Intestinal perforation from primary intra-abdominal fibromatosis

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A 60-year-old man with diffuse abdominal pain, nausea, and vomiting of 2 days' duration presented to our emergency department. He had no history of systemic disease, trauma, or abdominal complaints. Physical examination revealed diffuse abdominal tenderness and hypoactive bowel sounds. There were multiple air-fluid levels on plain abdominal radiography. The results of laboratory analysis performed at the time of admission were as follows: blood urea nitrogen, 44 mg/dL (normal range, 6-21 mg/dL); lactate dehydrogenase, 286 U/L (normal range, 100-210 U/L); leukocyte count, 13,200/mm^3 (89% polymorphonuclear); prothrombin time, 16.6 seconds (normal range, 11-14.5 seconds); international normalized ratio, 1.37 (normal range, 1-1.2); and C-reactive protein, 352 mg/L (normal range, 0-10 mg/L). Abdominal computed tomography showed a mesenteric mass that was adherent to small bowel loops and that exhibited minimal enhancement after the intravenous injection of contrast material. Free fluid near the mass and free air in the peritoneal cavity and perihepatic region were also noted (**Figure 1**). The primary diagnosis was small bowel perforation caused by the intestinal stromal or desmoid tumor. During laparotomy, diffuse pseudomembrane formation with minimal intra-abdominal fluid and an 8-cm mass that arose from the retroperitoneal area and involved the root of the mesentery were found. The tumor was in close association with the intestine from 150 cm distal to ligament of Treitz and was found to be conglomerated with jejunal loops. A perforation of approximately 1 cm was noted. The perforated segment and the conglomerated 40 cm of the jejunum were resected, and a primary anastomosis was performed. The mass was not completely excised because of the extensive involvement of the root of the mesentery and the great vessels. The findings from postoperative upper gastrointestinal endoscopy and colonoscopy were within normal limits, and the results of hormonal assays (estradiol, progesterone, total and free testosterone) and analyses for tumor markers (CEA, AFP, CA 19-9, CA 72-4) were within the reference range. Histologic examination of the mass revealed no signs of malignancy. Immunohistochemical studies showed diffuse expression of vimentin and the focal expression of actin. Staining for desmin and CD-34 was not significant, and the diagnosis was fibromatosis. The patient's postoperative

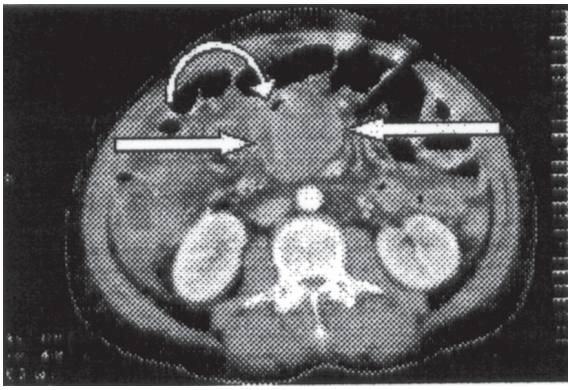

Figure 1 - Abdominal computed tomography demonstrated a well-demarcated slightly enhancing mass (left arrow), free fluid (right arrow) and free air (curved arrow) near the mass.

course was uneventful, and he was discharged from the hospital on the tenth postoperative day. The tumors of the mesentery and retroperitoneum are very rare. Intra-abdominal fibromatosis (IAF) is a benign mesenchymal tumor that originates from myofibroblasts and fibroblasts and can develop at any site of the gastrointestinal tract in patients of any age. Sporadic cases of IAF and the association of that disorder with Gardner's syndrome and familial adenomatous polyposis have been described.[1-4] Intra-abdominal fibromatosis can be solitary or multiple.[3] Intestinal obstruction and fistula formation are the most common presentations of that type of tumor, which can also cause symptoms from the invasion or compression of adjacent organs.[2,3] There are very few reports of small intestine perforation due to fibromatosis in the literature. No significant association of predisposing factors was detected in our patient, and the developments of IAF in this case was considered sporadic. Invasion and perforation of the intestine had occurred. The site of the tumor and the patient's presenting symptoms were unique. Because the clinical, radiologic, and histopathologic characteristics of IAF can mimic those of gastrointestinal stromal tumors, misdiagnoses can occur. Identifying the expression of CD-34 and S-100 via immunostaining may be helpful in the differentiation of those entities.[1,3] In our patient, the final diagnosis was also based on the results of histopathologic analysis via immunostaining. Complete surgical resection is the therapy of choice, but this often requires the resection of a segment of bowel, and resection along the mesenteric vessels significantly increases morbidity. Local recurrence of IAF is frequent, and a second excision may be required. With each subsequent surgery, mortality and morbidity increase. Patients with an unresectable IAF or a recurrent lesion may survive long-term, even though complications from the unresectable tumor occur.[2,4] In patients with Gardner's syndrome, an unresectable IAF; the involvement of the tumor with vital structures; the use of

hormone therapy, non-steroidal anti-inflammatory drugs, or chemotherapeutic medications; or radiation therapy may be preferable to surgical treatment.[2] However, the role of medical therapy is unclear.[2] Complete surgical resection is the mainstay of treatment for IAF, but is usually associated with increased rates of mortality and morbidity. The progression, stabilization, and regression of such tumors may occur,[2] and their clinical behavior seems to be more important than the treatment of choice.[5] Stable disease in 8 of 10 patients with an unresectable IAF has been reported.[4] In our patient, complete resection was not possible because of the proximity of the tumor to mesenteric vessels. No medical treatment with controversial efficacy was given to our patient, who remained asymptomatic during 6 months of follow-up. In conclusion, fibromatosis originating from the retroperitoneum or mesentery should be considered in the differential diagnosis of acute abdominal pain in patients with an intra-abdominal mass.

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References

1. Yantiss RK, Spiro IJ, Compton CC, Rosenberg AE. Gastrointestinal stromal tumor versus intra-abdominal fibromatosis of the bowel wall: a clinically important differential diagnosis. ***Am J Surg Pathol*** 2000; 24: 947-957.
2. Smith AJ, Lewis JJ, Merchant NB, Leung DH, Woodruff JM, Brennan MF. Surgical management of intra-abdominal desmoid tumours. ***Br J Surg*** 2000; 87: 608-613.
3. Colombo P, Rahal D, Grizzi F, Quagliuolo V, Roncalli M. Localized intra-abdominal fibromatosis of the small bowel mimicking a gastrointestinal stromal tumor: a case report. ***World J Gastroenterol*** 2005; 11: 5226-5228.
4. Burke AP, Sobin LH, Shekitka KM, Federspiel BH, Helwig EB. Intra-abdominal fibromatosis. A pathologic analysis of 130 tumors with comparison of clinical subgroups. ***Am J Surg Pathol*** 1990; 14: 335-341.
5. Reitamo JJ, Scheinin TM, Hayry P. The desmoid syndrome. New aspects in the cause, pathogenesis and treatment of the desmoid tumor. ***Am J Surg*** 1986; 151: 230-237.