CASE REPORT

An Unusual Cause of Progressive Chest Deformity in A Child

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ABSTRACT

Inflammatory myofibroblastic tumour (IMT) is a rare tumour with intermediate malignant potential and unclear aetiology. In most cases, the clinical presentation is non-specific. The liver and lung are the most common location for IMT in children, and mediastinum is an uncommon place. Symptoms vary depending on the location of IMT, thus making a definitive diagnosis to be even more difficult. Until the histological investigation can rule out IMT, it should be considered in the differential diagnosis of individuals with localised masses. We report here a case of an 8-year-old boy with right-sided chest pain and deformity before the diagnosis of mediastinal IMT was made through tissue biopsy.

INTRODUCTION

Chest wall deformities in children are due to congenital or acquired causes. The most common congenital chest wall deformities are pectus excavatum which accounts for 88% of the cases, and pectus carinatum, which contributes to the remaining 5%.[1] Acquired chest wall deformities may be caused by primary diseases of the chest wall itself, including tumours and infections, iatrogenic causes from previous surgery and secondary to post-traumatic deformities resulting from direct or indirect trauma to the trunk.[2] Inflammatory myofibroblastic tumour (IMT) is a rare neoplasm that mainly affects children and young adults. Thirty percent of lesions are asymptomatic, and the rest have non-specific features.[3]

Many of the cases were diagnosed incidentally during a routine examination. During a 10-year study period, a total of 19 cases of IMT were treated in a major hospital in China. The age of patients from their series ranged from four to 111-months and the location of the tumour varied, mostly abdominal in origin. Fourteen cases were abdominal IMT, two cases were thoracic and pulmonary IMT and one case was glottic IMT. Among these 19 patients, abdominal pain, abdominal mass, haematochezia and vomiting occurred in 12 cases, pulmonary and chest wall mass in one case, and symptoms such as cough, dyspnoea, and chest pain in four cases.[4] We report a case of an 8-year-old boy with IMT who presented with right-sided chest pain and progressive chest deformity.

CASE REPORT

MHH is an 8-year-old boy who was previously well. He was brought to a district hospital in Pahang due to worsening chest pain and anterior chest swelling that were present for four months. There was no respiratory distress or fever. Chest x-ray showed consolidation over the right middle zone, and he was diagnosed as having right lobar pneumonia.

He was advised for admission to that particular hospital, but his parent opted to bring him to our centre, Sultan Ahmad Shah Medical Centre @IIUM instead as his father works nearby. He had been experiencing right-sided chest pain for the past four months prior to presentation. It was intermittent and heavy in nature, with a pain score of one to two over ten. The pain was located over the right side of his chest with no radiation, but exaggerated by breathing. He was noticed to have a right anterior chest

swelling which was absent before, and it was not painful. The chest deformity has progressively become more prominent for the past three months. He was able to lie flat during sleep with no orthopnoea. He initially ignored the pain and swelling as there was no interruption in his daily activities. Subsequently, he started to feel more lethargic which was noticed by his teacher at school. He had a history of hospital admission twice at one year of age due to lower respiratory tract infection. Both were short hospital stays in which he was treated with intravenous antibiotic and discharged well. On admission, he was alert and not in pain. He had a lean body built and weighed 21 kg with a height of 126 cm.

No muscle wasting and no finger clubbing were observed. He was not in respiratory distress and not cyanosed. His chest was asymmetrical and more prominent on right antero-posterior (AP) (Figures 1A and 1B) view. There was a 1 cm x 4 cm non-tender right anterior chest swelling overlying the second until the fourth rib with a diffuse margin. There were no overlying skin changes or signs of inflammation. His chest expansion was reduced on the right side. There was no trachea deviation. There was dullness on percussion over the right middle zone and reduced tactile resonance over the right middle zone until the lower zone. Air entry was reduced with crepitations heard over the right middle zone.

The full blood count showed microcytic hypochromic anaemia, haemoglobin of 10.9 g/L with MCV 63 fl and MCH 19.6 pg and thrombocytosis of 658 x 14.6 10^9/L. Total white cell was elevated 14.6 10^9/L. Full blood picture showed left shift with no blast cell seen. The inflammatory markers were elevated with a CRP of 23.4 iu/L and an ESR of 73 mm/hr. *Mycoplasma pneumonia* serology was negative, and blood culture and sensitivity showed no growth. Chest x-ray showed ill-defined consolidation at the right perihilar region suggestive of a right mediastinal mass (Figure 2). Contrast enhanced computed tomography (CT) thorax revealed a heterogeneously enhancing anterior mediastinal mass with subtle calcifications and local infiltration to the right anterior chest wall. This mass appears to conform to an enlarged thymus gland (Figure3). He was referred to paediatric surgery and decided for biopsy. A percutaneous biopsy was performed, which showed evidence of inflammatory myofibroblastic changes with minimal lymphocyte and plasma cell and no evidence of granulation tissue or malignancy, suggestive of IMT. A surgical excision was advised, but the parents were not keen on surgery as his general condition improved, and his pain disappeared.

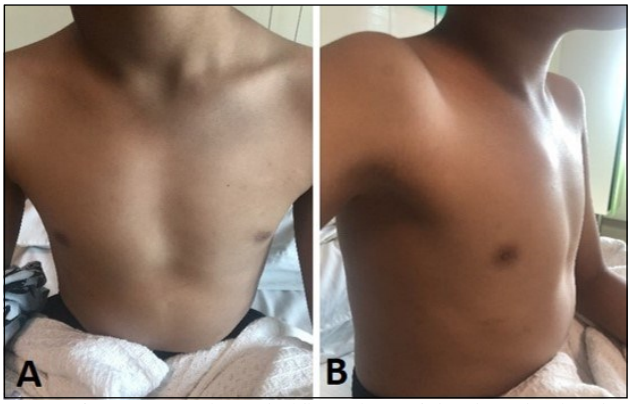

Figure 1A & B: Anterior and oblique views of the chest showing the swelling on the right anterior chest wall

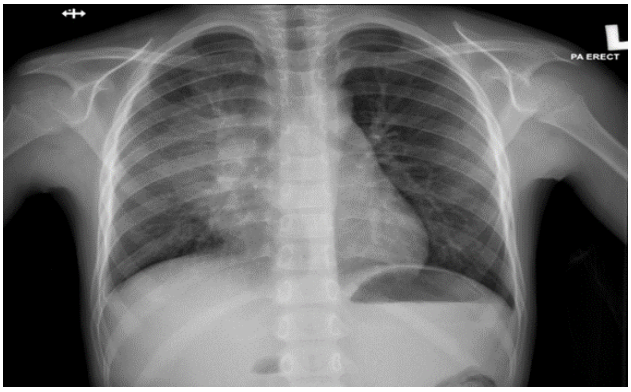

Figure 2: CXR showing an ill-defined consolidation at the right perihilar region. The rest of the lung fields were unremarkable

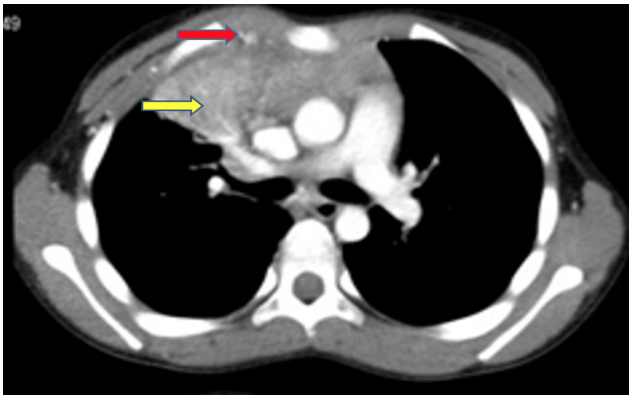

Figure 3: Contrast enhanced CT thorax showing a heterogeneously enhancing anterior mediastinal mass which seems to conform to an enlarged thymus gland. There is infiltration to the right anterior chest wall (red arrow). A collapsed consolidation of the right upper lobe is also observed (yellow arrow)

DISCUSSION

Inflammatory myofibroblastic tumour (IMT), also known as inflammatory pseudotumor, is rare and vary in its clinical presentation.[5] The anatomical and clinical manifestations of IMT are non-specific. However, it has been reported that lung and liver are the most commonly affected organs.[6] The manifestations of IMT can be divided into pulmonary and extrapulmonary manifestations. Unlike our patient, only 15% to 30% of patients generally present with an inflammatory syndrome manifested by fever, weight loss and malaise.[7] Other symptoms include cough, respiratory distress, anaemia, recurrent pulmonary infections and arthralgia, while 4% may be asymptomatic. Mediastinal IMT is rare and may occur either primarily or secondary to extrapulmonary involvement of a primary pulmonary lesion.[8]

Mediastinal IMT may present with symptoms of mass effect on the adjacent structures including chest pain (8%) as in our patient. Other symptoms are dyspnoea (40%), dysphagia and haemoptysis (4%) which are not present in this patient.[9] Extrapulmonary IMT shares many clinical and morphologic similarities with pulmonary IMT, but extrapulmonary IMT usually affect younger individuals, tends to be larger and often involves vital structures such as the heart and the great vessels.[8] CT thorax is crucial, although the diagnosis would not have been possible without histological evaluation. The laboratory tests associated with IMT are not specific. The affected patient could have microcytic anaemia, thrombocytosis, and elevated inflammatory markers such as erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP).

Hypogammaglobinaemia could also be present, likely due to overproduction of interleukin during inflammation.[7,10] IMT usually occurs insidiously and are discovered incidentally during physical examinations. It was previously described as a benign lesion, but due to its potential for recurrence and high rate of metastasis, it was later classified as an intermediate grade tumour.[10] Histologic analysis is required for diagnosis, as radiologic imaging alone is difficult to differentiate IMT from other neoplasms.[6] The recurrence rate varies from 2% to 25% for pulmonary and extrapulmonary IMT, respectively. Distant metastases, on the other hand, occurs in less than 5% of cases.[7] IMT is myofibroblastic in origin and consists entirely of spindle cells associated with a variable density polymorphic infiltration of mononuclear inflammatory cells.[9] The etiological factors responsible for IMT are not clearly understood. Some researchers believe that it is an immunological response to an infectious or non-infectious agent. The role of viral and bacterial infections has been reported, including human herpesvirus, Epstein-Barr virus, *Mycobacterium avium intracellular*, *Bacillus sphaericus*, and *Coxiella burnetti*.[10] The findings, however, were inconclusive. Molecular reports also suggest that anaplastic lymphoma kinase (ALK) and rearrangement of chromosome 2p23 enhanced tumour proliferation and metastatic features in 90% of IMT cases. ALK are highly specific for the lesions, but it is not a sensitive marker in children.[4]

Surgical treatment is the mainstay of therapy in the early stage of tumour growth, and complete removal of the tumour can prevent recurrences. Glucocorticoid therapy, which reduces the inflammatory markers and cytokines and radiotherapy or chemotherapy have adjuvant effects when surgery is contraindicated or if the mass is incompletely removed. These non-surgical treatments can shrink or even cause the tumour to disappear.[10] The parents were reluctant for the operation because they perceived the child's symptom was resolving. Thus, he is currently under follow up for any vicissitudes of the tumour.

CONCLUSION

IMT should be considered in the differential diagnosis of patients that came with localised masses, systemic symptoms or pulmonary lesions until excluded by histologic examination. IMT occurs in young children at various sites with corresponding clinical symptoms. Surgical resection remains the treatment of choice. For our patient whose parents were not keen on surgery, close monitoring is mandatory. Any alarming symptom should prompt for a reimaging and reconsideration of the surgery.

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