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CASE REPORT PEER REVIEWED | OPEN ACCESS

Autoimmune pancreatitis: A challenging diagnosis

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ABSTRACT

Autoimmune pancreatitis (AIP) is a rare form of chronic pancreatitis. However, due to its wide spectrum of clinical and radiological characteristics, it can mimic pancreatic malignancies that could expose patients to unnecessary therapies if it is misdiagnosed. Unlike pancreatic malignancies, AIP adequately responds to steroid therapies. Diagnosis is challenging as pathological samples are usually needed to confirm the diagnosis. A high index of suspicion is needed as the diagnosis of AIP can easily be missed. We present the case of a 54-year-old patient, who presented with weight loss, abdominal pain, and asymptomatic jaundice. A pancreatic malignancy was suspected after an abdominal computed tomography (CT). Nonetheless, after a careful examination, AIP was detected and treated.

Keywords: Autoimmune pancreatitis, IgG4, Pancreas, Pancreatitis, Steroids

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INTRODUCTION

Autoimmune pancreatitis is a rare kind of chronic pancreatitis which responds adequately to steroid therapy. This disease can be part of a multiorgan disorder that can manifest beyond the pancreas, called IgG4-related disease [1, 2]. Autoimmune pancreatitis can mimic pancreatic cancer, and its accurate diagnosis is important to avoid accidental surgery [1]. We present the case of a 54-year-old patient, for whom a pancreatic malignancy was suspected due to asymptomatic jaundice, weight loss, and a heterogeneous pancreas observed in a CT. Autoimmune pancreatitis was finally diagnosed and treated accordingly.

CASE REPORT

The patient is a 54-year-old male without any past medical history. He had a 6-month history of anorexia, severe weight loss (22% of previous weight) and intermittent episodes of mild abdominal pain. One week before presenting to the emergency room he noted a yellowing of his eyes and skin. On clinical examination, a malnourished patient with jaundice was encountered. The abdominal examination was unremarkable, and no lymph nodes or masses were identified. Laboratory exams revealed a cholestatic pattern with elevated conjugated bilirubin (21 mg/dL), gamma-glutamyl transferase (1359 U/L) and alkaline phosphatase (567 IU/L). Lipase and amylase were normal (56 and 102 U/L), yet Ca 19-9 and carcinoembryonic antigen (CEA) were mildly elevated (70 U/L and 25 ng/mL).

Due to this, an abdominal echography was performed and unveiled a normal gallbladder with a 13 mm common bile duct. An abdominal contrast-enhanced CT was requested, and it exposed a dilated common bile duct with a heterogeneous pancreas which was enlarged and surrounded by inflammatory tissue. However, no masses

or lymph nodes were identified. A magnetic resonance cholangiopancreatography (MRCP) was requested, revealing a narrowing of the bile duct near its distal end with a 0.25 mm pancreatic duct. Based on these findings, an endoscopic retrograde cholangiopancreatography was performed and confirmed the bile duct stricture in its distal end. A plastic stent was placed and several biopsies were taken from the head of the pancreas with the aid of endoscopic ultrasound.

Overall, the complementary images were insufficient to establish a definitive diagnostic. Due to the unavailability of immunohistochemistry assays or a pathologist's opinion on location, as the patient presented with severe weight loss and painless obstructive jaundice, he was misdiagnosed with a suspected pancreatic malignancy and surgical plans were discussed with the patient and his family. Because the patient was undecided about surgery, he sought a second opinion and came to our department. He provided us with the pathology samples and a new CT was performed (Figure 1). The pancreas appeared heterogeneous, surrounded by an inflammatory halo, yet no masses were found. Pathology reported pancreatic cells surrounded by severe inflammatory tissue. The pancreatic glands were enlarged, and yet the surrounding tissue was fibrotic and cells showed positivity for IgG4 (Figure 2). A type 1 autoimmune pancreatitis was suspected as serum IgG4 levels were high (over 400 mg/dL) and no evidence of malignancy was discovered on complementary exams. The patient underwent a 2-week steroids therapy (40 mg/day) which produced an observable clinical and radiological improvement (Figure 3). On follow-up controls and six months after initial diagnosis, the patient is performing well on steroid therapy (2.5 mg/day), jaundice has ceased (Total bilirubin 0.8 mg/dL), liver enzymes are normal and he has regained his previous weight.

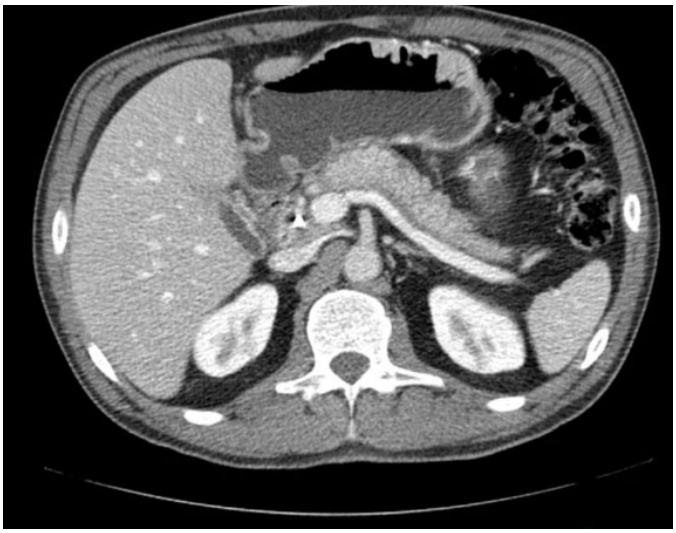

Figure 1: Contrast-enhanced CT, enlarged pancreas surrounded by inflammatory tissue.

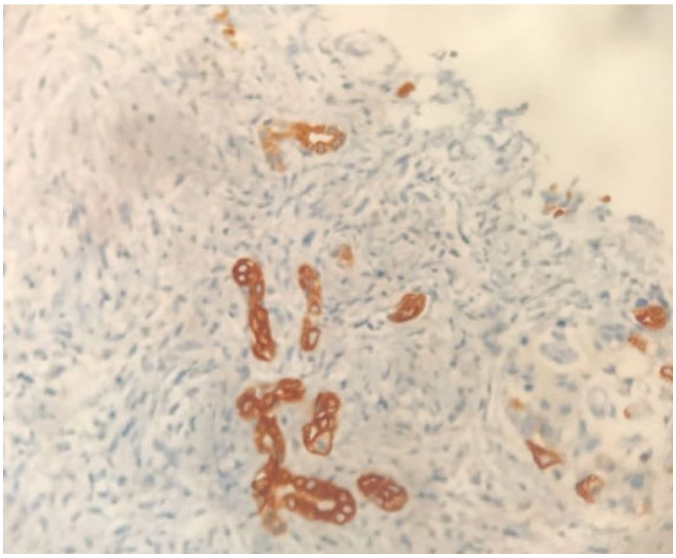

Figure 2: Some of the pancreatic glands showing positivity for IgG4 (immunohistochemistry, IgG4).

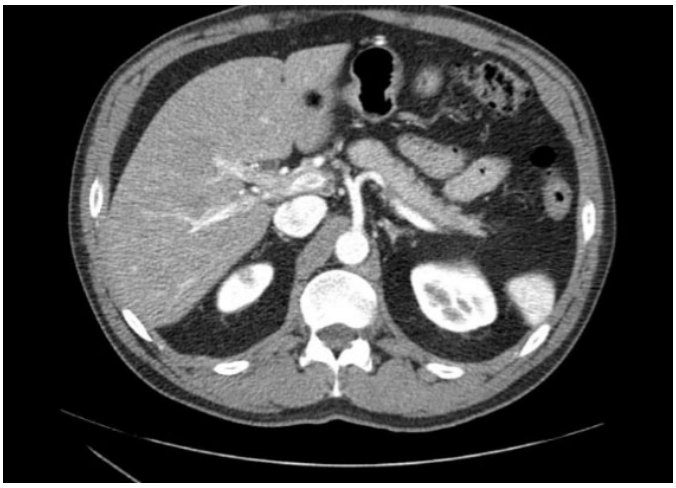

Figure 3: Contrast-enhanced CT after steroids, pancreas appears normal without inflammation.

DISCUSSION

Pancreatic cancer is the fourth leading cause of cancer-related deaths in western countries, with an overall 5-year survival rate of approximately less than 10%. The reasons for this poor survival rate are multifactorial including nonspecific symptoms, late diagnosis, and the close proximity of major blood vessels which can be invaded by this tumor. To this date, surgical resection is the only potential cure for pancreatic cancer, making early diagnosis a priority [1]. The term AIP was first proposed in 1995 by Yoshida et al. when they described a fibroinflammatory disease of the pancreas of possible autoimmune origin. Following investigations demonstrated that AIP is part of a multiorgan disorder called IgG4-related diseases [1, 2]. Autoimmune pancreatitis has been classified into two different kinds based on clinical, serological, radiological, and

pathological characteristics. In type 1 AIP, the pancreas is affected as part of a systemic IgG4-positive disease, also known as lymphoplasmacytic sclerosing pancreatitis. On the other hand, type 2 AIP is characterized by idiopathic duct-centric pancreatitis [3]. Patients with type 1 AIP usually present with a history of obstructive jaundice, abdominal pain, and acute pancreatitis; however, due to its wide spectrum of symptoms, it can also mimic a malignant disease [1, 4], as was thought for our patient. Since diagnosis is based on pathological findings, multiple guidelines have been developed to aid clinical diagnosis. These include invasive and noninvasive methods, such as imaging, serology, steroid therapy trialing, and biopsy [2, 5].

Pancreatic imaging irregularities are found in up to 85% of patients with AIP. Computed tomography and MRCP can reveal diffuse parenchymal enlargement with delayed enhancement [6]. Enlargement is usually accompanied by the effacement of the lobular contour of the pancreas, giving the gland a "featureless" or "sausage-shaped" appearance [2]. On certain occasions, a halo can be observed which is strongly suggestive of AIP. Endoscopic retrograde cholangiopancreatography (ERCP) and endoscopic ultrasound can also aid the diagnosis [2, 3]. Imaging is also critical in the evaluation of patients with suspected pancreatic cancer, however, unlike in AIP, CT usually shows a hypoattenuating pancreatic mass associated with pancreatic duct dilatation, atrophy of the upstream pancreas, yet the detection of pancreatic malignancies ranges from 76% to 96% for CT and from 83% to 93.5% for magnetic resonance imaging (MRI) [7, 8].

IgG4 typically accounts for less than 5% of all the total serum IgG in normal patients. Increases in IgG4 can be encountered in patients with AIP, yet IgG4 titers are not specific to AIP and can also be seen in patients with cancer, which is why serology data should be used together with clinical and radiological features for diagnosis [4]. To illustrate this, an IgG4 test has a sensitivity of 76% and a specificity of 93% for AIP when levels greater than 140 mg/dL are detected. However, the most important role of IgG4 testing should be monitoring the response to medical treatment [1, 2, 6]. Elevated serum IgG4 levels are characteristics of AIP. However, mild elevations in serum IgG4 are seen in up to 10% of subjects without AIP including pancreatic cancer [4]. In type 1 AIP, extrapancreatic involvement is common, as the biliary tree, lacrimal and salivary glands, as well as the kidneys, retroperitoneum, pituitary, and prostate can be affected. Diagnosis is based on histological features: in type 1 AIP, lymphoplasmacytic sclerosing pancreatitis must be present, obliterative phlebitis, storiform fibrosis, and abundant (>10 cells/HPF) IgG4-positive cells can also be found [3, 4, 6].

Unlike in malignancies, AIP responds to steroids, and a trial with these can be performed if a malignancy is ruled out beforehand. A positive test shows rapid improvement in clinical and radiological conditions that must be confirmed after the first two weeks [1, 2]. The management of AIP is fundamentally medical, as treatment is based on long-term use of corticosteroids with dose adjustments. Nonetheless, on discontinuation of steroids, relapses can occur in up to 31% of patients with type 1 AIP and 9% of patients with type 2 AIP. Immunomodulator therapy has shown promise in patients with AIP, yet more research is needed [2–4].

In our case, a malignancy was considered as painless jaundice and weight loss manifested. Nonetheless, after a thorough examination, an AIP diagnosis was finally reached. Differentiation from pancreatic cancer remains challenging and misdiagnosis can still occur. A high index of suspicion is needed as AIP is infrequent and can mimic multiple pancreatic pathologies. Also, in a unique way, this case illustrates that any diagnosis must be fully investigated before taking a surgical or clinical decision.

CONCLUSION

Every action or plan taken on a patient is a risk. Making an accurate diagnosis is a priority on any pathology. When treating AIP differentiation from pancreatic carcinoma is vital as AIP can be treated with steroids and does not require surgery. High clinical awareness and close follow-up are critical in AIP. As relapse can occur, a multidisciplinary team of gastroenterologist, surgeons, and radiologists is necessary to adequately treat these kinds of patients. In a unique way this case also highlights that physicians must always consider these rare pathologies to minimize risks and prevent unnecessary life-threatening procedures.

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Author Contributions

Frans I Serpa – Conception of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Christian A Armijos – Conception of the work, Design of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Marta L Cueva – Conception of the work, Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
C Estefania Burbano – Design of the work, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved
Gabriel A Molina – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

All co-authors have made a substantial contribution to the design, data collection and analysis of the research and the drafting of the manuscript and have reviewed and accepted the contents of the manuscript prior to its submission by the corresponding author.

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Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.