Inflammatory pseudotumour of the colon

I Spyrou, S Davakis, D Moris, E Lakiotaki, N Dimitrokallis, A Papalampros, D Schizas, E Pikoulis, E Felekouras

National and Kapodistrian University of Athens, Greece

ABSTRACT

INTRODUCTION A colorectal inflammatory myofibroblastic tumour (IMT) is a rare but benign entity masquerading as a malignant tumour. Although the lung is considered the most common site of occurrence, IMTs may arise in diverse extrapulmonary locations. We describe a case of a colonic IMT in a patient who presented in the emergency setting.

CASE HISTORY A 77-year-old man was admitted at our emergency department with acute abdominal pain. Physical examination revealed vague tenderness of the lower abdomen and non-palpable masses. Preoperative evaluation revealed a mass in the right lower quadrant of the abdomen, possibly originating from the terminal ileum, 1–2cm from the caecum. Owing to the clinical impression of a potentially malignant lesion, the patient underwent subtotal colectomy and omentectomy. The pathology report suggested the morphological and immunohistochemical features were more compatible with a colonic IMT.

CONCLUSIONS A colorectal IMT is a rare clinical entity that can easily mimic a highly malignant tumour and cannot be distinguished clinically or radiologically. An accurate diagnosis is based on histological examination and surgical resection is therefore usually required.

KEYWORDS

Inflammatory myofibroblastic tumour – Pseudotumour – Colon – Rectum – Treatment – Retroperitoneal

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CORRESPONDENCE TO

Demetrios Moris, E: dimmoris@yahoo.com

Formerly known as an inflammatory pseudotumour, an inflammatory myofibroblastic tumour (IMT) is a rare entity characterised by the proliferation of spindle cells and/or dendritic cells with a plethora of inflammatory cells, mimicking a malignant tumour in its clinical and radiological findings.¹ While the lung is considered the most common site of occurrence, IMTs may arise in diverse extrapulmonary locations (probably in every part of the body including the abdomen and retroperitoneum). Although the aetiology of IMTs is still unclear, there is evidence to suggest that these masses are often associated with infection, or follow minor trauma or surgery. In some cases, they can even be associated with viral infections or malignancies.¹

The clinical presentation of IMT is non-specific. Gastrointestinal and urinary symptoms may occur with an enlarging expansile mass.² There are no specific imaging features for IMT. For abdominal or retroperitoneal IMT, a solid mass abutting and compressing various organs is the usual presentation.²

Case history

A 77-year-old man was admitted to our emergency department with acute abdominal pain and fever (38°C) that had started 2 days earlier. The pain had been intermittent for

almost ten days before its acute phase. In the emergency department, the patient was stable but in agony (blood pressure 140/93mmHg, heart rate 103bpm, SpO₂ 95%). His medical history was unremarkable with no history of any kind of abdominal surgery. Physical examination revealed tenderness of the lower abdomen, rebound tenderness, present bowel sounds and non-palpable masses. Laboratory results were within normal limits except for leucocytosis (14.6 x 10⁹/l), thrombocytosis (554 x 10⁹/l) and elevated C-reactive protein (84mg/l). Upper gastrointestinal endoscopy revealed mild gastritis. Colonoscopy was inconclusive because of inappropriate bowel preparation. Nevertheless, the colonoscopy did reach the transverse colon and no lesion was found up to this point.

Computed tomography (CT) revealed a mass in the right lower quadrant of the abdomen, possibly originating from the terminal ileum, 1–2cm away from the caecum, accompanied by thickening of the ileal and caecal walls (8mm) as well as pericaecal fluid and inflammation in the pericaecal soft tissue (Fig 1). Owing to the clinical impression of a potentially malignant lesion, the case was discussed at the multidisciplinary team meeting and a laparotomy was suggested.

An exploratory laparotomy with a midline incision was performed. Intraoperatively, ascites was found without the

Figure 1 Abdominal computed tomography revealing a mass in the right lower quadrant of the abdomen, possibly originating from the terminal ileum, 1–2cm away from the caecum, accompanied by thickening of the ileal and caecal walls (8mm)

Figure 2 Gross specimen revealing a whitish solid mass (5.9cm in greatest dimension) in the pericolic adipose tissue alongside the ileocaecal valve and appendix

presence of pus or pseudomembranes. The caecum and the proximal part of the ascending colon were oedematous with a firm mass of approximately 5cm in diameter encompassing the terminal ileum and caecum. The mass infiltrated the mesosigmoid tissue and sigmoid colon. The intraoperative findings made a diagnosis of intestinal malignancy even more likely. As a result, a subtotal colectomy and omentectomy were performed, followed by an ileorectal end-to-side two-layer anastomosis. Multiple palpable paracaval and para-aortic lymph nodes were also found and resected.

The pathology report noted that gross inspection of the specimen revealed a whitish solid mass in the pericolic adipose tissue alongside the ileocaecal valve and appendix (Fig 2). Part of the mass extended to the mesosigmoid, reaching the sigmoid colon and invading the sigmoid colon wall. The appendix appeared normal and measured 4.2cm long. The maximum diameter of the dissected lymph nodes found in the specimen was 0.5cm.

Microscopically, the mass consisted of large cells with mild to moderate atypia and a small number of more atypical nuclei, which expanded to the adipose tissue in a diffuse pattern without cohesion. Many of these cells were spindle-shaped and formed bundles. They were intermingled with inflammatory cells (mainly plasma cells, eosinophils and neutrophils, and a small number of lymphocytes). These cells invaded and ruptured the muscle layer, expanding through the submucosa near the ileocaecal valve and focally reaching the mucosa (Fig 3).

Immunohistochemistry showed that most cells were PGM1, CD45 antigen, CD68, lysozyme, S100 and vimentin positive, and negative for CD1a, CD21, CD23, CD30, CD34, CD117, CD246, smooth muscle actin, HMB-45 and pankaratin. The lymphocytes comprised mostly T cells (all positive

Figure 3 Morphology on haematoxylin and eosin sections: Cellular infiltrates penetrating through the muscularis propria and focally reaching the intestinal mucosa (20× magnification) (A); spindle-shaped tumour cells in fascicular arrangement (40× magnification) (B); cells admixed with a large number of plasma cells, eosinophils, neutrophils and lymphocytes (200× magnification) (C); and cells showing mainly moderate cellular atypia with a few severely atypical nuclei (400× magnification) (D).

for CD5, some for CD4 and some for CD8) while B cells (positive for CD20) were scarce (Fig 4). Neither the mucosa of the ileum or the large bowel nor the appendix showed pathomorphological findings. All the lymph nodes were reactive. The morphological and immunohistochemical features were more compatible with those of an IMT of the colon than of a malignant lesion.

Figure 4 Immunohistochemical studies: PGM1 immunochemical staining showing intense granular positivity (200× magnification) (A); smooth muscle actin is focally positive (400× magnification) (B); tumour cells negative for desmin (40× magnification) (C); and tumour cells negative for anaplastic lymphoma kinase (200× magnification) (D).

The patient's postoperative recovery was uneventful and he was discharged on day 7. In the 36 months since his surgery, there has been no return of symptoms. Follow-up imaging at six months and yearly thereafter has not revealed any recurrence.

Discussion

A colorectal IMT is a rare but benign entity, with fewer than 30 cases being reported in the literature. First described in the early 1950s, the term 'inflammatory pseudotumour' of the colon is used to describe a non-malignant and rare lesion found most commonly in the lung and the orbit. However, IMTs can be found in any site of the body, including the abdominal cavity.⁵ Despite the relative uncertainty about its pathogenesis and aetiology, in some cases, it is thought to be the result of an inflammatory reaction following infection, autoimmune response or surgery.

As in our patient, the clinical and imaging findings of an IMT can mimic those of malignant tumours. Patnana *et al* therefore referred to it as 'the great mimicker'.⁴ Names that have been applied to this particular pathology over the years include inflammatory pseudotumour, plasma cell granuloma, inflammatory fibrosarcoma and plasma cell pseudotumour. This illustrates the fact that these lesions have complex histological characteristics and suggests that the

term 'inflammatory myofibroblastic tumour' represents a group of lesions that show non-specific chronic inflammatory changes.⁴

Various locations in the colon have been reported to be affected by IMTs although the descending colon and sigmoid colon seem to be the most frequently involved sites.⁵ Abdominal and retroperitoneal pseudotumours usually present with non-specific imaging features and should consequently be considered among the differential diagnoses of any abdominal mass. Owing to its non-specific radiological findings, it is almost impossible to diagnose an IMT based exclusively on preoperative imaging. As in our case, the presenting signs and symptoms could include abdominal pain, anaemia, a palpable mass or intestinal obstruction. The clinical findings depend mostly on the location of the lesion.

The behaviour and natural history of IMTs is usually benign, with recurrence noted in some cases. An IMT is therefore a neoplasm with 'intermediate biological potential', having a propensity for local recurrence but rarely metastasising. Histopathological features alone may not be sufficient for the prediction of malignant transformation as tumour size, cellularity, mitotic activity and presence of necrosis are not significantly correlated with risk of metastasis.⁵

Conclusions

Colorectal IMTs can easily mimic highly malignant tumours and cannot be distinguished clinically or radiologically. An accurate diagnosis is based on histological examination and surgical resection is therefore usually required. Surgeons should be familiar with this particular entity to avoid unnecessary radical therapy. Nevertheless, its behaviour is generally benign and surgical therapy is usually enough. Long-term follow-up review is certainly needed in all cases.

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