<Case Report>

Inflammatory pseudotumor of the appendix

Takamori NAKAYAMA[1], Kou SHIRAISHI[1], Shunji MORI[1], Kiyoshi ISOBE[1], Shouichi OHTSUKA[1] and Masao KASAHARA[2]

Department of Surgery[1] and Pathology[2]
Shizuoka Red Cross Hospital

Key words: inflammation, pseudotumor, appendix

Abstract

We report a rare case of inflammatory pseudotumor arising from the appendix. A 77-year-old man underwent surgery for a mass in the right lower quadrant of the abdomen. The lesion was widely resected since it macroscopically resembled a malignancy, and the pathological diagnosis was inflammatory pseudotumor. The biological behavior of inflammatory pseudotumor is interesting, because this entity straddles the border between inflammation and neoplasia. The most reasonable treatment seems to be complete resection.

Introduction

Inflammatory pseudotumor of the lung was first described in 1939[1], and extrapulmonary lesions have also been reported at a wide variety of sites[2]. Inflammatory pseudotumor is generally considered to be a benign condition. Adoption of the current name for this entity was proposed in 1954, since various terms had been used to describe it previously depending on the predominant cellular component[3]. We report a rare case of inflammatory pseudotumor arising from the appendix.

Case report

A 77-year-old man presented with a 2-week history of diarrhea and fever since January 30, 2003. On February 15, physical examination revealed a firm mass in the right lower quadrant of the abdomen. Routine laboratory tests showed no abnormalities, apart from leukocytosis (WBC : 10,180/ul) and elevation of the C-reactive protein level (CRP : 6mg/dl). Ultrasonography detected a round and solid mass, which was well demarcated and measured 5 × 5cm (Fig. 1A). Computed tomography demonstrated a tumor arising from the cecum (Fig. 1B). Barium enema revealed an extracolonic mass, with a normal smooth pattern of the colonic mucosa (Fig. 2).

From the results of these examinations, appendicitis with abscess formation was strongly suspected, while diverticulitis, gastrointestinal stromal tumor (GIST), colon cancer, and malignant lymphoma were also considered in the differential diagnosis. On February 17, laparotomy was performed and revealed a tumor arising from the appendix. The mass was firmly attached to the retroperitoneum and appeared to be malignant, so it was radically removed by ileocecal resection (Fig. 3).

The resected tumor was grayish, glistening, and firm. Although the lesion was well circumscribed, there was no capsule. An ulcer was seen at the orifice of the appendix, while bleeding and necrosis extended

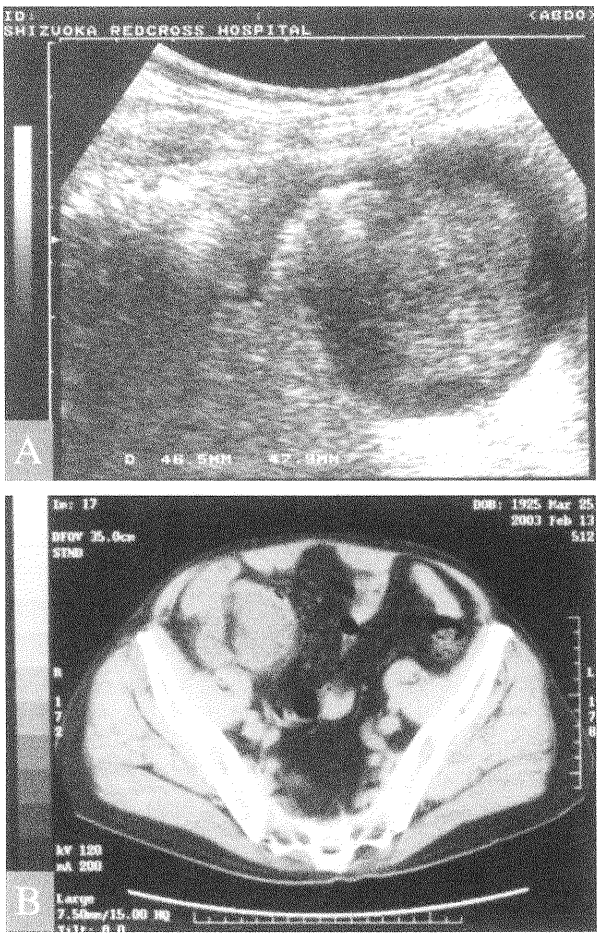

Fig. 1: A; Ultrasonography reveals a nonhomogenous hypoechoic mass with a peripheral hypoechoic band. B; Computed tomography shows a nonhomogenous soft tissue mass in the right lower quadrant of the abdomen and associated anteromedial displacement of the terminal ileum.

through the mucosa, muscular layer, and serosa. Cross-sections of the appendix revealed the presence of circumferential fibrosis (Fig. 4A). The mucosa and muscular layer were fused or absent throughout half of the distal body of the appendix, with the normal tissues being replaced by inflammatory granulomatous tissue that contained proliferating capillaries as well as infiltrating neutrophils, histiocytes, lymphocytes, and fibroblasts (Fig. 4B). This inflammatory tissue also widely involved the mesoappendix, mesentery, and retroperitoneum. There was a mixture of regions of

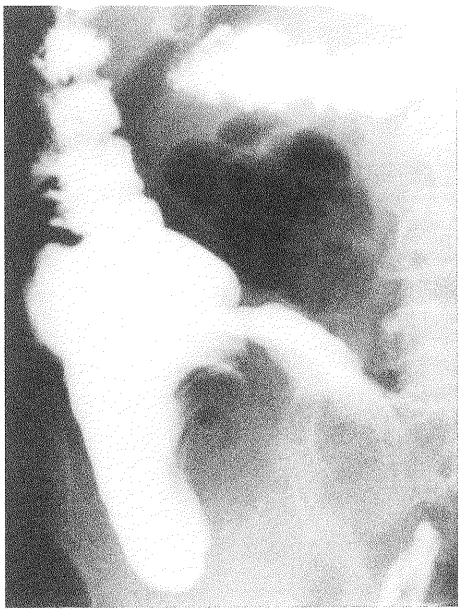

Fig. 2: Barium enema shows a large extraluminal mass on the medial aspect of the cecum.

severe fibrosis and abscess formation. In the remaining half of the appendix, the mucosa retained its normal architecture, but the serosa was buried in the surrounding fibrous tissue. The lymphoid follicles were atrophic, and no lymphocytes were detected. Fibrous septae were seen in the fatty tissue surrounding the appendix. Near the apex of the appendix, the inflammation was very mild (Fig. 4C). These findings indicated that transmural inflammation had arisen in the appendix and had spread to involve the surrounding fatty tissue, meeting the pathologic criteria for a pseudotumor originating from the appendix. The patient's postoperative course was uneventful, and there was no evidence of recurrence at 17 months after surgery.

Discussion

The etiology of inflammatory pseudotumor has been suggested to include trauma, surgery, infection, and autoimmunity[4]. Cases of inflammatory pseudotumor have been observed from early childhood to old age[5]. The lesion often pres-

Table 1 Inflammatory pseudotumor of the appendix

No	Age	Sex	Complaint	WBC	Size	Adhesion	Treatment	Ref.
1	10M	F	Mass	4600(/μl)	11(cm)	Loose	Appendectomy	6
2	8Y	M	Mass	7500	7	Loose	Appendectomy	7
3	65Y	M	Pain	7100	3	Loose	Right hemicolectomy	8
4	77Y	M	Mass	10180	5	Firm	Ileocecal resection	Our case

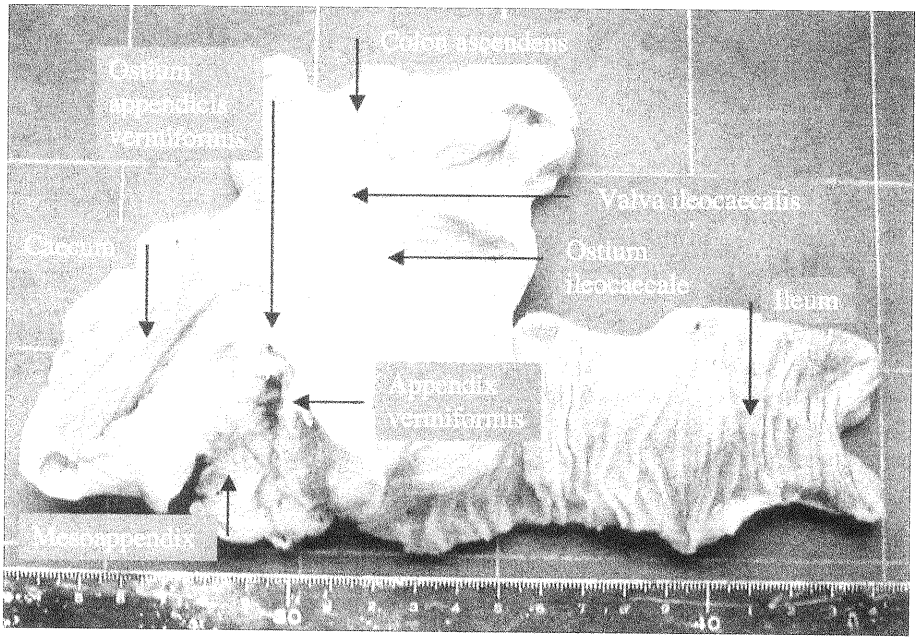

Fig. 3: The resected inflammatory pseudotumor of the appendix contains areas of hemorrhage and necrosis.

ents as an incidental mass without symptoms. In a subset of patients (15-30%), however, it is associated with a syndrome of unexplained fever, weight loss, anemia, and hypergammaglobulinemia or thrombocytosis[2].

The biological behavior of inflammatory pseudotumor is interesting because this entity straddles the border between inflammation and neoplasia. That is, inflammatory pseudotumor is a benign disorder that presents clinically in a manner simulating a neoplasm. It is a proliferative lesion composed of mature inflammatory cells without any evidence of neoplasia. However, these lesions have been reported to show the capacity for rapid growth, local recurrence, and malignant transformation[6].

Depending on the characteristics of the tumor, the most reasonable treatment seems to be complete resection[2], but it should be remembered that spontaneous resolution has also been reported, while steroid therapy achieves variable results[6]. Most of the previous reports have concerned inflammatory pseudotumors involving the lungs and bronchi, so it is unclear whether the lesion shows the same behavior at other less common locations. The limited information available suggests that the mechanism of development may vary with the organ involved, but inflammatory pseudotumor is difficult to classify in detail because of the small number of reported cases.

Inflammatory pseudotumor arising from the appendix is a rare condition, even though

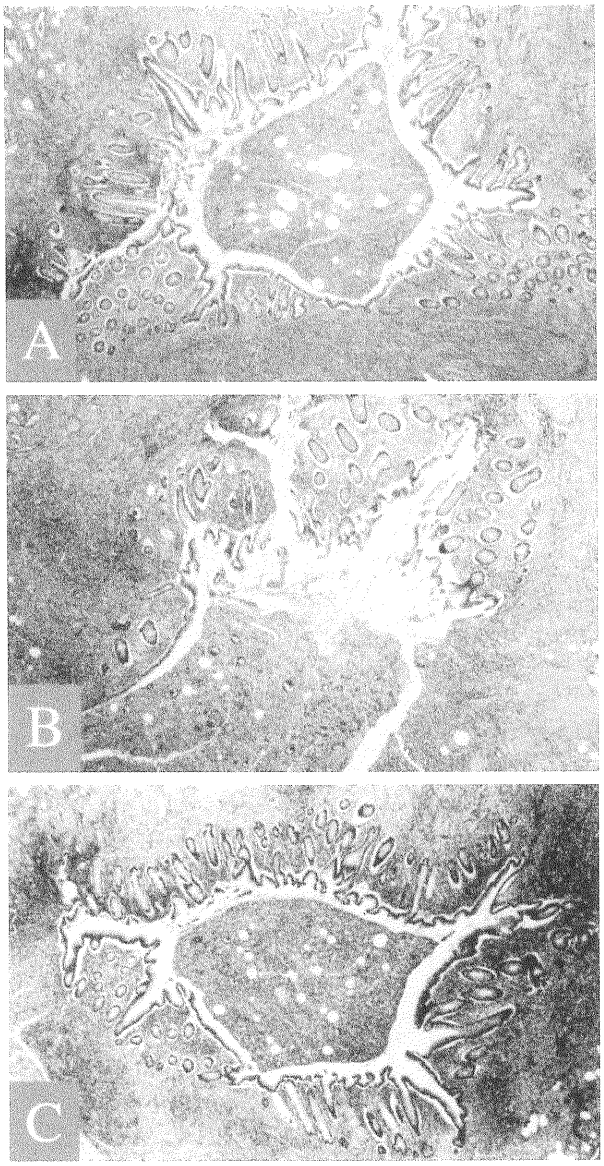

Fig. 4 : A ; Photomicrograph showing inflammation with abscess formation in the proximal part of the body of the appendix (hematoxylin and eosin stain, X2). B ; Photomicrograph showing fusion and disappearance of epithelium as well as infiltration with inflammatory cells in the distal body of the appendix. C ; Photomicrograph of the apex of the residual appendix.

the appendix is one of the most common sites to be affected by inflammation. Only three previous cases were revealed by an extensive review of the literature (Table 1)[6,7,8]. Appendectomy was performed in two of the reported patients, while radical resection was done in one because the macroscopic appearance mimicked a malignant tumor. In all reported cases, the postoperative course was uneventful and there was no recurrence. Our patient was the only one with leukocytosis and firm attachment of the tumor to the retroperitoneum.

Panniculitis of the mesoappendix follows the same clinical course as inflammatory pseudotumor[9], and needs to be included in the differential diagnosis. Mesenteric panniculitis has the histological features of fat necrosis with infiltrating histiocytes and phagocytes, so this condition was ruled out in our patient.

In conclusion, surgeons need to keep in mind the biological behavior of inflammatory pseudotumor. The most reasonable treatment seems to be complete resection while avoiding an aggressive procedure.

References

1) Brunn H : Two interesting benign lung tumors of contradictory histopathology ; Remarks on the necessity for maintaining chest tumor registry. J Thorac Surg 9 : 119-131, 1939.
2) Buell R, Wang NS, et al : Endobronchial plasma cell granuloma ; a light and electron microscopic study. Hum Pathol 7 : 411-426, 1976.
3) Umiker WO, Iverson L : Postinflammatory tumor of the lung. J Thorac Surg 28 : 55-62, 1954.
4) Wu JP, Yunis EJ, et al : Inflammatory pseudo-tumours of the abdomen ; plasma cell granulomas. J Clin Pathol 26 : 943-948, 1973.
5) Matsubara O, Tan-Liu NS, et al : Inflammatory pseudotumors of the lung ; progression from organizing pneumonia to fibrous histiocytoma or to plasma cell granuloma in 32 cases. Hum Pathol 19 : 807

-814, 1988.

6) Stringer MD, Ramani P, et al : Abdominal inflammatory myofibroblastic tumours in children. Br J Surg 79 : 1357-1360, 1992.

7) Narasimharao KL, Malik AK, et al : Inflammatory pseudotumor of the appendix. Am J Gastroenterol 79 : 32-34, 1984.

8) Sasaki S, Hanatate F, et al : Inflammatory pseudotumor of the appendix. Gastroenterological Surgery 12 : 519-522, 1989.

9) Lange JF, Morino M, et al : Mesenteric panniculitis of the mesoappendix. Int Surg 74 : 201-202, 1989.