CASE REPORT

Autoimmune pancreatitis with associated ulcerative colitis in a teenager

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SUMMARY

Autoimmune pancreatitis (AIP) is a rare entity that is extremely uncommon in children. Its diagnosis is also a clinical challenge. This form of chronic pancreatitis often presents itself with obstructive jaundice and/or a pancreatic mass and it is sometimes misdiagnosed as pancreatic cancer. We describe the case of a 13-year-old boy with obstructive jaundice and a 4 cm mass in the head of the pancreas that was diagnosed as AIP with associated ulcerative colitis.

BACKGROUND

Autoimmune pancreatitis (AIP) is an autoimmune disorder that occurs primarily in adults and it is extremely rare in children. This form of chronic pancreatitis may resemble pancreatic neoplasia and is frequently associated with obstructive jaundice, with or without a pancreatic mass.[1–3] Distinguishing between these two entities can prove challenging. While diagnostic criteria and management strategies for AIP have been developed for adults, no diagnostic criteria have been established for children. A current report established the characteristic features of paediatric autoimmune pancreatitis (P-AIP) based on a review of all children with AIP to date and, more recently, the same panel of experts developed recommendation statements to standardise the diagnostic and management approach of these patients.[3 4] As such, we report on this case to increase knowledge about this rare entity in the paediatric population.

CASE PRESENTATION

A 13-year-old boy presented with a 3-month history of asthenia, weight loss, abdominal pain and progressive obstructive jaundice 10 days before his admission to the hospital. Laboratory evaluation revealed elevated hepatobiliary enzymes, elevated total and direct bilirubin, sevenfold increased lipase and slightly elevated amylase. Viral panel was negative. Cancer antigen (CA) 19-9 levels were elevated and antinuclear antibody was positive (1/60). Alpha-fetoprotein and other autoantibodies were negative. Immunoglobulin levels were normal but subclass IgG_4 was elevated twofold (table 1).

Abdominal ultrasound, abdominal MRI and MR cholangiopancreatography (MRCP) revealed a 4 cm mass in the pancreatic head (figure 1), causing common bile duct (CBD) obstruction with significant intrahepatic and extrahepatic bile duct dilatation, distended gallbladder and hepatomegaly (figure 2). Endoscopic ultrasound with fine needle aspiration (EUS-FNA) revealed an enlarged pancreatic head and atrophic tail, suggestive of a pseudo-mass and regular CBD dilatation. Biopsy samples were insufficient for diagnosis. An endoscopic retrograde cholangiopancreatography (ERCP) with stent placement was performed, with clinical and laboratory improvement.

A second EUS-FNA showed pancreatic ducts and acini with lymphoplasmacytic infiltrate and some fibrosis patches that were compatible with an inflammatory process. There was no evidence of neoplastic cells. IgG_4 stain was also negative. After ruling out malignancy, the diagnosis of AIP was considered. Because there was a significant clinical and laboratory improvement with reduction of the pancreatic size after resolving biliary obstruction, treatment with steroids was not started.

OUTCOME AND FOLLOW-UP

In the following months, there was a complete normalisation of the laboratory findings, apart for the IgG_4 levels which remained elevated. A follow-up MRCP showed significant reduction of the pancreatic head and a subtle dilatation of the intrahepatic and extrahepatic bile ducts. A second ERCP was performed with new stent placement due to distal CBD stenosis. The stent was removed 16 weeks later with complete resolution of pancreatic enlargement and of biliary and pancreatic ducts dilatation. Although asymptomatic, the patient underwent colonoscopy a few months after AIP diagnosis due elevated calprotectin levels and recognised association between AIP and inflammatory bowel disease.

Ulcerative colitis (UC) was then diagnosed. Corticosteroid treatment was started and azathioprine was subsequently added.

A year later, the patient had a relapse of UC with the need to undergo therapeutic escalation with infliximab. Regardless, his AIP remained in remission at 3½ years of follow-up.

DISCUSSION

Although AIP is a well-known disease in adults, the experience in children is limited to a few cases described in existing literature.[1 3 5–9] Therefore, due to the disease's rarity in children, the diagnosis of AIP in paediatric population requires a high index of suspicion.

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Table 1 Laboratory data on admission (normal range)

Leucocytes, $\times 10^9$/L	11.2
Haemoglobin, g/dL	14.3
Platelets, $\times 10^{10}$/L	408
CPR (<5 mg/L)	9.5
ESR (mm/hour)	40
Lipase (U/L) (4–39)	295
Amylase (U/L) (8–51)	51
AST (U/L) (14–35)	125
ALT (U/L) (8–24)	330
Total bilirubin (mg/dL) (0.3–1.2)	7.91
Direct bilirubin (mg/dL)	4.62
GGT (U/L) (7–21)	406
Alkaline phosphatase (U/L) (127–517)	756
IgG (mg/L) (840–2013)	1470
IgG_2 (mg/L) (140–440)	555
IgG_4 (mg/L) (1–83)	268
Faecal elastase (<200 µg/g)	<30
AFP (ng/mL) (0.6–3.9)	0.8
CA 19.9 (U/mL) (<35)	192.7

AFP, alpha-fetoprotein; ALT, alanine aminotransferase; AST, aspartate aminotransferase; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; GGT, γ-glutamyl transferase.

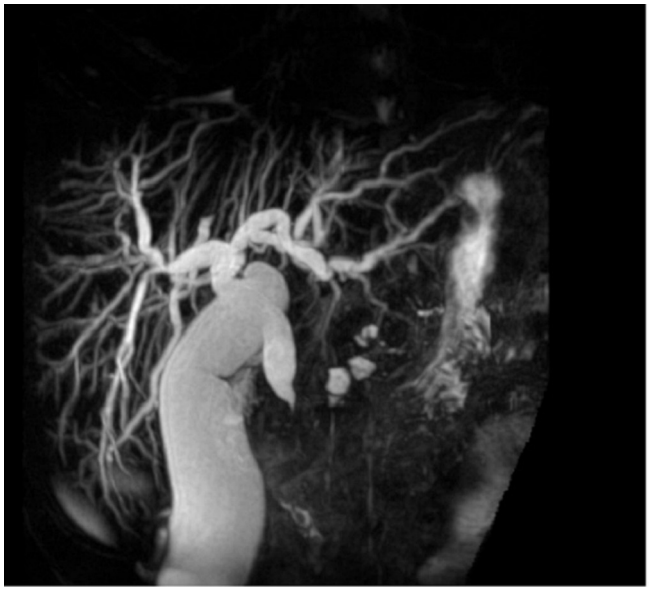

Figure 2 MR cholangiopancreatography (MRCP) showing narrowed distal common bile duct with markedly dilated intrahepatic and extrahepatic ducts.

Until recently there were no established guidelines in diagnosis and management for AIP in children and the approach was based on adult criteria.[2] However, it has been suggested that clinical presentation of AIP may be different in children and adults. As such, a recent report, which was based on data collected from previously reported cases and a large paediatric multicentre cohort study of pancreatitis (International Study Group of Pediatric Pancreatitis: In Search for a Cure—INSPPIRE), attempted to establish the characteristic features of AIP in children.[3] More recently, the same group of paediatric gastroenterologists developed clinical recommendations for the definition, diagnosis and treatment approach of P-AIP.[4]

Most children with AIP present with abdominal pain and/or obstructive jaundice in association with focal pancreatic enlargement, main pancreatic duct irregularities and distal CBD narrowing.[1 3–9] As in adults, there is a clinical challenge in distinguishing a neoplastic condition from AIP, since they benefit from different treatment approaches.

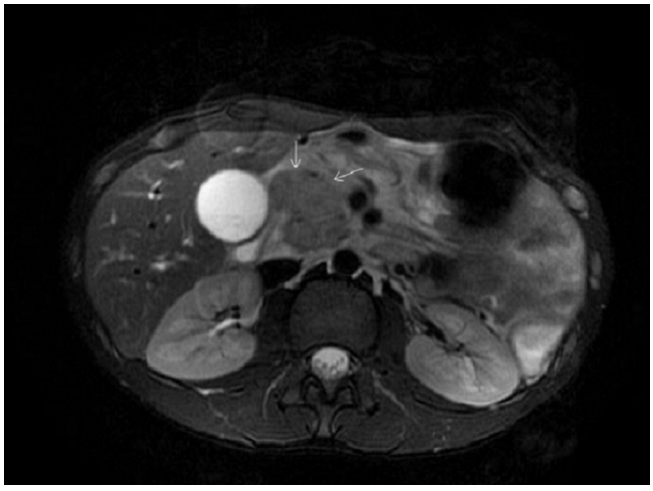

Figure 1 Axial T2-weighted abdominal MRI showing enlargement of the pancreatic head (arrows).

Two distinct subtypes of AIP have been identified in adults, each with differing associations, clinical courses, clinical profiles and histopathological patterns: type 1 or lymphoplasmacytic sclerosing pancreatitis and type 2 or idiopathic duct-centric pancreatitis.[2] It is now clear that P-AIP is a distinct subtype of pancreatitis that may present with histopathological features of both types. The pancreatic parenchymal changes in P-AIP includes lymphoplasmacytic and/or neutrophilic infiltrates and/or parenchymal fibrosis.[4] Even though histopathological findings are used as a diagnostic criterion for AIP, considering the barriers in obtaining pancreatic biopsies in children, P-AIP diagnosis may be based on clinical and imaging findings.[3 4]

High levels of serum IgG_4 may be suggestive of AIP in adults, specially of type 1, however this marker has limited value in children and can be non-specifically elevated in patients with other conditions, as well.[6 10]

While a clear association exists between AIP and concurrent immune/inflammatory diseases, especially UC, their exact relationship is still unclear.[4 5]

The imaging, clinical, analytical and histological evaluations in this case were indicative of P-AIP associated with UC. Pancreatic head enlargement and distal CBD narrowing associated with weight loss, abdominal pain and obstructive jaundice were present. The histological examination was negative for malignant cells and consistent with an inflammatory process, with no evidence of IgG_4 plasma cells on immunostaining.

The current standard treatment for P-AIP is steroid therapy, the same as adults. In some P-AIP cases, the main clinical finding for the disease diagnosis has been steroid-responsive pancreatic enlargement.[6 7] However, the current case saw a spontaneous regression of the pancreatic mass after relief of the biliary obstruction. Some reports have described spontaneous resolution of pancreatic masses, jaundice and strictures in the context of AIP, and some P-AIP patients have experienced disease resolution without any treatment.[3 4 11 12] In this case it was licit to have a wait-and-see approach based on the rapid clinical response and, so far, the patient has not relapsed.

Learning points

- Autoimmune pancreatitis (AIP) rarely occurs in children and is a distinctive subtype of pancreatitis.
- Paediatric autoimmune pancreatitis (P-AIP) should be taken into consideration in the differential diagnosis in children with a pancreatic enlargement.
- Most children with AIP present with abdominal pain and/or obstructive jaundice.
- The diagnosis of P-AIP can be made based on the combination of clinical presentation and imaging findings, possibly complemented by histopathological findings.
- First choice treatment in P-AIP are steroids. However, some patients may have symptom resolution without any treatment.

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