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Case Report

Biliary Obstruction Secondary to Autoimmune Pancreatitis Identified by Abdominal Ultrasound. A Case Report

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Abstract

We present an interesting case of biliary obstruction secondary to autoimmune pancreatitis diagnosed by means of ultrasound imaging in a young male patient who presented to our emergency department with a chief complaint of epigastric pain, fever and jaundice. On suspicion of biliary obstruction, an emergency ultrasound was performed, revealing homogeneously enlarged pancreas and dilatation of the intrahepatic biliary tree and common bile duct with abrupt sharpening of the latter in the intrapancreatic segment. These findings suggested pancreatic inflammation with secondary biliary obstruction. Since the patient was young and had no other medical history of interest, we suggested the diagnosis of autoimmune pancreatitis, which was confirmed on subsequent serologic workup (elevated IgG4 levels). This case highlights the importance of ultrasound to suggest this rare condition, with significant impact on patient management.

Keywords: *Autoimmune Pancreatitis; Biliary Obstruction; Ultrasound; Magnetic Resonance Imaging*

Abbreviations

ALT: Alanine Aminotransferase; AST: Aspartate Transaminase; GGT: Gamma-Glutamyl Transferase; AP: Alkaline Phosphatase; CRP: C-Reactive Protein

Introduction

Autoimmune pancreatitis (AP) was described for the first time in 1995 by Yoshida., *et al* [1]. It a rare disease and still a diagnostic challenge despite of increasing study in recent years [2]. According to the International Association of Pancreatology guidelines, AP is classified into two subtypes. Type 1 is recognized to be part of IgG4-related disease and type 2 is characterized by intraductal neutrophilic infiltration and no IgG4 elevation [3]. From an epidemiological perspective, there is wide variability in its prevalence according to the geographic area, with figures ranging from 1% to 38% [4-6]. This heterogeneity may be magnified by a myriad of factors, including but not limited to patient selection bias or diagnostic criteria used in the studies. It is more frequent in elderly males, but it may occur at any age. On the other hand, biliary obstruction is a rare clinical condition in young patients, with many potential causes. Although pancreatitis has been described to be one relatively common cause of all biliary obstruction cases, there are few reports to date describing biliary obstruction secondary to AP. Abdominal ultrasound imaging can be very helpful to identify the presence and cause of biliary obstruction and is particularly helpful in young patients and in an emergency setting. Other imaging techniques may be useful in follow-up.

Aim of the Study

The aim of this work is to describe a rare case of biliary obstruction secondary to AP in a young male patient who presented to our emergency department, and to illustrate the imaging findings obtained by abdominal ultrasound.

Case Presentation

A 19-year-old male patient with no medical history of interest presented to the emergency department of our institution with a chief complaint of epigastric pain radiating to both hypochondria of 1 month of evolution. Symptoms had become more intense over time, and fever (up to 38.5ºC), jaundice and acholia without pruritus were also present in the last days. The patient reported no other symptoms. Physical examination revealed only slight pain on epigastric palpation, and cutaneous and mucosal jaundice. Blood workup showed elevated total bilirubin (5.4 mg/dl), mainly direct bilirubin (4.14 mg/dl), ALT (582 U/L), and AST (260 U/L), GGT (1505 U/L), AP (507 U/L), amylase (160 U/L) and lipase (773 U/L), as well as increased ferritin (611 ng/ml), fibrinogen (492 mg/dl) and CRP (9.2 mg/L). The rest of the analytical profile was normal.

On suspicion of biliary obstruction, an abdominal ultrasound was performed, which showed dilatation of the intrahepatic and extrahepatic biliary tree with slight parietal thickening. Interestingly, an abrupt change in caliber at the beginning of the intrapancreatic segment of the common bile duct was observed. The pancreas was homogeneously increased in size, with diffusely decreased echogenicity and some echogenic punctate and linear images, without dilatation of the main pancreatic duct. Moreover, discrete peripancreatic inflammatory changes were observed, with no evidence of fluid infiltration or collections. In view of these findings in an otherwise healthy young male, suspicion of autoimmune pancreatitis was raised. Therefore, specific serologic workup including IgG4 fraction levels was obtained, revealing elevated levels (98.7 mg/dL) of IgG4.

After confirmation of the diagnosis, treatment with prednisolone was started. The patient improved after treatment with steroids and was discharged 1 week later. He has remained asymptomatic to date and follow-up imaging examinations, including magnetic resonance imaging (MRI), demonstrated complete resolution of the structural changes observed on ultrasound.

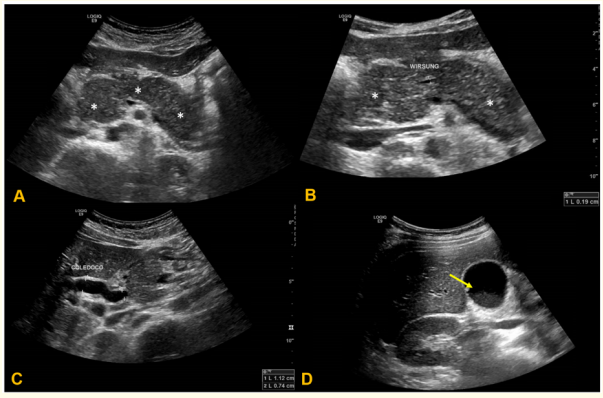

Figure 1: *Abdominal ultrasound. A. Homogeneous enlargment of the pancreas with associated slightly decreased echogenicity and some echogenic punctate and linear images (asterisks). B. No dilatation of the Wirsung's duct. C. Dilatation of the main bile duct with abrupt decrease in size at the intrapancreatic segment. D. Gallbladder slightly distended with echogenic material suggestive of biliary sludge (yellow arrow).*

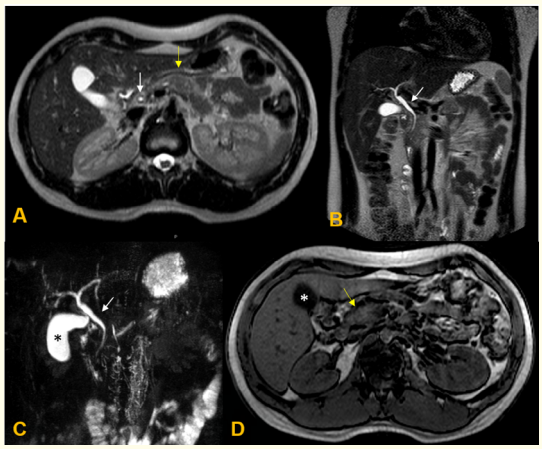

Figure 2: *Magnetic resonance imaging. A. T2-weighted single-shot image, axial view. B. T2-weighted single-shot image, coronal view. C. Cholangio-MRI, 3D TSE image. D. Out-of-phase T1-weighted image, axial view. Normal biliary tree (white arrows) and gallbladder (asterisk). Pancreas of normal size and signal intensity (yellow arrow). These findings indicate complete resolution of obstructive pancreatitis with no structural sequelae.*

Discussion

AP is a chronic fibroinflammatory disease, which currently represents both a localized disease as well as a manifestation of the systemic IgG4-related disease spectrum [7]. The usual symptomatology is obstructive jaundice, mild abdominal pain or recurrent acute pancreatitis. Biliary obstruction refers to the hindering of bile outflow due to obstructive anomalies [8], including intrinsic (e.g. by an intraluminal tumor or choledocholithiasis) or extrinsic compression. In the latter group, any mass or expansile lesion can be the cause. Accordingly, acute pancreatitis is a relatively common cause of biliary obstruction, as with the case presented. Significant pain is unusual and should suggest the possibility of malignancy. It responds rapidly to steroid treatment.

Regarding the diagnosis of AP, imaging examinations show diffuse or focal pancreatic enlargement; sometimes appearing like a mass and pancreatic ductal strictures, suggesting the diagnosis [2], which must be confirmed by serologic workup demonstrating elevated levels of IgG4. Of note, the sensitivity of IgG4 elevation varies between 44% - 95% in different settings, so that combined measurements of total serum IgG and IgG4 has been suggested to increase diagnostic sensitivity without sacrificing specificity [9,10]. Computed tomography (CT) and MRI play an essential role in the diagnosis of AP [2]. In an emergency setting, an appropriate imaging diagnostic approach is necessary. In young patients, abdominal ultrasound is an adequate imaging technique due to its accessibility, diagnostic yield for biliary conditions, and absence of ionizing radiation. It allows to rule out other more common causes of biliary obstruction (e.g. choledocholithiasis) as well as depicting structural changes that may suggest pancreatitis or extrinsic compression of the common bile duct, as was the case with our patient.

Pancreatic ductal adenocarcinoma is usually included in the differential diagnosis because acute pancreatitis presents as tumor-like mass in the pancreatic head with multifocal pancreato-biliary strictures presenting with obstructive jaundice in elderly males [3]. Other differential diagnoses should include any process causing pancreatic enlargement such as traumatic, infectious [11] and other autoimmune diseases [4]. Regarding treatment, AP is highly responsive to steroid treatment. To date, although the initial dose of steroid for induction of remission as 0.6 to 1 mg/kg per day is considered the mainstay, a steroid regimen has not been standardized, and there is no

consensus on the duration of induction, the tapering schedule, and the optimal dose and duration of maintenance therapy [2]. A recent systematic review and meta-analysis found that the rate of relapse tended to decrease with extended durations of glucocorticoid therapy up to 36 months [12]. Other drugs which are increasingly used to treat AP include monoclonal antibodies such as rituximab [13,14]. In fact, current trends in the spectrum of IgG4-related disease investigational treatment approaches have focused on targeting cells of the B-cell lineage, including B-cell-depleting agents (rituximab) and a non-depleting homodimer monoclonal antibody targeting CD19 and Fc-gamma RIIIb [15]. Good outcomes using anti-CD20 combined with targeted LTβR Inhibition have been reported in murine models of AP [16].

Finally, it is important to follow-up patients due to the risk of recurrence. In fact, a study showed that almost 50% of patients with AP experienced relapse during a median follow-up of 47 months [17]. In addition, the risk of developing malignant complications such as cholangiocarcinoma should not be overlooked, warranting patient follow-up [5].

Conclusion

Autoimmune pancreatitis is an uncommon condition that may rarely cause obstruction of the biliary tree in young male patients. Imaging findings are often enough specific to suggest the diagnosis, allowing to visualize pancreatic enlargement and progressive decrease in the diameter of the main bile duct. However, serologic workup with elevated IgG4 levels is required to confirm the diagnosis. Ultrasound is a fast, non-ionizing imaging examination that allows to rule out other more common causes of biliary obstruction and suggest the diagnosis in an appropriate clinical setting.

Ethical Statement

The authors declare that all their institution's protocols have been followed for this study, and that it complies with the ethical standards stated in the Declaration of Helsinki. Patient's informed consent was obtained.

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