

Autoimmune Pancreatitis - A Case Report

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ABSTRACT

Autoimmune pancreatitis (AIP) is a rare disease that has distinct histological, immunological, serological and radiological findings. It is characterised histologically by lymphoplasmacytic infiltrate, storiform fibrosis, obliterative phlebitis and presence of IgG4 positive plasma cells and lymphocytes. Elevated serum levels of IgG4 are also noted. It is usually misdiagnosed preoperatively as pancreatic cancer. It may involve extrapancreatic sites also and responds well to steroid therapy. Here, we share our experience of AIP in a 52-year-old male.

Keywords: IgG4, Pancreatic adenocarcinoma, Pancreatitis

CASE REPORT

A 52-year-old non alcoholic male gave a history of diffuse abdominal pain since 1 year. His blood sugar and LFT's were all under normal limits. A CECT scan was done and a necrotic mass in the superior aspect of pancreatic head partially compressing the portal vein and common bile duct extending caudally with near circumferential encasement was noted. Pancreatic CBD at the midpancreatic level associated with hydrops gall bladder, dilated supraduodenal common bile duct was seen [Table/Fig-1] and a diagnosis of mitotic necrotic pancreatic lesion was given and the patient was taken for surgery and a Whipple's procedure was done.

The specimen was received and histopathology slides were made. On H&E sections a dense lymphoplasmacytic infiltrate was seen in the pancreatic tissue. Obliterative phlebitis in the vessels was seen. There was dense storiform fibrosis seen in the stroma [Table/Fig-2]. Sections were taken for IHC. On IHC these cells were found to be IgG₄ positive [Table/Fig-3]. Hence, based on the H&E and IHC sections a diagnosis of AIP was made.

DISCUSSION

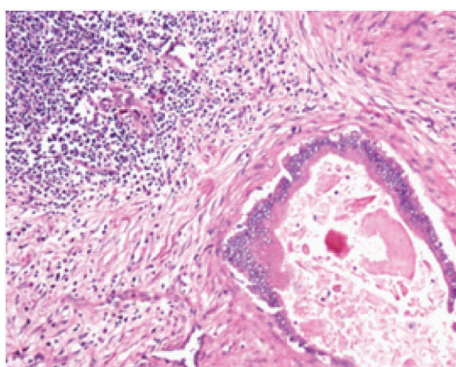
AIP is a very rare type of chronic pancreatitis that was first described in 1961 by Sarles et al., as primary inflammatory sclerosis of pancreas [1]. Various nomenclature were used to describe this disease. The term autoimmune pancreatitis (AIP) was coined by Yoshida et al., [2]. High serum IgG4 levels were noted in AIP patients [3] and this further led to the identification of infiltration of IgG₄ positive cells in the pancreas and also extrapancreatic organs [4]. AIP is a very rare form of chronic pancreatitis and is described as a pancreatic manifestation of systemic fibroinflammatory disease. Seen

predominantly in males over the age of 50 it is usually diagnosed in patients who undergo pancreatic resections for pancreatic cancers [5]. Various criteria were used to diagnose this entity with the Japanese Pancreatic Society criteria being described in 2006 [6]. Chari et al., proposed the Mayo Clinic (HISORT) criteria in 2006 which took into account the histology, imaging, serology, other organ involvement and response to therapy to reach a diagnosis of AIP [7]. Asian Diagnostic Criteria was proposed in 2008 [8]. Also known as lymphoplasmacytic sclerosing pancreatitis, chronic sclerosing pancreatitis, non alcoholic duct destructive pancreatitis, granulocytic epithelial lesion (GEL) positive pancreatitis AIP is a widely accepted terminology because the clinical, serological, histological and immunohistochemical findings point towards an autoimmune etiology. AIP could also show involvement of other organs such as the bile duct, liver, gall bladder, GIT, salivary glands, lacrimal glands, kidneys, retroperitoneum, lung, mesentery, breast, thyroid and the lymph node [9]. Histologically characterized by lymphoplasmacytic infiltrate, obliterative phlebitis, interstitial fibrosis and presence of IgG₄ positive lymphocytes and plasma cells, it responds well to steroid therapy [10]. Hamans et al., reported high serum IgG₄ levels in patients of AIP. Intense inflammation of IgG₄ positive cells was also noted in pancreas and a plethora of other organs by Kamisawa [4]. This was a turning point as it led to the conclusion that AIP was a part of systemic diseases known as IgG₄ associated disease (ISD) [4].

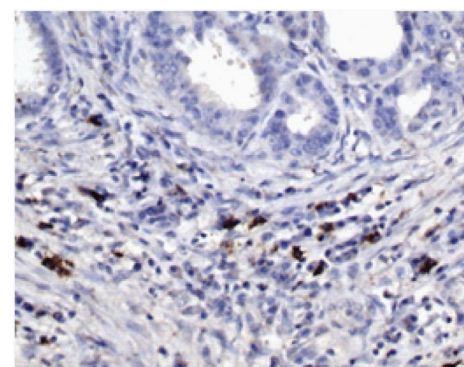
Seen mostly in males in the fourth and fifth decade, with abdominal pain, weight loss and jaundice being the clinical symptoms they mimic pancreatic cancers radiologically [9] and are diagnosed in patients on postoperatively on biopsy who have undergone



[Table/Fig-1]: CECT Scan



[Table/Fig-2]: H&E section



[Table/Fig-3]: IHC showing IgG4 positivity

pancreatoduodenectomy for suspected pancreatic cancers [5]. Our case was a 52-year-old male whose chief-complaint was a vague abdominal pain since one year. On CT scan a diagnosis of pancreatic cancer was made and he was taken up for resection. On H&E there was a dense lymphoplasmacytic infiltrate, obliterative phlebitis and storiform fibrosis. His H&E findings were classical of those seen in AIP [9]. To reinforce our diagnosis IgG₄ staining was done and IgG₄ positive lymphocytes and plasma cells were seen. A serum IgG₄ could not be done as a preoperative differential of AIP was not considered. HISORt criteria was applied [7]. Since our case showed lymphoplasmacytic sclerosing pancreatitis (LPSP) with more than 10 IgG₄ positive cell / hpf the group A histology criteria of HISORt was fulfilled and a diagnosis of AIP was made. Hence AIP is a form of chronic pancreatitis seen in males above the age of 50. It is a diagnostic challenge and it mimics pancreatic cancer. It forms a part of IgG₄ related systemic diseases [4].

CONCLUSION

Our aim was to make the pathologist, radiologist and the surgeon aware of this rare entity as it shows a dramatic response to steroid therapy and saves the patient from the unnecessary morbidity and mortality of pancreatic resections.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Mar 11, 2014**
Date of Peer Review: **May 27, 2014**
Date of Acceptance: **Oct 6, 2014**
Date of Publishing: **Feb 01, 2015**