tumor cells with prominent nucleoli bearing a resemblance to the Reed-Sternberg cells of Hodgkin's disease. Based on the two largest studies of abdominal and retroperitoneal lesions, it is clear that tumors in this location have a propensity for more aggressive behavior than their extra-abdominal counterparts, with recurrence rates of 23% to 37%.*

Keywords: *IMT, Multinucleated tumor cells, borderline malignancy.*

I. Introduction

Inflammatory myofibroblastic tumor is a histologically distinctive lesion that occurs primarily in the viscera and soft tissue of children and young adults. It is considered a tumor of borderline malignancy because of its tendency to recur locally (at least at certain sites) and its ability to rarely metastasize. It is composed of a variable mixture of collagen, inflammatory cells, and usually cytologically bland spindle cells showing myofibroblastic differentiation. Inflammatory myofibroblastic tumour has an equal sex distribution and occurs in all ages, though most occur in individuals less than 40 years. Inflammatory myofibroblastic tumour is the most common endobronchial mesenchymal lesion in childhood. [1]

II. Case Report

We report the case of a 22-year-old male patient, who had complained of weight loss, anorexia, low grade fever and breathlessness for 1 year.FNAC was reported as granulomatous inflammation favoring Kochs and true cut biopsy was reported as tubercular inflammatory lesion. Patient took antitubercular treatment for 8 months On examination patient has positive history of breathlessness. Basal crepitations and decreased respiratory sound were heard on left side. Chest computed tomography(CT) scan showed well defined rounded subtle heterogeneously enhancing lesion in anterior segment of left upper lobe [82(AP)x81(TR)x87(CC)mm] with splaying/bulging of left major fissure. The lesion is extending from left hilum to lateral chest wall with bulging of the bifurcation of left main bronchus and hilar vessels The CT features were in favor of Neoplastic (Germ cell) Vs Benign Lesion (Adenoma) Fibroma / Neurogenic.The resort to surgery was for diagnostic and therapeutic purposes and consisted of left pneumectomy. On gross examination, the tumour was 4x2x1 cm in size, firm, whitish and homogeneous. Microscopic examination revealed mass composed of spindle cells arranged in fascicles or showing storiform pattern. Spindle cells had oval or elongated nuclei with fine chromatin and inconspicuous nucleoli. Many lymphocytes with plasma cells, few histiocytes and occasional multinucleated giant cells were seen.

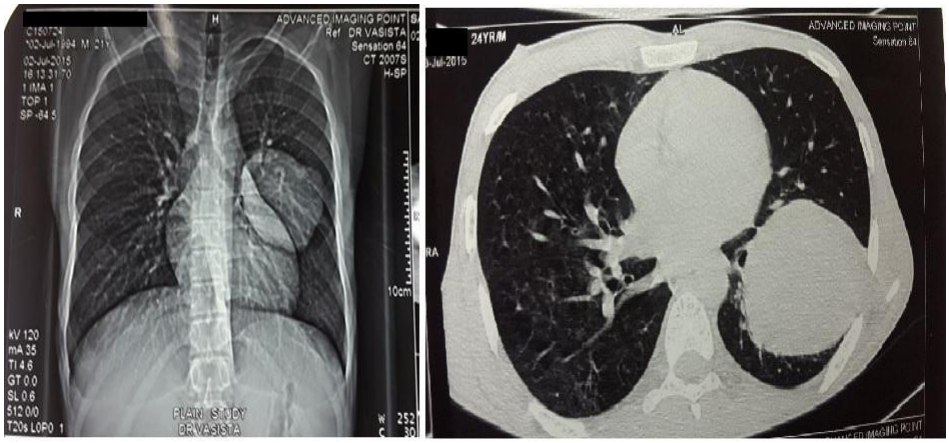

Figure 1 CT scan showing mass in the left lung

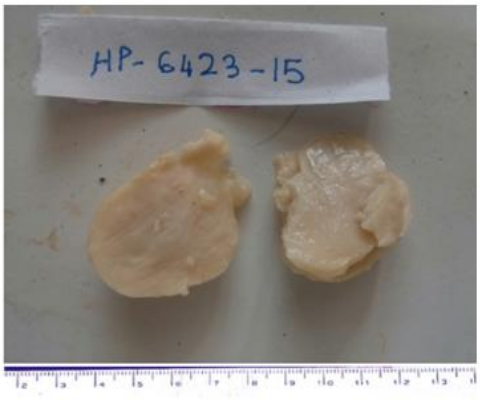

Figure 2: Gross appearance: 4 x 2 x 1 cm in size, firm, whitish and homogeneous.

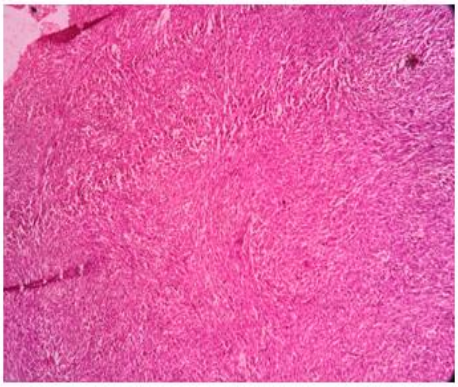

Figure 3: 10X view, showing spindle cells arranged in fascicles or showing storiform pattern.

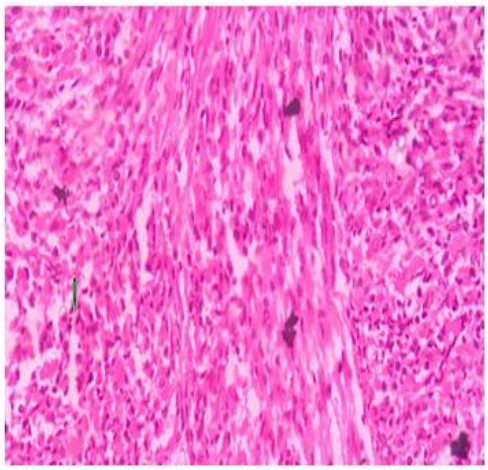

Figure 4: High power, showing spindle cells, many lymphocytes with plasma cells.

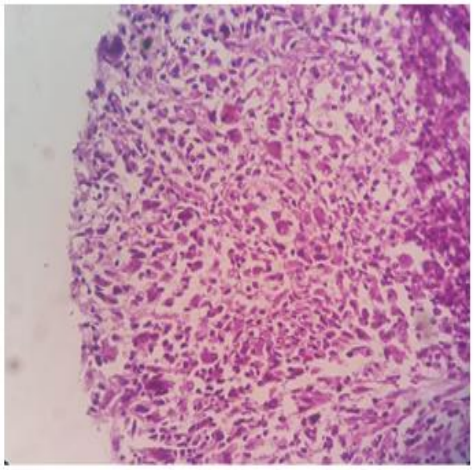

Figure 5: High power, showing histiocytes and multinucleated giant cells.

III. Discussion

The inflammatory myofibroblastic tumour is a very rare benign lesion representing 0.7% of all lung tumors. It was earlier called inflammatory pseudotumor, plasma cell granuloma, histiocytoma or fibroxanthoma. [2] Most of these tumours are discovered incidentally during radiological studies. There are no established decisive criteria for differential diagnosis. [3] The current histopathological definition of an IMT is a distinctive neoplasm composed of myofibroblastic mesenchymal spindle cells accompanied by an inflammatory infiltrate of plasma cells. [3] There are many uncertainties about the pathogenesis of IMT. Several hypotheses have been proposed such as an auto-immune mechanism or infectious origin. Indeed, 30% of cases are closely related to

recurrent respiratory infections which are caused by several microorganisms such as Mycoplasma, Nocardia, Actinomycetes, Epstein-Barr and human herpes virus. [2]Either way, tissue samples obtained by computed tomography–guided fine-needle or tru-cut biopsies, and even analysis of preoperative biopsies, are occasionally not enough to establish a diagnosis, [3] and there is no specific immunohistochemical staining for IMTs. As a result, the pathologist usually asks for the whole specimen.Patients are afflicted with nonspecific symptoms such as cough, dyspnea, hemoptysis, chest pain, fever and fatigue. Weight loss and anorexia are rare. [2] However, most patients are asymptomatic and the tumor is discovered incidentally on a chest X-ray performed for another reason. [2] The symptoms in our patient were breathlessness, weight loss, anorexia and low grade fever promoting a diagnosis of pulmonary tuberculosis.

Currently, surgery is the mainstay of the treatment for IMTs. Complete removal of the tumour generally provides resolution of all symptoms and laboratory abnormalities. However, tumours in intra- or retroperitoneal locations tend to invade adjacent structures, preventing curative resections and breeding local recurrences. [3] These patients require further management. Unfortunately, chemotherapy and radiotherapy are not successful in most patients. IMT has three histological subtypes: one is a richly vascularized and myxoid resembling fasciitis or granulation tissue; another is a compact fascicular spindle cell proliferation with variable collagenized regions and lymphoid nodules, resembling fibromatosis, and finally a very sclero-hyaline, slightly cellular pattern which looks like a desmoid tumor. [2]

The differential diagnosis of this lesion depends on the clinicopathologic setting, including the patient's age, gender, tumor location, and number of lesions. Differentiation from inflammatory leiomyosarcoma may pose a problem. However, the nuclei in leiomyosarcoma are cigar-shaped and arranged in a more regular fascicular growth pattern.Rare inflammatory myofibroblastic tumors have a conspicuous population of large multinucleated tumor cells with prominent nucleoli bearing a resemblance to the Reed-Sternberg cells of Hodgkin's disease. The immunohistochemical reactivity of the spindle and ganglion-like cells for actins and ALK and negativity for CD15 and CD30 assist in distinguishing these two entities. GIST may occasionally closely resemble an inflammatory myofibroblastic tumor, but GIST consistently stain for CD117 and DOG1 and are ALK negative.Although there are some histologic similarities between IMT and other inflammatory fibrosclerosing lesions, including sclerosing mediastinitis, idiopathic retroperitoneal fibrosis, and Riedel thyroiditis which can be differentiated by paying close attention to the clinical setting and gross and microscopic findings. [1] Finally, the question as to whether inflammatory myofibroblastic tumor and inflammatory fibrosarcoma are the same tumor, distinct entities, or represent a spectrum has been debated. It is not possible to make histologic distinctions between lesions reported by some authors as inflammatory fibrosarcoma and by others as inflammatory myofibroblastic tumor. There is also compelling evidence that these lesions are true neoplasms rather than pseudotumors. Many have been associated with aggressive local behavior that has resulted in patient death. In addition, as previously mentioned, some (but not all) tumors show aberrations of the ALK gene, supporting the evidence of a neoplastic process. [1]

Based on the two largest studies of abdominal and retroperitoneal lesions, it is clear that tumors in these locations have a propensity for more aggressive behavior than their extra-abdominal counterparts, with recurrence rates of 23% to 37%. [1]Immunohistochemistry shows reactivity for vimentin and smooth muscle actin. Immunohistochemical positivity for ALK is detectable in just over half of the cases with cytoplasmic staining, more rarely at the nuclear membrane. [2]

IV. Conclusion

Inflammatory myofibroblastic tumor is a rare benign tumor. Only histopathological and immunohistochemical study can confirm the diagnosis. It has a high possibility for recurrences. If the local invasion occurs, then complete resection is required.

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