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Sclerosing Mesenteritis Causing Chylous Ascites and Small Bowel Perforation

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest:

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None declared

Patient: Male, 80
Final Diagnosis: Sclerosing mesenteritis
Symptoms: Abdominal distension • abdominal tenderness
Medication: —
Clinical Procedure: Paracentesis
Specialty: Gastroenterology and Hepatology

Objective: Rare disease
Background:





Sclerosing mesenteritis (SM) is a rare idiopathic inflammation and fibrosis of the mesentery. Its etiology and pathogenesis remain unclear. It has been linked to abdominal trauma related to peritoneal dialysis, recent abdominal surgery, idiopathic inflammatory disorder, paraneoplastic syndrome, ischemia/infection, and autoimmunity. Abdominal CT is the most sensitive imaging modality, and diagnosis is usually confirmed by surgical biopsy. Patients most often present with abdominal pain, nausea, vomiting, diarrhea, and weight loss, and less commonly with chylous ascites and small bowel obstruction. Treatment is usually supportive; surgical intervention may be attempted for life-threatening complications such as bowel obstruction or perforation.

Case Report: This report describes an 80-year-old man with hypertension and end-stage renal disease (ESRD) presenting with increasing abdominal pain and tenderness over the past 5 months. Abdominal enhanced computed tomography (CT) revealed a fat-ring sign and peritoneal calcifications along the serosa surface of small bowel consistent with sclerosing mesenteritis. His hospital course was complicated by increasing ascites requiring multiple ultrasound-guided paracentesis, worsening leukocytosis, and persistent hypotension after dialysis, requiring pressor support. Ascitic fluid analysis was consistent with chylous ascites. The patient subsequently developed small bowel obstruction causing focal perforation, leading to the death of our patient. In this report, we review the clinical presentation, radiographic findings, treatment, and outcome in our patient and review the relevant literature.

Conclusions: Diagnosis of sclerosing mesenteritis is challenging due to its nonspecific clinical features. Sclerosing mesenteritis is a debilitating albeit self-limiting disorder that can rarely become fulminant, largely due to its complications.

MeSH Keywords: Abdominal Pain • Chylous Ascites • Intestinal Obstruction • Intestinal Perforation • Panniculitis, Peritoneal

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Background

Sclerosing mesenteritis (SM) is a rare idiopathic inflammation and fibrosis of the mesentery [1,2]. The first known series of sclerosing mesenteritis was published in 1924 and was categorized by the following subgroups: 'refractile mesenteritis', 'mesenteric lipodystrophy', 'mesenteric panniculitis', and 'mesenteric sclerosis' [3,4]. Most recently, sclerosing mesenteritis has been recognized to be associated with IgG4-related disease, and the term 'IgG4-related mesenteritis' has since been incorporated into this spectrum of disease [5,6]. Its etiology and pathogenesis remain obscure. A retrospective study of computed tomography (CT) images reports a prevalence of 0.54–0.90% in patients with a median age of 65 years [1,3,7]. Due to its nonspecific symptoms, diagnosis is often challenging. Various treatment regimens have been used, although reports are often anecdotal. Sclerosing mesenteritis is a debilitating albeit self-limiting disorder that can rarely become life-threatening (from severity of small bowel obstruction), epitomized by our patient [1]. We present a rare case of small bowel obstruction and chylous ascites due to sclerosing mesenteritis causing focal perforation of the small bowel, leading to the death of our patient.

Case Report

An 80-year-old African American man with a history of hypertension and end-stage renal disease (ESRD), who was on peritoneal dialysis, presented with increasing hematemesis, abdominal distention and tenderness over the past 5 months. He denied other gastrointestinal symptoms such as nausea, vomiting, melena, or hematemesis. Medications included 10 mg of Amlodipine daily and 1 tablet of Nephro-Vite daily. The patient had no significant past surgical history or allergies. On social history, he admitted to smoking 1 pack of cigarettes per day for approximately 10 years up to year 2001. On presentation, the patient was found to be hypotensive at 73/54 mm/Hg despite administration of intravenous fluids, eventually requiring vasopressor support. On physical examination, he appeared lethargic, somnolent, and oriented only to self. His abdomen was distended, tympanic to percussion, and had decreased bowel sounds. Testing was notable for hemoglobin of 7.3g/dL, white blood cell count (WBC) of $16.1 \times 10^9/L$, creatinine of 7.41mg/dL, pH of 7.27, lactate of 3.3 mg/dL on venous blood gas, and C-reactive protein of 25.6 mg/dL. IgG4 was negative. There were otherwise no other laboratory abnormalities. CT of the abdomen was performed, revealing fat-ring sign (Figure 1) and peritoneal calcifications extending along the serosal surfaces of small bowel (Figure 2), consistent with sclerosing mesenteritis. An initial nasogastric tube placed for decompression yielded melanic fluid and he was made NPO (nothing by mouth). Serial imaging confirmed the diagnosis of sclerosing mesenteritis given unchanged findings. Surgery



Figure 1. Fat-ring sign as preservation of fatty attenuation around vessels in the involved mesentery (arrows).

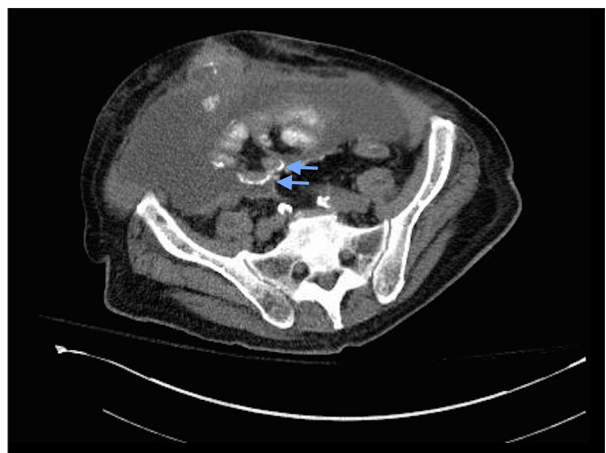


Figure 2. Abdominal CT imaging showing calcifications (arrows) along serosa surface of the small bowel.

was consulted, and the patient was deemed a poor surgical candidate given his hemodynamic instability. Unfortunately, corticosteroid treatment for the disease was deferred in light of a recent *Clostridium difficile* infection and he was started on total parenteral nutrition. His hospital course was complicated by increasing ascites requiring multiple ultrasound-guided paracentesis, worsening leukocytosis, and persistent post-dialysis hypotension requiring pressor support. Ascitic fluid analysis revealed a triglyceride level of 351 mg/dL, cell count of 533/uL, total protein of 4.0 g/dL, lactate dehydrogenase of 120 IU/liter, and calculated serum-ascites albumin gradient (SAAG) of 0.8 g/dL, consistent with chylous ascites. Repeat laboratory testing revealed a pH of 6.9, lactate of 7.0 mg/dL on venous blood gas, and WBC of $45.9 \times 10^9/L$. The patient became increasingly somnolent throughout the hospital stay. Interval imaging revealed focal perforations of his small bowel (Figure 3). The patient did not undergo surgical intervention as he was too unstable given his deteriorating clinical status and would likely not survive surgery. After careful discussion with the family, the decision was made to transfer the patient to the palliative care unit for comfort measures.

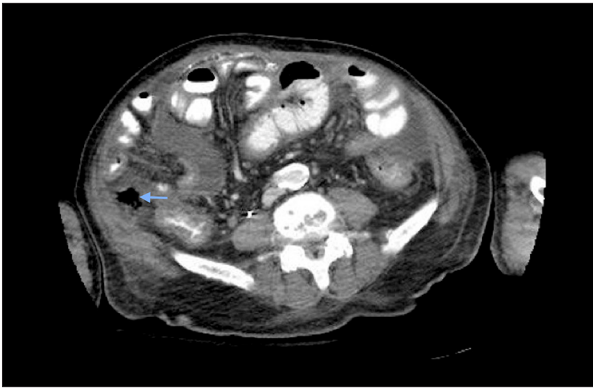


Figure 3. Abdominal CT imaging showing focal perforation of the small bowel (arrow) with a small pocket of extraluminal gas.

Discussion

Sclerosing mesenteritis is a rare fibrotic condition that is characterized by the presence of fat necrosis and chronic inflammation in the mesentery, primarily in small bowel mesentery in patients in Western countries [1,2,8]. In contrast to this finding, the disease often involves the large bowel mesentery in Japanese patients, in particular the sigmoid mesentery [8,9]. The reasons for this difference are not well understood. Ogden et al. reported a range of pathologic findings, and it is possible that the variations represent different stages of the disease, with features progressing from adipocyte necrosis to a chronic inflammatory state (mesenteric panniculitis) and finally fibrosis (sclerosing mesenteritis) [2]. In many cases, there may be a combination of any 2 or all 3 of these conditions. Hence, ‘sclerosing mesenteritis’ is the most semantically appropriate term in most cases [1–3].

Akram et al. demonstrated that the incidence of underlying neoplastic disease is about 56% [3]. The neoplastic diseases reported were lymphoma, melanoma, colorectal cancer, and prostate cancer [3,7,10]. Other causative factors were abdominal surgery and trauma, ischemia/infection, and autoimmune processes such as retroperitoneal fibrosis, sclerosing cholangitis, Riedel thyroiditis, and orbital pseudotumor. However, the actual pathophysiology is still unknown [1,4,11]. Some series demonstrated a high serum IgG4 level and infiltration of prominent IgG4-positive plasma cell in affected tissue in a subset of patients with the disease, suggesting that some cases are a manifestation of IgG4-related disease [3,5,8,12–14]. In the present case, IgG4 was negative and it was thought that peritoneal dialysis resulted in structural and functional damage to the peritoneal membrane, precipitating the process.

The association between sclerosing mesenteritis and diabetes mellitus has been described in the literature [15]. Histologically, sclerosing mesenteritis is characterized by fat degeneration

and necrosis, chronic inflammation, fibrosis, and macrophages infiltration [2]. Diabetes mellitus is an inflammatory disease, and in obese patients with insulin resistance, adipocyte necrosis and increased numbers of macrophages in adipose tissue are also present [16]. In a recent case series of 5 patients with sclerosing mesenteritis, all patients had diabetes mellitus [15]. Thus, it can be postulated that sclerosing mesenteritis and diabetes mellitus share a similar inflammatory process involving the fatty tissue. Further studies are needed to investigate this pathophysiological relationship.

Sclerosing mesenteritis often presents with abdominal pain, but symptoms may be nonspecific, which makes diagnosis extremely challenging [1–3,7,8]. In a study by Akram et al. in 92 cases of sclerosing mesenteritis, 70% had abdominal pain, 26% had bloating and distention, 25% had diarrhea, and 23% had weight loss [3]. In the present case, both abdominal pain and distention were present. There were no other intrabdominal abnormalities such as diverticulitis, peptic ulcer, or cholangitis that might have caused his abdominal pain.

Chylous ascites, seen in this patient, is an unusual presentation and was reported in 7% of all case reports and case series [13]. Chylous ascites is the accumulation of lipid-rich lymph in the peritoneal cavity, due to presence of thoracic or intestinal lymph in the abdominal cavity. Chylous ascites develops when there is a disruption of the lymphatic system secondary to traumatic injury or obstruction [17]. It can be defined as the presence of peritoneal fluid with a high triglyceride content, typically above 200 mg/dL [17]. Causes of chylous ascites include abdominal malignancy and cirrhosis in Western countries, and infectious etiologies, such as tuberculosis in developing countries [17,18]. Akram et al. reported that chylous ascites was found in 14% of the patients with SM [3]. It is believed that direct mechanical compression of the mass encasing the lymphatics results in chylous ascites [3]. Our patient had no history of cirrhosis, prior abdominal malignancy and the ascitic fluid was negative for acid fast bacilli.

Small bowel perforation is a late complication of sclerosing mesenteritis. CT remains the optimal modality for distinguishing the various types of mesenteric neoplasm, and can be essential in differentiating sclerosing mesenteritis from other tumors such as carcinomatosis, carcinoid tumor, lymphoma, and desmoid tumor, and mesenteric edema [10,11,13,19,20]. The CT appearance varies depending on the predominant tissue component and extent of inflammation and fibrosis [10,11,19,21]. The most common finding is a soft tissue mass in the small bowel mesentery, and the mass can be heterogeneous in the inflammatory phase or homogenous in the fibrotic phase. There may be preservation of fat around the mesenteric vessels, a phenomenon that is referred to as the “fat-ring sign” [9–11,19,21,22]. Published reports demonstrated that

pseudocapsule and increased attenuation of the mesenteric fat often described as ‘misty mesentery’ could help suggest the correct diagnosis [10,20]. In the study by Kipfer et al., a mass in the left upper quadrant was the most characteristic, and usually the only, physical examination finding [7]. Nevertheless, definitive diagnosis is usually established by histologic evaluation given the nonspecific radiologic features [1,2,7]. In the present case, surgical biopsy was not performed because the patient had already suffered from the complications of sclerosing mesenteritis and was provided comfort care.

There is no consensus regarding the optimal medical therapy for sclerosing mesenteritis as there are no clinical studies assessing different therapeutic approaches. Treatment is usually supportive and tailored towards the individual patient [3]. Asymptomatic patients may be managed without treatment [7]. Glucocorticoid has been reported to be beneficial in some case reports. Numerous other agents such as colchicine, azathioprine, cyclophosphamide, thalidomide, and tamoxifen have been used with varying success [3,4,13,20,22]. In the study by Akram et al., Tamoxifen and steroids were used in 20 patients, and 60% improved, suggesting that symptomatic patients may benefit from that regimen [3]. Surgery is reserved for obstructive complications, although resection may

be limited by vascular involvement [1,22]. Treatment of chylous ascites in sclerosing mesenteritis is limited to treating the primary cause, as there have been no specific recommendations made in the literature. Further longitudinal studies are warranted to fully understand and treat this unusual anomaly.

Conclusions

The nonspecific clinical features and rarity of this disorder makes diagnosing sclerosing mesenteritis extremely challenging. A high index of suspicion is essential for prompt detection, and the diagnosis of sclerosing mesenteritis requires careful correlation of clinical, radiologic, and histologic findings. We described a rare case of sclerosing mesenteritis leading to chylous ascites, small bowel perforation, and, subsequently, the death of our patient.

Conflict of interest

None.

Informed consent was obtained for this case report.

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