***Case report***

**INTRATHORACIC INFAMMATORY MYOFIBROBLASTIC TUMOR IN A NIGERIAN CHILD: A RARE PRESENTATION- CASE REPORT AND REVIEW OF LITERATURE**

**ABSTRACT**

**Background**

An inflammatory myofibroblastic tumor (IMT) is a rare, predominantly myofibroblastic neoplasm accompanied by inflammatory cells, including lymphocytes and eosinophils. It is predominantly found in children and adolescents. Extra-pulmonary sites for IMTs include the head and neck region. IMT of the thorax is rare, accounting for 0.04%-0.1% of all pulmonary tumors. Because the clinical presentation of IMT is nonspecific and rare, it poses a major diagnostic challenge. Hence, the need for IMT to be differentiated from other chronic infections and malignant lesions based on histopathologic findings and immunohistochemical analysis. The mainstay of care for localized tumors is a surgical resection. Spontaneous regression has also been reported in some people. A response rate of approximately 50% has been reported with the use of chemotherapy.

**Case presentation**

An 8-year-old male with complaints of generalized body swelling, which progressed to weakness of both lower limbs and later inability to walk, with associated urinary and fecal incontinence of about 8 months duration. However, 5 weeks before presentation, developed cough, difficulty with breathing, progressive weight loss, and drenching night sweats. An initial diagnosis of disseminated tuberculosis was made, but this was changed following a histology report of an inflammatory myofibroblastic tumor (IMT). Further treatment was not done as the child passed on.

An 8-year-old male who presented with a history of generalized body swelling, which progressed to weakness of both lower limbs and later inability to walk, with associated urinary and fecal incontinence of about 8 months duration. However, 5 weeks before presentation, developed cough, difficulty with breathing, progressive weight loss, and drenching night sweats.

**Conclusion**

Inflammatory Myofibroblastic Tumor is a rare tumor that is easily misdiagnosed. This case report contributes significant clinical insight into the presentation and diagnosis of a rare tumor in a low-resource tertiary healthcare setting. Its importance lies in highlighting the non-specific features at presentation and possible treatment approaches in the context of limited diagnostic infrastructure. Additionally, the case provides valuable insights into the management of IMT, informing clinicians practicing in similar environments.

Key words: Inflammatory Myofibroblastic Tumor (IMT), Anaplastic Lymphoma Kinase (ALK) gene, inflammation.

**INTRODUCTION**

Inflammatory myofibroblastic tumor (IMT) is a rare neoplastic disorder of intermediate biological potential, noted for its low likelihood of recurrence and metastasis.1 It is known by several names, such as pseudosarcoma, epithelioid inflammatory myofibroblastic sarcoma, and inflammatory pseudotumor.2 IMT typically occurs in the lungs which is the most common extra-soft tissue site, and in other soft tissues of children and adults. Nevertheless, the anatomic distribution differs, and a broad age range has been documented, with a low prevalence between 0.04% to 0.7% in the younger population.3,4 Although IMT is rare in the maxillofacial region, it has been identified in several locations, including the epiglottis, endolarynx, parapharyngeal space, maxillary sinus, orbits, submandibular area, and oral cavity.1,5 Although the etiopathogenesis of IMT remains unclear and controversial, several risk factors have been identified, including smoking, trauma, chronic inflammation, autoimmune diseases, and IgG4-related disease. 1,6 Some theories propose an abnormal immunological response to viruses or antigens (Human Herpesvirus-8, Human Immunodeficiency Virus, and Epstein-Barr virus are primarily implicated); however, the exact cause remains unknown. 7-9 Histologically, myofibroblasts are mesenchymal derived cells, consisting of myofibroblastic spindle cells distributed in a fascicular pattern, with prominent chronic inflammatory cell infiltration, spindle-shaped cells intermixed with lymphocytes, and plasma cells, showing vesicular nuclei and eosinophilic cytoplasm. Its biological behavior is not well defined, but it is generally regarded as benign; nonetheless, recurrences and metastasis have been reported in some studies. The only mesenchymal malignancy that specifically describes “myofibroblastic” differentiation is the so-called low-grade myofibroblastic sarcoma.10 In cases of malignant lesions, metastasis occurs in less than 5% of cases, with malignant transformation seen in 8-18% of cases. 2

The identification of recurrent anaplastic lymphoma kinase (*ALK)* gene rearrangements has significantly contributed to an enhanced understanding of this rare tumor. Anaplastic Lymphoma Kinase (ALK) is a receptor tyrosine kinase that was initially identified as a component of the nucleophosmin (NPM)-ALK fusion oncoprotein, which is present in anaplastic large cell lymphoma (ALCL).11 Additionally, the *ALK* rearrangement is less common in adults compared to children with IMT.12 There are multiple ALK fusions present, and a distinction is observed between pulmonary and extrapulmonary genes. The differing components of the tumor may explain the differences observed between these two age groups. Kinase fusions are vital to the biology of numerous IMTs and have been detected in approximately 80% of these tumors.13 This condition can be categorized into the more prevalent pulmonary variant and the less common extra-pulmonary variant. In children and young individuals, the pulmonary variant, known for its benign nature, is more frequently encountered. Conversely, the extra-pulmonary type, noted for its highly aggressive nature, is observed in the older population [after the 2nd decade].14,15

IMT CASE REPORT

An 8-year-old male who presented to us in a chronically ill state. Before presentation, he had developed a chronic cough, generalized body swelling, which progressed to weakness of both lower limbs, then inability to walk, with associated urinary and fecal incontinence of 8 months duration. A chest x-ray done revealed pleural effusion, which was drained and commenced on anti-kochs medications, which he had taken for 5 months, with resolution of cough, fecal and urinary incontinence, and regained milestone (standing with support).

Five weeks before presentation in our facility, he developed a cough, difficulty with breathing, and progressive weight loss. Cough was productive of yellowish, non-bloody, and non-foul-smelling sputum, with associated chest pain, fever, and drenching night sweats. On examination, he was chronically ill-looking, severely pale, with grade 3 digital clubbing, significant axillary and submandibular lymphadenopathy, dyspneic, tachypneic, and bilateral pitting pedal edema up to the knee. On chest examination, there was asymmetrical flattening of the correct upper lung zone, reduced chest expansion, air entry, vocal fremitus, and dull percussion notes over the correct lung zone. The abdomen was markedly distended, and the liver was 8cm below the right coastal margin, firm, non-tender, and ascites was present. There was marked muscle wasting, with grade 2 power in the lower limbs and grade 4 in the upper limbs. The tone was reduced in the upper limbs and increased in the lower limbs, and ankle clonus was present. A diagnosis of disseminated tuberculosis (pulmonary, Pott's disease) with suspected drug-resistant tuberculosis was made. He received blood and blood products, intravenous antibiotics, antifungal agents, anti-tuberculosis drugs, pyridoxine, prednisolone, and had deranged electrolytes corrected, as well as a pleural effusion drained.

A complete blood count showed leukocytosis (19,000), with neutrophilia (81%). The repeat count was 4,300 and 74%, respectively. The ESR was 10 mm/hr. Electrolytes showed hyponatremia (132 mmol/L) and acidosis (17 mmol/L) and were corrected; retroviral screening was negative. Chest x-ray showed right lung collapse with a pleural effusion. Ferritin 720.81ng/ml (normal range 10-150ng/ml). Gene expert test- negative for tuberculosis. Lactate dehydrogenase 325u/L (normal range 110-295u/L). Bone marrow aspiration (BMA)- normal. Liver function test- GGT 122(normal range <55U/L) and ALP 229 (normal range 0-211 U/L), albumin 23 (32-55g/L), total protein 49 (67-82g/L). Peripheral blood film (PBF) and bone marrow aspiration (BMA) were both normal. Bone marrow biopsy reported a gelatinous transformation of the marrow.

Chest Computed Tomography (CT) scan showed- a large paravertebral mass from T6 down to the abdomen, displacing the thoracic aorta anteriorly and to the left, displacing the right pleura, causing a collapse and displacement of the right lobe anteriorly, and significant bilateral pleural effusion. Pulmonary nodule (1x0.8cm) is seen in the anterior segment of the left upper lobe, with multiple para-aortic nodes in the abdomen displacing the aorta anteriorly; Diagnosis- a lymphoma with pleural and abdominal extension, and distant metastasis (bone, pulmonary).

Tru-cut biopsy of the right intrathoracic mass revealed a neoplastic lesion composed of spindle cells admixed with cells of eccentric nuclei and brightly eosinophilic cytoplasm, myxoid background with mixed inflammatory infiltrates comprising lymphocytes, plasma cells, and histiocytes. There are hyper- and hypo-cellular areas, thin and thick-walled blood vessels, and golden-brown pigmented areas, all within the lesion. Vessels with thrombus formation and skeletal muscle are seen. No mitosis or necrosis seen- Diagnosis –mesenchymal neoplasm, probably an inflammatory myofibroblastic tumor was made. Immunohistochemistry and molecular testing were advised for a definitive diagnosis, but could not be done due to financial constraints.

Due to the new diagnosis, the caregivers were counseled and treatment options explained in detail. However, further care was not done as the child passed on.

DISCUSSION

Inflammatory myofibroblastic tumor (IMT) found in the pulmonary area was first described in 1939 by Brunn and his colleagues. 10 The overall prevalence of IMT is 0.04–0.7%. It is typically seen in children and adolescents aged between 2 and 16 years, although a few cases have been reported in neonates. IMT has two types, the more common pulmonary variant and the less common extra-pulmonary variant.  14,15  The index child was an 8-year-old, which falls into the pulmonary variant, as seen from the history and examination findings; however, the clinical course did not appear benign, as some clinical features resembled those of the extra-pulmonary variant, known to have an aggressive course. In contrast, some other studies have found IMT located in different sites, including the umbilicus, jaw, brain, and the axilla, which were not found in the index child. The location of the lesion determines the symptoms at presentation. Usually, patients remain asymptomatic until the lesion becomes symptomatic.16 Symptoms may present as pain, and approximately 20% present with symptoms of generalized malaise, fever, and weight loss 17. Some of these findings were part of the presenting complaints of the index patient. Based on the duration of illness, our index child had a clinical feature suggestive of a chronic inflammation, which is a known risk factor for IMT, though viral screening was not done. In contrast, Neuhauser et al had positive EBV results in some patients with splenic IMT.18 It is also important to note that a negative viral screening does not preclude the possibility of a previous asymptomatic viral exposure.  Despite its classification as a benign or locally aggressive tumor, IMT can exhibit unpredictable behavior, necessitating careful diagnosis and management. 19

The radiological presentation of IMTs is diverse. On imaging, several masses in a single anatomic region may be visible, ranging from infiltrating lesions to well-delineated ones, with diverse inflammatory and fibrotic cells within the mass. This finding is in keeping with the reported child, where several nodes were found in both the thoracic and abdominal areas. On the CT scan, persistent and impeded contrast uptake has been reported in the fibrotic component of the IMT. Magnetic resonance imaging may reveal low signal intensity on T1- and T2-weighted images due to fibrosis, accompanied by restricted diffusion.20 Biologically, IMTs can induce inflammation with leukocytosis, neutrophilia, and elevation of C-reactive protein and erythrocyte sedimentation rate, and all these were evident in the laboratory investigations of the index child. 21 Historically, IMTs were considered to arise as a result of an exaggerated reactive or reparative process in response to tissue injury. Rising levels of γ-glutamyl transferase (γGT), alkaline phosphatase (ALP), aspartate transaminase (SGOT), and alanine transaminase (ALT) are pointers to obstructive cholestasis from hepatic infiltration; however, only γGT and ALP were found to be elevated in the index child, and this may explain the hepatomegaly.22,23 Additionally, imaging may suggest a mass present at specific sites; however, histological diagnosis is the mainstay in making a diagnosis of IMT. For the index case, a computed tomography (CT) scan suggested a lymphoma. Still, the histologic analysis aided the final diagnosis, thereby preventing a misdiagnosis and wrong management, as in the case of this child, who was previously managed for disseminated tuberculosis, until we had a histology report. 24

Various treatment modalities have been described in the management of IMT, including surgical excision, curettage, steroid therapy, radical surgery, and radiotherapy.5 The mainstay of treatment is surgical resection, with a very high recurrence rate (58%). However, complete resection significantly reduces this probability to around 2%. Recently, surgical excision has been supplemented with CO2 laser light.[5](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5670301/#bib0020) Radiation, chemotherapy (cyclosporine, azathioprine, methotrexate, and cyclophosphamide), ALK molecular targeted therapeutic drugs [crizotinib], and steroid therapy are used when the tumors are invasive, non-resectable, recurrent, show signs of malignancy/metastasis, or when surgical margins are positive.[5](https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5670301/#bib0020) In addition, NSAIDs are being suggested as a treatment option because of their anti-inflammatory and antiangiogenic effects.5 The use of NSAIDs resulted in tumor regression in older children, whereas others reported no response to NSAIDs.At the time of initial diagnosis, the index child was given steroids, and this could have contributed to some clinical improvement. However, with the new diagnosis of IMT, the child was scheduled for surgery and chemotherapy, but passed away before receiving any treatment. Further testing, such as immunohistochemistry and molecular genetic testing, could not be explored due to financial constraints.

CONCLUSION

IMT continues to be a diagnostic rarity and often surprises clinicians histologically, although the diagnostic features frequently become more apparent in hindsight. A multidisciplinary approach is crucial for managing IMT. Since recurrence is possible, prolonged follow-up and close monitoring can facilitate early detection. This report enriches the global data pool on IMT presentations, especially in African populations, where literature on such cases remains scarce.

DISCLAIMER

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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