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CASE REPORT | BILIARY

Sclerosing Mesenteritis Presenting as Hilar Cholangiocarcinoma Causing Recurrent Chylous Ascites and Gastric Outlet Obstruction

Thomas Hoang, MD[1], Adnan Karavelic, MD, FRCPC[2], Aswathy Sunil, MD[3], Timothy Murray, MD, FRCPC[3], Jennifer Telford, MD, FRCPC[4], Elizabeth Clement, MD, FRCSC[5], and Anthony Gador, MD, FRCPC[1]

[1]Department of Medicine, Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada
[2]Department of Pathology and Laboratory Medicine, Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada
[3]Department of Radiology, Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada
[4]Division of Gastroenterology, Department of Medicine, Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada
[5]Division of General Surgery, Department of Surgery, Faculty of Medicine, University of British Columbia, Vancouver, British Columbia, Canada

ABSTRACT

Sclerosing mesenteritis is a rare spectrum of disease that results in chronic inflammation and fibrosis of the abdominal mesentery. Its heterogeneous presentation, nondiagnostic imaging, and pathology findings make diagnosis challenging. In this study, we present the first reported case of hilar sclerosing mesenteritis resulting in chylous ascites, gastric outlet obstruction, and duodenal perforation. Diagnosis was made after extensive investigations including computed tomography imaging, magnetic resonance cholangiopancreatography, positron emission tomography scan, bidirectional endoscopy, endoscopic ultrasound, and diagnostic laparoscopy. While initially mistaken for hilar cholangiocarcinoma, the patient has significant clinical improvement with corticosteroids and now remains in symptomatic and radiographic remission on low-dose prednisone and tamoxifen.

KEYWORDS: sclerosing mesenteritis; cholangiocarcinoma; chylous ascites; gastric outlet obstruction

INTRODUCTION

Sclerosing mesenteritis (SM) is a rare benign condition marked by chronic inflammation and fibrosis of the intestinal mesentery.[1–4] Its etiology is mostly idiopathic, though risk factors include male sex, older age, autoimmune disease, and abdominal surgery.[5] Patients often present with abdominal pain, bloating, distension, and weight loss, but it can also be an asymptomatic incidental finding.[1] Complications occur in up to 24% of cases, including small bowel obstruction, chylous ascites, and mesenteric vein thrombosis.[1] SM in the hepatobiliary or peripancreatic areas can mimic cholangiocarcinoma or pancreatic cancer.[6,7] Diagnosis is challenging, as symptoms are nonspecific and imaging findings often unclear. Treatment remains sparse and is based on retrospective studies[5,8,9] In this study, we report a case of porta hepatis SM presenting as a hilar mass, mimicking cholangiocarcinoma.

CASE REPORT

A 59-year-old White man presented with 1 month of abdominal swelling, pain, and reduced oral intake, alongside a 50-pound weight loss over 10 months. His medical history included hypertension, dyslipidemia, alcohol use disorder in remission, and 3 prior episodes of resolved moderate-to-severe alcoholic pancreatitis.

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Correspondence: Thomas Hoang, MD (thomash1@student.ubc.ca).

Computed tomography (CT) revealed soft tissue thickening at the porta hepatis, large-volume ascites, and chronic superior mesenteric vein thrombosis (Figure 1). His abdomen was diffusely tender and distended, without palpable masses. Paracentesis yielded 500 mL of cloudy ascites. Fluid analysis showed a low serum albumin-ascites gradient, with 2,784 $\times$ 10^3 cells/L (57% neutrophils, 33% lymphocytes), lactate dehydrogenase (138 U/L), glucose (6.7 mmol/L), and lipase (5 U/L). Cultures and cytology were negative for malignancy. Gamma-glutamyltranspeptidase (85 U/L) and alkaline phosphatase (177 U/L) were mildly elevated; other tests were unremarkable.

Magnetic resonance cholangiopancreatography revealed ill-defined soft tissue thickening at the porta hepatis with mild biliary duct dilatation (Figure 1). Endoscopic ultrasound biopsy of the lesion was negative for malignancy but showed a chronic lymphohistiocytic infiltrate (Figures 1 and 2). Multiple IgG4 stains and serum IgG4 tests were negative. Large-volume paracenteses revealed no malignant cells, and autoimmune serologies and tumor markers were negative.

The patient developed significant gastric outlet obstruction within 1 month of his admission due to extrinsic compression by the hilar mass. Bidirectional endoscopy revealed no intraluminal pathology, but extrinsic duodenal compression leading to mucosal edema was noted. His portal pressure gradient (2 mm Hg) ruled out portal hypertension, and liver biopsy was negative for parenchymal disease. Positron emission tomography scan showed hypermetabolism at the porta hepatis lesion (Figure 1).

Initially diagnosed with a presumed cholangiocarcinoma, he was started on palliative dexamethasone 4 mg daily, leading to rapid symptomatic relief. A follow-up CT scan showed resolution of ascites and mass shrinkage. However, he developed a contained duodenal perforation adjacent to the lesion, which was managed nonoperatively.

Three months later, he was readmitted with recurrent gastric outlet obstruction after stopping steroids. CT imaging showed recurrence of the porta hepatis lesion, with no malignancy on endoscopic ultrasound-guided biopsy. Ascites fluid now showed chylous ascites with a lymphocytic-predominant nucleated cell count of 572 $\times$ 10^6 cells/L and elevated triglycerides/chylomicrons. Diagnostic laparoscopy confirmed large-volume grossly chylous ascites (Figure S1, http://links.lww.com/ACGCR/A38) and omental inflammation (Figure 2) and neovascularization. A tapering course of high-dose prednisone resulted in symptomatic resolution over 4 days. After excluding other causes, a diagnosis of hilar SM was made. At 9-month follow-up, the patient remained in remission on tamoxifen and low-dose prednisone at 2.5 mg daily.

DISCUSSION

In this study, we present a rare hilar variant of SM, mimicking cholangiocarcinoma and resulting in chylous ascites, gastric outlet obstruction, and small bowel perforation.

SM is a chronic fibroinflammatory disease of the abdominal mesentery that is histopathologically characterized by chronic inflammation, fibrosis, fat necrosis, and lipodystrophy.[1–3] It is

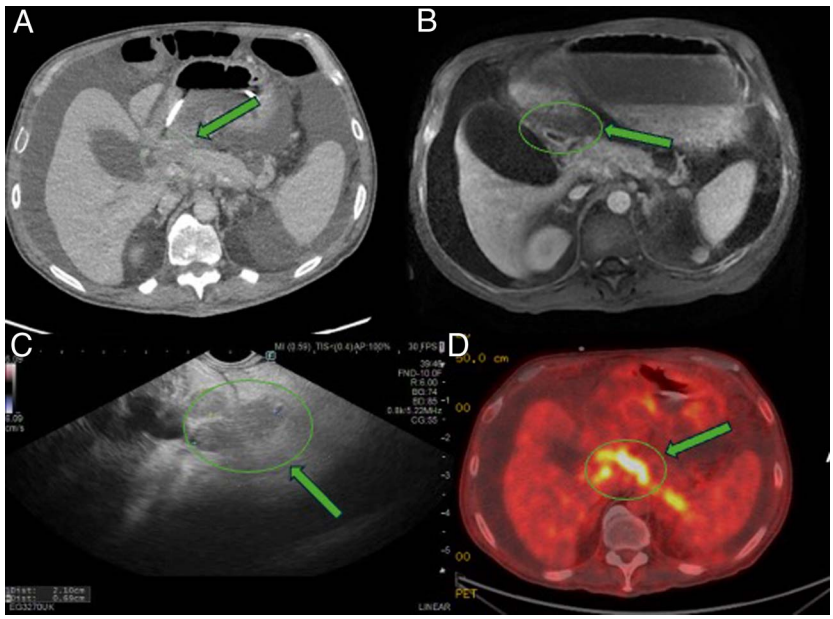

Figure 1. Cross-sectional imaging reveals an ill-defined soft tissue density (green arrow) around the porta hepatis on both (A) computed tomography and (B) magnetic resonance cholangiopancreatography. Nasogastric tube in situ for decompression of gastric outlet obstruction. Background large volume ascites is noted. (C) Endoscopic ultrasound reveals an ill-defined soft tissue mass with associated lymphadenopathy. (D) PET-computed tomography demonstrates overlapping fluorodeoxyglucose avidity. PET, positron emission tomography.

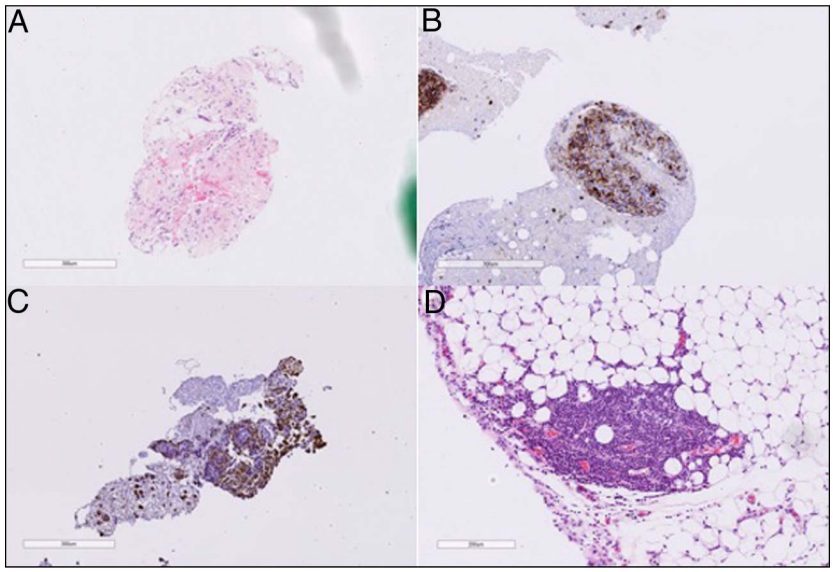

Figure 2. Endoscopic ultrasound-guided fine-needle biopsy at 10× magnification of the mass demonstrates (A) lesional tissue comprising a chronic lymphohistiocytic infiltrate (dark purple). The sample is highly positive for (B) $CD45^+$ and (C) $CD68^+$, markers of lymphocytes and histiocytes specifically (dark brown stain on immunohistochemistry). A similar infiltrate is observed in the (D) omental biopsy (dark purple), demonstrating chronic panniculitis.

rare, with an estimated incidence of 0.6%-1% and commonly affects men in the fifth to seventh decades of life, as demonstrated in our case.[3,10] Risk factors have included autoimmune disease and abdominal surgery, and we postulate that disease pathogenesis in this patient could have been triggered by immune/tissue repair dysregulation from his previous acute pancreatitis episodes.[2]

Depending on disease location, there is heterogeneity in clinical presentation. As in our case, abdominal pain, distension, and weight loss are common presenting symptoms.[1,10] In a previous review of 92 patients, 10% of cases were incidental findings.[1] While common in the small bowel (69%), previous reports have identified colon, omental, and pelvic involvement.[10]

Complications have included small bowel obstruction (24%), chylous ascites (14%), and superior mesenteric vein thrombosis (3%), all of which were present in our case.[1,11] Mechanistically, our case demonstrates that the extrinsic compression by the fibroinflammatory mass can result in gastric outlet obstruction. Although the hilum was the main region affected, the presence of chylous ascites suggests possible disease extension into the abdominal lymphatics. It is unclear what caused the bowel perforation, but we presume this was due to rapid regression of the infiltrate from the duodenum following steroids.

Though a benign process, SM is often misdiagnosed as a malignancy, resulting in significant patient distress and unnecessary invasive diagnostic interventions.[6,11] In a previous case series, 5 of 6 patients with peripancreatic SM were misdiagnosed with pancreatic cancer and 3 underwent partial resections.[6] Porta hepatis involvement is even rarer, and to our knowledge, we are only the second reported case of hilar SM.[7] In both cases, cholangiocarcinoma was the initial presumed diagnosis, though here we show for the first time that hilar SM can also result in chylous ascites and intestinal obstruction.

Previous retrospective studies have recommended tapering oral corticosteroids in conjunction with long-term tamoxifen as the mainstay of SM treatment.[1] In patients with tamoxifen contraindications, colchicine, azathioprine, and thalidomide have also been recommended, based on limited evidence, as alternative steroid-sparing agents.[1–3,12] Recent case reports have also highlighted steroid-free remission with tumor necrosis factor-a and IL-23 blockade.[8,9] Although the disease can be asymptomatic and self-limited, patients are at high risk of complications and should be closely monitored for progression.[2]

In conclusion, hilar SM represents a rare presentation of a rare disease that easily mimics malignancy resulting in heterogeneous complications. Our study highlights the need for broader recognition and supplements existing data on medical therapy, helping clinicians appropriately diagnose and manage this disease process.

DISCLOSURES

Author contributions: T. Hoang drafted the manuscript and reviewed the literature. A. Gador is the article guarantor, and conceptualized/supervised the study. A. Sunil, T. Murray, A. Karavelic, E. Clement, and J. Telford provided images, corresponding interpretation, and consultative expertise. All authors provided critical revisions of the manuscript.

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Informed consent was obtained for this case report.

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REFERENCES

1. Akram S, Pardi DS, Schaffner JA, Smyrk TC. Sclerosing mesenteritis: Clinical features, treatment, and outcome in ninety-two patients. *Clin Gastroenterol Hepatol.* 2007;5(5):589–24.
2. Saha B, Tome J, Wang XJ. Sclerosing mesenteritis: A concise clinical review for clinicians. *Mayo Clin Proc.* 2024;99(5):812–20.
3. Green MS, Chhabra R, Goyal H. Sclerosing mesenteritis: A comprehensive clinical review. *Ann Transl Med.* 2018;6(17):336.
4. Sharma P, Yadav S, Needham CM, Feuerstadt P. Sclerosing mesenteritis: A systematic review of 192 cases. *Clin J Gastroenterol.* 2017;10(2):103–11.
5. Saha B, Tome J, Wang XJ. Sclerosing mesenteritis: A concise clinical review for clinicians. *Mayo Clin Proc.* 2024;99(5):812–20.
6. Scudiere JR, Shi C, Hruban RH, et al. Sclerosing mesenteritis involving the pancreas: A mimicker of pancreatic cancer. *Am J Surg Pathol.* 2010;34(4): 447–53.
7. Medina-Franco H, Listinsky C, Mel Wilcox C, Morgan D, Heslin M. Concomitant sclerosing mesenteritis and bile duct fibrosis simulating Klatskin's tumor. *J Gastrointest Surg.* 2001;5(6):658–60.
8. Rothlein LR, Shaheen AW, Vavalle JP, et al. Sclerosing mesenteritis successfully treated with a TNF antagonist. *BMJ Case Rep.* 2010;2010: bcr0720103145.
9. Byriel B, Walker M, Fischer M. Sclerosing mesenteritis complicated with mesenteric lymphoma responsive to ustekinumab. *ACG Case Rep J.* 2022; 9(5):e00757.
10. Emory TS, Monihan JM, Carr NJ, Sobin LH. Sclerosing mesenteritis, mesenteric panniculitis and mesenteric lipodystrophy: A single entity? *Am J Surg Pathol.* 1997;21(4):392–8.
11. Lim HW, Sultan KS. Sclerosing mesenteritis causing chylous ascites and small bowel perforation. *Am J Case Rep.* 2017;18:696–9.
12. Ginsburg PM, Ehrenpreis ED. A pilot study of thalidomide for patients with symptomatic mesenteric panniculitis. *Aliment Pharmacol Ther.* 2002; 16(12):2115–22.