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CASE REPORT

Peritoneal Sarcoidosis
An Unrecognized Cause of Sclerosing Peritonitis

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Sclerosing peritonitis is a rare syndrome characterized by abdominal pain, intestinal obstruction, and thickening of the peritoneum with massive adhesions (1). Only eight undisputed cases of peritoneal sarcoidosis have been reported (2–7). We report a patient with a history of ocular and pulmonary sarcoidosis who developed sclerosing peritonitis, with none of the known causes, and recovered promptly on corticosteroid therapy. We therefore suggest that peritoneal sarcoidosis is an unrecognized cause of sclerosing peritonitis and that, unlike the other causes of this severe syndrome, it is curable by corticosteroid therapy.

CASE REPORT

A 53-year-old Caucasian woman was admitted to Saint Lazare Hospital in September 1990 with abdominal pain, vomiting, and ascites. Sarcoidosis had been diagnosed 12 years earlier when she presented with uveitis, which was controlled with a six-month course of oral prednisone. In 1989, pulmonary sarcoidosis was diagnosed on the basis of dyspnea, pulmonary interstitial infiltrate, restrictive syndrome, an abnormally high lymphocyte count in bronchoalveolar lavage fluid, and noncaseating epithelioid granulomas within bronchial and transbronchial biopsies. Thoracic computed tomography showed no lymph nodes but revealed a small pleural effusion and minimal ascites; no paracentesis was done. Serum angiotensin-converting enzyme was normal but the tuberculin skin test was positive. Repeated cultures of gastric juice, bronchial secretion, and bronchial biopsy specimens were negative for mycobacteria and fungi. Dyspnea and pulmonary function tests improved spontaneously within six months. The patient had no history of previous abdominal surgery and was taking no medication.

In May 1990, she developed abdominal pain, vomiting and abdominal distension. A pelvic ultrasound scan revealed a left ovarian tumor (diameter: 6 cm) and ascites. Ascitic fluid protein was 4.5 g/dl, with a white cell count of 140/μl, 45% mesothelial cells, 55% lymphocytes, and a red blood cell count of 60,000/μl.

On June 20, 1990, the patient underwent a first laparotomy: the peritoneal cavity contained 4 liters of ascitic fluid and ovarian examination showed a bilateral tumor; the small bowel had thickened and was covered by a dense whitish membrane suggesting a diagnosis of sclerosing peritonitis. Hysterosalpingooophorectomy and omentectomy were performed. Microscopic examination revealed benign ovarian cystadenofibroma. Total parenteral nutrition was started because the postoperative persistence of vomiting preventing oral nutrition. A small bowel follow-through showed a fixed, shrunken small bowel and separated loops with a kinked terminal ileum (Figure 1). Upper digestive fibroscopy and colonoscopy were normal. Histologic examination of duodenal and colonic per-endoscopic biopsies revealed normal mucosa and submucosa.

On August 27, 1990, a second laparotomy was performed. The peritoneal cavity still contained 3 liters of ascitic fluid, small bowel involvement was similar to that observed previously, and the mesentery and retroperitoneum were still normal. Several biopsies of the peritoneum, small bowel, and liver were performed, as well as appendicectomy and gastrostomy. Microscopic examination of liver tissue was normal. Microscopic examination of a surgical specimen of the small bowel external wall showed a thickened subserosa with collagen fibrosis sticking to the muscularis propria, which contained a few fibroblasts and very few lymphocytes; neovascularization was seen between this thick membrane and the muscularis propria (Figure 2). There was no evidence of the presence of any foreign body under polarized light. Despite a large number of careful explorations, no granuloma was found. Special stains and cultures were negative and strongly so for mycobacteria. Macroscopic and histological findings were therefore consistent with the diagnosis of sclerosing peritonitis.

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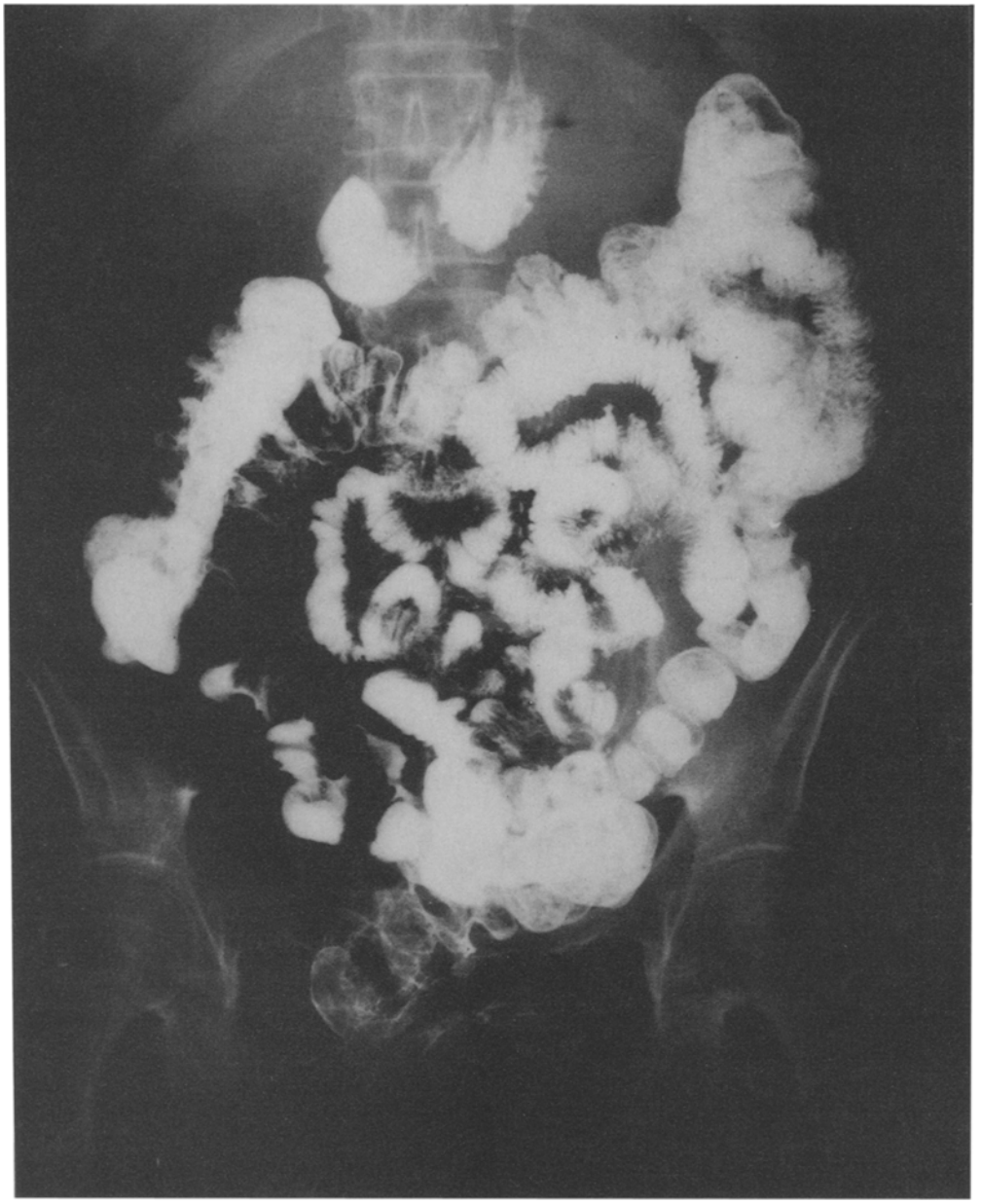

Fig 1. Barium study of the small intestine: Separated loops with kinked terminal ileum.

After surgery, not only was the patient still unable to eat, but vomiting promptly recurred when the gastrostomy tube was clamped. On September 18, 1990, the patient was transferred to Saint Lazare Hospital. Physical examination revealed weight loss of 5 kg, ascites, and pleural effusion. Laboratory work-up showed 3000 × 10^9/liter leukocytes, with a normal differential count, an erythrocyte sedimentation rate of 70 mm/hr, 12.4 g/dl hemoglobin, 236 × 10^9/liter platelets, normal ALAT and ASAT, 168 IU/liter alkaline phosphatase (normal < 90), total bilirubin of 3 μmol/liter, serum albumin of 2.7 g/dl, normal serum gamma globulin and serum calcium, pleural fluid protein of 3.6 g/dl, a pleural fluid white cell count of 150/μl with 53% lymphocytes and a red blood cell count of 59,000/μl. Culture of pleural fluid was negative.

On the basis of a provisional diagnosis of sclerosing peritonitis due to peritoneal sarcoidosis, intravenous corticosteroid treatment was started, comprising 1 g/day methylprednisolone for two days and then 60 mg *per os* once a day. A striking improvement occurred, as two days after the treatment started, the gastrotomy tube was clamped without recurrence of vomiting, and one week afterwards the patient was able to eat normally. Parenteral nutrition was then progressively discontinued and the gastrostomy tube removed. Five weeks after the start of the treatment, the patient had gained 5 kg, a barium study

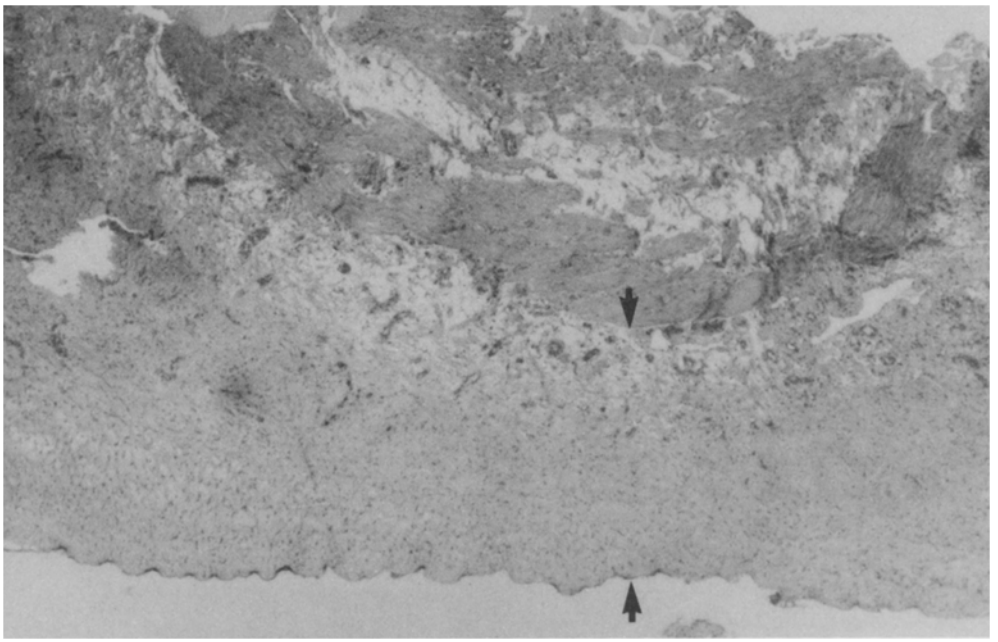

Fig 2. Surgical small bowel specimen: Collagen fibrosis thickening subserosa (arrows) and sticking to the muscularis propria. (H&E, ×120).

of the small intestine was normal, and computed tomography showed no ascites, although the small pleural effusion persisted. After one month of treatment, corticotherapy was tapered and after six months, the patient remained asymptomatic on 15 mg/day of oral prednisone.

DISCUSSION

According to Bender, "Sclerosing peritonitis is a rare entity in which there may be peritoneal changes, from peritoneal thickening to the development of a thick, firm white grayish tissue which encases the small intestine in a cocoon" (1). Clinically, there is abdominal pain, vomiting, and intestinal obstruction; histologically, there is nonspecific focal inflammation, granulation tissue and a peculiar laminated fibrosis. All these features were found in our patient. Sclerosing peritonitis probably constitutes a form of peritoneal reactivity to a variety of stimuli (1). Well-known causes of sclerosing peritonitis are bacterial infection (8), especially mycobacteria (9); ingestion of adrenergic-blocking drugs such as practolol (10, 11); an indwelling foreign body, such as that used in continual ambulatory peritoneal dialysis (12, 13) or peritoneovenous shunt (14, 15); and peritoneal contamination by fibrogenic foreign materials (16). The possibility of prior surgical peritoneal fibrosis can be excluded in our patient, as initial peritoneal macroscopic changes were seen in the absence of any prior surgery and no surgical contaminant was found. Elsewhere, anecdotal cases of sclerosing peritonitis have been described in association with abdominal trauma (17), asbestos exposure (18), carcinoid syndrome (19), and familial Mediterranean fever (20). None of these reported causes was present in our patient. There are also 20 other reported cases of idiopathic sclerosing peritonitis for which no cause was found; this syndrome affects young girls, predominantly in tropical countries (21–28). In sclerosing peritonitis, ascites is unusual and has only been reported in cases of continual ambulatory dialysis (29), peritoneovenous shunt (14), tuberculosis (9), and in three of the 20 above cases of idiopathic sclerosing peritonitis (25, 27, 28). Meigs' syndrome is defined by the combination of fibroma of the ovary, transudate ascites, and pleural effusion, which disappear in less than three weeks when the fibroma is removed (30). This syndrome can be ruled out in our patient, since she had exudate effusion that persisted for four months after tumor removal. Our patient had the typical clinical and histological spectrum of sclerosing peritonitis and a previous history of systemic sarcoidosis.

A positive tuberculin skin test and normal serum angiotensin-converting enzyme activity are not uncommon features in sarcoidosis, as seen in 10–45%

of cases (31) and about 50% of cases (32, 33). The combination of a blood-stained exudative lymphocytic pleural fluid like that of our patient with lung sarcoidosis is highly characteristic of pleural sarcoidosis (34, 35), especially in the absence of mycobacteria. Under these conditions, percutaneous pleural biopsy is not always helpful in diagnosing pleural sarcoidosis (35).

Only 13 cases of peritoneal sarcoidosis have been reported (2–7, 36–39). In the well-documented cases, diagnosis was based upon the combined findings of systemic sarcoidosis features, peritoneal epithelioid granulomas, and the absence of another cause of granulomatous disease, especially tuberculosis (2–7). Exudative lymphocytic ascites, with or without blood, was present in seven cases and healed spontaneously or after steroid therapy. In our patient we were unable to demonstrate the presence of peritoneal epithelioid granuloma. However, the history of lung and pleural sarcoidosis, followed one year later by exudative, lymphocytic, bloody ascites with no mycobacteria or fungi, prompt us to consider that our patient's peritoneal involvement, typical of sclerosing peritonitis, was caused by sarcoidosis. To our knowledge, peritoneal sarcoidosis has never been described as a cause of sclerosing peritonitis. However, Robertson and Cunningham (7) recently reported a patient with primary peritoneal sarcoidosis, in whom "the serosa over the small bowel and the stomach were thickened and bowel loops were matted together." Even though the latter patient had no clinical sign of sclerosing peritonitis, these histological changes are probably typical of those seen in this syndrome. Lastly, in our patient, a dramatic improvement occurred on corticosteroid treatment. This is an additional strong argument for incriminating sarcoidosis as a hitherto undetected cause of sclerosing peritonitis, since such improvement, which is usual in peritoneal sarcoidosis, has not been described in other secondary or idiopathic forms of sclerosing peritonitis.

SUMMARY

In a Caucasian woman with a history of ocular and pulmonary sarcoidosis, the occurrence of sclerosing peritonitis with exudative ascites but without any of the well-known causes of this syndrome prompts us to consider that sclerosing peritonitis is a manifestation of sarcoidosis. The dramatic improvement that occurred on corticosteroid therapy adds credibility to this previously unreported association.

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