Inflammatory myofibroblastic tumor of the small bowel

N. Abbes Orabi[1,3], H.M. Paterson[1], E. Danse[2], C. Remue[1], D. Léonard[1], A. Kartheuser[1]

(1) Colorectal Surgery Unit, Cliniques universitaires St-Luc ; (2) Department of Radiology, Cliniques universitaires St-Luc, Brussels, Belgium ; (3) Department of Abdominal Surgery, C.H.R Mons, Clinique St-Joseph, Mons, Belgium.

To the Editor,

First described in the lung, inflammatory myofibroblastic tumors (IMT) are pseudosarcomatous lesions that occur in the viscera and soft tissue of children and young adults. Intraabdominal and retroperitoneal locations have been associated with higher degrees of cellular atypia, more aggressive clinical course, higher local recurrence and even distant metastases (1). A 23-year-old woman presented with abdominal pain, fatigue, night sweats and weight loss for six weeks. On physical examination, a large mass was palpable in the lower abdomen. MRI confirmed the presence of a 7 × 8 × 9 cm solid lesion involving the small bowel (Figs. 1 and 2). A Pfannenstiel laparotomy was performed and a tumor was found arising from the mesenteric border of the terminal ileum. The tumor was completely resected with primary ileo-caecal anastomosis. Histopathology revealed an inflammatory myofibroblastic tumor (IMT) with proliferation of spindle-shaped cells in fascicular growth patterns and lymphocytic inflammatory infiltrate. Immunohistochemistry was highly positive in tumor cells for actin alpha (Figs. 3a and 3b). Postoperative recovery was uncomplicated and the patient left hospital five days after surgery.

The most common sites of extrapulmonary IMT lesions are mesentery and omentum. Small bowel location is particularly rare (1). Females are affected slightly more commonly than males. Besides abdominal symptoms, some patients present systemic manifestations, including fever, night sweats, weight loss, and malaise. Blood analysis can show an inflammatory syndrome, anemia, thrombocytosis and hypergammaglobulinemia which often resolve with excision of the lesion. Coffin *et al*. reported a study of 53 patients with extrapulmonary lesions. They observed a 25% recurrence rate from 1 to 24 months after initial excision (2). Aggressive tumor behaviour was related to close proximity with vital structures and multinodular presentation, which compromised complete surgical excision. The etiology of IMT remains unclear. Infective organisms have been suggested in the tumorigenesis, including *Human herpes virus 8, Mycobacterium avium-intracellulare, Corynebacterium equi, Campylobacter jejuni, Bacillus sphaericus, Coxiella burnetti, Escherichia coli and Epstein-Barr virus*, but none proven. IMT is also thought to arise as an aberrant inflammatory response to tissue injury. However, clonal abnormalities of ALK (anaplastic lymphoma kinase) have been associated with tumorigenesis because of the high frequency of ALK staining in IMT. ALK positivity is associated with a better prognosis in anaplastic large cell lymphomas but its prognostic significance in IMT is unclear (3,4,5). However, ALK staining was not contributive for our patient.

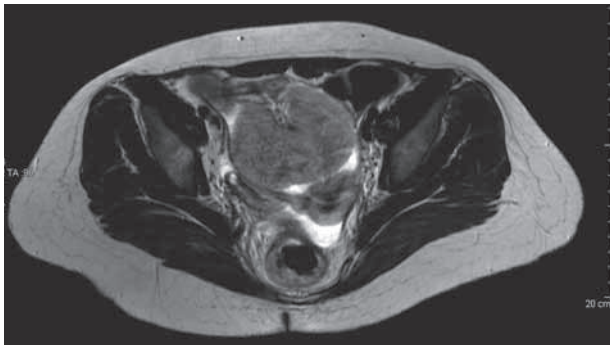

Fig. 1. — MRI showing an heterogenous 8 × 7 × 9 cm anterior pelvic mass.

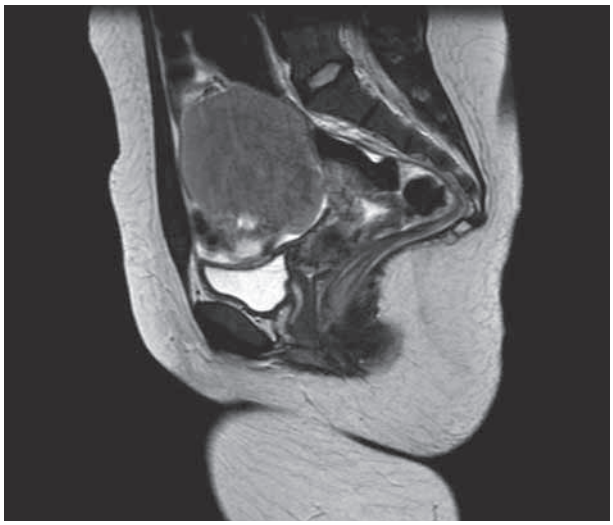

Fig. 2. — MRI showing the lesion above the bladder

Correspondence to : Pr. Alex Kartheuser, University Hospital St Luc, Colorectal Surgery Unit, 10 Avenue Hippocrate, B-1200, Brussels, Belgium.
E-mail : alex.kartheuser@uclouvain.be

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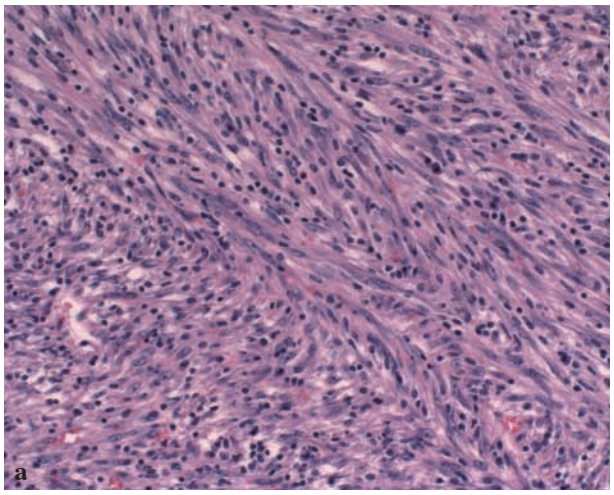

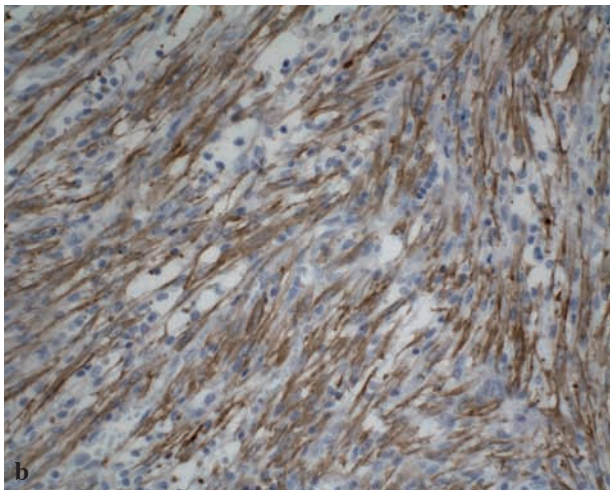

Fig. 3a-b. — Proliferation of spindle cells and lymphocytic inflammatory infiltrate with positive Actin staining (courtesy of Ch. Sempoux and A. Jouret-Mourin)..

IMT must be considered in the differential diagnosis of gastrointestinal masses. IMT is now classified as a neoplasm due to its potential for local recurrence, infiltrative growth, vascular invasion and malignant transformation. Long-term follow-up to detect recurrence is recommended (1,3). The treatment is surgical resection with reexcision of recurrent tumors, the benefit of chemotherapy and radiotherapy remains unproven (3,6,7).

Conflicts of interest

The authors have no conflict of interest.

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