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Anaplastic Lymphoma Masquerading as Sclerosing Mesenteritis: A Case Report

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Abstract

Introduction—Sclerosing mesenteritis is a rare fibroinflammatory disease of unknown pathogenesis and etiology. Although more commonly benign in its clinical course, sclerosing mesenteritis can mimic certain malignant disorders, such as lymphoma.

Case Report—This report describes the case of a previously healthy young woman who develops an acute illness thought to be related to sclerosing mesenteritis following an extensive diagnostic work-up including biopsy sampling, which was negative for malignancy. However, despite aggressive medical care, the patient passed away. Autopsy revealed extensive inflammation secondary to anaplastic lymphoma.

Conclusion—The case very graphically highlights an important clinical observation that lymphoma can masquerade as sclerosing mesenteritis.

Keywords

Lymphoma; Sclerosing mesenteritis

Introduction

Sclerosing mesenteritis is a rare fibroinflammatory disorder of unknown pathogenesis and etiology. Although more commonly benign in its clinical course, sclerosing mesenteritis can also mimic more aggressive diseases. We describe a case of lymphoma-related mesenteritis

and highlight the importance of diligent investigation and awareness of disease associations when sclerosing mesenteritis is suspected.

Case Presentation

The patient was a previously healthy 29-year-old woman who presented with a 6 month history of worsening jaundice, diffuse abdominal pain, and significant weight loss. Her initial work-up was unremarkable, however her symptoms persisted. Soon thereafter, she developed worsening fatigue and early satiety. Due to her worsening symptoms, a colonoscopy and esophagogastroduodenoscopy (EGD) were performed to further investigate intraluminal pathology. Her colonoscopy was visually unremarkable, and her EGD revealed grade one non-bleeding esophageal varices, gastritis, and duodenal sweep edema. Further work-up included CT scan of her abdomen which showed a soft tissue prominence surrounding the pancreatic head and duodenum. Further characterization with MRI/MRV of the abdomen and pelvis revealed extensive inflammation of the periduodenal and peripancreatic fat with a hypo-enhancing lesion surrounding the pancreatic head and duodenum in addition to a chronic thrombus within the portal vein. Attempts were made to sample intra-abdominal tissue via CT and endoscopic ultrasound-guided aspiration, yet these were unsuccessful due to extensive inflammation and narrowing of the duodenum, respectively. She therefore underwent open laparotomy with biopsies obtained from her duodenum, gastrohepatic ligament, and retrogastric lymph nodes. These samples were negative for malignancy but did show evidence of acute inflammation and extensive amounts of fibroadipose tissue and fat necrosis. Given the unclear etiology of her symptoms, she was transferred to our tertiary care center for further evaluation. Initial work-up was extensive and included a multitude of rheumatologic, autoimmune, and infectious data, which were unremarkable. Extensive oncologic data was non-specific (Table 1). Given this non-specificity with image and pathological findings consistent with a fibroinflammatory disorder, sclerosing mesenteritis was our leading diagnosis. She was started on steroid treatment (prednisone 40 mg every day). We were unable to use tamoxifen in the setting of known clot.

The patient's symptoms of abdominal pain, decreased appetite, and yellowing of her skin persisted despite our therapy. Further imaging revealed worsening extensive abdominal inflammation in addition to gastrohepatic, periceliac, peripancreatic, portocaval, and mesenteric lymphadenopathy (Figure 1). She subsequently developed candida tropicalis and vancomycin-resistant enterococcus (VRE) infection in the blood and was treated appropriately with antimicrobials. She continued to decompensate and on day 36 of her hospitalization developed acute respiratory failure with cardiovascular collapse requiring intubation and maximum dose vasopressor agents. Shortly thereafter, the patient expired.

Autopsy revealed multiple intra-abdominal adhesions involving the visceral organs and peritoneum. Anaplastic lymphomatous involvement of abdominal soft tissue, adrenal glands, thoracic/abdominal/retroperitoneal lymph nodes, liver, portal vein, pancreas, duodenum, colon, lungs, and heart was identified. A remarkable amount of fibrotic tissue encased the first part of the duodenum as it entered the retroperitoneum, the pancreas, portal vein, and bile duct. There was also peri-aortic fibrosis extending from the level of the renal arteries to the bifurcation of the iliac vessels, causing obstruction of bilateral renal vessels, ureters, superior and inferior mesenteric arteries, and inferior vena cava.

Discussion

Although the majority of documented cases of sclerosing mesenteritis appear to be idiopathic in nature [1, 2], various associations have been noted. Among these, malignancy has been observed on numerous instances. Specifically, lymphoma is often implicated [3–5].

Furthermore, the CT and gross appearance of sclerosing mesenteritis and malignant processes such as lymphoma can be identical. In the largest reported patient series, one quarter of cases of sclerosing mesenteritis had findings indistinguishable from malignancy on imaging and gross inspection [1]. Therefore, sclerosing lymphoma can be misdiagnosed as a non-malignant fibroinflammatory disorder, such as sclerosing mesenteritis [5–7], and initial treatment with steroids could mask the proper diagnosis. With these similarities in mind, the importance of histologic examination becomes clear. However, insufficient tissue sampling could be misleading. In our patient, additional tissue sampling was not performed due to her tenuous clinical status. Extensive duodenal narrowing precluded endoscopic biopsies. Indeed, initial attempts at EUS and CT-guided biopsy were unsuccessful in our patient. There are limited reports of a possible role for positron emission tomography (PET) in distinguishing between malignancy and benign fibroinflammatory disorders such as retroperitoneal fibrosis [8, 9]. No PET scan was performed in our patient.

Despite lack of co-morbidities, patients with anaplastic lymphoma can deteriorate rather quickly. This stems from the ability of lymphoma to invade multiple organ systems. During her hospitalization, our patient developed liver, pulmonary, and cardiac disease secondary to her underlying malignancy, in addition to VRE and fungal infection in the blood likely related to malignancy-associated immunosuppression. This further highlights the need for early initiation of appropriate therapy.

In sum, our patient was a previously healthy 29 year old with a fatal progressive intra-abdominal fibroinflammatory process that mimicked sclerosing mesenteritis. Her final diagnosis of sclerosing anaplastic lymphoma was obtained by autopsy despite thorough evaluation during her clinical course. Gross findings on autopsy revealed a remarkable amount of intra-abdominal and pelvic fibrosis involving multiple organs, viscera, and blood vessels. Our patient's case highlights the diagnostic challenges and pitfalls that may be involved with fibroinflammatory disorders such as sclerosing mesenteritis and describes an exceptionally severe presentation of a malignant sclerosing lymphoma. While imaging characteristics may be relatively sensitive for sclerosing mesenteritis, their specificity is less clear. Ultimately, tissue specimens remain imperative for accurate and timely diagnosis, while recognizing that sample error may be misleading. This is especially true for a patient not responding to adequate treatment for the presumed diagnosis. Diligent investigation and awareness of the association and clinical similarities to malignancy is essential.

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Fig. 1.

CT scan evidence of extensive inflammation and adenopathy. **a** Axial CT image through the mid abdomen shows diffuse fat stranding in the root of the mesentery (*yellow arrows*) and enlarged mesenteric lymph nodes (*yellow arrowhead*). **b** Higher cut through the porta hepatis shows extension of the mesenteric fat stranding into the region of the falciform ligament insertion site (*long yellow arrow*) and enlarged lymph nodes in the portocaval region (*short yellow arrow*). Thrombosed main portal vein (*yellow arrowhead*). Biliary stent in the CBD (*short white arrow*)

Table 1

Data against and supporting a diagnosis of malignancy

Parameter (against malignancy)	Patient's value
CA 19.9	8 (wnl)
CEA	<1 (wnl)
Bone marrow aspirate	Non-diagnostic
JAK2 mutation	Negative
BCR-ABL mutation	Negative
Tissue biopsies	Negative for malignancy
Parameter (supporting malignancy)	Patient's value
White blood cell count	28,000 (elevated)
Hemoglobin	9.3 (iron studies consistent with anemia of chronic disease)
Platelet count	826 (elevated)
CA 125	55.3 (elevated)
LDH	1775 (elevated)
Image findings	Mass, lymphadenopathy, and portal vein thrombus

Wn/ within normal limits