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Diagnostic Conundrum in a Patient with Haemoptysis - Tuberculosis *vs* Inflammatory Myofibroblastic Tumor: Case Report

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ABSTRACT

A 37 year old health care worker presented with haemoptysis, night sweats and cough. Computed tomography (CT) scan of her thorax showed a right upper lobe cavitating lesion. She was treated for tuberculosis despite negative sputum cultures for Acid Fast Bacilli as the history and radiological findings appear classical for tuberculosis. She continued having persistent haemoptysis despite being on treatment and was referred to a thoracic surgeon for a surgical opinion. She underwent right video assisted thoracoscopic right upper lobe wedge resection and histological assessment of the tissue showed inflammatory myofibroblastic tumor of the lung.

Keywords: *Haemoptysis, Tuberculosis, Inflammatory Myofibroblastic Tumor, Computed Tomography*

Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare pathological condition that can affect the spleen, breast, maxillary sinus, central nervous system, maxillary sinus and soft tissue (Coffin *et al*., 2007; Zhang *et al*., 2009). The most common location of IMT would be in the lung and it tends to occur in children or adolescents.

Clinical symptoms and radiological features of pulmonary IMT are non-specific making it is difficult to diagnose before surgical resection. It is difficult to differentiate IMT and tuberculosis in the lung purely by imaging without histology. Especially in countries with high burden of tuberculosis, patients may be treated presumptive for tuberculosis (TB). In this case, the patient was treated with tuberculosis medications for 6 months while breast feeding despite negative laboratory test for TB as her history and radiological findings were highly suggestive of tuberculosis.

Due to the proliferation of myofibroblastic cells and its potential malignant behavior, IMT is

typically treated with resection and patients will be closely monitored for recurrence.

In this case, surgery was diagnostic and therapeutic for the patient. Without surgery, the patient would have been treated wrongly for tuberculosis and the IMT would have been left untreated.

Case Presentation

A 37-year-old lady who works as an occupational therapist in a healthcare institution with no significant past medical history presented to the Emergency Department with 3 days history of fever, night sweats and haemoptysis (5-10ml).

On examination, she was febrile at 38 degrees but was otherwise stable and had no other significant findings. Her lungs were clear on auscultation and there was no cervical or axillary lymphadenopathy

Investigations

Biochemical and haematological investigations were within normal range. Initial Chest X-ray showed an opacity in the right upper lobe (Fig. 1a).

Thoracic computed tomography (CT) scan showed a thin-walled cavity in the right upper lobe. Sputum was also taken for Acid fast bacilli smear and culture (Fig. 1b).

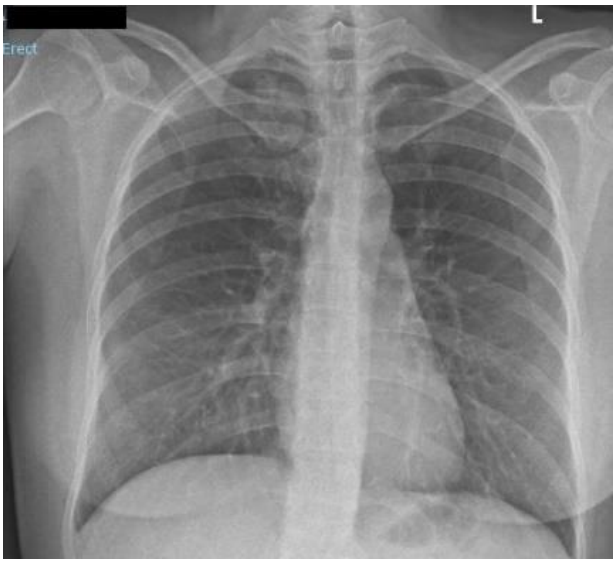

Figure 1a: CXR.

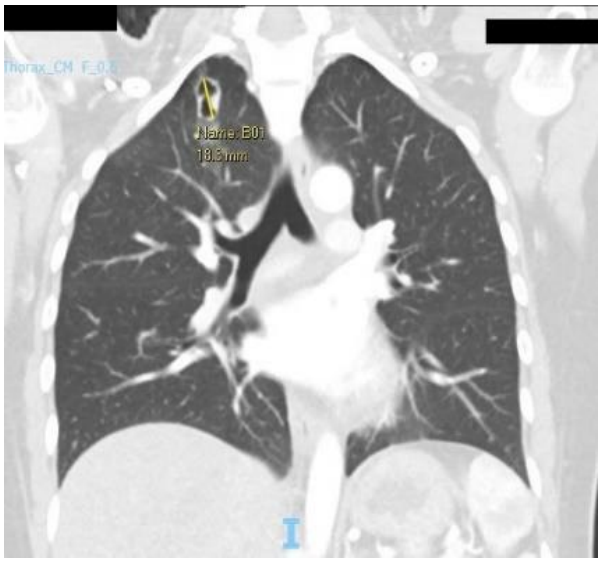

Figure 1b: Initial CT thorax demonstrating right upper lobe lunge lesion.

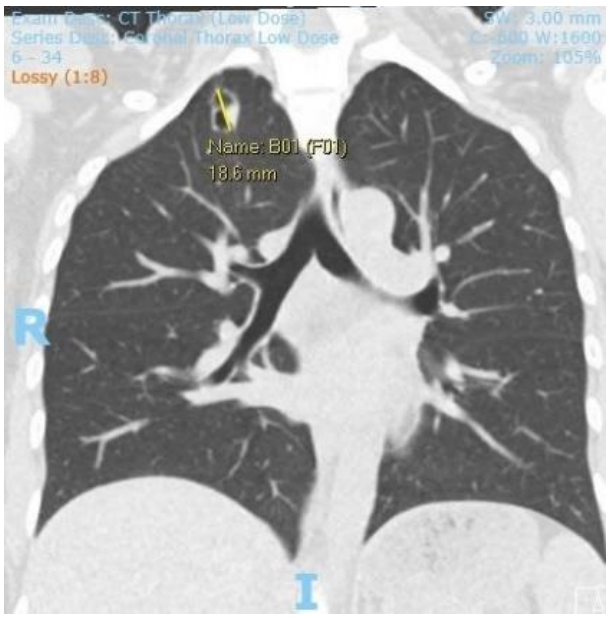

Figure 1c: CT Thorax 3 months later which showed that the right upper lobe lung lesion remained stable.

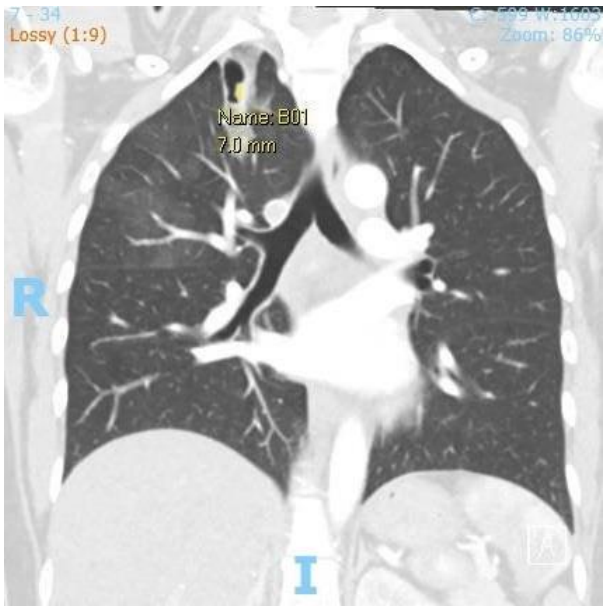

Figure 1d: CT Thorax 6 months after initial CT thorax which showed the right upper lobe lung lesion remained unchanged.

Treatment

She was started immediately on anti-tuberculosis treatment based on the history and radiological findings. Sputum was taken for AFB smear and culture prior to initiating treatment. Her baby was also started on pyridoxine supplements in view of the TB medications.

Outcome and Follow-Up

She was followed up 2 months later and was noted to still have persistent haemoptysis intermittently. The repeated thoracic computed tomography showed stable right upper lobe cavitating lesion (Fig. 1c). She was reassured and continued on TB medications.

She presented again 2 months later with increasing frequency and quantity of haemoptysis. Computed tomography scan again showed a stable right upper lobe cavitating lesion (Fig. 1d) and the initial sputum culture returned as negative for AFB. She was referred to the thoracic surgery unit for an opinion and underwent right video assisted thoracoscopic wedge resection of the right upper lobe lung lesion. Prior to the surgery, a bronchoscopic evaluation was performed which showed normal mucosa and no active bleeding.

She recovered well and was discharged on post-operative day 2. She had no further haemoptysis. Histological finding reviewed that the lesion was an inflammatory myofibroblastic tumor (Fig. 2a-b).

Anaplastic lymphoma kinase (ALK) gene fusion was also detected.

She will be followed up with yearly thoracic computed tomography as IMT is known to have a potential to recur (Fig. 3).

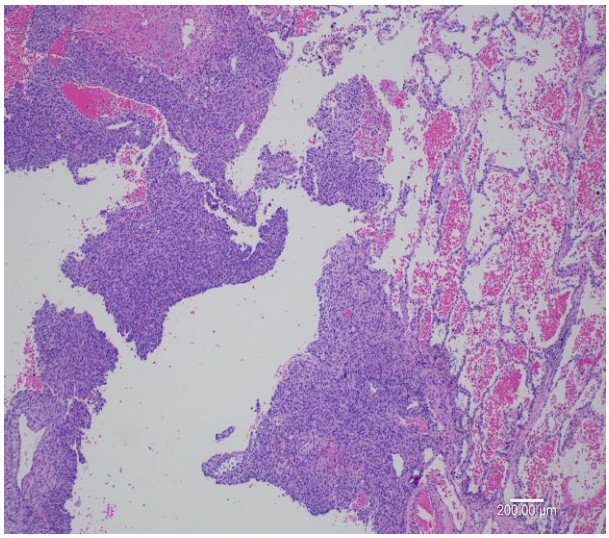

Figure 2a: H&E 4x: lung tissue with hemorrhage and a cavitary lesion composed of a proliferation of spindle cells with mixed inflammatory cells.

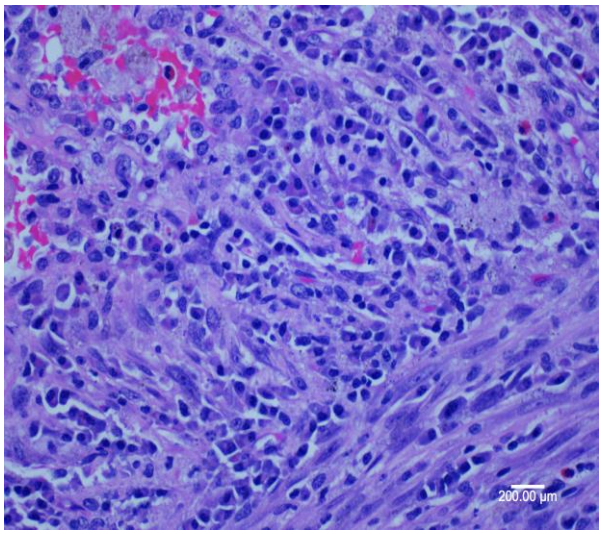

Figure 2b: H&E 40x: spindle cell proliferation with a whorling or storiform pattern with interspersed foamy macrophages, lymphoplasmacytic cells, polymorphs and some eosinophils.

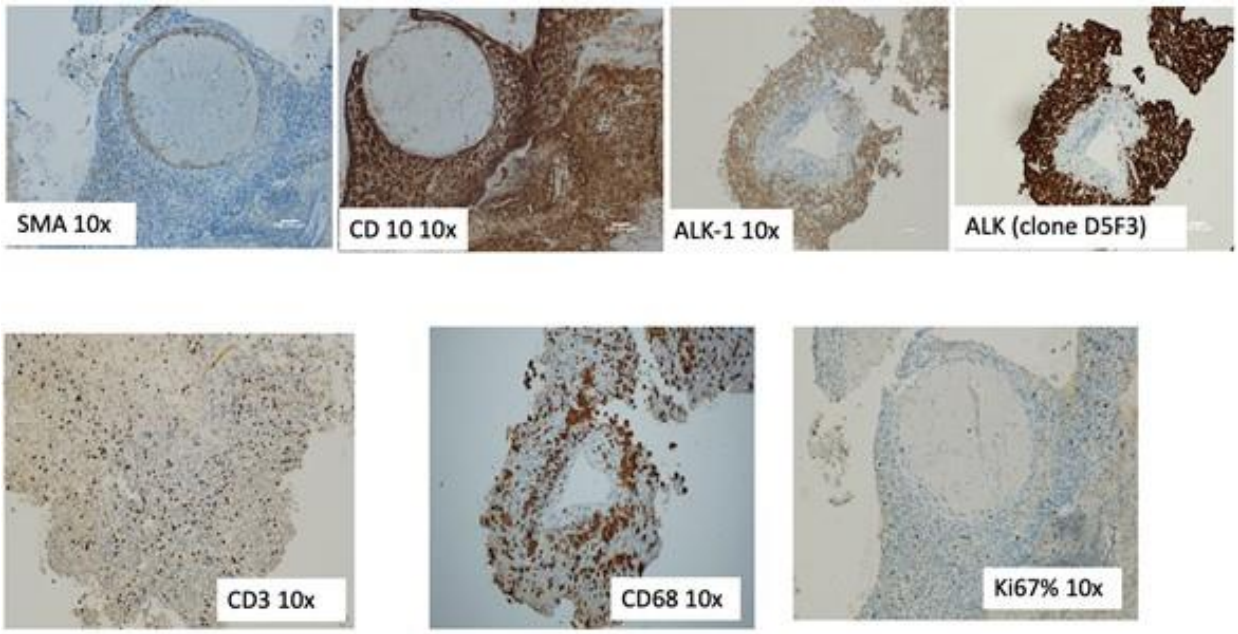

Figure 3: Immunohistochemistry staining of right lung lesion. SMA shows focal weak reactivity in tumor cells. CD3, CD 10, CD68 immunohistochemical stain showed diffuse reactivity in spindled cells. This elicited a possible diagnosis of metastatic stromal sarcoma of the endometrium which are CD10+, but this Is not specific of endometrial stromal sarcomas since CD 10 is common in stromal cells accompanying diverse carcinom. ALK-1 and ALK (clone D4 5F3) showed positive reaction in tumor cells. Ki 67 shows proliferative rate up to 12% in the most proliferative areas.

Discussion

Inflammatory Myofibroblastic Tumor is defined by the World Health Organization (WHO) in 1994 as a soft tissue tumor composed of fibrous tissue, myofibroblasts and inflammatory infiltration, predominantly histocytes and plasma cells (İçmeli *et al*., 2014).

It can arise in many places but the most common location would be in the lung. The cause of IMT in lung is unclear but it has been thought to be associated with recurrent infection (İçmeli *et al*., 2014).

Around 50% of IMTs show a chromosomal translocation of the anaplastic lymphoma kinase (ALK) gene with a partner gene locus with potentially aberrant kinase expression (Schweckendiek *et al*., 2015). Half of the IMTs has a clonal translocation that activates the anaplastic lymphoma kinase (ALK)-receptor tyrosine kinase gene located at 2p23 locus (Griffin *et al*., 1999). As a result, ALK protein is overexpressed and can be detected at the immunohistochemical level. It is hence used as a diagnostic marker for this disease. ALK is a receptor-type protein tyrosine kinase, which is thought to be oncogenic due to gene fusion, such as in anaplastic large cell lymphoma, lung cancer and IMT.

Inflammatory myofibroblastic Tumor in the lung can occur at any age but they are more common in children and adolescents. Hammas and Camela have described IMTs in children (Hammas *et al*., 2012; Camela *et al*., 2018). IMTs however are not commonly described in adults. The common presentation with pulmonary IMT would be cough, fever, haemoptysis and lethargy. Radiological findings of pulmonary IMT are variable and nonspecific such as calcification, cavity, necrosis and pneumonia (Barbetakis *et al*., 2006; Zhang *et al*., 2020). These findings are similar to lung cancer and tuberculosis.

Histopathologically, IMTs are divided into 3 patterns: Organizing pneumonia, histiocytic and lymphohistiocytic. The most common would be histocytic pattern which is characterized by spindle shaped myofibroblasts arranged in whorls, including myofibroblasts and fibroblasts arranged in a storiform manner with chronic inflammatory cell infiltration (Griffin *et al*., 1999). This is demonstrated in the histology of the specimen obtained from the patient (Fig. 2a-b).

Tuberculosis is the top 10 causes of death worldwide in 2015 and most of the new cases occur in Asia. Sometimes, IMT in the lung are misdiagnosed as tuberculosis and this is especially common in countries with high burden of disease for tuberculosis (Tam and Lai, 2019). In this case, the lady was an allied health worker working in a mental institution in Singapore which would make her high risk of contracting tuberculosis. IMT of the lung are treated with surgical resection and it is reported that 6.6-13% of pulmonary OMT following resection may recur locally (Hussain *et al*., 2005). As such, patients should be followed up closely with serial imaging to monitor recurrence.

Learning Points/Take Home Messages

- We report a rare case of haemoptysis as the primary presentation for IMT in the lung.
- The patient was misdiagnosed with tuberculosis and was treated with antituberculosis medication for 6 months.
- Despite the high incidence of tuberculosis in our population, we should consider the possibility of pulmonary IMT to prevent misdiagnosis and administering the wrong treatment.

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