Abdominal inflammatory myofibroblastic tumor: Review of the literature by means of a case report

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Inflammatory myofibroblastic tumor occurring at intraabdominal sites in children has rarely been described. In this paper, an 18-month old girl with intraabdominal inflammatory myofibroblastic tumor who presented with fever, anemia, constipation, weight loss and an abdominal mass is reported. A comprehensive review of the literature is also documented. [Turk J Cancer 2002;32(1):28-31]

Key words: Inflammatory myofibroblastic tumor, abdominal mass, children

Inflammatory myofibroblastic tumors (IMT) are rare proliferative lesions clinically resembling a malignant neoplasm. Their classification is controversial and confusing. IMTs are well described in the lung and upper respiratory tract of young adults and children; but may occur at any age and affect any organ system (1,2,3).

Typically, it is a circumscribed but nonencapsulated lesion containing spindle cells proliferating in a background of fibrosis, with lymphocytes, plasmacytes, histiocytes, foamy macrophages, and occasionally eosinophils and neutrophils nuclear pleomorphism and atypical mitoses are absent (4,5).

At one time thought to be reactive in nature, the inflammatory myofibroblastic tumor has come to be considered a true neoplasm with the potential for recurrence and multifocality; clonal cytogenetic findings support this view. Complete excision is necessary to avoid local recurrence (6,7).

Case Report

An 18-month old girl with a 35-day history of intermittent fever, constipation and profound weight loss was referred for investigation of a large form, mobile, left-sided abdominal mass. Investigations revealed microcytic hypochromic anemia (haemoglobin level 5.3 g/dl) and thrombocytosis (platelet count 1106x10⁹/L). Ultrasonography and abdominal CT demonstrated an 8x10 cm mass adjacent to the left kidney. After a short period of investigations, a
laparotomy was performed. A large lobulated mass was found arising from the small bowel mesentery. A laparotom ic biopsy was reported as an IMT. Microscopically, the tumor was composed of an admixture of proliferating spindle cells and chronic inflammatory cells embedded in variably vascular stroma that ranged from being loosely myxoid to densely collagenous (Figure 1). The spindle cells stained with vimentin but were uniformly negative with desmin, leukocyte common antigen (LCA) and neuron specific enolase (NSE). There was no evidence of pleomorphism or hyperchromasia and no abnormal mitotic figures. The inflammatory cells were a polymorphous mixture of plasma cells and T and B-lymphocytes with scattered eosinophils and mast cells (Figure 2). Immediately after pathological diagnosis; the tumor was excised without any complication. She recovered with resolution of her symptoms by two weeks after her surgery. It is currently 22 months after surgery without evidence of recurrence.

![Image](image.jpg)

Fig 1. IMT, arranged of spindled myofibroblast in myxoid, oedematous matrix (H&E x40)

**Discussion**

IMT is the most frequent benign primary lung tumor in childhood. Due to confusion about the histogenesis of the lesion, various synonyms have been used (such as inflammatory, pseudotumor, plasma cell granuloma e.g.) In the last few years, IMT with borderline biological course, has been described in which myofibroblasts represent not only a reactive mechanism but also a true neoplastic component. It has been regarded as a benign and reactive disorder for a long time. Only in recent reports it has been demonstrated that, in spite of an apparently benign morphological pattern, some cases of IMTs have a malignant course (3,4,7).
The myofibroblast was eventually recognised as the principal spindle-cell type in this tumor, which led to the new term IMT. Myofibroblasts are spindle cells having ultra-structural features in common with smooth muscle cells and fibroblasts. The important function of the myofibroblast in tissue repair is consistent with the hypothesis that an aberrant response to tissue injury is the pathogenesis of IMT, however in most cases there is no identifiable precipitating factor (2,6).

Evidence to support a directly infectious etiology is scanty but an immunological pathogenesis remains possible. The role of cytokines, particularly interleukin-6 (IL-6) in pathogenesis and the possibility for a specific therapeutic approach has been described. Many authors postulate a post-inflammatory process; although the presence of clonal chromosomal abnormalities suggests a neoplastic process (1,5,7).

Until 1992, a total of 21 children, predominantly female, with IMTs affecting intra-abdominal sites have been reported in the literature. These tumors are often large. Multi-centric lesions are rare. There are no reports of malignancy arising in an IMT; nevertheless, the clinical, radiological and histological features of these may cause confusion with malignant lesions. Most patients presented with fever, anemia, trombocytosis, hyperglobulinemia and weight loss. These systemic features resolve after tumor excision but may enable the diagnosis to be suspected before operation. Misdiagnosis has led some patients to be inappropriately treated with chemotherapy and radical surgery (2,4).
In IMTs, high cellularity with large, plump, active myofibroblasts with prominent nucleoli can cause confusion with malignancy, in particular rhabdomyosarcoma. However the lack of atypia, hyperchromasia and abnormal mitotic figures are pointers toward a benign lesion. IMT should be diagnosed by routine staining because special stains and immunocytochemistry can be misleading. Preliminary biopsy and full histological evaluation is recommended in cases where resection may be particularly hazardous (5,6,7).

Other reasons for confusion of IMTs with malignant neoplasms include their capacity for local tissue infiltration, occasional rapid growth and the development of local recurrence. Mediastinal and esophageal involvement from adjacent pulmonary pathology appears to be particularly aggressive. Their course is complicated, which ranges from spontaneous regression through gradual enlargement to rapid growth with local invasion. Tumor resolution or regression has been reported after radiotherapy, chemotherapy and steroid therapy. Local recurrences after incomplete excision are recognised, may occur many years later and may be fatal. This underlines the importance of complete surgical resection whenever possible (1,3).

References