A pediatric tuberculosis with recurrent fever confirmed to be inflammatory myofibroblastic tumor

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Case Report

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Abstract

Background: Symptoms of inflammatory myofibroblastic tumor (IMT) are atypical, and histopathological misdiagnosis of IMT is still inevitable. Here we present a pediatric case that an eight-year-old boy with recurrent fever for fifteen months, received anti-tuberculosis therapy for six months and was ultimately confirmed to be abdominal IMT.

Case presentation: An eight-year-old boy had a recurrent fever for 15 months, accompanied by cough, vomiting, meteorism, night sweating, and emaciation. The histopathological characteristic of intestinal and greater omentum implied fibrous tissue hyperplasia, with eosinophil and lymphocyte infiltration. The patient was diagnosed with tuberculosis, and symptoms were relieved partially after anti-tuberculosis treatment. Four months later, the symptoms aggravated again and histopathology of the second sample of greater omentum revealed IMT. Eventually, the patient recovered well after receiving regular chemotherapy.

Conclusions: The clinical course of IMT is variable, and pediatricians should pay attention to distinguishing IMT from tuberculosis.

Background

The inflammatory myofibroblastic tumor (IMT) is a rare lesion of unclear etiology and variable clinical course, consisting of a proliferation of fibroblasts and myofibroblasts mixed with inflammatory cells. It is an ultra-rare tumor classified as a neoplastic disease of intermediate biological potential, given the low risk of recurrence and metastatic potential\(^1\). Symptoms of IMT are atypical and pathological misdiagnosis and missed diagnoses of this disease are still common. Here we present a pediatric case of tuberculosis that was verified to be IMT nearly two years later.

Case Presentation

An eight-year-old male patient was admitted to the hospital for recurrent fever for fifteen months. His personal history, past history, and family history were not special. He had no history of tuberculosis exposure.

The child had a fever of 40 °C, accompanied by cough, chest pain, abdominal distension, vomiting, night sweat, and emaciation fifteen months ago. The patient was admitted to the local hospital nearly once or twice a month for recurrent fever, and symptoms were alleviated temporarily after antibiotic treatments. The laboratory test revealed a rise of white blood cells (WBC, as high as 18.05 × 10 ^ 9/L), eosinophils (as high as 14.5%), C-reactive protein (CRP, up to 187 mg/L), and erythrocyte sedimentation rate (ESR, up to 115 mm/h). Thoracoabdominal enhanced computer tomography (CT) revealed multiple enlargements of lymph nodes in the thorax, abdomen, and armpit and a bit of bilateral pleural effusion. Histopathologic features of inguinal lymph nodes displayed reactive hyperplasia, and bone marrow aspiration demonstrated increased eosinophils and a few cells of unknown origin. The greater omentum was
discovered to be covered by massive miliary nodules by laparoscopic examination, and the histopathologic characteristics of the mesentery and greater omentum indicated massive proliferation of fibrous tissue, combined with many eosinophils and lymphocytes infiltration. The patient was diagnosed with intraperitoneal tuberculosis, and the symptoms did not improve after treatment with isoniazid and rifampicin for five months.

Physical examinations were recorded. The patient was anemia and emaciation. The scar of the bacillus Calmette Guerin vaccine existed on the right upper arm. Enlarged lymph nodes of the bilateral side of the neck, armpit, and groin were palpable, with the maximum one at the size of 1.0 centimeter (cm) × 1.0 cm. The abdomen was slightly distended, and a mass of about 5 cm × 5 cm can be touched at the umbilicus. The other examinations were normal.

The laboratory tests revealed the elevation of leukocyte (maximum: 24.5 × 10^9/L), eosinophils (maximum: 31%), platelet (maximum: 856 × 10^9/L), C-reactive protein (maximum 106 mg/L), and decrease of hemoglobin (minimum: 69 g/L). Tests for tuberculosis yielded normal results, and radionuclide images of the bones were normal. Thoracoabdominal enhanced CT revealed bilateral pulmonary inflammation, pleural effusion, and pleural thickening. Many enlarged lymph nodes were found in the mediastinum, bilateral hilar, axilla, and abdominal cavity, with partial necrosis and obvious enhancement. The intestinal wall of the abdominal cavity was thickened and swollen, and there was a little fluid in the abdominal cavity and pelvis (Fig. 1).

The patient was diagnosed with a fever with enlarged lymph nodes of unknown origin. Disseminated tuberculosis and lymphoma were considered.

The children received isoniazid (H), rifampicin (R), pyrazinamide (Z), ethambutol (E) (HRZE) for anti-tuberculosis treatment, and meropenem for antibacterial. One week later, the child's temperature gradually returned to normal, and abdominal distension and appetite ameliorated. During the following four months of receiving HRZE therapy, the patient remained afebrile for one month, with weight increase, improvement of abdominal distention and vomiting, and relief of lesions on the thoracoabdominal CT. When the regiment was adapted to HR, symptoms including fever, abdominal distension, and appetite loss aggravated. The superficial lymph nodes of the whole body were enlarged, and several masses were formed in the abdomen, with a maximum of about 3 cm × 4 cm.

Exploratory laparotomy revealed many miliary nodules attached to the abdominal wall, greater omentum, and intestinal wall. Histologic features of abdominal mass, stained by hematoxylin and eosin, represented that spindle-shaped cells proliferated in patches or nodules with variable fibrosis and hyaline degeneration, and mitosis was difficult to find. There was obvious infiltration of inflammatory cells, mainly eosinophils, as well as lymphocytes, plasma cells, and histiocytes in the background. Immunohistochemical staining of abdominal mass displayed that the spindle cells were negative for EMA, Des, s-100 protein, CD34, aK, CD117, DOG-1, and EBER1/2-ish. The Ki-67/mib-1 proliferation index was 20% and S were partially positive. The FISH test did not detect the ALK gene and ROS1 gene translocation. After six times of fierce discussions directed by pathologists and hematologists, the patient
was eventually diagnosed as an inflammatory myofibroblastic tumor. The patient was ultimately transferred to the department of hematology and oncology for chemotherapy and recovered well after regular chemotherapy.

**Discussion And Conclusions**

An inflammatory myofibroblastic tumor (IMT) is a mesenchymal neoplasm of intermediate biological potential, which may recur and rarely metastasize. IMT is usually composed of myofibroblastic and fibroblastic spindle cells accompanied by inflammatory cells consisting of lymphocytes and eosinophils. IMT usually arises in children and young adults, and the etiology and pathogenesis are not fully understood [2, 3]. IMTs can occur in any location, and the primary site is the abdominal cavity, especially in the mesentery, retroperitoneal, and omentum. Patients with MIT can present atypical symptoms, including fever, emaciation, fatigue, abdominal pain, anemia, thrombocytosis, polyclonal hyperglobulinemia, and an increase in erythrocyte sedimentation [3]. The children in the case have atypical symptoms including fever, emaciation, fatigue, abdominal pain, thoracoabdominal enhanced CT and initial pathological lesions of the intestinal canal, and greater omentum, implying tuberculosis. However, the anti-tuberculosis treatment fails to cure.

The MIT diagnosis is based on pathological alternations, which may be challenging. IMT often presents as a circumscribed nodular mass, but multinodular lesions have also been described [4]. IMT has diverse morphological changes, including a paracellular spindle cell proliferation set in a predominantly hyalinized and chronically inflamed background, and a highly cellular myofibroblastic proliferation, as well as atypical neoplastic elements. Most tumors are composed of spindle cells, accompanied by fibrosis, hyaline degeneration, calcification, or necrosis. The inflammatory component may be variable, usually comprised of plasma cells, lymphocytes, eosinophils, and neutrophils [4, 5]. There was a report that gene alterations, such as fusion of the gene encoding anaplastic lymphoma kinase (ALK) on chromosome 2p23 and tropomyosin 3 (TPM3), are detected in 30% of pediatric IMT cases [6].

Surgery is the standard of treatment for local disease, with a high chance of cure in completely resected cases. As for advanced disease, the therapy is not precisely defined. Chemotherapy regimens for MIT are disputed. Some researchers in support of chemotherapy suggest that IMT, especially from mesentery and peritoneum, may be potentially malignant by means of local infiltration or distant metastasis [7].

In conclusions the clinical characteristics of IMT are not typical, and the pathological diagnosis is also challenging. IMT may easily be confused with tuberculosis. Pediatricians should pay attention to distinguishing IMT from tuberculosis.

**Abbreviations**

IMT : inflammatory fibroblastic tumor; WBC: white blood cells;

CRP  C-reactive protein; ESR: erythrocyte sedimentation rate ;
CT: computer tomography; CM: centimeter

H: isoniazid; R: rifampicin; Z: pyrazinamide; E: ethambutol.

**Declarations**

**Ethics approval and consent to participate**

Not applicable.

**Consent for publication**

Written informed consent was obtained from the patient’s parents for publication of their child’s personal or clinical details, along with any identifying images in this study. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

**Availability of data and materials**

All data are contained within the article.

**Competing interests**

The authors declare that they have no competing interests.

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**Authors’ contributions**

YL conducted the data collection, performed literature review, drafted and edited the manuscript. YW revised the manuscript and performed literature review. All authors read and approved the final manuscript.

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**References**


**Figures**

![Figure 1](image)

**Figure 1**

Thoracoabdominal enhanced CT. (A) bilateral pulmonary inflammation, pleural effusion, and pleural thickening, enlarged lymph nodes in the mediastinum. (B) thickened and swollen intestinal wall of the abdominal cavity.