

## A Rare Case of Inflammatory Myofibroblastic Tumor of the Lung Related to Hodgkin Lymphoma: Case Report and Literature Review

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### Abstract

### Case Report

Inflammatory myofibroblastic tumor (IMT) is a rare affection. The diversity of its cell-composition implies the existence of various clinical, biological and radiological expressions. Only histopathologic studies enable to confirm the diagnosis. Long considered a reactional wound, recent studies tend to refer towards a neoplastic nature. Complete surgical resection is the most effective care. It minimizes the risk of recurrence and is sometimes back up by radiotherapy. To our knowledge, this case report is the first of its kind to describe a Hodgkin lymphoma-related IMT.

**Keywords:** Inflammatory myofibroblastic tumor, pseudotumor, Hodgkin Lymphoma, histopathology, immunohistochemistry, surgical removal.

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### INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare condition, as it accounts for only 0, 7 % of all lung tumors [1]. It is known by several terms given its cell-composition variability: indeed, it can either be inflammatory pseudotumor, xanthogranulomatous one or even fibrous histiocytoma. The most recent cytogenetic studies indicate that it is genuinely a neoplasm rather than a reactional lesion [2]. Such growths may clinically and radiologically occur as malignancies. The definitive diagnosis is secured by anatomical pathology tests. The association of Hodgkin's disease with IMT has not been reported yet in literature.

### CASE REPORT

Our study is about a 29-year-old toxic habit free male patient, followed-up since 2016 for a nodular sclerosis Hodgkin's lymphoma for which he benefited from radiochemotherapy. Being in remission within one year, the young man exhibited for the last three months a productive cough combined with low abundance haemoptysis and a dyspnoea ranked grade 2 according to the mMRC scale (modified medical research council), the whole evolving in a context of preservation of his overall condition. The patient was eupneic on clinical assessment, at rest and in ambient air, with a SaO<sub>2</sub> = 97%. Pleuropulmonary examination revealed an apical left condensation syndrome and thoracic x-ray showed a well-marginated apical left

opacity, of an alveolar-like appearance, associated to a borderline widened superior mediastinum. Thoracic contrast-enhanced CT scan has highlighted a mediastino-pulmonary tissue mass of 33x26x27mm with poorly circumscribed lobulated contours, encompassing the upper lobe bronchus and causative of downstream atelectasis of the left upper lobe (figure 1, 2). The so-called mass was associated to round mediastinal adenopathies of regular outlines, whose targets are sitting at the pre-vascular 3A nodes where they measure 18x17 mm, and at the lower paratracheal 4R nodes with 10x9mm dimensions (figure 2). The supplementary carried out C-spine CT has also shown a ganglionic relapse, as adenitis were found in the left lymph node chain IV -measuring 24x12mm- and in the right chain III where their size is 15x11mm.

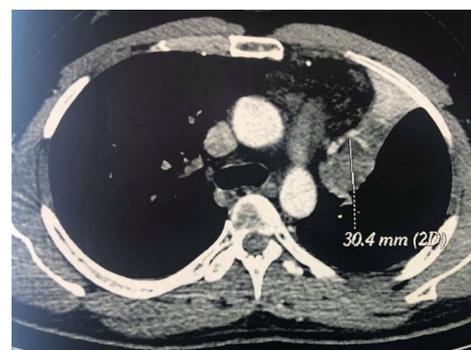
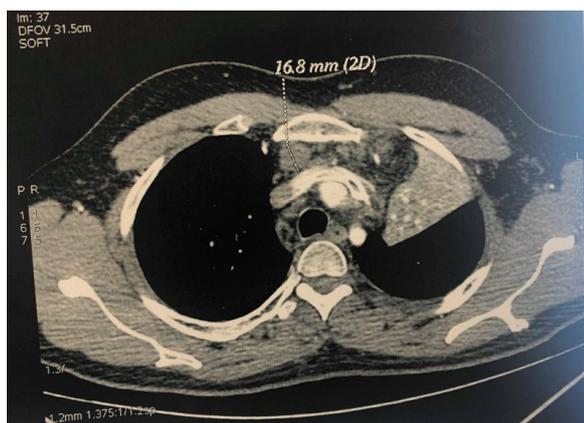


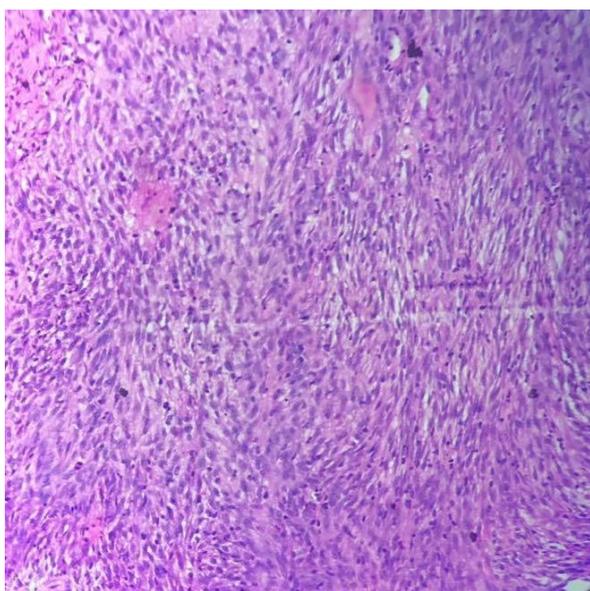
Fig-1: Chest CT scan showing a mediastino-pulmonary tissue mass with poorly circumscribed lobulated contours, encompassing the upper lobe bronchus



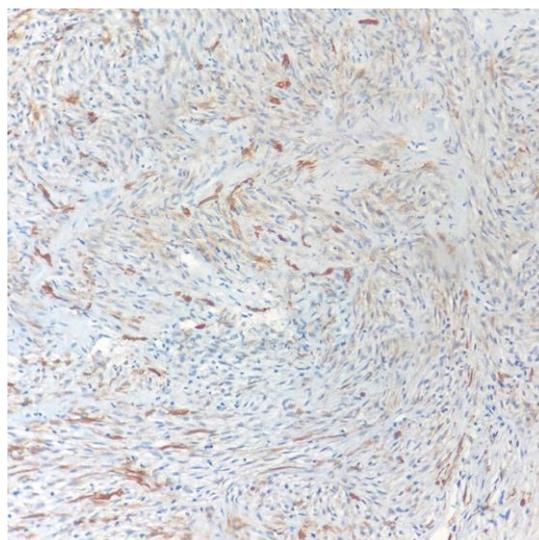
**Fig-2: Chest CT scan showing a atelectasis of the left upper lobe with mediastinal adenopathies (chain 3A)**

Meanwhile the abdomino-pelvic scan did not point to any peculiarity. Laboratory tests, for their part, displayed normocytic normochromic anaemia with a hemoglobin value at a level of Hb = 11g / dl and a white blood cell (WBC) count at 9600. Besides, C-reactive protein (CRP) level was at 67. The additional achieved assessments showed no abnormalities.

The patient benefited in our unit from bronchoscopic examination. It enabled us to identify a mucosal bud that completely clogs the left upper lobe bronchus, wherein we realized the biopsy. The histological study disclosed an infiltration of bronchial mucosa by a fusocellular tumorous proliferation which immunohistochemical analysis concluded to inflammatory myofibroblastic tumor (IMT) with positive marking to smooth muscle anti-actin by said proliferation (figure3, 4). The subject was addressed to surgery department for a potential bilobectomy.



**Fig-3: High magnification of a histological section of inflammatory myofibroblastic tumor**



**Fig-4: The immunohistochemical investigations showed positive staining for smooth muscle actin**

## DISCUSSION

Inflammatory myofibroblastic tumors are not often encountered. They arise among adolescents below the age of 16 in at least one half of the cases, showing no sex predilection since they equally affect boys and girls [3].

They have only been disclosed for the first time in the lung in 1939, even though other extrapulmonary locations been reported, as was the case with Völker and al [5]. Indeed, they outlined an IMT of the larynx. By comparing it to spindle cell carcinoma, morphologic similarities were such that only immunohistochemical analysis has led to establish the final and correct diagnosis. Lekas and al [6], for their part, described a urinary bladder site IMT that they primarily mistaken for low grade leiomyosarcoma of myxoid subtype. On another hand, the neoplasm that Al-Jabri [7] detected in the liver shared similar appearances at imaging to those of metastatic liver lesion, which brought them to rely on histological examination to establish a proper diagnosis. A wide range of other localizations have been noted in the literature such as spleen, lymph nodes, esophagus, stomach, salivary glands, breast, epididymis, central nervous system, and soft tissues [4].

In our examinee, from a pathophysiological perspective, inflammatory cells appear to be subject of dysregulated growth. It has been postulated that the IMT development may be related to a primary infection, that being supported by the fact that several microorganisms were retrieved, including Mycoplasma, Nocardia, Actinomycetes, Epstein-Barr and Human Herpes Virus [8].

Meanwhile, further works stipulate that it may be question of an actual neoplasm on the grounds that, in the myofibroblastic component, a fusion gene was identified, evolving the ALK gene, a tyrosine kinase

oncogen positioned on chromosome 2p23, which is at the origin found disposed in anaplastic large cell lymphomas. A merge like this originates constitutive overexpression of the ALK, generating the proliferation of cells [4, 9]. Worth noting that in our case, ALK overexpression by the tumor was not immunohistochemically detected.

When our patient suffered from chronic cough, others can also experience, hemoptysis, pleuritic chest pain, stridor or else post-obstructive pneumonia. Symptoms are thus generally non-specific and can even absent as it was for up to 40% of reported cases [3].

As imaging also gives non-specific results, inflammatory pseudotumors (IPT) remain evoked as a diagnosis of exclusion. In fact, those growths are embodied in the form of well-circumscribed solitary opacity of 1 up to 10 centimetres in diameter, localized at the peripheral area of the lung and foremost on the inferior lobes [10]. Further less common lesions have been outlined, such as calcifications, pleurisy and multiple nodules in respectively in 15, 10 and 5 % of the cases. The presence of hilar and mediastinal lymph nodes with wound extension towards the mediastinum, as in the case among our subject, was reported in less than 7% of patients [10].

The positive diagnosis of IPT draws on pathology data and usually requires surgical biopsy, whereas in our case it is an endobronchial biopsy that was performed given the exceptional endobronchial tumour spread.

From the histopathological point of view, IPT is constituted by an inflammatory infiltrate incorporating plasma cells, B and T cell lymphocytes, histiocytes and macrophages at times Xanthomatous. A population of fibroblasts and/or myofibroblasts is also found within the greater or lesser hyaline collagen fiber [12]. This twofold component explains why IPT have a wide range of designations, knowing that the two most encountered in the literature are “inflammatory pseudotumors” (IPT) and “Inflammatory myofibroblastic tumor” (IMT).

There is no standardized histological classification of IPT. Matsubara and al [3] still managed to distinguish three histological kinds: the organized pneumonia type (44%), the organized histiocytoma (44%) and the lymphoplasmacytic proliferation type (12 %) [3]. The latter type is the one we found in our case. Cerfolio and al [12], for their part, depict two varieties of IPT, invasive and non-invasive one. There also exist a number of aggressive and recurring IPTs that are histopathologically characterized by the presence of at least 3 of the following criteria: vascular invasion, hotbeds of necrosis, nuclear atypia, numerous mitosis, significance of inflamed cellularity, and the presence of numerous non-standard giant cells, leading to the

classification of the neoplasm in an intermediary framework between benign fibrous histiocytoma (an actual inflammatory pseudotumor) and malignant one. Hence, such intermediate forms must be regularly monitored, since they may possibly evolve towards secondary malignancy.

The standard treatment is the fullest possible surgical removal, either out of fear of neoplasia in absence of prior diagnosis, or else of local invasion by contiguity that sometimes occurs at the detriment of vascular or even mediastinal structures [13].

Major Mediastinal disorders may warrant a preoperative radiation therapy. Further remedies, such as corticosteroids, antibiotics or else chemotherapy have been attempted with varying degrees of success, inter alia, on multiple, relapsing, or non-operable forms [11].

The tumor rarely recurs when surgically removed with an interval ranging from 6 months to 11 years [11]. When this happens, it is mainly related to its infiltrative forms.

## CONCLUSION

It is still surprising when the diagnosis of inflammatory pseudotumor is arisen with histologic examination that remains, along with immunohistochemical study, the only way to assert the nature of such lesion.

Although it is a benign tumor, its local invasion and recurrence potential are sufficient to justify its complete surgical excision.

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