CASE REPORT

Autoimmune pancreatitis mimicking Klatskin tumour on radiology

Yousaf Bashir Hadi,¹ Abdul Malik Amir Humza Sohail,² Zishan Haider³¹Medical College, Aga Khan University, Karachi, Pakistan²Aga Khan University, Karachi, Pakistan³Department of Radiology, The Aga Khan University, Karachi, Pakistan

Correspondence to

Yousaf Bashir Hadi,
yousaf.hadi@gmail.com

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SUMMARY

Autoimmune pancreatitis (AIP) is categorised into two distinct types, AIP type 1 and 2. Although there can be multisystem involvement, rarely, the cholangitis associated with AIP can present radiologically in a manner similar to that of Klatskin tumour. We present the case of a 65-year-old man who was almost misdiagnosed with a Klatskin tumour because of the similarity in radiological features of the two aforementioned clinical entities. The patient presented with a history of jaundice, pruritus and abdominal pain, and work up showed deranged liver function tests, elevated cancer antigen 19-9 levels and positive antinuclear antibodies. CT scan of the abdomen showed findings suggestive of Klatskin tumour but due to diffuse enlargement of the pancreas and surrounding low-attenuation halo found on a closer review, a diagnosis of AIP was performed. The patient was started on standard corticosteroid therapy and responded well, with complete resolution of the radiological findings.

BACKGROUND

Autoimmune pancreatitis (AIP) is a rare yet increasingly recognised type of chronic pancreatitis with characteristic histopathological and radiological manifestations. Based on associated clinical findings and presentation, AIP is categorised into two distinct types, AIP type 1 and 2. AIP type 1 is typically associated with IgG4-related autoimmune disease, which can involve multiple organs including the liver, salivary glands, retroperitoneum, bile ducts and the pancreas, etc.¹ Type 2 has features of granulocytic infiltration, generally occurs in young individuals and is less often associated with high IgG levels.² Rarely, the cholangitis associated with AIP can present radiologically in a manner similar to that of a cholangiocarcinoma or Klatskin tumour. Differentiation of these two disease entities can be difficult but is of particular importance in order to avoid unnecessary surgeries and allow early initiation of medical treatment of AIP.

We present the case of a 65-year-old man who was almost misdiagnosed with a Klatskin tumour because of the similarity in radiological features of the two aforementioned clinical entities.

CASE PRESENTATION

A 65-year-old man presented to the gastroenterology clinic with a 3-month history of jaundice, pruritus and abdominal pain. The patient had no comorbidities, was a non-smoker and had no history of alcohol consumption. On examination, he was vitally stable and all systemic examinations

were unremarkable. Laboratory findings revealed a total serum bilirubin level of 3.6 mg/dL with a direct bilirubin concentration of 2.5 mg/dL. γ -Glutamyl transferase (GGT), serum glutamyl pyruvate transaminase (SGPT) and alkaline phosphatase levels were elevated to 437, 271 and 465, respectively. Screening for hepatitis A, B, C, D and E turned out to be negative. Cancer antigen (CA) 19-9 levels were 97.9.

CT scan of the abdomen showed soft tissue thickening in the common hepatic duct at the region of confluence causing wall thickening, measuring approximately 1.7×0.6 cm in size, and luminal obliteration at this level. There was poor contrast enhancement noted. Mild to moderate intrahepatic biliary dilation was seen in the absence of any significant portal or para-aortic lymphadenopathy. These findings were suggestive of a neoplastic lesion in the common hepatic ducts in the region of confluence causing biliary channel obstruction at this level and could be a result of cholangiocarcinoma/Klatskin tumour.

On further review, evidence of diffuse enlargement of the pancreas with a surrounding low-attenuation halo was seen (figure 1). All these changes can also be appreciated on figures 2 and 3, which are postcontrast images. Minimal peripancreatic fat stranding was also noted. The low-attenuation halo was also surrounding the adjacent splenic vein, which appeared significantly attenuated.

There were multiple wedge shaped hypoattenuating areas in bilateral renal cortices most likely

Figure 1 Axial postcontrast CT showing mild intrahepatic biliary dilation with soft tissue thickening at the confluence of right and left hepatic duct (arrow). Swollen pancreas with subtle halo can also be seen.

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Figure 2 Postcontrast coronal CT image, showing diffuse soft tissue thickening of entire visible common bile duct (arrow). Note the swollen pancreas showing loss of lobulation and the surrounding hypodense halo.

representing renal infarcts. Overall, these findings were suggestive of an autoimmune process involving the pancreas and kidneys. The common hepatic duct wall thickening and obliteration were thought to be a part of this autoimmune process. However, the possibility of a malignant lesion in the duct at the region of confluence could not be entirely excluded.

On further laboratory testing, antinuclear antibodies were found to be homogeneously positive and a diagnosis of AIP was performed.

Figure 3 Postcontrast axial CT image demonstrating swollen, sausage-shaped pancreas with surrounding hypodense halo (arrow) typical of autoimmune pancreatitis. Note diffuse thickening of common bile duct walls.

DIFFERENTIAL DIAGNOSIS

Klatskin tumour/cholangiocarcinoma versus AIP.

TREATMENT

The patient was started on standard corticosteroid therapy for AIP.

OUTCOME AND FOLLOW-UP

At 4 weeks' follow-up, complete resolution of symptoms was reported by the patient. Laboratory findings showed a total bilirubin level of 1.2 and a direct bilirubin level of 0.7. GGT, SGPT and alkaline phosphatase levels were found to be 83, 63 and 110 IU/L, respectively. CA 19-9 levels dropped to 8.22.

An MRI was performed showing almost complete resolution of the swollen pancreas with peripancreatic inflammatory changes and the intrahepatic biliary dilation. A normal common bile duct was noted. The previously present abnormally enhancing soft tissue swelling at the porta hepatis was not found, as seen in [figure 4](#). The kidneys were also normal.

The patient is doing well at a 6-month follow-up.

DISCUSSION

AIP accounts for 5–6% cases of chronic pancreatitis.³ It was initially described by Yoshida *et al*⁴ as a disease entity with characteristic histopathological and imaging findings. As described above, two types of AIP have been described. Most recently, the International Association of Pancreatology developed the International Consensus Diagnostic Criteria (ICDC) for AIP, which laid emphasis on five features of prime importance in AIP. These features included the characteristic radiological findings of the pancreatic parenchyma and duct, pancreatic histology, response to corticosteroid therapy, elevated serum IgG4 levels and other multisystem involvement with IgG4 related disease.⁵ It is to be noted, however, that elevated serum IgG4 levels may not always be found even in type 1 AIP, with Krasinskas *et al*⁶ reporting that elevated serum IgG4 levels were only found in 44% of patients with both types of AIP.

Multisystem involvement can be seen in IgG4-associated autoimmune disease. The cholangitis associated with the disease process can present with either a diffuse sclerosing pattern or a

Figure 4 Coronal MR showing T2-weighted images after 4 weeks of steroid therapy showing marked improvement in common bile duct thickening (arrow) and pancreatic changes.

pseudotumour pattern.⁷ Typically, this cholangitis has concentric bile duct thickening with multiple foci, smooth strictures, minimal proximal dilation and absence of diverticula with preferential involvement of large bile ducts.⁸

In extremely rare cases, this cholangitis has been reported to present as a Klatskin tumour on radiology,⁸ as in our case, where a characteristic lesion resembling a Klatskin tumour was seen. Imaging modalities such as CT, positron emission tomography scan and MRI have not been found useful in differentiating a hilar cholangiocarcinoma from IgG4 cholangitis with high certainty.⁸ However, as in our case, the concurrent involvement of the pancreas should raise a high suspicion of an autoimmune aetiology for the lesion as opposed to a neoplastic one. However, cases of IgG4-related cholangitis with no pancreatic involvement have been described in the literature.⁶ Therefore, it

may be important to keep an autoimmune differential in mind while diagnosing Klatskin tumour on radiology.

It has been suggested that because of a lower relapse rate of patients on steroid therapy, early initiation of steroid therapy may be advantageous for autoimmune cholangitis and autoimmune cholangitis patients.⁹ In our case, steroid therapy resulted in complete resolution of pancreatic and bile duct lesions. We therefore conclude that the earliest possible initiation of corticosteroids, as in our case, may help improve disease outcome, at least by avoiding unnecessary surgical and diagnostic procedures.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Rarely, IgG4-related autoimmune pancreatitis can radiologically present as Klatskin tumour.
- ▶ In radiographs presenting with typical features of Klatskin tumour, signs of autoimmune disease should be explored. Laboratory investigations may provide further information that may be helpful in differentiating the two disease entities.
- ▶ If signs of Klatskin tumour and autoimmune pancreatitis coexist on radiology, standard corticosteroid therapy for autoimmune pancreatitis may be the best initial step, as it can resolve both the pathologies if the signs of Klatskin tumour are a result of autoimmune cholangitis.

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