Acta Scientific Gastrointestinal Disorders (ISSN: 2582-1091)

Volume 7 Issue 6 June 2024

Case Report

Autoimmune Pancreatitis in a 9-Year-Old Boy

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DOI: 10.31080/ASGIS.2024.07.0632

Received: May 10, 2024

Published: May 31, 2024

Abstract

Introduction: Autoimmune pancreatitis (AIP) is a rare and distinct form of chronic pancreatitis with autoimmune manifestations. The diagnosis of AIP is suggested by a combination of clinical features, imaging modalities of pancreatic parenchyma and ducts, and serology. We present the case of a pediatric male patient with recurrent bouts of painless jaundice.

Case: A 9-year-old male patient was admitted to the emergency department with painless jaundice and scleral icterus. Ct scan of the abdomen revealed diffuse enlargement of the pancreas with featureless borders and loss of definition of the pancreatic clefts, revealing a "sausage-like" appearance. Those findings are highly suggestive of Autoimmune Pancreatitis (AIP). Autoimmune serology revealed normal levels of immunoglobulin IgG4 and other immunologic markers. A trial of prednisone for six weeks was initiated and yielded a remarkable resolution of the patient's symptoms.

Discussion: The pathophysiology of autoimmune pancreatitis is not well understood in the pediatric age group. Adults usually experience painless obstructive jaundice and weight loss, while children are more likely to present with acute abdominal pain and obstructive jaundice, along with other findings like weight loss, fatigue, and vomiting. In our case, the patient presented an atypical presentation of pediatric AIP with a painless and obstructive pattern of jaundice similar to the adult presentation.

Conclusion: Autoimmune pancreatitis is a rare condition, predominantly seen in adults but has also been described in children. Clinicians should keep this disease on the differential in patients presenting with obstructive jaundice.

Keywords: Autoimmune Pancreatitis; Sausage Shape Pancreas; Corticosteroids

Introduction

Autoimmune pancreatitis (AIP) is a rare and distinct form of chronic pancreatitis with autoimmune manifestations. It is a T-cell-mediated disease with lymphoplasmatic infiltration of pancreatic tissue and resultant parenchymal fibrosis. The diagnosis of AIP is suggested by a combination of clinical features, imaging modalities of pancreatic parenchyma and ducts, and serology. Serum immunoglobulins like IgG4 and other markers have been linked to this disease, but the lack of specific biomarkers hinders the diagnosis of AIP [1]. This disease can present with mild abdominal pain, painless jaundice, or recurrent acute pancreatitis. AIP can be confused clinically and on imaging with pancreatic ductal adenocarcinoma (PDAC). It is crucial to differentiate between the two entities, as treatment and prognosis are completely different [2]. We present the case of a pediatric male patient with recurrent bouts of painless jaundice.

Case Report

A 9-year-old male patient was admitted to the emergency department with painless jaundice and scleral icterus. Past medical history includes a previous episode of painless jaundice with an obstructive pattern on liver function testing. MRCP initially revealed narrowing of the distal portion of the common bile duct. A stent was placed, and no further evaluation was done.

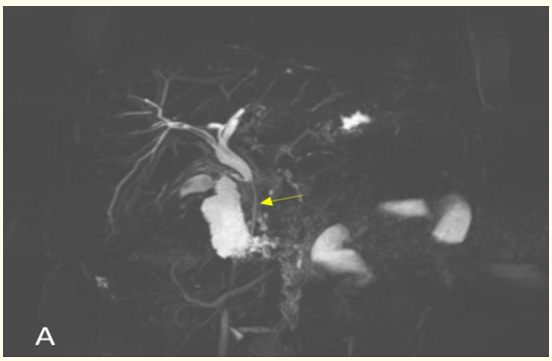

Figure 1: MRCP showing a stent in the common bile duct (yellow arrow) with a long segment of narrowing in the head of the pancreas without dilatation of the pancreatic duct.

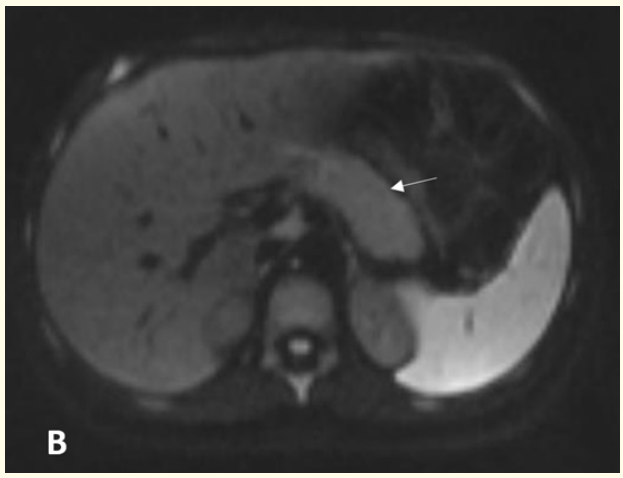

Figure 2: Mild edema of the pancreas with a featureless appearance (white arrow).

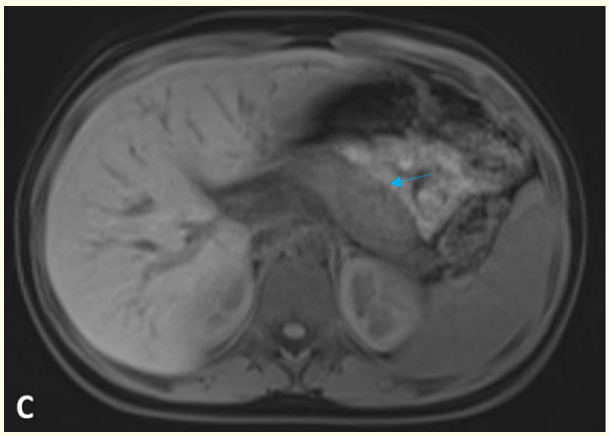

Figure 3: T1 with fat saturation shows absence of the normal high signal intensity of the pancreas (blue arrow).

On this presentation, the patient seemed calm and noted that he was not in pain. Physical examination revealed no epigastric or right upper quadrant tenderness. Vitals were stable, and family history was not pertinent. Laboratory testing of liver function revealed a cholestatic pattern with marked elevations in alkaline phosphatase (ALP), gamma-glutamyl transferase (GGT), and direct and indirect bilirubin. Lipase and C-reactive protein (CRP) levels were normal. Contrast-enhanced Computed Tomography (CT) of the abdomen revealed diffuse enlargement of the pancreas with

featureless borders and loss of definition of the pancreatic clefts, revealing a "sausage-like" appearance. Those findings are highly suggestive of Autoimmune Pancreatitis (AIP). Magnetic resonance cholangiopancreatography (MRCP) without contrast showed narrowing of the pancreatic duct as well in the distal common bile duct, extending 3 cm along the intra-pancreatic portion (Figures 1, 2, 3). Diffuse enlargement of the pancreas with delayed enhancement was noted, and there was no evidence of a mass. Those findings further supported a diagnosis of AIP. EUS was done to confidently rule out the presence of any sub-millimetric pancreatic tumor. Autoimmune serology revealed normal levels of immunoglobulin IgG4 and other immunologic markers. A trial of prednisone for six weeks was initiated and yielded a remarkable resolution of the patient's symptoms. A follow-up MRCP was conducted, revealing the complete resolution of pancreatic enlargement as well as pancreatic and common bile duct narrowing. The patient's previously placed stent was removed, and he was discharged after making a full recovery.

Discussion

AIP was first described by Yoshida., et al. in 1995 as an autoimmune disease responsive to steroid treatment [3]. It is a relatively rare condition, accounting for 2–6% of all chronic pancreatitis cases in adults [4]. It has also been reported in pediatric populations, albeit rarely. Based on the International Consensus Diagnostic Criteria of AIP, diagnosis is based on a combination of five features: histology and imaging findings, serological markers, response to steroids, and presence of systemic organ diseases [5]. Those features can also help distinguish between the two types of AIP: type 1 AIP and type 2 AIP. Type 1 AIP is known as the IgG4-related subtype; on histology, it is characterized by lymphoplasmacytic sclerosing pancreatitis (LPSP) with igG4 plasma cell infiltration [2]. Systematic manifestations of type 1 AIP include retroperitoneal fibrosis, sclerosing cholangitis, chronic peri-aortitis, and Riedel's thyroiditis [6]. On the other hand, Type 2 AIP is the non-IG4-related subtype; it is relatively uncommon and seen mostly in young patients. On histology, it is characterized by idiopathic duct-centric pancreatitis with granulocytic epithelial lesions [2]. Moreover, type 2 AIP is associated with ulcerative colitis in 15–30% of cases [6].

The pathophysiology of autoimmune pancreatitis is not well understood in the pediatric age group. Physicians follow the adult criteria for the management and diagnosis of AIP in children, as there are still no established guidelines directing the diagnostic and therapeutic workup of AIP in the pediatric age group [7]. Symptoms can also differ between adults and children. Adults usually experience painless obstructive jaundice and weight loss, while children are more likely to present with acute abdominal pain and obstructive jaundice, along with other findings like weight loss, fatigue, and vomiting [5]. Concerning serology, children were found to present with normal IgG4 levels in 80% of the cases, with an increase in amylase and lipase levels in only half of the patients. Given these findings, children are considered to follow the presentation of Type 2 AIP, with other studies suggesting that children may follow a distinct disease pattern not yet recognized [5].

In our case, the patient presented an atypical presentation of pediatric AIP with a painless and obstructive pattern of jaundice similar to the adult presentation. The diagnosis of AIP was made based on imaging findings of "sausage-shaped" pancreas on MRCP along with edematous hypoechoic pancreas with narrowing of the pancreatic duct, all of which are typical characteristics of autoimmune pancreatitis [2]. Moreover, our diagnosis was further confirmed by the significant and rapid response to corticosteroids therapy, which is consistent with the diagnostic criteria for AIP outlined in the Mayo Clinic guidelines [8].

Conclusion

Autoimmune pancreatitis is a rare condition, predominantly seen in adults but has also been described in children. It is an IgG-4-mediated sclerosing disease of the pancreas but can involve other organs as well. Clinicians should keep this disease on the differential in patients presenting with obstructive jaundice.

Bibliography

1. Nettleton J., *et al*. "Scrotal necrosis and no Fournier's in sight: a rare case of juvenile gangrenous vasculitis". *BMJ Case Report* 12.2 (2019): e226530.

2. Chen W and Plewig G. "Lipschütz genital ulcer revisited: is juvenile gangrenous vasculitis of the scrotum the male counterpart?" *Journal of the European Academy of Dermatology and Venereology* 33.9 (2019): 1660-1666.

3. Caputo R., *et al*. "Juvenile gangrenous vasculitis of the scrotum: Is it a variant of pyoderma gangrenosum?" *Journal of the American Academy of Dermatology* 55 (2006): S50-53.

4. Marcos-Pinto A., *et al*. "Juvenile gangrenous vasculitis of the scrotum: A rare cause of genital ulcer". *Australas Journal of Dermatology* 61.4 (2020): e424-e426.

5. Satoh M and Yamamoto T. "Genital pyoderma gangrenosum: report of two cases and published work review of Japanese cases". *Journal of Dermatology* 40.10 (2013): 840-843.

6. Jimenez-Cauhe J., *et al*. "Juvenile gangrenous vasculitis of the scrotum: the male counterpart of Lipschütz ulcer?" *Journal of the European Academy of Dermatology and Venereology* 33.12 (2019): e459-e461.