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Early Onset of Pediatric Autoimmune Pancreatitis at the US-Mexican Border

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Case Report

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Abstract

Autoimmune pancreatitis (AIP) is an uncommon cause of pancreatitis that favorably responds to steroid therapy. Its true epidemiology remains unclear because only few cases of pediatric AIP have been reported. According to the adult criteria, AIP can be classified into 2 types: Type 1 or AIP without granulocyte epithelial lesions (GELs), and type 2 or AIP with GEL. Weexperienced two cases of pediatric AIP with unusually early age onset compared to previously reported data. With limited exceptions, our reported cases have a similar presentation, laboratory findings, radiographic findings, and positive response to steroid therapy compared to the previously reported cases. The main difference consisted in an early onset of disease (3-4 years of age) compared to the average age of onset of 11.9 years. A review of the current available reports demonstrated that more than 70 % of the cases of pediatric AIP could not be conclusively classified into the two types of AIP, raising the concern for a possibility of different pathophysiology between pediatric and adult AIP and the need for a pediatric-specific diagnostic criteria and treatment guideline for AIP.

Keywords: Autoimmune pancreatitis; Pancreatic head; Pancreatography; Ultrasonography

Introduction

Pancreatic disease in children is uncommon but comprises of diverse etiologies and carries significant morbidity and mortality [1]. Although it is rare, acute pancreatitis is the most common pediatric pancreatic disease and recent literatures have shown an increasing incidence in children [1-5]. Autoimmune pancreatitis (AIP) in the pediatric population is thought to be an uncommon cause of acute and chronic pancreatitis but the true epidemiology remains unclear and more cases have been recently recognized [6-9]. AIP can be classified into 2 types: Type 1 or AIP without granulocyte epithelial lesions (GELs), and type 2 or AIP with GEL. Type 1 is more commonly seen in elderly males with clinical presentation of obstructive jaundice, extra pancreatic involvement, and elevated serum immunoglobulin (Ig)G4. Pancreatic histopathology typically shows periductal dense infiltration of plasma cells and lymphocytes; storiform fibrosis; venulitis with lymphocytes and plasma cells frequently causing destruction of the affected vein; and profound IgG4 positive plasma cells (> 10 cells per high-power field [HPF]) [10]. Type 2 is more common in younger patients with equal sex distribution [10,11]. Unlike type 1, patients do not have elevated serum IgG4 or other organ involvement (OOI), and they are more

prone to present with abdominal pain, and have lower tendency to present with obstructive jaundice [10,12]. Type 1 and 2 can often be distinguished by serum IgG4 level, OOI, and histology. Both types have similar radiologic pancreatic imaging findings. Computed tomography (CT) or magnetic resonance imaging (MRI) is a commonly used method for pancreatic imaging. Diffusely enlarged pancreas (especially with a capsule-like rim) without pancreatic ductal dilatation or pancreatic low density mass is highly suggestive of AIP [10]. In comparison to adults, there have been no reported cases of AIP definitive type 1 in children and radiological findings seem to be less intense [6]. Early recognition of the disease is important since the few reports in the literature available for pediatric AIP shows a drastic response to steroid therapy, which leads to avoiding unnecessary invasive procedures in children [6-8,12-25]. Although there is a favorable response to steroids, some reported cases describe other autoimmune diseases later in life, particularly inflammatory bowel disease in AIP type 2 [6,17,25]. Up to our best knowledge, there are no defined guidelines for diagnosis and treatment of AIP in children. We will describe a small case series of early onset pediatric AIP and its comparison to the other reported cases in the literature.

Case 1

A previously healthy 4 year-old Hispanic male, presented with a four-day history of worsening severe epigastric pain and persistent emesis after being hit in the stomach by his friend. Past medical history and family histories were insignificant. Blood tests showed amylase 102 IU/L (normal range 25 – 115 IU/L), lipase 1208 IU/L (normal range 36 – 285 IU/L), and total bilirubin 0.5 mg/dL. Abdominal ultrasound (US) revealed a normal gallbladder, biliary tree, and pancreas. Magnetic resonance cholangiopancreatography (MRCP) showed peripancreatic, perihepatic and pericolic fluid, normal bile duct, and partially visualized pancreatic duct; the pancreatic duct in the region of the head was not visualized. The patient was treated with conservative management and was started on a low-fat diet after resolution of the abdominal pain, which the patient tolerated well. Since the pancreatic duct was not fully visualized, a repeat MRCP was scheduled 1 month later but the patient was lost to follow up.

Two years later, at the age of six, the patient re-presented with a four-day history of epigastric pain. He denied recent trauma and had been doing well during the interim. Blood tests showed amylase 88 IU/L, lipase 525 IU/L (normal range 36 – 285 IU/L), total bilirubin 0.4 mg/dL, HDL 37 mg/dL, triglyceride 89 mg/dL, cholesterol 148 mg/dL, normal liver function tests, ANA positive with titer of 1:160, and normal IgG4 level. Abdominal US revealed focal swelling of the head of the pancreas. MRCP showed mildly enlarged pancreas with mass-like effect at the pancreatic head causing partial obstruction of the distal common bile duct and pancreatic duct (Figure 1). An ultrasound guided percutaneous pancreatic core-biopsy showed benign inflammatory fibrosis with lymphoplasmacytic infiltration, frequent eosinophils, and parenchymal acinar destruction without any neoplastic process. Granulocytic epithelial lesions (GEL) causing duct disruption was not found and IgG4 stain was negative. The patient was started on daily oral Prednisone 20 mg. Resolution of the patient's symptoms was noted in less than 48 hours and a diagnosis of AIP probable type 2 was confirmed. At 1 month follow up, the patient's symptoms resolved and serum lipase normalized. A repeat abdominal ultrasound at that time showed resolution of the pancreatic inflammation and normalization of the pancreatic anatomy. Oral prednisone was continued for 6 months and then slowly tapered without reoccurrence of symptoms.

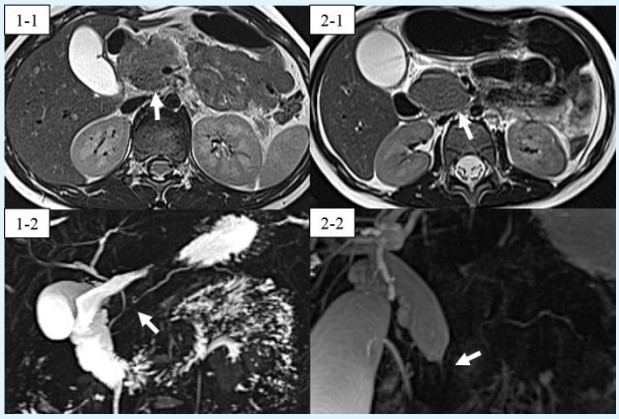

Figure 1: MRI results for case 1 and 2. 1-1 and 2-1 show a low T2 intensity mass-like effect at the pancreatic head (arrow). 1-2 shows partially obstructed pancreatic duct and distal common bile duct at the pancreatic head (arrows). 2-2 shows narrowing of the distal common bile duct at the level of the pancreatic head due to the mass-like effect (arrow).

Case 2

A previously healthy 3 year-old Hispanic female presented with a seven-day history of worsening epigastric pain, non-bloody non-bilious emesis, and mild scleralicterus. The family denied abdominal trauma, past medical history, and family history of gastrointestinal

pathology. Blood tests showed lipase 30IU/L (normal range 36 – 285 IU/L), total bilirubin 7.0 mg/dL, direct bilirubin 4.9 mg/dL, AST 122IU/L, ALT 232IU/L, ALP 1118IU/L, GGT 978IU/L, ANA positive with a titer of 1:160, and IgG4 level was normal. Studies for CMV, EBV, and hepatitis A, B, and C were negative. Liver-kidney microsomal antibody and smooth muscle antibody were negative. Abdominal ultrasound showed diffuse swelling of the pancreas with markedly dilated common bile duct; mild central intrahepatic biliary ductal dilatation; no pancreatic ductal dilatation; and no evidence of stones and cholecystitis. MRCP showed enlarged pancreas with mass-like effect at the pancreatic head causing partial obstruction of the common bile duct and pancreatic duct (Figure 1). AIP type 2 was highly suspected but the family refused pancreatic biopsy. The patient was started on Methylprednisolone 20 mg IV, which was eventually changed to daily oral Prednisone 20 mg. After 48 hours of steroid therapy, there was remarkable improvement in the patient's symptoms and laboratory markers and a diagnosis of AIP - not otherwise specified (NOS) was made. At 1 month follow up, the patient's symptoms had completely resolved and laboratory markers normalized. Abdominal ultrasound repeated at that time showed normal pancreatic anatomy without inflammation. Prednisone was gradually tapered after 6 months of therapy without reoccurrence of abdominal pain or symptoms at 12 months follow up.

Discussion

Autoimmune pancreatitis (AIP) is a rare entity among children and there are no well-established guidelines for its diagnosis and length of therapy. There are two types of histological patterns to AIP defined by the International Consensus Diagnostic Criteria (ICDC): lymphoplasmacytic sclerosing pancreatitis (LPSP) or AIP without granulocyte epithelial lesions (GELs) and idiopathic duct-centric pancreatitis (IDCP) or AIP with GEL. The term AIP type 1 and type 2 are used for clinical profiles of LPSP and IDCP, respectively, because pancreatic histology often is not obtained. AIP type 1 is an IgG4-related disease characterized by elevated serum IgG4 levels, involves other organs with infiltration of IgG4-positive plasma cells, and is more common in elderly Asian males [10,17,19]. Type 2 is more common in the younger population with no gender preference, has a normal serum IgG4 level, and is commonly associated with inflammatory bowel disease, especially ulcerative colitis [10,12,15,17,19]. Because type 2 does not have serological markers, in order to make a definite diagnosis, histological confirmation of IDCP is necessary [10]. Clinically, patients with type 2 are more likely to present with abdominal pain and are less likely to present with obstructive jaundice [17]. Of all the cases that have been reported for children, none have had "AIP definitive type 1" (Table 1) [6]. The treatment for AIP is steroid administration and generally patients show good response to treatment.

Table 1: Details of presentation, diagnosis, and treatment of AIP cases.

Reference number	Patient number	Age (years)	Sex	Symptoms at initial presentation			Serum Amylase / Lipase	Serum ANA	Serum IgG4 (mg/dL)	Radiologic imaging	Pathology		ICDC classification	Treatment			Comorbid conditions
				Abdominal pain	Jaundice	Weight loss					Method of Biopsy / Histology of the pancreas	IgG4 stain	GEL		Method	Response to steroid	Relapse to steroid	
# 6	1	11	M	+	+	+	WNL / ND	–	WNL	CT/ERCP: Diffusely enlarged pancreas with stricture of lower bile duct and pancreatic duct.	CT - guided / Lymphoplasmacytic infiltration with neutrophils infiltrating the lining epithelium of pancreatic ducts	Scarce	+	Definitive AIP-2	Steroid therapy	+	–	None
	2	14	F	ND	+	ND	WNL / ND	–	190	CT: Mass-like enlargement of the pancreatic head with lower bile duct stricture and caliber irregularity of the pancreatic duct.	CT - guided / Lymphoplasmacytic infiltration, periductal and lobular neutrophilic infiltation	–	+	AIP	Endoscopic papillotomy and biliary stent insertion.	No steroid therapy		UC 1 year later
# 7	3	10	M	+	+	+	WNL / WNL	–	WNL	CT: Pancreatic head mass and dilated common bile duct	PD / Ductocentriclymphoplasmacytic inflammation and fibrosis	31 cells/HPF	+	AIP	PD	No steroid therapy		None
	4	15	M	ND	+	+	WNL / WNL	–	WNL	CT: Pancreatic head mass with dilated common bile duct and pancreatic duct	Transduodenal laparoscopic TCB / Ductocentriclymphoplasmacytic inflammation and venulitis	10 cells/HPF	ND	Probable AIP-1	Steroid therapy	+	–	Developed Celiac disease 1 year later

	5	11	F	+	–	ND	High / High	–	Elevated	US: Mildly prominent pancreatic head and body, and dilated pancreatic duct ERCP: Irregularity and beading of pancreatic duct	EUS-FNA / Inconclusive	ND	ND	ProbableAIP -1	Observation initially but started steroids later for relapse.	+	+	None
# 8	6	14	F	+	ND	ND	High / ND	–	WNL	CT/MRCP: Pancreatic head enlargement. Dilated pancreatic duct with narrow distal portion.	Biospy not performed			AIP - NOS	Steroid therapy	+	+	None
# 9	7	11	M	+	–	–	WNL / High	–	WNL	MRCP: Focal swelling of the pancreatichead, delayed contrast enhancement, irregular pancreatic duct.	Laparotomy / Periductal fibrosis with lymphocytic and plasma cell infiltration.	ND	ND	AIP	ND			ND
#12	8	9	F	+	–	+	WNL / High	–	WNL	CT / MRCP / EUS: Pancreatic head enlargement and pancreatic duct dilatation, beaded appearance.	EUS - FNA / Lymphocytic infiltration, marked fibrosis, and acinar atrophy	–	–	Probable AIP -2	Initially treated with observation but relapsed. Steroid Therapy.	+	–	None
# 13	9	10	M	+	–	+	High/ WNL	–	WNL	US / MRCP: Enlarged pancreas, multiple stenoses of the pancreatic duct, and dilatation of biliary tract.	Not performed			AIP - NOS	Steroid therapy	+	–	None
# 14	10	7	M	+		ND	WNL / High	ND	WNL	CT / MRCP: Pancreatic head mass with irregular pancreatic duct.	CT - guided / Duct centric pancreatitis	ND	ND	AIP - NOS	Steroid therapy	+	–	None
#15	11	16	M	–	+	+	WNL / WNL	ND	WNL	CT/MRI: Enlarged head of the pancreas and dilated biliary tract	Laparotomy / Periductal lymphoplasmocytic infiltration	ND	ND	AIP - NOS	Steroid therapy	+	–	Diabete Mellitus
# 16	12	15	F	+	–	ND	ND / ND	ND	ND	Method ND:No pancreatic swelling. Main pancreatic duct stenosis.	Distal pancreatectomy / Idiopathic duct-centric pancreatitis	ND	ND	Definitive AIP-2	Distal pancreatectomy .	No steroid therapy		None
# 17	13	14	M	+	–	+	ND / WNL	+	Elevated	US : Bulky pancreatic head and body. MRCP: High T2 and low T1 signal surrounding the head and neck of the pancreas with common bile duct dilatation.	Laparotomy / ND	90 cells /HPF in lymph node biopsy	ND	Probable AIP -1	Steroid therapy	ND	ND	Developed UC 6 months later
	14	11	M	+	–	+	ND / High	+	Elevated	US / MRCP: Bulky pancreas. Biliary tract dilatation.	ND / ND	12 cells /HPF in liver biopsy	ND	Probable AIP -1	Steroid therapy	+	+	Developed IBD a few weeks later.
	15	7	M	+	–	ND	ND / High	–	WNL	US / MRCP: Diffusely enlarged pancreas.	ND / ND	> 10 cells /HPF in liver biopsy	ND	Probable AIP-1	Steroid therapy	ND	ND	Possible IBD
# 18	16	10	F	+	–	ND	High / High	–	WNL	MRCP: Diffusely enlarged pancreas and irregular main pancreatic duct.	Not performed			AIP - NOS	Steroid therapy	+	–	None
# 19	17	13	F	+	+	ND	WNL / WNL	–	WNL	CT / EUS / ERCP: Multiple pancreatic masses, stricture at the distal common bile duct, and biliary dilatation.	EUS - FNA / Lymphocytic inflammatory infiltrate	ND	ND	Probable AIP-1	Initially treated with pancreatic stent insertion but relapsed and started steroid therapy.	+	+	Fibrosing mediastinitis 3 months later

# 20	18	8	F	+	ND	+	WNL / WNL	ND	WNL	MRCP: Small volume pancreas and low-grade inflammation throughout. Collapsed pancreatic duct.	EUS - FNA / Insufficient tissue for analysis	ND	ND	AIP - NOS	Steroid therapy	+	–	None
# 21	19	15	F	+	+	–	ND / ND	–	WNL	US / CT / EUS / MRCP: Pancreatic head mass with delayed contrast enhancement. Enlarged and irregular main pancreatic duct.	EUS - FNA / Periductal lymphocytic infiltration and obliterative phlebitis.	> 10 cells /HPF in liver biopsy	ND	Probable AIP-1	Steroid therapy	+	–	None
# 22	20	10	M	ND					WNL	ND	ND / ND	–	+	Definitive AIP-2	Steroid therapy	+	ND	Chronic glomerulonephritis , Evan's syndrome
# 23	21	12	M	+	+	ND	WNL / ND	–	WNL	MRCP: Enlarged pancreatic head with obstruction of the common bile and pancreatic duct.	US guided percutaneous pancreatic corebiopsy / Fibrous tissue with lymphocytic and plasma cell infiltration	ND	ND	AIP	Biliary stent placement with pancreatic enzyme supplementation	No steroid therapy		None
# 24	22	10	M	+	+	+	ND / ND	ND	WNL	CT / MRCP / EUS: Pancreatic head mass, dilatation of the distal pancreatic duct, and dilated common bile duct with a taper at the level of the pancreas.	PD / Lymphoplasmacytic infiltration without out evidence of malignancy.	ND	ND	AIP	PD	No steroid therapy		None
# 25	23	9	ND	+	+	ND	ND / ND	ND	WNL	MRI / EUS: Diffusely enlarged pancreas with biliary stricture.	EUS - TCB / Features consistent with AIP	ND	+	Definitive AIP-2	Steroid therapy	+	–	None
	24	9	ND	+	+	ND	ND / ND	ND	WNL	MRCP / EUS: Diffusely enlarged pancreas and irregular pancreatic duct	EUS - TCB / Features consistent with AIP	ND	+	Definitive AIP-2	Steroid therapy	+	–	None
	25	14	ND	–	–	ND	ND / ND	ND	274	CT / EUS: Diffusely enlarged pancreas and irregular pancreatic duct.	EUS - TCB / Features consistent with AIP	ND	+	Definitive AIP-2	Observation	No steroid therapy		Crohn's disease
	26	16	ND	–	–	ND	ND / ND	ND	WNL	MRI: Diffusely enlarged pancreas. EUS: Irregular main pancreatic duct.	EUS - TCB / Features consistent with AIP	ND	+	Definitive AIP-2	Steroid therapy	+	–	None
	27	17	ND	–	–	ND	ND / ND	ND	WNL	CT / EUS / ERCP: Diffusely enlarged pancreas and irregular main pancreatic duct.	EUS - TCB / Features consistent with AIP	ND	+	Definitive AIP-2	Steroid therapy	+	–	None
	28	17	ND	–	–	ND	ND / ND	ND	WNL	EUS: Irregular main pancreatic duct.	EUS - TCB / Likely AIP	ND	+	Definitive AIP-2	Observation	No steroid therapy		None
	29	18	ND	–	–	ND	ND / ND	ND	WNL	EUS: characteristic features of AIP.	EUS- TCB / Nondiagnostic sample	ND	ND	Probable AIP -2	Observation	No steroid therapy		Crohn's disease
#26	30	14	F	Details unknown										AIP	Details Unknown			
	31	17	F											AIP				
Case 1	32	6	M	+	–	–	WNL / WNL	+	WNL	MRCP: Mildly enlarged pancreas with masslike effect at the pancreatic head causing partial obstruction of the distal common bile duct and pancreatic duct.	US guided percutaneous pancreatic corebiopsy / Benign inflammatory fibrosis with lymphoplasmacytic infiltration, frequent eosinophils, and parenchymal acinar destruction	–	–	Probable AIP -2	Steroid therapy	+	–	None

Case 2	33	3	F	+	+	–	NP / WNL	+	WNL	MRCP: Enlarged pancreas with mass-like effect at the pancreatic head causing partial obstruction of the distal common bile duct and pancreatic duct.	NP	AIP - NOS	Steroid therapy	+	–	None

ANA: Antinuclear antibody, Elevated: More than 2 folds of upper limit of normal, ERCP: Endoscopic retrograde pancreatography, EUS: Endoscopic ultrasonography, FNA: Fine needle aspiration, High: More than 3 folds of normal upper limit, HPF: High power field, IBD: Inflammatory bowel disease, MRCP: Magnetic resonance cholangiopancreatography, ND: Not described, NP: Not performed, PD: Whipple pancreaticoduodenectomy, TCB: Trucut biopsy, UC: Ulcerative colitis, US: Ultrasound, WNL: Within normal limit (Including less than 3 folds of normal upper limit for serum amylase and lipase, and less than 2 folds of normal upper limit for IgG4).

Table 1: Brief description of presentation, diagnosis, and treatment of previous cases of AIP and two cases we experienced.

*ANA: Antinuclear antibody, Elevated: More than 2 folds of upper limit of normal, ERCP: Endoscopic retrograde pancreatography, EUS: Endoscopic ultrasonography, FNA: Fine needle aspiration, High: More than 3 folds of normal upper limit, HPF: High power field, IBD: Inflammatory bowel disease, MRCP: Magnetic resonance cholangiopancreatography, MRI: Magnetic resonance imaging, ND: Not described, NP: Not performed, PD: Whipple pancreaticoduodenectomy, TCB: Trucut biopsy, UC: Ulcerative colitis, US: Ultrasound, WNL: Within normal limit (Including less than 3 folds of normal upper limit for serum amylase and lipase, and less than 2 folds of normal upper limit for IgG4).

An extensive review of 33 cases of AIP in children that were reported in English, including the two cases that we experienced, was performed (Table 1). The average age at diagnosis was 11.9 years and there was no apparent sex predilection. The most common symptoms encountered were abdominal pain followed by jaundice and weight loss. Only about half of the patients with reported laboratory results had serum amylase or lipase elevation of more than 3 fold increase from normal upper limits. ANA and serum IgG4 seems to be negative in majority of the cases. Regarding imaging, MRCP or computed tomography (CT) seems to be the most frequent ordered test that shows diffuse pancreatic swelling, mass at the pancreatic head, or both. Few cases performed endoscopic retrograde pancreatography (ERCP) and they showed irregular pancreatic duct. Endoscopic ultrasonography (EUS) was the most common method of biopsy, followed by CT-guided biopsy and percutaneous biopsy. Most patients were treated with steroids, show in grapid response to treatment, and only few patients had relapse. The dosage and length of therapy were variable. Most of the cases used oral Prednisone or Prednisolone 1 mg/kg/day or 20 – 40 mg/day for 3 – 6 months. Although AIP significantly responds to steroid therapy and pancreatic neoplasm is rare in children, 3 patients either received partial pancreatectomy or whipple pancreaticoduoden ectomy (PD) due to difficulty differentiating AIP from malignancy. Inflammatory bowel disease (IBD) either before or after presenting with AIP was the most common comorbid condition.

The present case series showed two different presentation of AIP in children. Both cases had similarities with other reported cases in the literature except for the age. Up to our best knowledge, there are no other cases of AIP that have been reported in children less than 7 years of age. As reported in most of the other pediatric cases of AIP, our patients had normal serum IgG4 levels. Both cases showed improvement after 48 hours of steroid therapy. Previously reported cases recommended the use of Prednisone or Prednisolone for 3-6 months. In our patients, we were able to wean off steroids at 6 months without recurrence in symptoms. Both of our patients have not presented with other autoimmune diseases most likely due to their young age and recent onset of the AIP. For case 1, following the ICDC, we were able to confirm a diagnosis of AIP probable type 2 based on imaging, histology, response to steroids, and no IBD association. For case 2, the patient most likely had AIP type 2 based on age, no OOI, and negative serology, but because the pathology was not available, she was diagnosed as AIP - NOS based on imaging, response to steroids, and no IBD association.

Conclusion

In conclusion, any patient with focal inflammation of the pancreatic head causing a mass-like effect should have AIP as part of the differential diagnosis. Although it may be a challenge to differentiate between AIP and malignancy, it is important to understand about AIP in the pediatric population to prevent invasive procedures, start anti-inflammatory therapy early, and to know the

association with other autoimmune diseases (i.e. inflammatory bowel disease) to decrease the morbidity when followed as an outpatient. Also, approximately 70% of the cases reported could not be clearly classified as either type 1 or type 2, and treatment methods differed depending on the reported cases, raising the question that there may be a need to create a pediatric-specific diagnostic criteria and treatment guideline for AIP.

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