JOP. J Pancreas (Online) 2007; 8(3):335-343.

CASE REPORT

Autoimmune Pancreatitis: Medical and Surgical Management

Desmond P Toomey[1], Niall Swan[2], William Torreggiani[3], Kevin C Conlon[1]

[1]Professorial Surgical Unit, [2]Department of Pathology, and [3]Department of Radiology, The University of Dublin, Trinity College, Adelaide and Meath Hospitals Incorporating the National Children's Hospital, Tallaght. Dublin, Republic of Ireland

ABSTRACT

Context Autoimmune pancreatitis is characterised by a lymphoplasmacytic infiltrate consisting in part of plasma cells that produce large amounts of IgG4. It can manifest as focal or diffuse enlargement of the pancreas with associated strictures of the pancreato-bilary tree giving rise to symptoms including abdominal pain, weight loss and obstructive jaundice; thus it can be extremely difficult in both presentation and investigation to distinguish from pancreatic carcinoma. Recent advances now facilitate preoperative diagnosis and effective medical management, including steroid treatment of autoimmune pancreatitis so preventing major surgical intervention.

Case report Two cases of autoimmune pancreatitis are described, each of which presented with obstructive jaundice and a relatively painless pancreatic mass, one with vascular involvement. They each had elevated serum CA 19-9 and ultimately required surgical exploration to definitely exclude malignancy before embarking on non operative treatment. The first case settled spontaneously while the second rapidly improved with steroid treatment.

Conclusion These two cases illustrate the difficulties in diagnosing this condition, the efficacy of steroid therapy and the role of surgical intervention in unresponsive cases or those where a diagnostic dilemma remains.

INTRODUCTION

Approximately 2% of patients operated on for an apparently malignant pancreatic mass are found instead to have a subtype of chronic pancreatitis characterised by a lympho-plasmacytic infiltrate [1, 2]. The classical risk factors of alcohol excess and familial pancreatitis are absent in these cases [3]. Previously it has had many different classifications such as "non alcoholic duct destructive pancreatitis"[4], however this condition is currently referred to as "lymphoplasmacytic sclerosing pancreatitis" or in reference to its likely pathogenesis "autoimmune pancreatitis" (AIP) [2, 5, 6]. Due to remarkably similar clinical and radiological profiles, AIP can be extremely difficult to distinguish from malignancy, in many cases provoking major pancreatic resection [6]. Recently, steroids have emerged as an effective treatment and, with a preoperative diagnosis, can prevent surgical intervention [7, 8, 9]. We present two Irish patients with AIP and review the literature, to clarify the salient issues surrounding its diagnosis and management, in particular to highlight the features that distinguish it from cancer and the need for surgical exploration in ambiguous cases.

CASE REPORT

Case 1

A 47-year-old lady presented with a 3 day history of epigastric pain, new onset pale stools and dark urine and 14 kg weight loss in

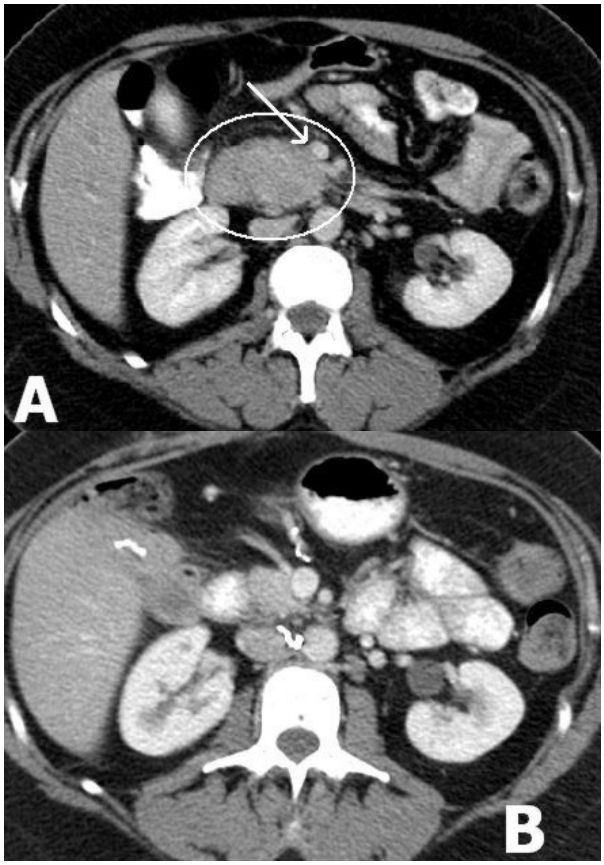

Figure 1. CT of Case 1 at presentation (**a.**) showing mass in the head of the pancreas (circle) which involved the superior mesenteric vein (arrow). This mass had resolved 5 months post operatively (**b.**).

the previous 8 months. Her past history is unremarkable apart from a partial colectomy for a diverticular bleed. She was a non smoker and did not drink alcohol. On admission she was noted to be jaundiced, her liver function tests had an obstructive pattern (Table 1) and her CA 19.9 was elevated at 444.7 U/mL (reference range: 0-60 U/mL).

Abdominal ultrasound demonstrated a dilated common bile and intrahepatic ducts with a possible mass in the head of her pancreas. Endoscopic retrograde cholangiopancreatography (ERCP) delineated a tapered stricture of the distal common bile duct consistent with a pancreatic neoplasm. The patient was transferred to a tertiary centre where computed tomography (CT) confirmed intra- and extra-hepatic duct dilatation although the pancreatic duct was obscured. There was also a 1.5 cm mass in the head of the pancreas which involved the superior mesenteric vein. This was highly suspicious for ("but not definitely") pancreatic adenocarcinoma (Figure 1a).

As carcinoma could not be adequately excluded, the patient proceeded to staging laparoscopy, which showed no sign of metastatic disease. Open examination of the pancreas then revealed that the organ was grossly inflamed throughout and unresectable however it was questionable whether any mass or tumour was present. Pancreatic head biopsies were taken and a choledocho-jejunostomy was fashioned.

Histological examination of the biopsies showed no evidence of malignancy but demonstrated all four cardinal features of AIP (Figure 2). These histological findings alone fulfilled the Mayo Clinic diagnostic criteria [10] and when combined with the ERCP and CT images also satisfied the Japanese criteria [11]. Following biliary bypass the patients jaundice receded and her CA 19-9 returned to normal levels (17.4 U/mL). As the patient's symptoms had settled while awaiting histology results, steroids were not com-

Table 1. Liver function tests of Case 1 at selected timepoints.

	Pre-operative	1 month post-operative	2nd presentation (hepatitis)	29 months post-operative
Bilirubin (reference range: 0-17 μmol/L)	162	13	6	5
Alanine aminotransferase (ALT) (reference range: 7-35 IU/L)	313	71	843	68
Alkaline phosphatase (reference range: 35-104 IU/L)	523	179	309	131
Gamma glutamyltransferase (GGT) (reference range: 5-40 IU/L)	446	139	122	33

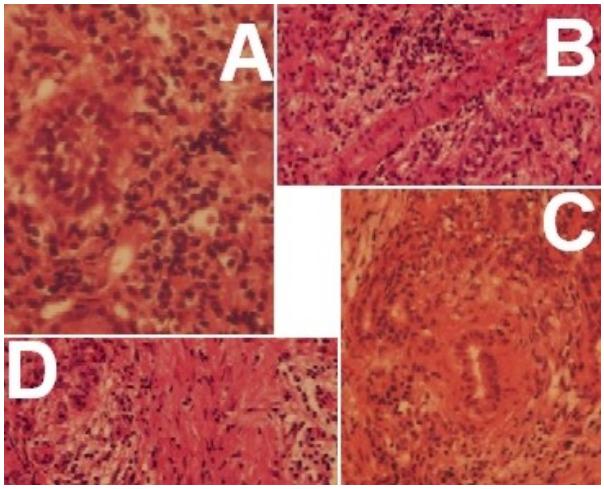

Figure 2. Pancreatic biopsies from Case 1 showing the four cardinal features of autoimmune pancreatitis-lymphoplasmacytic infiltrate (**a.**), vein with periphlebitis (**b.**), periductal distribution of inflammation (**c.**) and band of interstitial fibrosis (**d.**).

menced and she was discharged home with tramadol hydrochloride for any recurrent pain and oral pancreatic enzyme supplements for pancreatic exocrine insufficiency.

She re-presented 5 months post operatively complaining of right upper quadrant pain. CT showed marked resolution of the pancreatic abnormalities (Figure 1b). She was not jaundiced however her liver function tests were abnormal (Table 1) with a hepatitic pattern prompting a liver biopsy which showed a mild cholestatic and portal lobular inflammation suggestive of mild chronic hepatitis. This was reported as consistent with several possible diagnoses including common bile duct obstruction, primary sclerosing cholangitis and primary biliary cirrhosis. A slightly raised IgG at 16.7 g/L (reference range: 6-16 g/L) (IgG4 quantification was not available at that time), and positive auto-antibodies (namely anti-nuclear antibody: 1:80, homogenous), anti-dsDNA antibody and anti-smooth muscle antibody raised the suspicion of autoimmune hepatitis and a diagnosis of overlap syndrome (metachronous or synchronous autoimmune pancreatitis and hepatitis) was made. Her symptoms settled spontaneously and her liver function tests now show only mild derangement (Table 1).

She was subsequently assessed for a complaint of dry mouth and dry eyes and, based on a salivary gland biopsy and positive anti-salivary gland antibodies, a diagnosis of Sjögren's syndrome was made for which she was prescribed artificial tears and saliva.

She is currently 2.5 years post laparotomy and continues to do well. Her pain returns episodically and is managed with simple analgesia, she has a good appetite and has gained weight.

Case 2

A 59-year-old lady, with a background history of hypercholesterolaemia, was referred for a tertiary opinion with an eight week history of slight epigastric discomfort, unquantified weight loss and jaundice, which had not resolved despite halting statin therapy. Her biochemical investigations suggested a hepatitic picture (Figure 3) which was supported by ultrasound imaging showing a diffusely echogenic pattern with periportal distribution. However, both ultrasound and subsequent computed tomography of her abdomen delineated a solid mass in the body of her pancreas with no evidence of biliary tree or pancreatic duct dilatation (Figure 4a). Her CA 19-9 was elevated at 426 U/mL.

Hepatitis A, B and C serology, as well as liver auto-antibodies, were negative and she denied any excessive alcohol consumption. Fine needle aspiration cytology from the pancreatic mass showed some atypical epithelial cells, moderately suggestive of malignancy, and occasional inflammatory cells. A differential

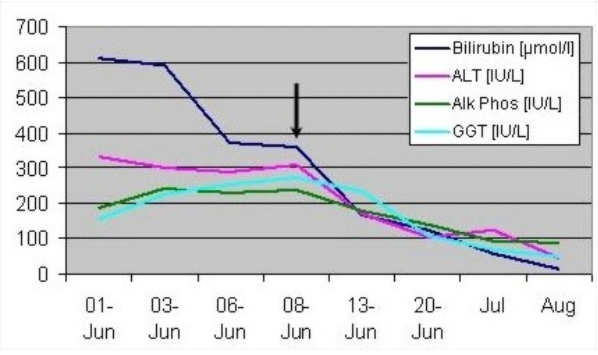

Figure 3. Chart showing response of Case 2 liver function tests to steroid therapy. Arrow indicates commencement of steroids. (ALT: alanine aminotransferase; Alk Phos: alkaline phosphatase; GGT: gamma glutamyltransferase).

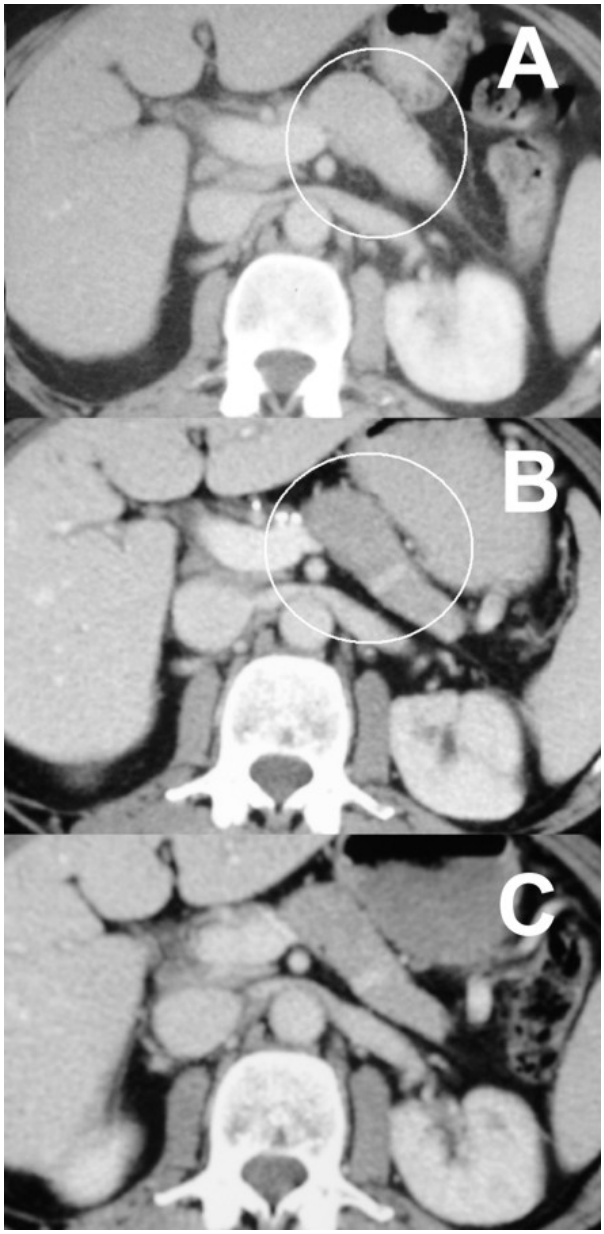

Figure 4. CT of Case 2 at presentation (**a.**) showing mass (circle) in the body of the pancreas, residual fullness after 4 weeks of steroid therapy (**b.**), and resolution 4 months later (**c.**).

diagnosis of hepatotoxicity secondary to rosuvastatin with concomitant pancreatic mass, possibly carcinoma, or autoimmune pancreatitis and biliary sclerosis was made.
The patient underwent a diagnostic laparoscopy with laparoscopic ultrasound which demonstrated no evidence of malignancy, a segmental hyperaemia of the pancreas with a diffusely enlarged pancreatic neck and no ductal dilatation. Histology of an intra-operative liver biopsy suggested cholestatic hepatitis consistent with drug induced injury, primary biliary cirrhosis or primary sclerosing cholangitis while hepatic artery lymph node biopsies showed reactive changes and not malignancy. IgG4 level was within the normal range (0.535 g/L; reference range: 0.045-1.635 g/L).
Although not all diagnostic criteria for AIP, either American [10] or Japanese[11] criteria were satisfied, malignancy had been adequately excluded and so oral prednisolone therapy (40 mg daily followed by reducing dose over 8 weeks to zero) was commenced based on a working diagnosis of AIP. There was resolution of the pancreatic mass on follow up CT, four weeks later (Figure 4b), with some residual fullness of the body, which coupled with a marked improvement of the patient's jaundice and a reduction in CA 19-9 to 118 U/mL, supported this diagnosis. Repeat CT 4 months later (Figure 4c) showed resolution of this fullness and the patient continues to do well 6 months post operatively.

DISCUSSION

Two atypical cases of AIP are presented which demonstrate the role of surgical exploration when all diagnostic criteria cannot be fulfilled. AIP has a spectrum of morphology from a focal, tumorifactive process, most often in the head of the pancreas, but also in the body and tail, to a disease that is diffusely distributed throughout the organ [2, 12, 13]. It can be difficult, particularly in the former scenario, to confidently rule out carcinoma before initiating medical treatment [14].
In Case 1, histology was required to enable a diagnosis. All four cardinal histological features of AIP, namely lymphoplasmacytic infiltration of the pancreas with a periportal distribution of inflammation, interstitial fibrosis and periphlebitis (Figure 2), are necessary for a diagnosis [6]. The lymphoplasmacytic infiltrate consists of lymphocytes and plasma cells (often producing high levels of IgG4) [7] with occasional macrophages, neutrophils and eosinophils. The resultant interstitial fibrosis,

centres on the medium and large intralobular ducts [4, 6, 15, 16, 17] but may involve the smaller ducts in advanced cases and generally results in luminal narrowing without the distal dilatation seen with malignant lesions [18]. The vasculitic element is usually limited to a phlebitis of the venules with arterial sparing [5, 15, 17, 19]. In advanced disease the acini can become inflamed and sclerotic with loss of the normal architecture [2, 4].

Similar infiltration and fibrosis of the biliary tree, which is strongly associated with focal disease of the pancreatic head, also occurs and culminates in obstructive jaundice, a common presenting symptom. As seen in Case 2, where the mass is distant from the common bile duct, obstructive jaundice is likely due to this duct wall infiltration and not ductal compression by the mass. Follicular hyperplasia, present in both patients, may also result in local lymphadenopathy [2, 4, 5, 15, 17, 19].

Case 1 later presented with metachronous hepatitis and Sjögren's syndrome, both autoimmune diseases associated with AIP. Similarities between AIP and primary sclerosing cholangitis suggest an autoimmune aetiology and numerous series and case reports link AIP with different autoimmune diseases (Table 2) [2, 3, 4, 5, 11, 16, 19, 20, 21, 22, 23, 24]. In many cases a similar lymphoplasmacytic infiltrate has been found in other symptomatic organs [23]. A significant proportion of patients produce auto-antibodies against carboanhydrase II, nuclear antigen, lactoferrin and smooth muscle and this, in combination with the disease regression invoked by steroids, is suggestive of an autoimmune origin [2, 16, 18, 19, 22].

Table2. Diseases associated with autoimmune pancreatitis in the literature.

Crohn's disease
Sjögren's syndrome
Primary biliary cirrhosis
Primary sclerosing cholangitis
Autoimmune hepatitis
Ulcerative colitis
Interstitial pneumonia
Systemic lupus erythromatosis
Sialoadenitis
Reidel's thyroiditis
Retroperitoneal fibrosis
Inflammotory pseudotumour
Prostatitis
Tubulointerstitial nephritis

AIP most commonly presents in men in the 6^{th} and 7^{th} decade however cases as young as 10-year-old have been reported [1, 6, 15, 18, 19, 25]. The incidence of this condition remains obscure but seems to account for 4 to 11% of chronic pancreatitis cases [14, 26].

Both patients had a classical presentation of mild abdominal pain, obstructive jaundice, anorexia and weight loss [4, 5, 6, 9, 15, 19], notably similar to the presenting complaint in patients with pancreatic cancer. By definition, AIP occurs in the absence of the normal risk factors for pancreatitis. Up to 40% of cases have a clinical history of other autoimmune diseases [3, 4, 6, 22]. Other fibrotic diseases, including members of the multifocal idiopathic fibrosclerosis family of diseases and inflammatory pseudotumour, may also occur synchronously or metachronously with AIP [5, 11, 23, 24].

Serum IgG4 levels can distinguish between masses due to AIP and pancreatic cancer with levels corresponding with disease activity. Although, according to Hamano *et al*. [23], a cut-off level of 135 mg/dL was 95% sensitive and 97% specific for distinguishing between AIP and pancreatic cancer, results that are supported by Hirano *et al*. [27]; serum IgG4 was either unavailable or unhelpful in both these patients, demonstrating the need for expansive diagnostic criteria. Quantification of serum autoimmune antibodies and rheumatoid factor may demonstrate elevated levels but their specificity is low and they may be indicative of an autoimmune comorbidity [28]. Serum CA 19-9, a marker usually associated with cancer, may be elevated in AIP [12, 15] as seen in both these patients, however, this may be associated with jaundice rather than the specific disease process.

Depending on the morphology of the disease in a given patient, CT can delineate a diffuse enlargement of the pancreas or a mass/focal

fullness [3, 6, 15]. The latter are most common in the head and may be multiple but both frequently resemble malignancy [13]. Furthermore, there may be bilary tree dilatation with thickening of the bile duct walls. Distinguishing CT features of AIP include minimal/absent peripancreatic stranding, an organ with a sharp outline and homogenous attenuation, absence of vascular encasement and a halo of low attenuation secondary to fibro-inflammatory changes in peripancreatic adipose tissue [3, 15, 18, 21, 28]. In addition, we have found that, unlike invasive ductal carcinoma, there is no pancreatic ductal dilatation associated with lesions of the pancreatic neck, body and tail. Unusually, the mass in Case 1 involved the superior mesenteric vein and neither patient demonstrated the classical halo.

Although magnetic resonance cholangio-pancreatography can show primary sclerosing cholangitis-like, intra-hepatic duct beading, or stricture of the pancreato-biliary tree it is inferior to endoscopic retrograde cholangio-pancreatography and percutaneous trans-hepatic cholangiography [18, 19, 28, 29].

Transabdominal ultrasound is relatively non specific in AIP; however, the ongoing development of contrast enhanced ultrasonography, which characterises lesions by dynamic imaging of vascularity and perfusion patterns, is a promising new imaging tool for differentiating these masses from cancer [30, 31]. Endoscopic ultrasound (EUS) may complement CT findings by imaging diffuse or focal organ enlargement and altered echotexture [3]. Guided fine needle aspiration cytology, as utilized in Case 2, has a role limited to detection of malignant cells as histology is required for a pathological diagnosis of AIP. The false negative rate of core biopsy (possibly due to the focal nature of this disease) may be minimised by use of transabdominal ultrasound or EUS guided core biopsy [18, 32]. The technique of using IgG4 positive lymphoplasmacytic infiltrates from biopsies of other involved organs to support the diagnosis of AIP[10] was not available in either case as they had no synchronous disease.

Recent advances in the treatment of AIP has produced alternatives to surgical resection. Improved preoperative diagnosis allows for a trial of medical treatment with close follow up to ensure adequate disease regression. There are isolated reports of spontaneous resolution of these pancreatic masses, similar to Case 1 [33]; however, recent evidence supports the use of steroids, as in Case 2. Steroids can induce sustained and often complete, clinical and radiological remission with normalization of laboratory results, improvement or disappearance of pancreato-biliary strictures and restoration of organ function [7, 8, 9, 12, 18, 22, 34, 35, 36]. However, a significant proportion of patients experience relapses of pancreatitis, duct strictures and extra-pancreatic manifestations with corresponding fluctuations in IgG4 levels [7]. Also steroids

Table 3. Diagnostic algorithm for diagnosing autoimmune pancreatitis. (Mayo Clinic experience; modified from Chari *et al*. [10]).

Any one group required for diagnosis	
One of:	• All 4 characteristic histological features on pancreatic biopsy; • Ten, or more, IgG4 positive plasma cells/high powered field on pancreatic biopsy.
OR	
All of:	• Diffuse pancreas enlargement and rim of low attenuation on CT/MRI; • Irregular pancreatic duct on ERCP; • Serum IgG4 above normal level.
OR	
All of:	• Unexplained pancreatic disease after workup excludes all other causes including malignancy; • Non pancreatic biopsy showing infiltrate with abundant IgG4 positive plasma cells or serum IgG4 above normal level; • Marked response of disease manifestations with steroids.

have been successfully used to treat recurrent strictures in cases where previous resection has provided a histopathological diagnosis [29].

Several groups have composed diagnostic criteria in an effort to avoid surgical exploration without risking misdiagnosing malignancy. Previous publications from Japan relied quite heavily on radiological criteria [11] but a recent audit from the Mayo Clinic has suggested an algorithm which has a strong emphasis on clinical, serological and histological factors with less weighting of radiological results (Table 3) [10]. Their use of steroids as a diagnostic modality is controversial and care must be taken to ensure that malignancy is not inappropriately treated [10]. We generally will employ a short trial of steroid treatment in cases where a clinical and radiological or histological suspicion of AIP is combined with a raised IgG4 level allowing improving symptoms, imaging and serology to verify the diagnosis. However, the described cases illustrate failings in the existing criteria and the need to maintain a low threshold for surgical exploration, either laparoscopic or open, where diagnostic doubts exist or disease is unresponsive or progressive.

Surgical resection, when required, is more treacherous than that for cancer [21]. We have found that a very vigorous peripancreatic, fibrotic reaction distorts surgical planes and strongly adheres the organ to the peripancreatic vessels making dissection difficult in significantly more cases resulting in higher blood losses and longer operating times. Furthermore, these cases tend towards higher overall complication rates [21].

In conclusion, AIP has a significant prevalence and can be difficult to distinguish from malignancy but it is being recognised more frequently than before. Although it often shows a good response to steroids, surgical exploration or resection are still required in cases where any doubt remains about a possible diagnosis of malignancy or when symptoms fail to respond to medical treatment.

Received January 24th, 2007 - Accepted April 3rd, 2007

Keywords Autoimmune Diseases; Immunoglobulin G; Pancreatic Neoplasms; Pancreatitis; Steroids

Abbreviations AIP: autoimmune pancreatitis

Correspondence
Kevin C Conlon
Professorial Surgical Unit
The Trinity Centre for Health Sciences
The Adelaide and Meath Hospitals
Incorporating the National Children's Hospital
Tallaght
Dublin 24
Ireland
Phone: +353-1.896.3719
Fax: +353-1.896.3788
E-mail: profsurg@tcd.ie

Document URL: http://www.joplink.net/prev/200705/12.html

References

1. Abraham SC, Cruz-Correa M, Argani P, Furth EE, Hruban RH, Boitnott JK. Lymphoplasmacytic chronic cholecystitis and biliary tract disease in patients with lymphoplasmacytic sclerosing pancreatitis. Am J Surg Pathol 2003; 27:441-51. [PMID 12657928]

2. Kloppel G, Luttges J, Sipos B, Capelli P, Zamboni G. Autoimmune pancreatitis: pathological findings. JOP. J Pancreas (Online) 2005; 6(1 Suppl):97-101. [PMID 15650292]

3. Sahani DV, Kalva SP, Farrell J, Maher MM, Saini S, Mueller PR, et al. Autoimmune pancreatitis: imaging features. Radiology 2004; 233:345-52. [PMID 15459324]

4. Ectors N, Maillet B, Aerts R, Geboes K, Donner A, Borchard F, et al. Non-alcoholic duct destructive chronic pancreatitis. Gut 1997; 41:263-8. [PMID 9301509]

5. Klimstra DS, Adsay NV. Lymphoplasmacytic sclerosing (autoimmune) pancreatitis. Semin Diagn Pathol 2004; 21:237-46. [PMID 16273942]

6. Weber SM, Cubukcu-Dimopulo O, Palesty JA, Suriawinata A, Klimstra D, Brennan MF, Conlon K. Lymphoplasmacytic sclerosing pancreatitis: inflammatory mimic of pancreatic carcinoma. J Gastrointest Surg 2003; 7:129-37. [PMID 12559194]

7. Nishino T, Toki F, Oyama H, Shimizu K, Shiratori K. Long-term outcome of autoimmune pancreatitis after oral prednisolone therapy. Intern Med 2006; 45:497-501. [PMID 16702740]

8. Tanaka S, Kobayashi T, Nakanishi K, Okubo M, Murase T, Hashimoto M, Takeuchi K. Corticosteroid-responsive diabetes mellitus associated with autoimmune pancreatitis. Lancet 2000; 356:910-1. [PMID 11036899]

9. Erkelens GW, Vleggaar FP, Lesterhuis W, van Buuren HR, van der Werf SD. Sclerosing pancreato-cholangitis responsive to steroid therapy. Lancet 1999; 354:43-4. [PMID 10406367]

10. Chari ST, Smyrk TC, Levy MJ, Topazian MD, Takahashi N, Zhang L, et al. Diagnosis of autoimmune pancreatitis: the Mayo Clinic experience. Clin Gastroenterol Hepatol 2006; 4:1010-6. [PMID 16843735]

11. Okazaki K, Kawa S, Kamisawa T, Naruse S, Tanaka S, Nishimori I, et al. Clinical diagnostic criteria of autoimmune pancreatitis: revised proposal. J Gastroenterol 2006; 41:626-31. [PMID 16932998]

12. Kamisawa T, Egawa N, Nakajima H, Tsuruta K, Okamoto A, Kamata N. Clinical difficulties in the differentiation of autoimmune pancreatitis and pancreatic carcinoma. Am J Gastroenterol 2003; 98:2694-9. [PMID 14687819]

13. Ohana M, Okazaki K, Hajiro K, Kobashi Y. Multiple pancreatic masses associated with autoimmunity. Am J Gastroenterol 1998; 93:99-102. [PMID 9448184]

14. Pearson RK, Longnecker DS, Chari ST, Smyrk TC, Okazaki K, Frulloni L, Cavallini G. Controversies in clinical pancreatology: autoimmune pancreatitis: does it exist? Pancreas 2003; 27:1-13. [PMID 12826899]

15. Kawamoto S, Siegelman SS, Hruban RH, Fishman EK. Lymphoplasmacytic sclerosing pancreatitis with obstructive jaundice: CT and pathology features. AJR Am J Roentgenol 2004; 183:915-21. [PMID 15385281]

16. Okazaki K, Uchida K, Ohana M, Nakase H, Uose S, Inai M, et al. Autoimmune-related pancreatitis is associated with autoantibodies and a Th1/Th2-type cellular immune response. Gastroenterology 2000; 118:573-581. [PMID 10702209]

17. Zamboni G, Luttges J, Capelli P, Frulloni L, Cavallini G, Pederzoli P, et al. Histopathological features of diagnostic and clinical relevance in autoimmune pancreatitis: a study on 53 resection specimens and 9 biopsy specimens. Virchows Arch 2004; 445:552-63. [PMID 15517359]

18. Kim KP, Kim MH, Kim JC, Lee SS, Seo DW, Lee SK. Diagnostic criteria for autoimmune chronic pancreatitis revisited. World J Gastroenterol 2006; 12:2487-96. [PMID 16688792]

19. Okazaki K, Chiba T. Autoimmune related pancreatitis. Gut 2002; 51:1-4. [PMID 12077078]

20. Barthet M, Hastier P, Bernard JP, Bordes G, Frederick J, Allio S, et al. Chronic pancreatitis and inflammatory bowel disease: true or coincidental association? Am J Gastroenterol 1999; 94:2141-8. [PMID 10445541]

21. Hardacre JM, Iacobuzio-Donahue CA, Sohn TA, Abraham SC, Yeo CJ, Lillemoe KD, et al. Results of pancreaticoduodenectomy for lymphoplasmacytic sclerosing pancreatitis. Ann Surg 2003; 237:853-8. [PMID 12796582]

22. Yoshida K, Toki F, Takeuchi T, Watanabe S, Shiratori K, Hayashi N. Chronic pancreatitis caused by an autoimmune abnormality. Proposal of the concept of autoimmune pancreatitis. Dig Dis Sci 1995; 40:1561-8. [PMID 7628283]

23. Hamano H, Kawa S. Are there any other organs in which autoimmune pancreatitis associated lesions remain to be identified. Intern Med 2006; 45:883-4. [PMID 16946568]

24. Yoshimura Y, Takeda S, Ieki Y, Takazakura E, Koizumi H, Takagawa K. IgG4-associated prostatitis complicating autoimmune pancreatitis. Intern Med 2006; 45:897-901. [PMID 16946571]

25. Bartholomew SV, Zigman A, Sheppard B. Lymphoplasmacytic sclerosing pancreatitis presenting as a pancreatic head mass in a child: case report and management recommendations. J Pediatr Surg 2006; 41:E23-5. [PMID 16677871]

26. Finkelberg DL, Sahani D, Deshpande V, Brugge WR. Autoimmune Pancreatitis. N Engl J Med 2006; 355:2670-6. [PMID 17182992]

27. Hirano K, Kawabe T, Yamamoto N, Nakai Y, Sasahira N, Tsujino T, et al. Serum IgG4 concentrations in pancreatic and biliary diseases. Clin Chim Acta 2006; 367:181-4. [PMID 16426597]

28. Okazaki K, Uchida K, Chiba T. Recent concept of autoimmune-related pancreatitis. J Gastroenterol 2001; 36:293-302. [PMID 11388391]

29. Marrache F, Hammel P, O'Toole D, Cazals-Hatem D, Vullierme MP, Sibert A, et al. Severe cholangitis following pancreaticoduodenectomy for pseudotumoral form of lymphoplasmacytic sclerosing pancreatitis. Am J Gastroenterol 2005; 100:2808-13. [PMID 16393239]

30. D'Onofrio M, Zamboni G, Tognolini A, Malago R, Faccioli N, Frulloni L, Pozzi Mucelli R. Mass-forming pancreatitis: value of contrast-enhanced ultrasonography. World J Gastroenterol 2006; 12:4181-4. [PMID 16830370]

31. Kitano M, Kudo M, Maekawa K, Suetomi Y, Sakamoto H, Fukuta N, et al. Dynamic imaging of pancreatic diseases by contrast enhanced coded phase inversion harmonic ultrasonography. Gut 2004; 53:854-9. [PMID 15138213]

32. Levy MJ, Reddy RP, Wiersema MJ, Smyrk TC, Clain JE, Harewood GC, et al. EUS-guided trucut biopsy in establishing autoimmune pancreatitis as the cause of obstructive jaundice. Gastrointest Endosc 2005; 61:467-72. [PMID 15758927]

33. Ozden I, Dizdaroglu F, Poyanli A, Emre A. Spontaneous regression of a pancreatic head mass and biliary obstruction due to autoimmune pancreatitis. Pancreatology 2005; 5:300-3. [PMID 15855829]

34. Czako L, Hegykozi E, Palinkas A, Lonovics J. Autoimmune pancreatitis: functional and morphological recovery after steroid therapy. World J Gastroenterol 2006; 12:1810-2. [PMID 16586561]

35. Horiuchi A, Kawa S, Akamatsu T, Aoki Y, Mukawa K, Furuya N, et al. Characteristic pancreatic duct appearance in autoimmune chronic pancreatitis: a case report and review of the Japanese literature. Am J Gastroenterol 1998; 93:260-3. [PMID 9468255]

36. Horiuchi A, Kawa S, Hamano H, Hayama M, Ota H, Kiyosawa K. ERCP features in 27 patients with autoimmune pancreatitis. Gastrointest Endosc 2002; 55:494-9. [PMID 11923760]