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CASE REPORT | PANCREAS

Lupus Pancreatitis Masquerading as Pancreatic Cancer: A Rare Clinical Presentation

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ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune multisystemic inflammatory disease. SLE-associated pancreatitis is uncommon, and pancreatic cancer in SLE is very rare. Imaging findings in SLE with pancreatitis can mimic malignancy. Endoscopic ultrasound with fine-needle aspiration/biopsy can guide in the accurate diagnosis and management of SLE-associated pancreatitis.

KEYWORDS: systemic lupus erythematosus; endoscopic ultrasound-guided fine-needle aspiration; endoscopic retrograde cholangiopancreatography; pancreatitis; pancreatic cancer

INTRODUCTION

Systemic lupus erythematosus (SLE)-associated pancreatitis is rare. SLE can cause vasculitis of pancreatic vessels, which further leads to acute or subclinical pancreatitis.[1] Acute pancreatitis can occur several months or years after the diagnosis of SLE and can be the first presentation of SLE.[2–4] SLE-associated pancreatitis mimicking malignancy is very rare. This article will discuss the clinical presentation, diagnosis, and management of SLE-associated pancreatitis.

CASE REPORT

A 49-year-old Hispanic woman with a 6-year history of SLE, which presented with a malar rash and Hashimoto thyroiditis, was referred to the gastroenterology clinic for evaluation of abdominal pain with abnormal imaging. She described the abdominal pain as located in the epigastrium and burning in nature, with a pain score of 9 out of 10. The pain radiated to the back, and there were no specific aggravating or relieving factors. She had lost 90 pounds in the past year. She denied fever, nausea, and vomiting. She was admitted to the hospital for further evaluation and management of her symptoms. She reported that she was diagnosed with SLE approximately 6 years ago, but she stopped taking medications for SLE because of financial constraints. Social history included chronic tobacco use (15 pack-years), no alcohol use, and no illicit drug use. She had no family history of SLE. Surgical history includes cholecystectomy. She had no prior endoscopic retrograde cholangiopancreatography. Physical examination showed moderate-to-severe epigastric tenderness without any rebound tenderness. Laboratory results showed a white blood cell count of 4.5 k/μL (4–11 k/μL), hemoglobin 9.3 g/dL, platelet count 147 k/μL (165–400 k/μL), alkaline phosphate 346 IU/L (35–129 IU/L), aspartate transaminase 17 IU/L (5–37 IU/L), alanine transaminase 15 IU/L (5–41 IU/L), lipase 37 IU/L (13–60 IU/L), cholesterol 119 mg/dL (<200 mg/dL), low-density lipoprotein 61 mg/dL (<100 mg/dL), triglyceride 73 mg/dL (50–200 mg/dL), and calcium 9.4 mg/dL (8.8–10.5 mg/dL). Tumor markers, including alpha-fetoprotein and carcinoembryonic antigen, were normal, and CA 19-9 was elevated at 112 (<35 units/mL). Autoimmune markers showed positive for anti-nuclear antibody, anti-Smith antibody, and antinuclear ribonucleoprotein antibody. Anti-double-stranded DNA antibodies and rheumatoid factor were negative. The C3 complement level was 27 mg/dL (90–180 mg/dL), the C4 complement level was 5 mg/dL (10–40 mg/dL), C-reactive protein was 3.7 mg/dL (0–0.5 mg/dL), and serum immunoglobulin G4 (IgG4) was 33.4 (4–86 mg/dL). Computed tomography pancreatic protocol showed a 1.1 cm pancreatic head mass resulting in a dilated main pancreatic duct measuring up to 1.4 cm, suggesting pancreatic neoplasm with no evidence of gallstone (Figure 1). Endoscopic ultrasound (EUS) showed pancreatic atrophy, an isoechoic, heterogeneous, regular round lesion measuring 27.6 × 27.4 mm noted at the head of the pancreas with upstream dilation of the pancreatic duct. The fine-needle aspiration and biopsy (FNA/B) of the pancreatic

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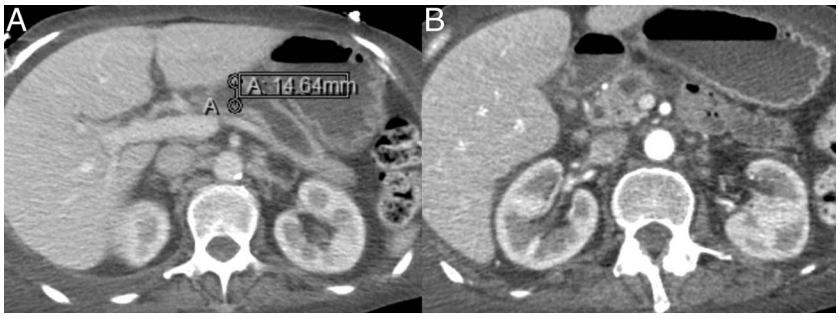

Figure 1. (A) CT pancreatic protocol showed a severely atrophic pancreas with a dilated main pancreatic duct measuring 1.4 cm in diameter. (B) CT pancreatic protocol showed a hypodense lesion at the pancreatic head measuring 1.1 cm. CT, computed tomography.

mass was performed (Figure 2). Pathology showed acute and chronic inflammation of pancreatic tissue and stained negative for IgG4. These results did not support autoimmune pancreatitis or malignancy. Endoscopic retrograde cholangiopancreatography (ERCP) showed a 2 cm-long moderate to high-grade stricture of the pancreatic duct at the head and neck region with marked dilation of the pancreatic duct in a saccular form with filling defects in the pancreatic duct at the body of the pancreas. The pancreatic duct was explored endoscopically using the SpyGlass (Boston Scientific) direct visualization system, but could not be advanced beyond the stricture. Pancreatic ductal brushing using a cytobrush was performed. A transpapillary plastic stent was placed into the main pancreatic duct across the stricture. The cytopathologic evaluation revealed only acute inflammatory debris.

Rheumatology was consulted, and she was started on prednisone 1 mg/kg (60 mg daily), and this dose was tapered over 7 months. The patient also received hydroxychloroquine and mycophenolate mofetil. She was then discharged home with the above-mentioned medicines and continued on weight-based dosing of pancrelipase. At 2-month follow-up, her abdominal pain improved and her C-reactive protein decreased to 0.6 mg/dL (0–0.5 mg/dL). A repeat EUS performed 3 months later showed a normal pancreatic duct with moderate pancreatic atrophy. A repeat ERCP showed marked improvement of the pancreatic duct stricture with resolution of upstream dilation of the pancreatic duct. It was decided to replace the pancreatic duct stent to prevent the recurrence of the stricture. A repeat computed tomography pancreatic protocol performed at 3 months also showed marked improvement of pancreatic duct dilation and complete resolution of the mass at the head seen on prior imaging (Figure 3).

DISCUSSION

Acute pancreatitis occurs in approximately 2%–8% of patients with SLE and usually in the setting of active SLE.[5] The prevalence of chronic pancreatitis with SLE is not known. The pathogenesis of pancreatitis in SLE is not clearly understood. It is hypothesized that vasculitis from autoantibodies, vascular occlusion from immune complex precipitation, antiphospholipid antibodies resulting in microthrombosis, viral infections, and drug-induced toxicity can precipitate pancreatitis in SLE.[6]

The risk of pancreatic cancer with SLE is not known.[7] The plausible mechanism for the association of SLE and pancreatic

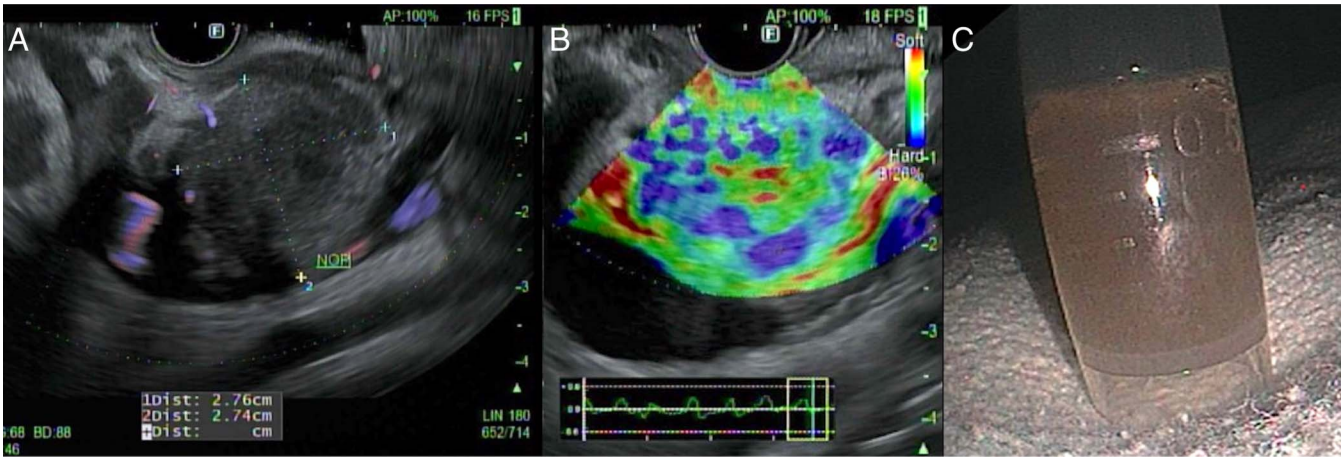

Figure 2. (A) EUS showed an isoechoic, heterogeneous, regular round lesion measuring 27.6 × 27.4 mm at the head of the pancreas. (B) Elastography showed heterogeneous necrotic tissue with septations. (C) Fine-needle aspiration of the fluid showed turbid color suggestive of an inflammatory process. EUS, endoscopic ultrasound.

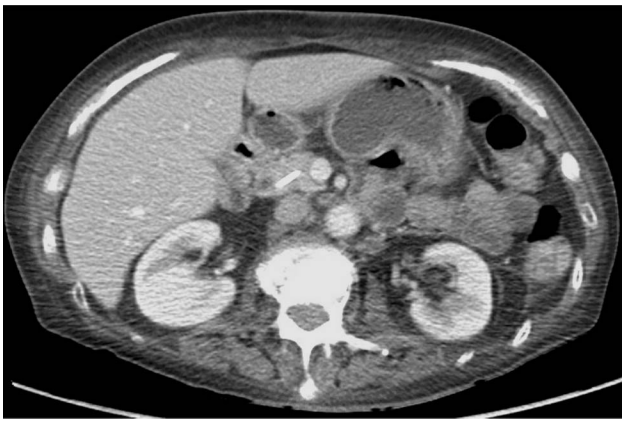

Figure 3. Follow-up CT in 3 months showed a pancreatic duct stent in place and no pancreatic head mass. CT, computed tomography.

cancer is chronic inflammation.[8] Another possible mechanism is an excess autoantibody effect in SLE, which can lead to DNA damage and impairment of DNA repair and further influence the risk of cancer.[9] Several case reports showed an association between SLE and pancreatic cancer (Table 1).[10,11]

To our knowledge, there is only 1 prior case report of SLE with chronic pancreatitis-mimicking malignancy.[16] Abdominal computed tomography showed a dilated common bile duct because of compression from an enlarged pancreatic head mass. EUS with FNA of the pancreatic head mass revealed a benign inflammatory process. ERCP for biliary stenting was complicated by hemoperitoneum. Follow-up contrast-enhanced ultrasound of the pancreas showed a dilated pancreatic duct. He was treated with pancrelipase and low-dose methylprednisolone. It was unclear whether the dilated PD improved with SLE treatment.

In our patient, computed tomography pancreatic protocol showed a pancreatic head mass with the dilated pancreatic duct, but there was no biliary duct dilation. Other causes of pancreatitis, such as alcohol, gallstones, hypercalcemia, hyperparathyroidism, hypertriglyceridemia, were ruled out. EUS-FNA/B showed acute

Table 1. Reported case of SLE associated with chronic pancreatitis

Author (yr)	Age, sex	Duration of SLE (yrs)	Imaging	EUS/ERCP findings	Outcome
Borum et al[1] (1993)	24, F	9	CT showed multiple pancreatic calcification	ERCP showed multiple filling defects consistent with stones in a diffusely dilated pancreatic duct	Puestow procedure for relief of pain
Borum et al[1] (1993)	34, F	11	CT showed a 3 cm pseudocyst pancreas and inflammatory changes	Not performed	Distal pancreatectomy with splenectomy
Hortas et al[12] (1995)	61, F	1	CT showed mild dilation of the pancreatic and bile duct	ERCP showed mild dilation of the pancreatic and bile duct	No surgery
Hortas et al[12] (1995)	51, F	14	CT showed pancreatic calcification	ERCP showed dilatations and stenoses of the main pancreatic duct, with normal bile ducts	No surgery
Penalva et al[13] (2003)	14, F	8	CT scan showed the presence of pancreatic calcifications and small pseudocysts in the head and tail of the gland	ERCP showed an irregular main pancreatic duct with total occlusion at the union between the body and tail of the pancreas	Splenopancreatectomy
Izzedine et al[14] (2005)	59, F	9	CT showed pseudocyst in the tail of pancreas and calcification	ERCP showed an enlarged head of the pancreas and mildly dilated pancreatic duct	Supportive care
Gutierrez et al[15] (2008)	26, F	3	Magnetic resonance cholangiopancreatography (MRCP) showed an irregular and dilated Wirsung duct and 3 pseudocysts	Endoscopic ultrasonography (EUS) confirmed the findings of chronic pancreatitis	External drainage of the pelvic pseudocyst
Ardesia et al[16] (2014)	42, F	Not mentioned	Abdominal CT showed an enlarged head of the pancreas, dilated CBD, and intrahepatic biliary dilation	EUS FNA histology consistent with the inflammatory process. ERCP complicated by hemoperitoneum	Supportive care with pancrelipase and steroids
Our case	49, F	6	CT pancreatic protocol showed a 1.1 cm pancreatic head mass resulting in a dilated main pancreatic duct	ERCP showed 2 cm-long moderate to high-grade stricture at the pancreas duct of the head and neck region	PD stent was placed. Resumed SLE meds

CBD, common bile duct; CT, computed tomography; EUS, endoscopic ultrasound; ERCP, endoscopic retrograde cholangiopancreatography; F, female; PD, pancreatic duct; SLE, systemic lupus erythematosus.

and chronic inflammation and was negative for IgG4. ERCP-directed pancreatic ductal brushings showed acute inflammatory debris. Follow-up imaging showed marked improvement of pancreatic duct dilation and resolution of the pancreatic head mass. The patient was continued on pancrelipase with meals and was counseled on compliance of SLE medications.

SLE-associated pancreatitis should be considered in the differential diagnosis of pancreatic mass with or without biliary and pancreatic duct dilation. EUS with FNA/B and ERCP with or without pancreatoscopy and stent placement can aid in the accurate diagnosis and effective management of SLE-associated pancreatitis. A multidisciplinary team-based approach is the key to the appropriate and accurate diagnosis and management of SLE-associated pancreatitis.

DISCLOSURES

Author contributions: G. Lanke and B. Songtanin wrote the original draft. K. Das reviewed and edited the manuscript. B. Songtanin is the article guarantor.

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