INFLAMMATORY MYOFIBROBLASTIC TUMOR WITHIN ILEAL INTUSSUSCEPTION AS THE CAUSE OF RECURRENT ABDOMINAL PAIN IN A 57-YEAR OLD PATIENT

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The study presented a rare case of inflammatory myofibroblastic tumor (IMT) in a 57-year old male patient who presented with recurrent abdominal pain. He was diagnosed to have a tumor of the small intestine within ileal intussusception, demonstrated by CT enterography. The patient underwent surgery to remove the tumor with a margin of healthy tissue. Histopathological and immunohistochemical examination results enabled to diagnose IMT. IMT is a rare tumor that occurs mainly in children and young adults. Its etiology remains to be fully understood. Due to the differentiated histology it can be found in many organs and soft tissues, being responsible for different, non-specific clinical and radiological symptoms. Due to the rarity of this tumor a clear treatment protocol has not yet been established. However, given the tendency to recur with possible distant metastases an important element of treatment consists in long-term clinical patient observation. 

Key words: inflammatory myofibroblastic tumor, intussusception, small intestine.

Inflammatory myofibroblastic tumor (IMT) is a rare soft tissue tumor of unknown etiology, varied anatomical location, and diverse clinical course. Most reported cases were observed in the lungs and abdominal cavity organs. Incidence mainly referred to children and young adults with a predominance of female patients. The study presented an atypical case of a 57-year old male patient diagnosed with inflammatory myofibroblastic tumor within the intussuscepted ileum, being responsible for recurrent episodes of gastrointestinal obstruction.

CASE REPORT

A 57-year old male patient was admitted to the Department of General, Gastroenterological, Colorectal, and Oncological Surgery in September, 2012, with clinical symptoms of gastrointestinal obstruction. On admission, the patient complained of nausea, and vomiting after every meal, stool retention, and increasing hypogastric pain. Additionally, the patient complained of general weakness and loss of body weight (20 kg during a period of 5 months). Urgent abdominal pain occurred periodically (once a week), since December, 2011, and resolved spontaneously or after relaxing drugs. Abdominal pain was accompanied by nausea and vomiting. At that time the patient was repeatedly diagnosed in the ER and outpatient clinic. The physical examination, laboratory, endoscopic, and ultrasound results showed no pathology. After conservative treatment clinical symptoms regressed.
The only disturbing symptom was gradual weight loss and iron deficiency anemia.

In order to thoroughly evaluate the small bowel the patient underwent CT enterography at the Department of Radiology and Diagnostic Imaging. The examination showed ileal intussusception, approximately 13 cm long. Within the intussusception the examination revealed the presence of a nodular mass, 54x47x63 mm in size, subject to contrast enhancement (fig. 1, 2).

On the basis of the clinical picture and imaging examinations, following a surgical consultation, the patient was qualified for surgery with tumor suspicion of the ileum. The intraoperative examination revealed the presence of a tumor of the ileum with intussusception into the distal colon. Resection of 40 cm of the ileum and the tumor was performed with a 50 mm healthy tissue margin, followed by endo-to-side anastomosis. The perioperative and postoperative period proved uneventful, the patient remains under ambulatory observation.

The microscopic examination revealed the presence of histologically homogenous fibroblast and myofibroblast cells arranged in bundles associated with chronic inflammatory infiltration composed of plasmocytes and eosinophils. The immunohistochemical examination showed a positive reaction to the presence of vimentin, smooth muscle actin, polyclonal desmin, and keratin (fig. 3).

**DISCUSSION**

In the presented study case IMT was diagnosed in a 57-year old male patient within the small intestine. IMT is most often diagnosed in children and young adults with a female predominance (1, 2, 3). Due to the diverse histology of the tumor it may be found in many organs and soft tissues. Depending on its location IMT may be responsible for different, non-specific clinical symptoms and radiological signs. IMT located in the abdominal cavity

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*Fig. 1. Frontal section of the small intestine tumor*

*Fig. 2. Cross section of the tumor and intussusception of the small intestine*

*Fig. 3. A micrograph of the inflammatory miofibroblastic tumor cells*
Inflammatory myofibroblastic tumor within ileal intussusception often achieves quite a considerable size and can be felt as a pathological mass upon palpation. Other possible clinical symptoms include atypical abdominal pain, weight loss, fever, and in selected cases gastrointestinal obstruction (4). In such cases the plain abdominal X-ray study demonstrated an atypical intestinal loop distribution, eventually calcifications within the tumor. The abdominal ultrasound and CT examinations might show an ambiguous solid mass, usually well-demarcated. Considering our study case the patient complained of malaise, loss of appetite, and significant weight loss during a short period of time. Abdominal CT showed ileal intussusception over several centimeters with a tumor 5.5 cm in size within the intussusception.

Laboratory results also showed abnormalities: microcytic anemia and elevated ESR. These abnormalities were rarely observed by other Authors (1, 5). Due to the diverse structure and unpredictable clinical course final diagnosis of inflammatory myofibroblastic tumor (IMT) is possible following histological and immunohistochemical examinations. Histologically the tumor is composed of spindle cells derived from myofibroblasts, inflammatory infiltration cells (plasmocytes, lymphocytes, eosinophils), and histiocytes. Considering immunohistochemistry the characteristic, but not pathognomonic feature of IMT is the positive reaction to the presence of vimentin, smooth muscle actine (SMA), CD34 antigen, polyclonal desmin and keratin (7). We also showed the presence of IMT short arm chromosome 2 aberrations in case of large-cell anaplastic lymphomas. Region 23 of the short arm of chromosome 2 is the locus of the gene coding anaplastic lymphoma kinase (ALK). Due to chromosomal disturbances in as many as 8-63% of IMT cases one may observe ALK protein expression, which can be detected by means of anti-ALK antibodies: ALK1 and p80 (8, 9). Unfortunately, this is not characteristic of IMT. Similar abnormalities were observed in case of selected sarcomas, malignant neurilemmoma, or sympathoblastoma (7).

The question of the presence of ALK protein overexpression in the inflammatory myofibroblastic tumor has prognostic significance. Chan et al. demonstrated that ALK positive tumors gave no recurrence or distant metastases, thus, were of favorable prognosis (10). Mergan et al. came to similar conclusions (11).

The presented study case was diagnostically difficult. For six months recurrent clinical symptoms of mild intensity, rapidly regressing after conservative treatment, normal physical and additional examination results delayed clinical diagnosis. Only gradual weight loss and iron deficiency anemia were arguments for the continuous search of a malignancy.

One should emphasize the role of CT enterography, which gave rise to the correct diagnosis and surgical management. Unfortunately, depending on the location of the tumor radical surgery is not always possible. Thus, when considering IMT treatment one should think of other methods, such as chemotherapy, radiotherapy, and sytemic steroid therapy (12). Bearing in mind the tendency of IMT to recur and give distant metastases, an extremely important element of treatment consists in the long-lasting clinical observation of patients. Due to the rare occurrence of the inflammatory myofibroblastic tumor the therapeutic protocol has yet to be established. It is believed that complete excision of the tumor is the basic and best therapeutic method. In the presented study case, radical resection of the ileum was performed together with the tumor and healthy tissue margin, followed by end-to-side anastomosis. The patient remains under ambulatory control without disease symptoms.

REFERENCES


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