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Case Report

Autoimmune pancreatitis

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ABSTRACT

We report the case of a 70-year-old male with chronic abdominal pain, who presented with increased intensity of the pain, and was initially diagnosed and treated for acute biliary pancreatitis. However, the symptoms persisted after hospital discharge, and he was subsequently studied with cholangio-MRI, abdominal CT scan, and ERCP, which revealed dilation of the biliary tract and a mass in the head of the pancreas. An excisional biopsy of the lesion showed chronic inflammation with severe sclerosing fibrosis and a lymphoplasmacytic infiltrate. A diagnosis of autoimmune pancreatitis was made, and the patient was initiated on azathioprine with good clinical response.

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Pancreatitis autoinmunitaria

RESUMEN

Presentamos el caso de un varón de 70 años con dolor abdominal crónico, que se presenta con aumento de este; se le diagnostica y se lo trata inicialmente como cuadro de pancreatitis biliar. A pesar del manejo inicial, los síntomas persisten después de la primera hospitalización, y se estudia con tomografía computarizada de abdomen, colangiorresonancia y colangiopancreatografía retrograda endoscópica, estudios en los cuales se detecta dilatación de la vía biliar y masa en la cabeza del páncreas. Se llevó a cabo biopsia excisional, que demuestra inflamación crónica con intensa fibrosis e infiltrado linfocitario. Se hace diagnóstico de pancreatitis autoimmunitaria y se inicia azatioprina, con adecuada respuesta.

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Introduction

Autoimmune pancreatitis is a rare disease that presents with clinical, serologic, and histologic characteristics of a defined autoimmune process that usually responds to the use of steroids.¹ Yoshida introduced the term in 1995, replacing others that did not properly define the disease such as primary chronic pancreatitis, sclerosing chronic pancreatitis, duct destructive chronic pancreatitis, and sclerosing lymphoplasmacytic pancreatitis.²⁻⁶

Most information comes from Japan, where the number of documented cases has increased, probably due to improvements in diagnostic techniques and greater awareness of the disease. Cases have been reported in Europe, USA, and

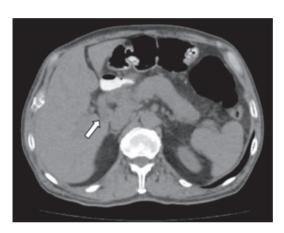


Figure 1 – Abdominal computed tomography scan with contrast. Notice mass in the head of the pancreas (arrow).

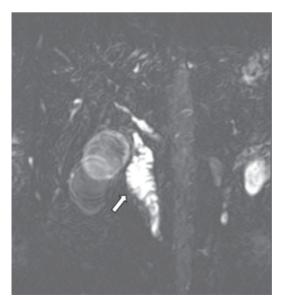


Figure 2 – Cholangio-magnetic resonance imaging showing dilation of the pancreatic duct (arrow).

South Korea, which suggest that autoimmune pancreatitis has a worldwide prevalence. This disease may present as an isolated primary entity or it can be associated to other autoimmune diseases such as primary sclerosing cholangitis, primary biliary cirrhosis, retroperitoneal fibrosis, rheumatoid arthritis, sarcoidosis and Sjögren's syndrome.^{7,8}

Case report

We report the case of a 70-year-old male with a 10 month history of upper abdominal pain, jaundice, dark urine, pale stools, night sweats, generalized pruritus and weight loss of 28kg and no history of autoimmune disease. An initial diagnosis of biliary pancreatitis was made, and an endoscopic retrograde cholangiopancreatography (ERCP) was attempted, without success. Labs showed normal transaminase levels, direct hyperbilirubinemia, and elevated levels of alkaline phosphatase (972 UI/L), amylase (174 U/L) and lipase (1314 U/L). Hepatobiliary ultrasound showed a distended gallbladder with thickened walls, dilated intra and extra-hepatic ducts, a 17 mm dilation of the common bile duct, and a distal image that suggested biliary sludge associated with a diffuse growth of the pancreas. A new ERCP was attempted without success so a computed tomography (CT) scan was performed. The scan showed dilation of the biliary tree and a mass in the head of the pancreas (figure 1). The main differential diagnoses that were initially considered were cholangiocarcinoma and pancreatitis (Balthazar B). Subsequently, a cholangio-magnetic resonance imaging (MRI) revealed an obstruction of the biliary duct at the level of the intra-pancreatic tract, which could correspond to a multi-focal cholangiocarcinoma (figure 2). The patient was finally taken to surgery for an exploratory laparotomy, and frozen section biopsy revealed a chronic inflammatory process. Further histological studies showed destruction of glandular architecture with severe sclerosing fibrosis and lymphoplasmacytic infiltrates (figure 3). The patient was started on azathioprine, with good response.

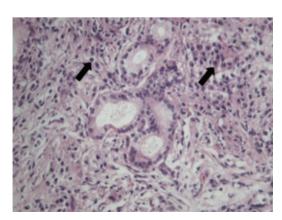


Figure 3 – H&E staining showing lymphoplasmacytic infiltrates in pancreatic parenchyma.

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