Case Series

Auto-immune pancreatitis with unusual presentations – A case series

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Abstract

Auto-immune pancreatitis (AIP) is a rare benign disease commonly presented with painless obstructive jaundice and biliary obstruction with rare complications like pseudocyst. We present a case series of two patients of AIP with unusual presentations; one case presented with periorbital swelling, jaundice, and pseudocyst, and the other case presented with abdominal pain and biliary obstruction without jaundice; both showed good response with steroids.

Keywords: Auto-immune pancreatitis, periorbital swelling, pseudocyst, steroids

Introduction

Autoimmune pancreatitis (AIP) is a benign disease and a eccentric form of chronic pancreatitis with autoimmune etiology,[1] and association with pseudocyst was relatively rare.[2,3] AIP was classified into two types. Type 1 AIP was associated with extra-pancreatic manifestations and IgG4-related disease, whereas type 2 is related to the granulocytic epithelial lesion. AIP accounted for 5–6% of all patients with chronic pancreatitis.[3] The common presentation is painless obstructive jaundice with an enlarged sausage-shaped pancreas on imaging. Uncommon presentations of this disease are further rare with plausible difficulty in suspicion and diagnosis. Nowadays, endoscopic ultrasound availability has made pancreatic biopsy feasible and so is the diagnosis of auto-immune pancreatitis. Auto-immune pancreatitis is a rare disease and may have unusual presentation like pseudocyst.

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Case Series

Case 1

A 36-year male was referred to our department with painless obstructive jaundice and fever. There was no history of weight loss and vomiting. On examination, the patient had bilateral periorbital swelling left > right [Figure 1]. Laboratory analysis showed an increased total bilirubin of 6.0 mg/dl, alanine aminotransferase (ALT) of 698 U/L, aspartate aminotransferase (AST) of 320 U/L, alkaline phosphatase (ALP) of 781 IU/L, aan elevated serum IgG4 concentration of 40.2 g/L (0.03 to 2.01 g/L), and an elevated total leucocyte count of 14400/μL. CA 19-9, CEA, and viral markers were normal. Computed tomography (CT) revealed a diffusely enlarged pancreas, with adjacent edema and hypo-enhancing rim. MRCP showed dilated CBD and intra-hepatic biliary radicals, with distal CBD stricture [Figure 2]. The patient underwent endoscopic retrograde cholangiopancreatography (ERCP) biliary stenting, where the cholangiogram showed distal CBD stricture with no

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calculi. Endoscopic ultrasound (EUS) showed a bulky pancreas with no mass, and the patient underwent a pancreatic biopsy. It showed storiform fibrosis and lymphoplasmacytic infiltration with IgG4-positive plasma cells 18/HPF [Figures 3 and 4]. After this, the patient lost follow-up for 10 months and was off any treatment. The patient later came to our hospital with clinical symptoms of mild abdominal pain and a lump in the epigastric region with a further increase in periorbital swelling. There was no evidence of fever or vomiting. Blood tests showed a total bilirubin of 1.6 mg/dl, AST of 125 U/L, ALT of 134 U/L, and ALP of 320 IU/L. Serum IgG4 was elevated to 38.7 g/L [Table 1]. CT showed a pseudocyst of size 100 × 66 mm in lesser sacs [Figure 5]. We started prednisolone at 40 mg once a day. At 6 weeks of follow-up, the patient improved symptomatically, periorbital swelling decreased, and the lump was not palpable. Review imaging with ultrasound abdomen showed that the pseudocyst decreased to size 2 cm.

Figure 1: Left eye > right eye periorbital edema (yellow arrow)

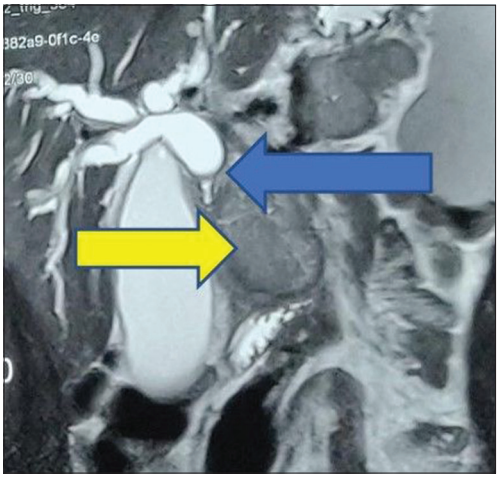

Figure 2: Magnetic resonance imaging showing the bulky head of the pancreas (yellow arrow) dilated proximal CBD, IHBRD, cystic duct, and distal CBD stricture (blue arrow) in the head region of the pancreas

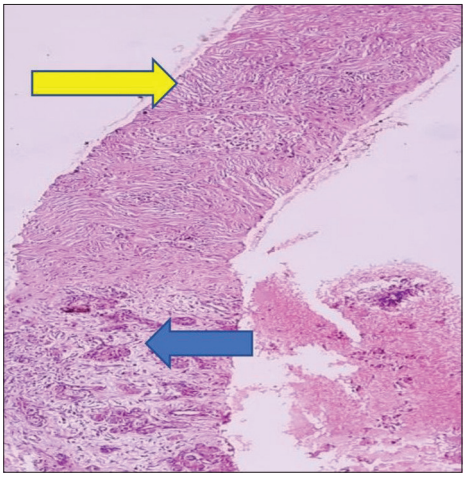

Figure 3: Histology showing normal pancreatic acini and duct (blue arrow) with storiform fibrosis (yellow arrow)

Case 2

A 66-year male was referred to our department with clinical symptoms of abdominal pain, post-prandial fullness, anorexia, and weight loss. There was no history of jaundice, fever, and pruritus. Laboratory analysis showed normal bilirubin (0.7 mg/dL), AST of 18 U/L, ALT of 11 U/L with elevated ALP (427 IU/L), and serum IgG4 of 7.85 g/L [Table 1]. CA 19-9 and CEA were normal. CT showed dilated CBD with abrupt termination at the pancreatic portion and bulky pancreas in the head region. Endoscopic ultrasound showed a heterogeneous lesion involving the head region of the pancreas along with a bulky pancreas throughout the body and the tail part. EUS-guided fine needle aspiration biopsy (FNAB) was performed and showed lymphoplasmacytic infiltration with IgG4-positive plasma cells >20 cells/HPF. We started steroids, and the patient showed improvement clinically with weight gain and a decrease in pancreatic size at 8 weeks.

Discussion

AIP is a benign disease with a good response to steroid therapy. AIP is commonly presented with biliary obstruction but rarely presents with unusual symptoms and rarely has complications like pseudocyst or carcinoma pancreas. The mechanisms of cyst formation are mostly because of the highly active inflammatory process and narrowing of the pancreatic duct and its branches.[4,5] They are only a few case reports and a series of AIP associated with pseudocyst, which are summarized in Table 2. Not all studies diagnosed and explained types of AIP associated with pseudocysts as our understanding of AIP has evolved only over the past 2 decades. The clinical presentation in our case series was painless obstructive jaundice with unusual manifestations like

Table 1: Investigations

Investigations	Case 1	Case 2
Total leucocyte count	14400/µL	-
Total bilirubin	6.0 mg/dl	0.7 mg/dl
Alanine aminotransferase	698 U/L	11 U/L
Aspartate aminotransferase	320 U/L	18 U/L
Alkaline phosphatase	781 IU/L	427 IU/L
IgG4	40.2 g/L	7.85 g/L
After treatment		
Total bilirubin	1.6 mg/dl	-
Alanine aminotransferase	134 U/L	-
Aspartate aminotransferase	125 U/L	-
Alkaline phosphatase	320 IU/L	-
IgG4	38.7 g/L	-

Table 2: Comparing studies of Autoimmune pancreatitis presenting with pseudocyst formation and its characteristics and treatment outcome

Study	Age	Sex	Symptom	Cyst location	Cyst size	Serum IgG4 levels	Initial treatment	Outcomes
Sohn *et al.*[1]	47	Male	Abdominal pain	Tail	20mm	228mg/dl	Steroids	Disappearance
Donet *et al*[2] Cases 17	54.8±10.9	Male -12, Female-5	Abdominal pain -15/17 Jaundice -3/17	Solitary , Head -4 Tail -6 Multiple -7	Small <3cm-4 cases Large >3cm-13 cases	14/17 cases-Elevated	Small Cysts – 4 cases –steroids Large cysts -13 cases-10/13-steroids 9 cases-2/9-Endoscopic drainage 7/9 cases-Surgery	Disappearance. Partial response-4/10 cases-large cyst
Yamamoto *et al*[3] Case 1	61	Male	Abdominal pain	Tail	60mm	1050mg/dl	EUS guided drainage	Resolved
Case 2	54	Male	Abdominal pain	Tail	100mm	290mg/dl	EUS guided drainage	Resolved
Case 3	63	Male	Abdominal pain	Tail	40mm	706mg/dl	Steroids -No response Surgery	Resolved
Case 4	64	Male	Abdominal pain	Tail	130mm	605mg/dl	EUS guided drainage	Resolved
Case 5	56	Female	Abdominal pain	Tail	25mm	184mg/dl	Steroids	Disappearance
Hesse *et al* [4]	73	Male	Painless jaundice	Body, tail	60×80mm	1190mg/dl	Steroids	Disappearance
Zhang *et al* [5]	48	Male	Abdominal pain	Tail	40×30mm	181mg/dl	Surgery	Resolved
Xu *et al* [6]	48	Female	Abdominal pain	Tail	30×20mm	165mg/dl	Surgery	Resolved
Chang *et al.*[7]	53	Male	Abdominal pain, Jaundice	Head, Body	45mm	4040mg/dl	Steroids and EUS guided drainage	Disappearance
Gompertz *et al.*[8]	63	Male	Jaundice, Pruritis	Head	40mm	804mg/dl	Steroids-No response. Surgery	Resolved
Takita *et al* [9]	26	Female	Abdominal pain	Tail	Approx 40mm	231mg/dl	Steroids-No response. Surgery	Resolved
Cui *et al* [10]	46	Male	Abdominal pain	Tail	23×17mm	1500mg/dl	Steroids	Disappearance
Nijs *et al*[11]	47	Male	Abdominal pain, Jaundice	Head	-	-	Steroids	Disappearance
Koizumi *et al.*[12]	75	Male	Abdominal pain	Body , Tail	15cm	820mg/dl	EUS guided drainage Steroids	Resolved
Matubayashi *et al* [13] 13 cases	62.2±11.9	Male-8, Female -5	Abdominal pain – 6 Jaundice - 4	Body , tail	Mean size >10mm	Mean >135mg/dl – 11 cases	Steroids	Response -6 cases, Partial response-1 case No response - 3 cases
Kubota *et al*[14] 12 cases	64.5	Male-11, Female-1	Abdominal pain -5/7 Jaundice – 4/8	Body, tail	Mean size-50mm		Corticosteroids -10/12 cases-Response 2/12 cases - Surgery	Disappearance
Ishikawa *et al*[15] 4 cases	63.2±13.5 y	Male -4	Abdominal pain	Body , tail	-	-	Corticosteroids - 3 cases-Response One case - Surgery	Disappearance
Kitoh *et al* [16]	66	Male	Abdominal pain	Tail	20mm		Steroids	Disappearance
Okamoto *et al* [17]	63	Male	Abdominal pain	Tail	-	-	Steroids	Disappearance
Nishimura *et al.*[18]	51	Female	Abdominal pain	Body , tail	80mm	410mg/dl	Steroids	Disappearance
Muraki *et al* [19]	53	Female	Abdominal pain	Head	35×25mm	227mg/dl	Steroids	Disappearance
Muraki *et al* [19]	55	Male	Abdominal pain	Body , tail	32×42mm	758mg/dl	Steroids	Disappearance
Muraki *et al* [19]	62	Male	Abdominal pain	Body , tail	25×20mm	265mg/dl	Steroids	Disappearance
Welsch *et al* [20]	64	Male	Abdominal pain	Head	50mm	-	Surgery	Resolved
Falck VG *et al.*[21]	59	Male	Abdominal pain	Tail	-	281mg/dl	Surgery	Resolved
Kawakami *et al* [22]	63	Male	Abdominal pain	Tail	40×30	1160mg/dl	Surgery	Resolved
Nishimura *et al.*[23]	47	Female	Abdominal pain	Tail	Large	623mg/dl	Steroids	Disappearance
Total cases -71		Total Males-54 Females -17	Abdominal pain-53 Jaundice-15 Pruritis-1	Head -9 Body-9 Tail-29			Steroid response -39 cases. EUS Drainage-8 cases Surgery-17 cases	

periorbital inflammation and pseudocyst in one and abdominal pain in another. In a few cases reports,[5-7] abdominal pain is the most common symptom, but isolated presentation with pain in the abdomen without jaundice is rare as in our second case.

In most of the studies of AIP with pseudocyst, the most common cyst location was the tail, followed by the body and head of the pancreas, whereas in our study, the location of the cyst was the body of the pancreas. In our case series serum, IgG4 levels are

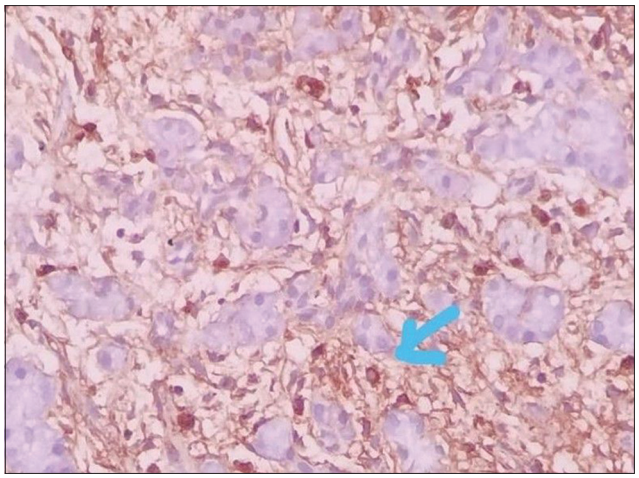

Figure 4: Immunohistochemistry showed IgG4-positive lymphoplasmacytic cells (sky blue arrow)

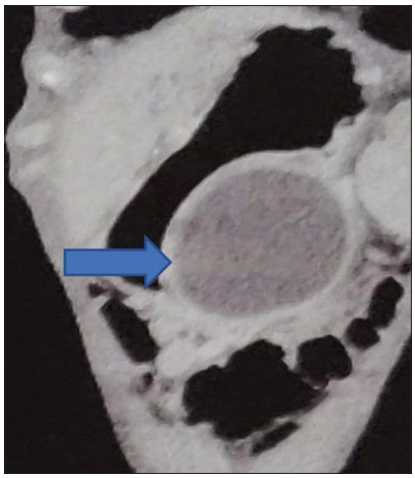

Figure 5: Contrast-enhanced computed tomography coronal view image showing fluid collection of approx. size 100 × 66 mm (blue arrow) with an enhancing wall seen in lesser sac anteroinferior to the proximal body of pancreas

elevated in both patients. The size of the cyst was 61 × 31 mm. From a therapeutic point of view, in a few instances, pseudocyst responded completely with conservative management with steroids, whereas in a few cases, there was no response to steroids and they underwent surgery. In one study, treatment response was found to be dependent upon the size of the cyst with size <3 cm responding completely to steroids, whereas a cyst size >3 cm responded with surgery. Despite the large size of the cyst in our patient, the pseudocyst resolved with conservative management with steroids. A few have used EUS-guided cyst drainage as a treatment option effectively along with steroids. There are no proper guidelines for the treatment of AIP with cyst formation. The steroids overall show clinical, serological, radiological, and extra-pancreatic lesion response of AIP in both our patients on follow-up. Presentation as only abdominal pain along with anorexia and weight loss as in one of our cases has been a standalone presentation. Maybe the patient has presented earlier in course of disease and eventually would have developed jaundice given that there was imaging evidence of biliary obstruction. This is a small case series, so generalization of findings is not possible.

Conclusion

AIP is a rare disease complicated with pseudocyst. Most of the studies suggested that surgical resection or EUS-guided drainage was the effective treatment for pseudocyst associated with AIP; a few studies recommended early initiation of steroids for better outcomes. More studies are required regarding the long-term prognosis and appropriate duration of steroids for AIP associated with pseudocyst. Our study showed good response with steroids.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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