Inflammatory Myofibroblastic Tumor with Extensive Involvement of the Bowel in a 7-Year-Old Child

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We present a case of unusual localization of inflammatory fibroblastic tumor in the terminal ileum, cecum, and ascending colon in a 7-year-old child. Segmental resection of the terminal ileum, cecum, and ascending colon with a tumor mass up to 6 cm in diameter was performed. Pathohistological examination of biopsy specimen was performed on routine hematoxylin-eosin sections, as well as immunohistochemically with primary antibodies to CD3, CD20, CD68, factor VIII, vimentin, smooth muscle actin, desmin, cytokeratin and S-100 protein, and κ and λ light chains. The tumor was composed of highly vascularized tissue with interlacing fascicles of elongated spindle cells admixed with plasma cells, histiocytes, lymphocytes, and eosinophils. The diagnosis of inflammatory myofibroblastic tumor was confirmed by immunohistochemistry. Inflammatory myofibroblastic tumor cannot be distinguished clinically from highly malignant neoplasm or some other conditions. Surgical resection and careful pathohistological analysis are needed, and a long-term follow-up is recommended.

Key words: abdominal neoplasms; blood vessel tumors; child; fibroblast; immunohistochemistry; neoplasms, fibroepithelial

Inflammatory myofibroblastic tumor (IMT) of the ileum is a rare lesion that arises in the submucosa (1-3). It is usually solitary and polypoid, and often presents clinically with symptoms and signs of bowel obstruction (1-3). However, multifocal, transmural IMT affecting a regional noncontiguous lymph node was also described (3). IMT is characterized by rapid growth, local invasiveness, and recurrence (4,5) and therefore could be erroneously regarded as a highly aggressive process radiologically, at time of surgical intervention, and even by well-experienced pathologists.

Several terms have been used to describe the lesion: inflammatory pseudotumor (3,6,7), inflammatory fibroid polyp (1,2,4,8), pseudosarcomatous fibromyxoid tumor (9,10), and inflammatory fibrosarcoma (11-15). Similar tumors have been described in numerous locations including trachea (16), lung (17,18), bladder (9,10), spleen (19), skin (20), ileum (1-4), and lymph nodes (6,7,21).

IMT is increasingly recognized as part of a spectrum of inflammatory myofibroblastic proliferations (12). It is a potentially locally aggressive tumor that occurs predominantly in the mesentery of children and young adults. No reliable morphological features have been identified, which could predict prognosis (12).

We report a case of inflammatory myofibroblastic tumor in a 7-year-old girl with unusual, extensive involvement of the ileum, cecum, and ascending colon. The tumor had no polypoid component but involved noncontiguous regional lymph nodes.

Case Report

A 7-year-old girl was admitted to the emergency room with abdominal pain. There was history of a variable degree of abdominal cramping and postprandial pain for the last two months, without nausea, vomiting, or bowel disturbance.

Physical examination revealed a temperature of 36.9°C, blood pressure 110/70 mmHg, pulse rate 85/min and respiratory rate 20/min. The patient had abdominal distension and increased bowel sounds. Skin turgor was adequate, and there was no evidence of dehydration.

Laboratory tests detected hemoglobin concentration of 14.3 mmol/L and hematocrit 26%. The total and differential counts of leukocytes were normal. The stool was negative for occult blood. Microbiological cultures performed on appropriate specimens of blood and urine were negative.

Ultrasound of the abdomen showed a large mass in the low right quadrant measuring up to 6 cm in diameter (Fig. 1). The lesion was suggestive of a malignant neoplasm.
Surgical exploration revealed an ileocecal obstruc-
tion due to a malignant lesion of the ileum, cecum,
and ascending colon. Enlargement of two non con-
tiguous regional lymph nodes was also observed. Segmental
resection of the terminal ileum, cecum, and right
hemicolon was performed with end-to-end anastomosis.
The postoperative course was uneventful, and the patient
was discharged after 8 days. The patient did not receive
any further therapy; she was well and without recurrence
two years after surgery.

Pathological Findings

The gross specimen measured up to 15 cm in length
and included the terminal ileum, cecum, and ascending
colon. An intramural circumferential mass filled the ce-
cum and completely reduced the lumen of the terminal il-
ium and ascending colon to 0.5 and 1 cm, respectively.
The attached ileal mesentery was edematous and con-
tained two white lymph nodes up to 1 cm in diameter.
The appendix was attached to the mass by the
mesoappendix. The cut surface of the tumor was tan,
firm, trabeculated, and knobby, without necrosis, hemor-
hage, or cyst formations.

Microscopic examination revealed highly
vascularized fibrillar tissue with interfacing fascicles of
elongated spindle cells and loosely, haphazardly arranged,
plump, fusiform or stellate myxoid cells intermingled
with plasma cells, histiocytes, lymphocytes, and eosinophils. The nuclei of spindle cells were oval and ve-
sicular with occasionally prominent nucleoli. Mitoses
were rare. Some spindle cells showed well-defined cyto-
plasmic borders with elongated, tapering, bipolar pro-
cesses. Others contained moderately abundant
eosinophilic cytoplasm with rounded borders (Fig. 2).
Lymph nodes showed minimal changes in architecture,
with spindle cell fibroblastic/myofibroblastic prolifera-
tion in the subcapsular and trabecular sinuses (not shown).
The inflammatory infiltrate was predominantly composed
of histiocytes, small lymphocytes, and plasma cells. The
appendix was unaffected. Microorganisms, including
acid-fast bacilli and fungi, were not observed in histologic
sections stained with hematoxylin-eosin, PAS, and Ziehl-
Nielsen methods.

Discussion

Inflammatory myofibroblastic tumor of the ileum is
an uncommon process that often cannot be differentiated
clinically or radiologically from other more aggressive
neoplasms of the small or large intestine. It was initially
regarded as an aberrant inflammatory response despite its
gross and histological features of a spindle cell tumor
(13,14). In our patient there was no evidence of systemic
infection, and microbiologic and serologic examinations
were negative for viruses, fungi, bacteria, and mycobac-
teria. In the largest reported series of IMT from all sites,
61 of 84 lesions were in the abdomen, retroperitoneum,
or pelvis; 12 cases in the head and neck region, including
upper respiratory tract; 8 cases in trunk; and 3 cases on
the extremities (11). The patients with abdominal tumors
present with abdominal mass, fever, weight loss, abdom-
inal pain, vomiting, constipation, and intussusception.
The lesions ranged in size from 1 to 17 cm and the age of
the patients from 3 months to 46 years. Overall, the le-
sion was more common in women than men. One or more
recurrences were noted in thirteen patients, but distant
metastases were not documented. Myint et al (3) re-
ported a case of IMT with unusual, multifocal involve-
ment of the ileum and reviewed the literature on the sub-
ject. The majority of published IMT cases in the ileum
were polypoid, measuring from 2 to 17.5 cm. The pa-
tients’ age ranged from 20 months to 86 years. We ob-
served several unusual features in our patient. Firstly,
the lesion was not polypoid. Secondly, the lesion was pre-
dominantly intramural. Also, the lesion occluded the ter-
minal ileum, cecum, and part of ascending colon; regional lymph nodes were involved.

Currently, IMT is regarded as a benign, non-metastasizing proliferation of myofibroblasts with a potential for recurrence and persistent local growth (12-14). All tumors are infiltrative with focal myxoid change. Those occurring in gastrointestinal tract frequently show transmural and mesenteric involvement (15). Histologically, they may resemble three pathohistological changes: 1) nodular fascitis; 2) fibrous histiocytoma; and 3) desmoid or scar (11). However, no reliable morphological features have been identified that predict prognosis (12). Also, prognostic markers such as Ki-67, PCNA, bcl-2, and p53 did not correlate with local recurrences, metastases, or tumor deaths (12).

Because IMT of the intestine is invasive and composed of spindle cells, it should also be differentiated from mesenchymal neoplasms including rhabdomyosarcoma, leiomyosarcoma, and malignant fibrous histiocytoma (11-14, 22). Unlike IMT, leiomyosarcoma, malignant fibrous histiocytoma, and rhabdomyosarcoma consist of atypical mesenchymal cells with atypical mitoses. Immunohistochemistry may also be of substantial help (3,11). In our case, we observed no involvement of the mesentery; there were few mitoses and no cytological atypia of the cells.

The differential diagnosis of IMT of the ileum also includes malakoplakia, eosinophilic gastro- enteritis, Crohn's disease, and idiopathic sclero- sing mesenteritis (3,13,14). Features of these diseases could not be found in our patient.

Inflammatory fibrosarcoma and inflammatory myofibroblastic tumor are the proposed terms for such tumors because of inadequacy and inaccuracy of the conventional names such as plasma cell granuloma, inflammatory pseudotumor, or inflammatory fibroid polyp. Pseudotumor is a negative expression and carries little scientific meaning. If tumor denotes a mass according to its original definition, this lesion is indeed a true tumor, because it may reach an alarmingly ponderous size. One of the most popular terms was inflammatory fibrous polypl, suggested by Shimer and Helwig (2), but all lesions are not polyoid, especially if they are located at other sites, such as the spleen (19) or lymph nodes (7). The term inflammatory myofibroblastic tumor is descriptive, unassuming and noncommittal, and may be recommended for this lesion.

In conclusion, IMT of the ileum, cecum, and ascending colon cannot be distinguished clinically from highly malignant neoplasm or some other processes. The inability to make this clinical distinction requires surgical resection. It is obvious that fibroblastic and myofibroblastic proliferation with inflammatory infiltration should be regarded as potentially malignant lesion. A long-term follow-up is certainly needed.

Acknowledgments

This work was supported in part by a grant No. 009193 from the Ministry of Science and Technology of the Republic of Croatia.

References

10 Young RH. Pseudoneoplastic lesions of the urinary bladder and urethra: a selective review with emphasis on recent information. Sem Diag Pathol 1997;14:133-46.
19 Thomas RM, Jaffe ES, Zarate-Orsorno A, Medeiros LJ. Inflammatory pseudotumor of the spleen: a clinic- patho-


Received: July 8, 1999
Accepted: September 27, 1999

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