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(CASE REPORT)

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Autoimmune pancreatitis associated with Sjögren's syndrome: A case report

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Abstract

Autoimmune pancreatitis (AIP) is a rare but increasingly recognized entity. It is characterized by diverse diagnostic criteria, including pancreatic and extrapancreatic involvement, immunological abnormalities, and a therapeutic response to corticosteroids.

We report the case of Mrs. R.E.B, a 68-year-old woman followed for decompensated cirrhosis of autoimmune origin, specifically primary biliary cirrhosis, treated with ursodeoxycholic acid. She was admitted for acute epigastric pain associated with vomiting. The diagnosis of acute pancreatitis was made based on clinical presentation, elevated lipase levels, and abdominal CT findings consistent with pancreatitis. As part of the etiological workup, an abdominal ultrasound and a metabolic phospholipid profile were performed, both of which were negative. The investigation of an autoimmune etiology led to a workup including IgG4 levels, which were found to be three times the normal value. This was followed by MR cholangiopancreatography (MRCP), which revealed a "sausage-shaped" pancreas, supporting the diagnosis of autoimmune pancreatitis. The search for other systemic involvement revealed the presence of xerophthalmia and xerostomia upon questioning—symptoms the patient had previously overlooked. A labial salivary gland biopsy confirmed lymphocytic sialadenitis, consistent with Sjögren's syndrome. The suggestive findings on MRI, positive serology, and the association with Sjögren's syndrome led to the diagnosis of autoimmune pancreatitis (AIP). Corticosteroid therapy was initiated alongside symptomatic treatment for acute pancreatitis, leading to a remarkable clinical and biological improvement.

Keywords: Autoimmune Pancreatitis; Extra-Pancreatic Features; IGG4; Sjögren's Syndrome

1. Introduction

Autoimmune pancreatitis (AIP) is a rare but increasingly recognized condition. Clinical, serological, radiological, and particularly histological features allow for the differentiation between the two types of AIP. In addition to its pancreatic manifestations, AIP is often associated with other rare autoimmune diseases. In this review, we present a case of type 1 autoimmune pancreatitis, focusing on the various clinical and pathophysiological aspects of the disease, as well as the challenges in its diagnosis and management, particularly when it is associated with other rare autoimmune conditions.

2. Patient and Observation

A 68-year-old woman, followed since 2022 for cirrhosis secondary to primary biliary cirrhosis (PBC), was diagnosed based on the following criteria: Clinical: jaundice and pruritus; Biological: cholestasis; Immunological: positive anti-mitochondrial antibody serology. She had decompensated with ascites, treated with Aldactone 75 mg, and had hemorrhagic decompensation while on Carvedilol 6.25 mg. She was also receiving ursodeoxycholic acid at a dose of 13

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mg/kg/day. She was hospitalized in our department for epigastric stabbing pain, typical of pancreatitis, along with postprandial vomiting. General examination revealed a jaundiced patient who was mildly tachycardic, with abdominal examination showing epigastric tenderness and dullness in the flanks. Initial tests revealed a lipase level seven times above normal, a biological inflammatory syndrome, and liver function abnormalities, including cytolysis three times the normal and cholestasis six times the normal, with hypoalbuminemia. A CT scan (C+ C-) revealed an enlarged pancreas with loss of lobulations (Figure 1).

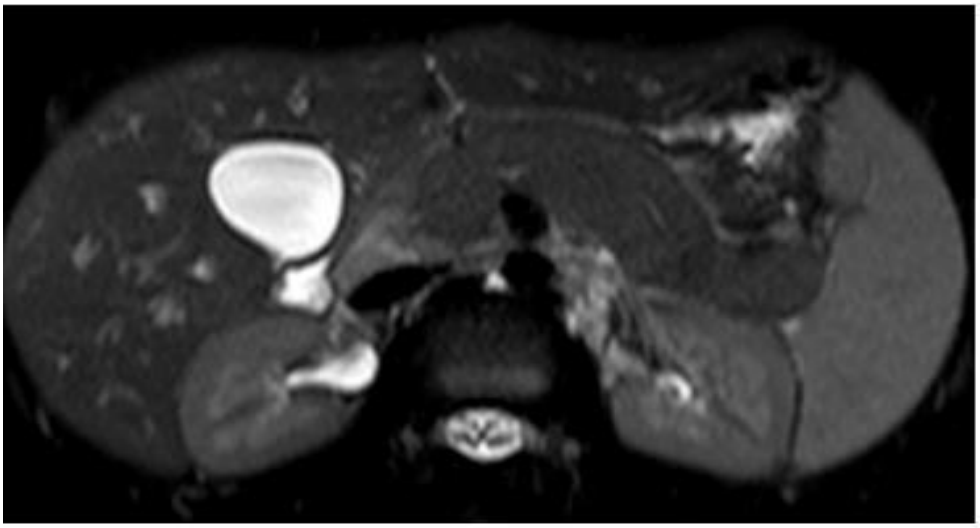

Figure 1 CT scan showing an enlarged pancreas with loss of lobulations.

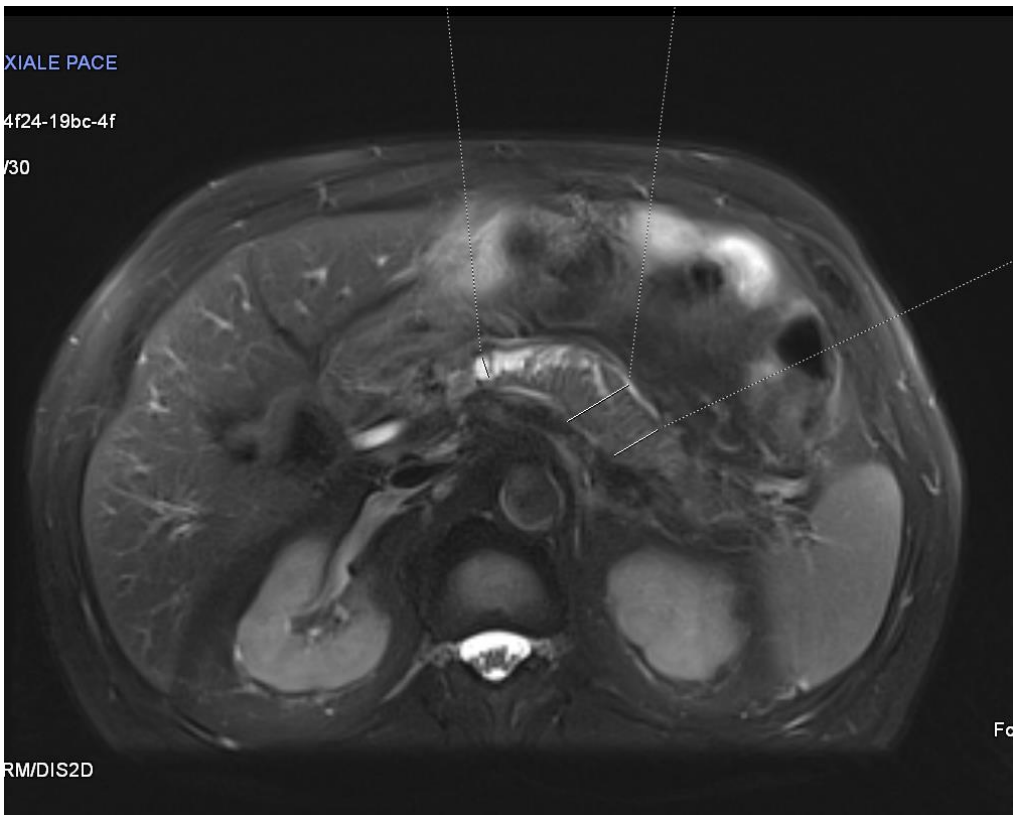

Figure 2 MRCP demonstrating a sausage-like appearance of the pancreas.

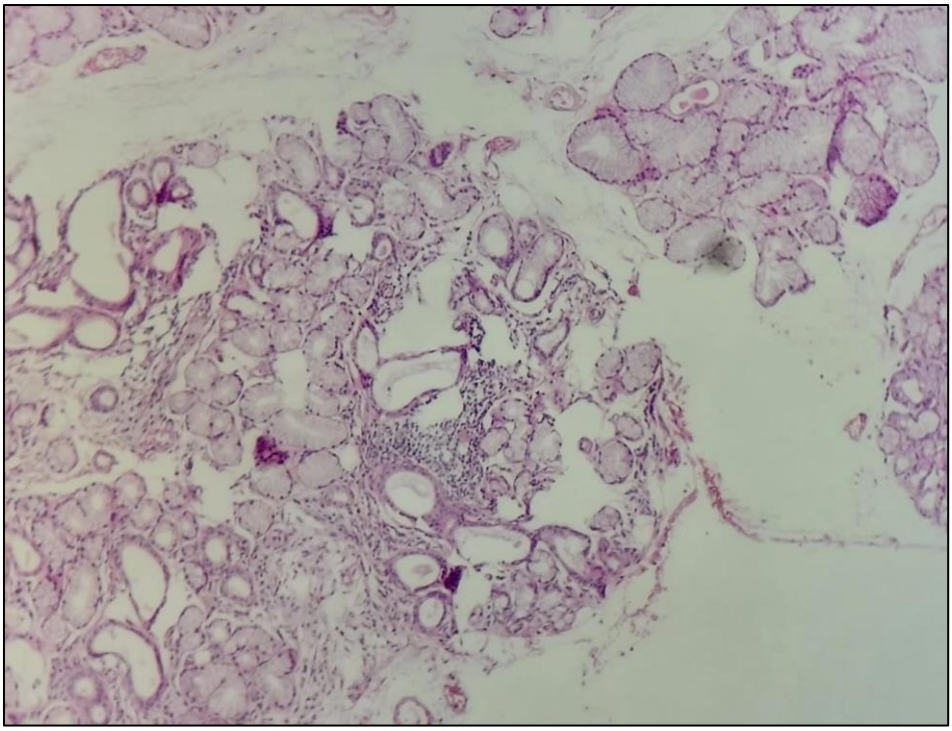

Figure 3-a Histological image of salivary gland biopsy's patient zoom x5

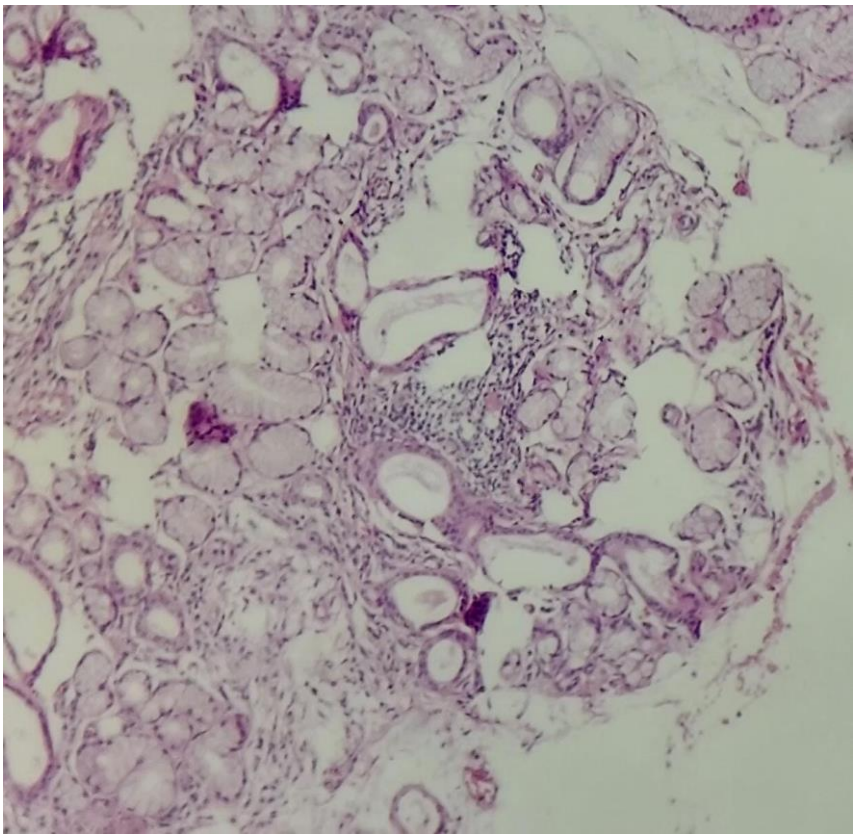

Figure 3-b Histological image of salivary gland biopsy's patient zoom x10

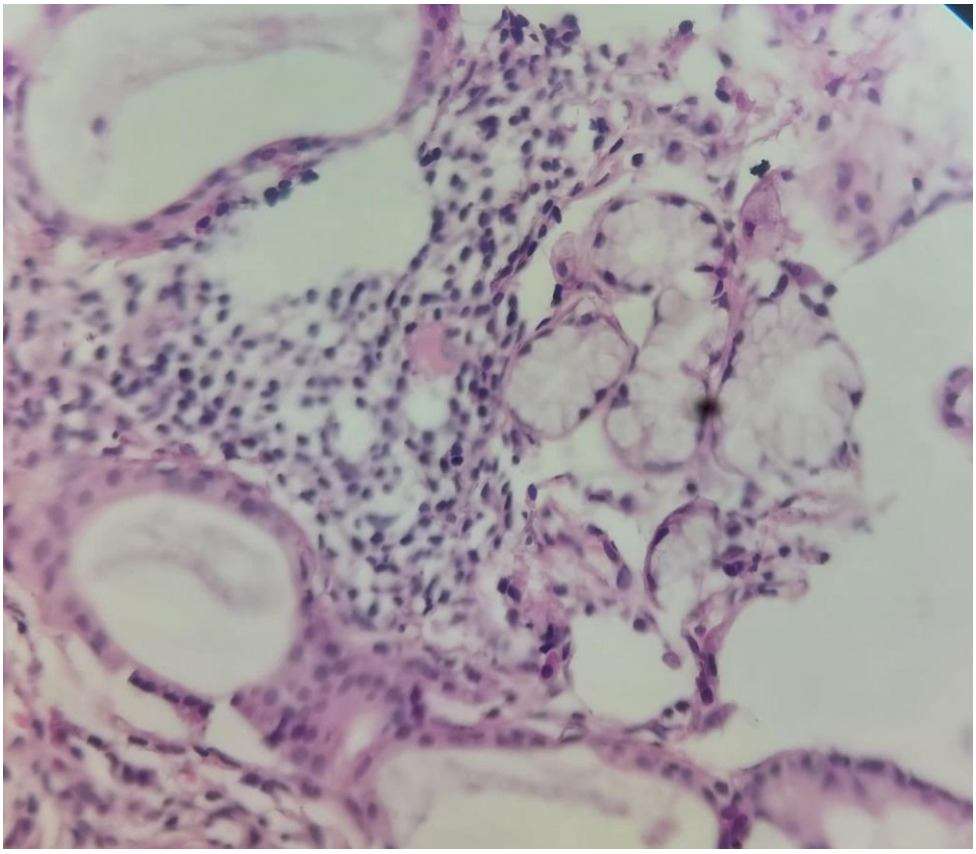

Figure 3-c Histological image of salivary gland biopsy's patient zoom x40

Figure 3 Histological image of the salivary gland biopsy showing an interstitial lymphocytic focus without ductal metaplasia made up of more than 50 lymphocytes ; (a- Z x5), (b- Z x 10), (c- Z x40)

The Balthazar score was stage C. The diagnosis of acute pancreatitis was confirmed. The patient was kept fasting, received cautious rehydration with albumin therapy, pain management according to the EVA pain scale, a single dose of injectable omeprazole, and preventive anticoagulation. The etiology workup revealed no signs of lithiasis on ultrasound, no history of alcoholism or iatrogenic causes, and no metabolic abnormalities (calcium-phosphate balance or triglycerides). Given the suspicion of primary biliary cholangitis, an autoimmune origin was considered, and IgG4 levels were found to be three times the normal value. This serology was complemented by an MRCP (figure 2), which showed an enlarged pancreas with loss of lobulations and a subtle T2 hypointense halo, giving it a sausage-like appearance.

The search for other systemic involvement revealed the presence of xerophthalmia and xerostomia upon questioning—symptoms the patient had previously overlooked. A labial salivary gland biopsy confirmed lymphocytic sialadenitis, consistent with Sjögren's syndrome (figure 3). The remaining investigations, including TSH levels and colonoscopy to screen for inflammatory bowel disease, were unremarkable. The suggestive findings on MRI, positive serology, and the association with lymphocytic sialadenitis led to the diagnosis of autoimmune pancreatitis (AIP). Corticosteroid therapy was initiated, resulting in a remarkable clinical and biological improvement. Follow-up at three and six months showed no signs of disease relapse.

Corticosteroid therapy was initiated for one month with an initial dose of 40 mg of prednisolone, followed by a taper of 5 mg per week over three months. This treatment resulted in significant clinical and biological improvement, notably the resolution of pancreatic pain and normalization of liver function tests. Follow-up assessments at 3 and 6 months showed no signs of relapse, with a favorable response according to the Paris II criteria.

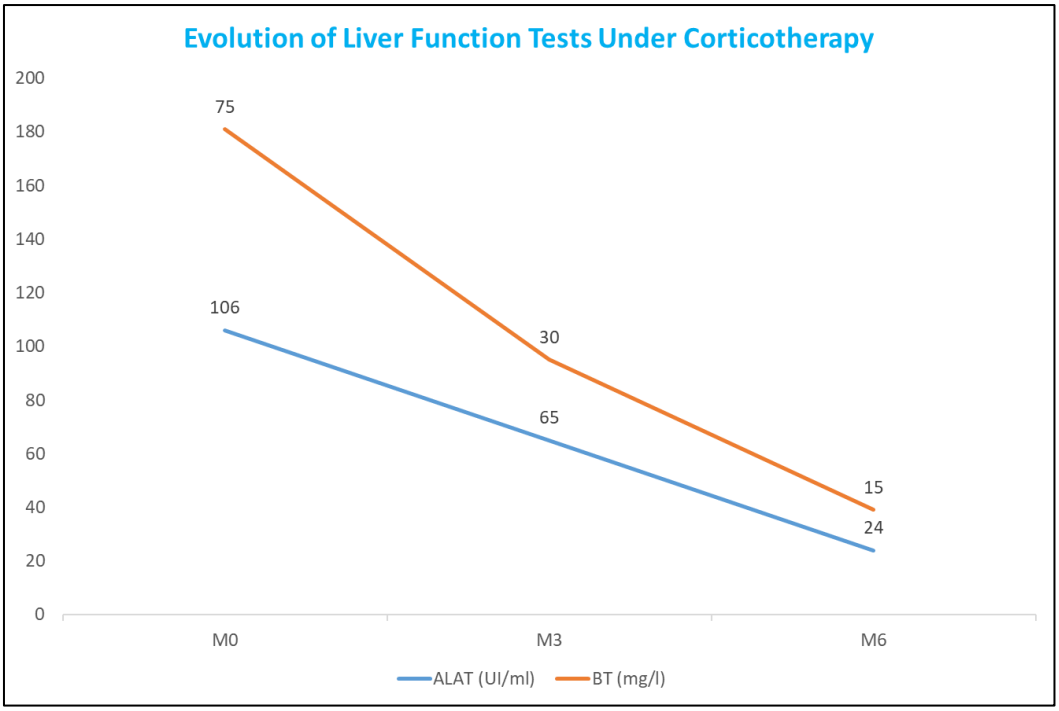

Figure 4 Follow-up liver function tests showing normalization post-treatment.

3. Discussion

The incidence of autoimmune pancreatitis (AIP) remains poorly established [1], although several international efforts have been made to establish precise diagnostic criteria to avoid underdiagnosis. A survey conducted in Japan, where AIP is more common, reports a prevalence of 4.6 per 100,000 people and an incidence of 1.4 per 100,000 [2]. Among 245 pancreatic resections, 11% revealed an undiagnosed AIP [3]. Two forms of AIP are currently recognized, each with distinct clinical and histological profiles. Type I, the most common form, accounts for about 80% of cases and is associated with elevated IgG4 levels, thus being referred to as "IgG4-related disease." Our case corresponds to the type I AIP. It is histologically characterized by lymphoplasmacytic sclerosing pancreatitis (LPSP)[4]. Type II, less common (about 20% of cases), is often associated with inflammatory bowel diseases (IBD) and is distinguished by its duct-centric idiopathic chronic pancreatitis (IDCP) histopathological subtype [5].

The clinical manifestations of AIP are nonspecific and primarily include obstructive jaundice, often with a pancreatic mass, seen in 60-75% of type I AIP cases [6]. Pancreatic-type pain may also be present, generally mild in type I AIP but more severe in type II AIP[7]. Other symptoms may include diabetes, extrapancreatic manifestations, fatigue, and weight loss. IgG4 levels are the most diagnostically valuable serological test for AIP, with a threshold of 135 mg/dL and a sensitivity of 75% and specificity of 97%. However, it is important to note that 25% of AIP cases may be seronegative [8].

AIP is frequently associated with extra-pancreatic involvement, which is supported by common histological abnormalities such as lymphoplasmacytic infiltration, IgG4-positive plasma cells, obliterative phlebitis, and storiform fibrosis. A positive response to steroid treatment is also a key feature, which was the case of our patient. Extra-pancreatic organ involvement can be synchronous or metachronous and includes sclerosing cholangitis (65-85%), sialadenitis (14%), retroperitoneal fibrosis (10%), Riedel's thyroiditis (8%), as well as interstitial lung disease and tubulointerstitial nephritis (8%)[9].

Imaging is an essential tool for diagnosing AIP, allowing the detection of parenchymal, capsular, or ductal abnormalities. However, a normal imaging study does not exclude the disease. In the early stages, AIP is characterized by hypovascularity, seen as hypointensity on CT or MRI, with delayed enhancement specific to AIP, which helps differentiate it from pancreatic cancer [10], [11]. Endoscopic ultrasound (EUS) can also provide crucial information, revealing a homogeneous, hypoechoic pancreas with a reticular or "tortoise-shell" pattern, along with a hypoechoic band at the periphery, distinguishing it from chronic pancreatitis and pancreatic cancer [9].

Diagnostic criteria, such as the revised Japanese criteria of 2011, updated in 2018, are widely used to establish a formal diagnosis of AIP [12]. These include clinical elements (diffuse or focal pancreatic hypertrophy, ductal narrowing), serological findings (elevated IgG4 levels), and histopathological features (lymphocytic infiltration, presence of IgG4-positive plasma cells, and storiform fibrosis). Extra-pancreatic involvement and a positive response to corticosteroid therapy are also key diagnostic criteria.

Regarding therapeutic management, initial flare treatment generally relies on corticosteroids, which are effective in about 98% of cases. A multicenter Japanese study reported significantly higher remission rates in AIP patients treated with steroids compared to those untreated [13]. However, relapses are common, particularly after corticosteroid therapy is discontinued in type I AIP, with relapse rates ranging from 30 to 50% within the first two years [14]. In the case of relapse, immunosuppressors, such as azathioprine, are often used in combination with corticosteroids. For rare cases of steroid-refractory AIP, rituximab, a monoclonal antibody, is an effective alternative to avoid the side effects of corticosteroids.

4. Conclusion

AIP remains a complex and difficult-to-diagnose condition, requiring heightened awareness to avoid diagnostic errors. Early recognition of clinical signs and the use of a multimodal diagnostic approach, including clinical, serological, and histological criteria, are essential for optimal disease management. Corticosteroid therapy remains the cornerstone of treatment, but relapse management strategies, particularly the use of immunosuppressors and rituximab, are necessary to address frequent relapses.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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