Pediatr Radiol
DOI 10.1007/s00247-008-1132-2

CASE REPORT

Autoimmune pancreatitis in an 11-year-old boy

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Received: 9 September 2008 / Revised: 20 November 2008 / Accepted: 17 December 2008

Abstract We report a case of histopathologically proven autoimmune pancreatitis in an 11-year-old boy. Abdominal US and MRI showed a focal swelling of the pancreatic head, the latter also showing delayed contrast enhancement. There was diffuse irregular pancreatic duct narrowing, compression of the intrapancreatic common bile duct, and mild proximal biliary dilatation on MR cholangiopancreatography. Laboratory results revealed normal serum IgG and subclass 4 with negative autoimmune antibodies, and slightly elevated carbohydrate antigen 19-9. This highlights the differentiation of autoimmune pancreatitis from pancreatic head cancer and, to a lesser extent, other forms of pancreatitis in children.

Keywords Pancreas · Autoimmune pancreatitis · Ultrasonography · MRI · Child

Introduction

Although autoimmune pancreatitis (AIP) is not a common disease, it is being increasingly recognized. It is important to be aware of this disease because AIP can clinically appear as pancreatic cancer or other types of pancreatitis. Moreover, AIP is an impressive disease to clinicians as it responds dramatically to oral steroid therapy, in contrast to other types of chronic pancreatitis [1].

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Case report

An 11-year-old boy presented to the emergency department with a history of nausea and vomiting for the previous few days associated with anorexia, diarrhea and dull aching epigastric abdominal pain that was not related to meals, not relieved by defecation and not accompanied by fever or weight loss. He denied any history of abdominal trauma and there were no previous similar episodes or relevant medical or surgical history. Although the patient's family history was positive for pancreatic cancer, it was negative for pancreatitis. On general examination, he had normal vital signs, was afebrile and was not jaundiced. There was epigastric tenderness but no organomegaly.

Laboratory data revealed elevated serum amylase (134 IU/L; normal range 28–100 IU/L) and serum lipase (141 IU/L; normal <37 IU/L), and normal blood glucose, direct bilirubin and other liver function tests. Autoimmune markers, including antinuclear antibody, anti-soluble liver antigen/liver-pancreas antibodies and antibodies for autoimmune hepatitis, primary biliary cholangitis, and primary sclerotic cholangitis were negative, with normal levels of serum immunoglobulins E, M, A, G and subclass 4 (IgG4), and slightly elevated carbohydrate antigen 19-9 (CA 19-9).

Abdominal US revealed an enlarged hypoechoic pancreatic head measuring 2.8×2.4 cm with no abnormalities of the other abdominal organs (Fig. 1). There was no abdominal or pelvic fluid collection. Abdominal MRI confirmed the pancreatic head mass (Fig. 2); it demonstrated a hypointense surrounding rim in T2-W images (Fig. 3) and showed delayed contrast enhancement on the dynamic

Published online: 04 February 2009

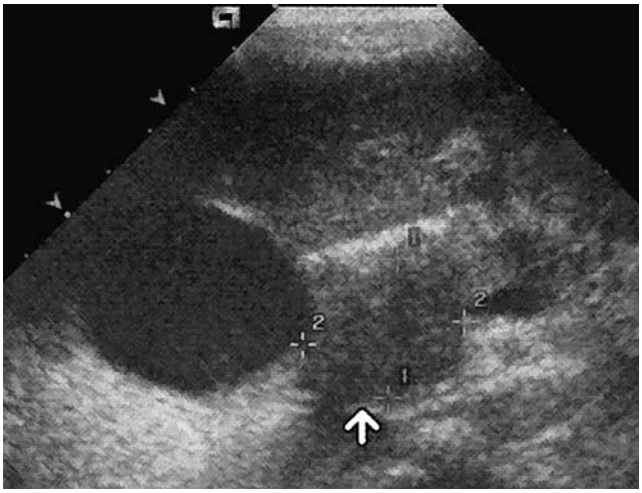

Fig. 1 Transverse abdominal US image shows an enlarged hypoechoic pancreatic head measuring 2.8×2.4 cm (*arrow*)

images (Fig. 4). MR cholangiopancreatography (MRCP) showed diffuse irregular narrowing of the main pancreatic duct, disappearance of the right-angled branches, narrow intrapancreatic portion of the common bile duct and consequent mild dilatation of the intrahepatic biliary radicals (Fig. 5).

The imaging findings were suggestive of AIP [2], but pancreatic biopsy was necessary to differentiate AIP from pancreatic cancer as the management differs significantly. At diagnostic laparotomy, the pancreatic head mass was identified and multiple biopsies taken. Histology showed extensive pancreatic tissue destruction with marked fibrosis that was periductal in distribution. The pancreatic parenchyma was richly infiltrated with lymphocytes and multiple plasma cells. There was no evidence of malignancy. The appearances were characteristic of AIP [3].

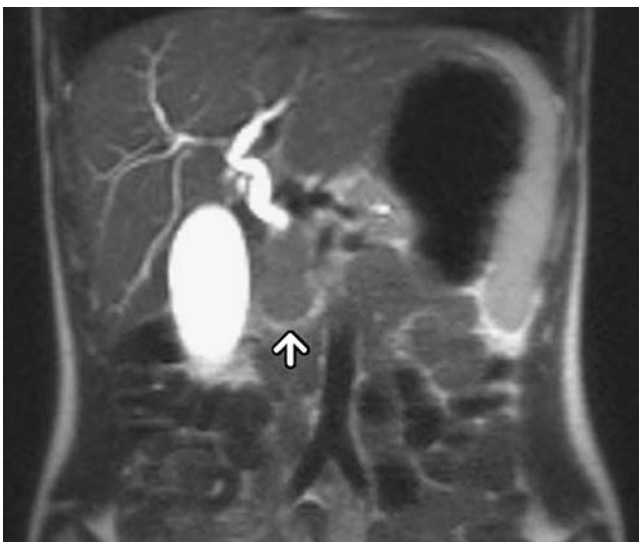

Fig. 2 T2-W HASTE coronal MR image shows focal enlargement of the pancreatic head (*arrow*)

Discussion

It is difficult to estimate the true incidence of pancreatitis in childhood because most of the literature comprises case reports or small series of patients. Although pancreatitis is not seen as commonly in children as in adults, it is generally underdiagnosed and requires a high index of clinical suspicion. Childhood pancreatitis differs from pancreatitis in adults in its etiology [4], usually being caused by blunt abdominal trauma, drugs, or viral infection. Chronic pancreatitis in children has been classified into two morphological forms: calcific and non-calcific varieties. Calcific chronic pancreatitis is usually due to juvenile tropical pancreatitis or hereditary pancreatitis, while non-calcific chronic pancreatitis is unusual and is generally due to congenital or acquired lesions of the pancreatic duct or trauma [5].

With mounting evidence suggesting an underlying autoimmune mechanism, the term AIP was originally introduced by Yoshida et al. [6]. AIP likely accounts for a significant proportion of cases previously classified as idiopathic pancreatitis [7]. A set of diagnostic criteria has been proposed by the Japan Pancreas Society in an attempt to differentiate AIP from other forms of pancreatitis and pancreatic cancer. The criteria are based on a combination of the findings of imaging, laboratory testing, and histological analysis [2].

In our patient, the diagnosis of AIP was based on the MR and MRCP imaging findings and histological examination of the pancreas, although negative autoimmune antibodies and normal levels of immunoglobulins specially IgG and subclass 4 were obtained. These results are similar to those in the patients presented by Toomey et al. [3] who diagnosed AIP based on the abdominal US and CT imaging features of a solid mass in the pancreatic body despite normal serum IgG and elevated CA 19-9 levels. We agree with Finkelberg et al. [2] who state that diagnosis may be made in the absence of diagnostic laboratory findings. If histological findings of an autoimmune process are obtained, a diagnosis of AIP can be confirmed [2]. Serum IgG4 levels can distinguish between masses due to AIP and pancreatic cancer with levels corresponding with disease activity. Although a cut-off level of 135 mg/dl was 95% sensitive and 97% specific for distinguishing between AIP and pancreatic cancer [3], serum IgG4 was unhelpful in our study and in other cases [3].

In this patient with AIP, although there were some imaging similarities with those of pancreatic cancer, in the form of decreased T1 signal intensity and delayed enhancement after gadolinium administration, as well as obstruction

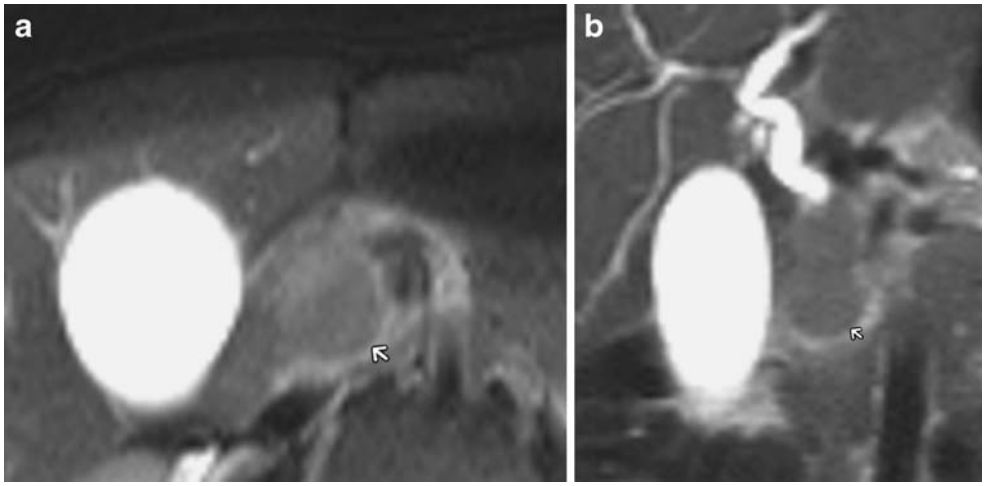

Fig. 3 Zoomed T2-W MR images. Axial TSE image (**a**) and coronal HASTE image (**b**) show the distinct hypointense capsule-like rim that surrounds the pancreatic head (*arrow*)

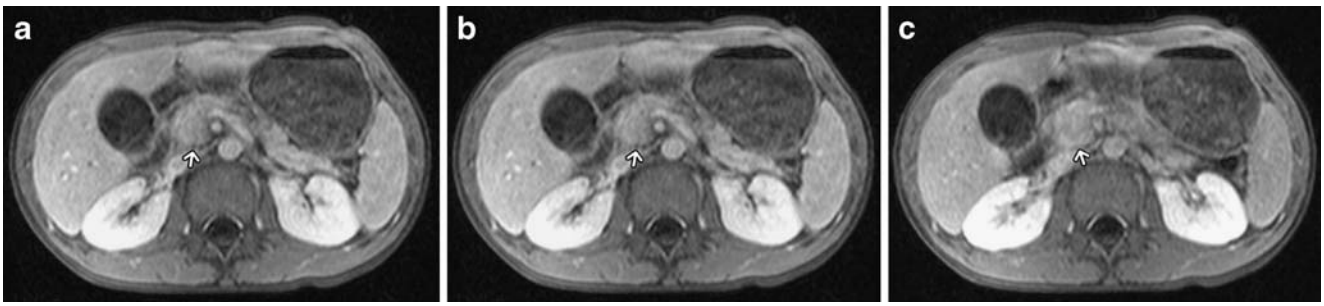

Fig. 4 Dynamic contrast-enhanced axial T1-W MR images (**a**) 1 min, (**b**) 2 min, and (**c**) 3 min after contrast medium injection show delayed enhancement of the pancreatic head (*arrow*)

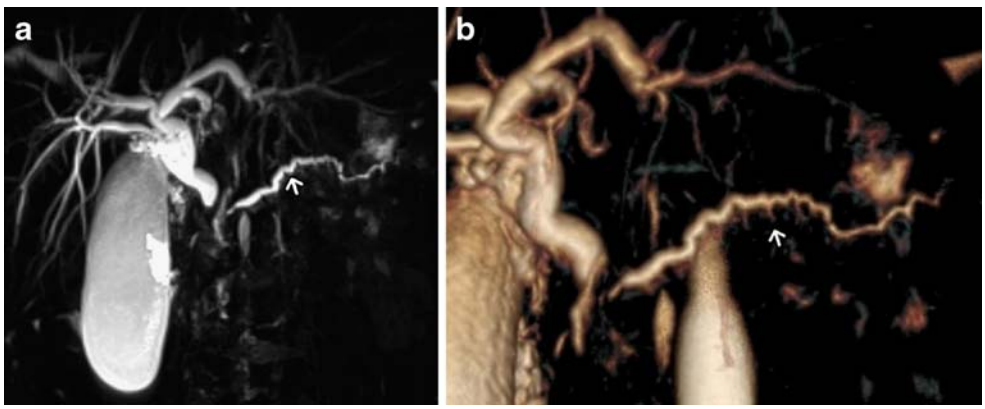

Fig. 5 MRCP. MIP image (**a**) and 3-D volume-rendered image (**b**) show diffuse irregular narrowing of the main pancreatic duct (*arrow*), absence of the right-angled branches and a focal stricture in the distal portion of the common bile duct with mild dilatation of the intrahepatic biliary radicals

of the pancreaticobiliary ducts, differentiation could be achieved from the appearance of the pancreatic duct. Irregularity of the pancreatic duct favours the diagnosis of chronic pancreatitis. In contrast, a smoothly dilated pancreatic duct with an abrupt interruption favours the diagnosis of cancer. Additional findings, such as retroperitoneal lymphadenopathy and obliterated perivascular fat planes may suggest underlying malignancy [8].

As shown in this case report, the diagnosis of AIP can be suggested from the imaging features. These features include the hypointense rim on T2-W images, the MRCP findings of an irregularly narrowed main pancreatic duct, the disappearance of the right-angled branches, and the narrowed intrapancreatic common bile duct. In children with AIP the clinical differential diagnosis, other than pancreatic cancer, includes acute pancreatitis and the other two most common forms of childhood chronic pancreatitis – juvenile tropical pancreatitis and hereditary chronic pancreatitis. Their differentiation can be achieved based on the clinical presentation and radiological findings.

Patients with juvenile tropical pancreatitis have diabetes and chronic abdominal pain with an atrophic pancreas, dilated ducts and pancreatic calcifications. AIP is differentiated from hereditary chronic pancreatitis on the basis of the negative family history and the radiological findings. The morphological features of chronic hereditary pancreatitis are pancreatic atrophy and large spherical calcifications that occur in half of patients. Radiologically, severe acute pancreatitis in children often manifests as heterogeneous pancreatic enlargement, a poorly defined pancreatic contour, peripancreatic fluid and, typically, a smooth normal-calibre pancreatic duct [5].

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