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Autoimmune pancreatitis with multiple extrapancreatic manifestations

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Abstract

Autoimmune pancreatitis is a rare but increasingly recognised entity, which may account for up to 7% of cases of idiopathic chronic pancreatitis. There are several characteristic imaging features associated with the disease however it often mimics pancreatic malignancy both clinically and radiologically, and may result in unnecessary surgery being performed. While autoimmune pancreatitis may involve the pancreas in isolation, almost half of these patients have manifestations of the disease in other organs. These additional sites of involvement may be identifiable radiologically and, when present, help confirm the diagnosis. We present a case of autoimmune pancreatitis with multiple extrapancreatic manifestations and review the available literature.

Keywords: Autoimmune pancreatitis; Renal pseudotumor; Retroperitoneal fibrosis; Tomography; X-ray computed

1. Introduction

Autoimmune pancreatitis is a rare but increasingly recognised entity, which may account for 5–7% of cases of idiopathic chronic pancreatitis [1,2]. Patients most commonly present with obstructive jaundice and some have mild epigastric pain. While there are several characteristic imaging features associated with the disease, in many cases autoimmune pancreatitis can mimic pancreatic malignancy both clinically and radiologically, leading to unnecessary surgery [3]. This condition can affect the pancreas alone or present as a systemic, multi-organ autoimmune disorder. We report the case of a male patient with autoimmune pancreatitis, not suspected clinically, whose abdominal CT showed typical pancreatic abnormalities in addition to involvement of the kidneys, peripancreatic lymph nodes and retroperitoneum, which strongly supported the diagnosis.

2. Case report

A 66-year-old man presented to his family physician with a history of recent onset of vague epigastric pain. He had no other symptoms, although he reported a deterioration of his long-standing diabetes mellitus, which now required insulin. Suspecting cholelithiasis and biliary colic, he had initially been referred for ultrasonography, at which time several enlarged peripancreatic lymph nodes, up to 2 cm in diameter, were identified. No biliary tree abnormality was identified. While the pancreas appeared normal sonographically, the question of pancreatic malignancy was raised and an abdominal CT was arranged.

The CT showed a diffusely enlarged, 'sausage-shaped' pancreas, with loss of the normal pancreatic clefts (Fig. 1). A low-attenuation rim surrounded the pancreas. There was no peripancreatic fat stranding. Several mildly enlarged peripancreatic nodes were identified. Also noted were multiple, ill-defined low-attenuation rounded lesions in both kidneys (Fig. 2). In addition, abnormal soft tissue was demonstrated anterior to the abdominal aorta, extending inferiorly along the common iliac arteries (Fig. 3).

Based on these imaging findings, a diagnosis of autoimmune pancreatitis with multiple extrapancreatic manifestations was made.

An autoimmune antibody work-up was performed; the patient's only significantly elevated parameter was his IgG4 level. Treatment with oral corticosteroids was initiated. Following 3 months of therapy, the patient's symptoms had resolved however his insulin-dependent diabetes was stable. A follow-up CT showed complete resolution of the pancreatic abnormalities, lymphadenopathy and periaortic soft

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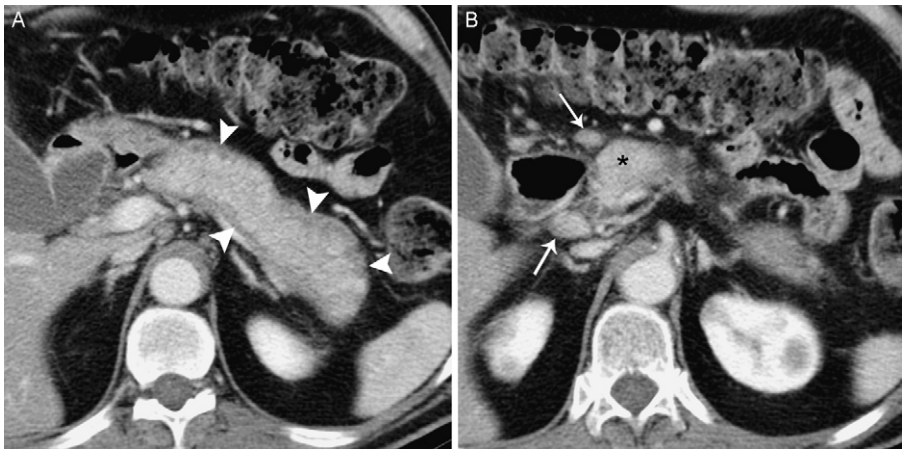

Fig. 1. (A) Transverse, contrast-enhanced CT image of the upper abdomen shows diffuse enlargement of the pancreas with a peripheral rim of low attenuation (arrowheads), the characteristic finding in autoimmune pancreatitis. There is no stranding of the peripancreatic fat. (B) Transverse image at the level of the uncinate process (asterisk) shows enlarged peripancreatic lymph nodes (arrows).

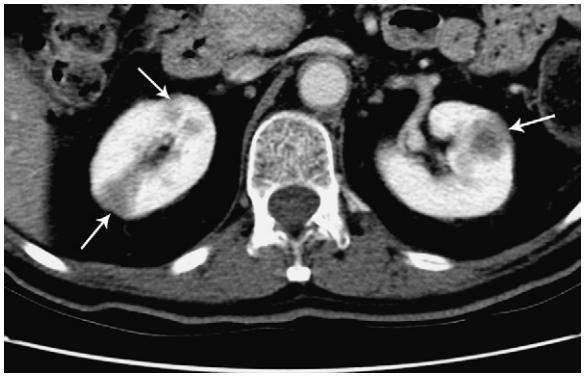

Fig. 2. Multiple rounded, ill-defined, low-density solid lesions (arrows) are demonstrated in both kidneys, and show typical appearances for inflammatory pseudotumors.

tissue, with partial resolution of the renal pseudotumors (Fig. 4).

3. Discussion

The concept of a form of chronic pancreatitis caused by an autoimmune mechanism was reported by Sarles et al. [4], however the term 'autoimmune pancreatitis' (AIP) was first proposed by Yoshida et al., who suggested that this should be considered a disease in its own right in 1995 [5]. Since then, AIP has increasingly been recognised as a cause of 'idiopathic' chronic pancreatitis: it has been reported to cause between 5 and 7% of cases of chronic pancreatitis [1,2]. The majority of patients are aged over 50 years, and it is approximately twice as common in men [1].

Patients with AIP typically have no history of alcohol abuse or of attacks of acute pancreatitis; the most common symptom is painless, obstructive jaundice, which occurs in 63–86% of patients [1,6]. About one-third of patients will report mild epigastric pain, and a similar proportion has some manifestation of pancreatic insufficiency such as weight loss, steatorrhoea or new onset of diabetes mellitus [6]. Because of these symptoms, pancreatic malignancy is often suspected clinically and patients are commonly referred for imaging, hence it is important that radiologists be familiar with AIP and its potential appearances on cross-sectional imaging modalities.

Typically, CT demonstrates diffuse enlargement of the pancreas, which is often described as 'sausage-shaped' [7]. The pancreas is usually sharply defined but featureless due to absence of the normal pancreatic clefts. A characteristic finding is the

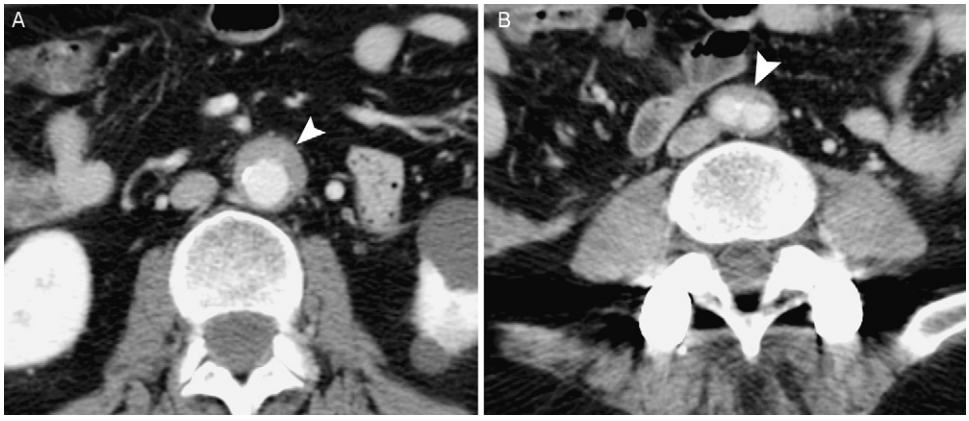

Fig. 3. (A) and (B) show abnormal periaortic soft tissue (arrowheads) which extended inferiorly along the common iliac arteries.