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Autoimmune Pancreatitis: A Case Report

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Abstract

Autoimmune Pancreatitis is an uncommon form of chronic inflammation of the Pancreas. It is an autoimmune disease and so it can be associated with other autoimmune disorders. There are 2 types of Autoimmune Pancreatitis (AIP) with type 1 being the more common form. We reported a case of Autoimmune Pancreatitis where the patient presented with recurrent episodes of acute Pancreatitis managed conservatively until he was diagnosed with autoimmune pancreatitis type 2 based on clinical picture, lab investigations and radiological findings.

Keywords: Autoimmune Pancreatitis, AIP, Acute Pancreatitis, Magnetic Resonance Imaging, MRI, Magnetic resonance cholangiopancreatography, MRCP, Computed Tomography, CT.

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1. Introduction

Autoimmune pancreatitis is an uncommon cause of chronic and recurrent pancreatitis and is likely an under-diagnosed condition. It is reported to account for up to 2% of all cases of chronic pancreatitis, with a prevalence of less than 1 per 100,000 population [1]. There are two major classifications of AIP that are defined by unique features, Type 1 AIP, also known as lymphoplasmacytic sclerosing pancreatitis (LPSP), typically present in late adulthood with a mean age of diagnosis of 50 years and older and affects males three times more commonly than females [2]. Type 2 AIP, also known as idiopathic duct-centric pancreatitis (IDCP), affects males and females equally and has a younger mean age of diagnosis (43 years) compared to Type 1 AIP [3]. Radiographically, the morphology of AIP can appear like pancreatic cancer, if untreated, AIP can lead to pancreatic insufficiency, fibrosis, and other complications [4]. Immunoglobulin G4 (IgG4) is used as a biomarker of AIP especially for the first type of AIP but IgG4 levels could be normal in AIP type 2 and elevated in other cases like in pancreatic cancer [5]. Clinical features of AIP could painless jaundice, abdominal pain and recurrent episodes of acute Pancreatitis but it could also be asymptomatic [6].

2. Case Report

YKH is a 27-year-old male, previously healthy man who presented to the hospital on 14th March 2021, with 3 days history of gradual epigastric pain, radiating to the back, associated with nausea and anorexia. There was no history of fever, or change in urinary or bowel habits. The patient has never smoked or consumed alcohol and does not take any recreational drugs. The patient denies any history of previous hospitalization for any reason. He has an insignificant surgical and medical history. He has family history of Diabetes.

On physical examination, the abdomen was soft with mild epigastric tenderness. On the ultrasound of the abdomen, there was no cholelithiasis and no intrahepatic bile duct dilatation with a normal common biliary duct. There was mild altered pancreatic echotexture. The patient was then diagnosed with Acute Pancreatitis. He was treated conservatively, kept on a fat-free diet plan and his pain was managed. The patient was discharged after two days on 16th March on analgesics and Creon capsules.

On 30th March 2021, YKH was seen in the surgical clinic as a follow-up for his previous complaints. He had no current complaints. His laboratory test results revealed normal White Blood Cell count at 7.8^3/uL, normal Haemboglobin level of 14.1g/dl, normal platelet count of 236^3/uL, high C-Reactive protein of 5.6 mg/L and elevated Lipase of 387 U/L. All other lab investigations were normal.

On 17th July 2021, YKH presented to the emergency department complaining of abdominal pain of moderate severity which started one day before admission. This was the patient's third visit with the same complaints. This abdomen was soft and lax with mild epigastric tenderness. His laboratory test results this visit showed normal renal function with Creatinine of 0.8mg/dl, eGFR of 122.4

mL/min/1.73m(2) and urea of 9 mg/dl. His C-reactive protein was elevated at 23.6 mg/L. His white blood cell count was normal at 8.4 ^3/uL, his hemoglobin was low at 12.1g/dl. He had normal GGT level at 12 U/L, elevated Lipase at 472 U/L, normal Amylase at 134 U/L and normal Total Bilirubin at 0.7 mg/dl. All other lab tests were normal. The patient received analgesia and Intravenous fluids and although he was advised to stay for continued hospital care, he refused admission.

On 23rd July the patient returned to the emergency due to abdominal pain that was mild in severity that started 3 days back and associated with diarrhea complaining of watery stool 4 times that day. He had abdominal tenderness in the epigastric area and periumbilical areas. He was admitted with acute pancreatitis. An MRI of Pancreas (Figure 1) was performed showing diffuse edematous pancreas with diffusion restriction involving body and tail, interval progression and minimal prominent pancreatic duct at the body and tail region. An Endoscopic Ultrasound was then performed showing bulky parenchyma of pancreas that is sausage shaped with multiple small benign-looking lymph nodes that are in the peripancreatic area. A fine needle aspiration was taken from the pancreas but was inconclusive on histopathology.

On 21st September 2021, on the follow-up clinic appointment YKH was still complaining of recurrent abdominal pain and gaseous distention radiating to his back. He was avoiding food because of pain leading to a weight loss of about 13 Kg during the last two months. There was no history of nausea or vomiting. His Lipase was elevated at 537 U/L, hemoglobin was low at 13.4 mg/dl and white blood cell count was normal at 7.9x10^9/L. All other lab investigations were normal. Taking in consideration of the clinical picture, normal IgG4, diffusedly enlarged pancreas on imaging and inconclusive EUS, a diagnosis of type 2 autoimmune pancreatitis was concluded. YKH was then started on Prednisolone 40 mg for one month then tapering the dose. Afterwards, he had no recurrence of abdominal pain and his lab investigations improved.

Due to non-compliance, the treatment was interrupted and there was recurrence of symptoms. The patient travelled to Thailand, where he was seen by a Gastroenterologist, the Fine Needle Aspiration of the pancreas was done, and the result was confirmatory of Autoimmune Pancreatitis, so he was kept on Prednisolone in addition to Azathioprine. 2 weeks after arrival in UAE he developed an episode of acute pancreatitis, which was considered as an adverse effect of Azathioprine. After stopping Azathioprine and only continuing Prednisolone, his symptoms improved and he had no further attacks of pancreatitis. The MRI of Pancreas done in February 2022 (Figure 2) has shown dramatic improvement in the pancreatic morphology, and Prednisolone tapering was continued as he is presently on Prednisolone 5 mg daily with no symptoms and his recent laboratory investigations were all normal.

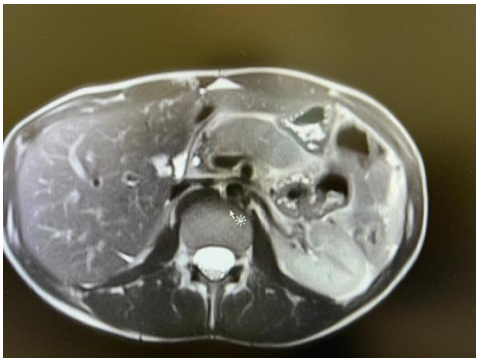

Figure 1: YKH's MRI of Pancreas before treatment, showing diffuse edematous pancreas with diffusion restriction involving body and tail, interval progression and minimal prominent pancreatic duct at the body and tail region

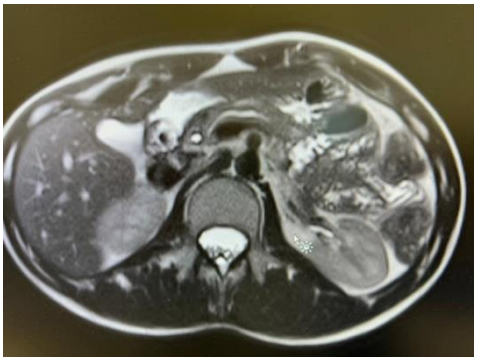

Figure 2: MRI of Pancreas after Glucocorticoids treatment showing improvement in the pancreatic morphology

3. Discussion

There are two widely utilized diagnostic criteria for Autoimmune pancreatitis. The first criteria include three components namely imaging, serology, and histology [7]. The Mayo Clinic criteria consist of five components, including Histology, Imaging, Serology, Organ involvement, and Response to steroid therapy [8].

Clinically, autoimmune pancreatitis can manifest with symptoms related to biliary or pancreatic involvement, Patients can present with recurrent episodes of abdominal pain, with or without attacks of pancreatitis, Obstructive jaundice is a common presentation that can be accompanied by non-specific symptoms such as nausea, vomiting, loss of appetite, or weight loss depending on the degree of biliary or pancreatic strictures and involvement [9].

The clinical picture of both types of AIP are similar but the second type of AIP is confined to the Pancreas but may also display an elevation in liver tests resembling Cholestasis causing an elevation in IgG4 levels [10]. Type 2 AIP can be associated

with IBD. While Type 1 AIP can present as an IgG4-related disease with other organ involvement including sclerosing cholangitis, Sclerosing sialadenitis and Sclerosing dacryoadenitis [11].

Magnetic resonance cholangiopancreatography is recommended for evaluation of the pancreatic duct in patients with suspected AIP [12]. As compared with abdominal CT, MRI has a higher sensitivity but a similar specificity in distinguishing AIP from pancreatic ductal adenocarcinoma [13]. Imaging should include the head, neck, thorax, abdomen, and pelvis to evaluate for involvement of other organs associated with IgG4-related disease (IgG4-RD) [14].

Endoscopic ultrasonography could be performed and a biopsy could be sent for diagnostic histology to rule out Pancreatic cancer and could also help in diagnosis of both types of AIP [15]. On Histology, in type 2 AIP there could be Idiopathic duct-centric pancreatitis (IDCP) with a granulocytic epithelial lesion in the pancreatic duct with minimal IgG4-positive cells in the pancreatic parenchyma [16].

Corticosteroid therapy are given to most patients, which leads to improvement of symptoms as well as a reduction in complications of AIP, in patients presenting with obstructive jaundice and sclerosing cholangitis, early initiation of corticosteroids is recommended [17]. It has been suggested that a lack of improvement with corticosteroids could hint towards the possibility of alternate diagnoses such as pancreatic malignancy, the lack of response to a glucocorticoid trial within two to three weeks warrants referral for surgical exploration for pancreatic cancer [18].

Relapse of AIP or failure to wean steroids may occur in up to half of the patients, especially in IgG4-related AIP requiring long-term steroids or use of steroid-sparing agents. Azathioprine has been shown to be effective in patients to help in weaning steroid therapy [19]. Rituximab can be utilized for both induction and maintenance therapy and is the only choice for patients who are intolerant or resistant to steroids and immunomodulators. Induction involves either 4 weekly doses of 375 mg/m2 body surface area (BSA) or 2 doses of 1000 mg each administered 2 weeks apart [20].

The long-term prognosis of AIP is not yet unknown, many patients with AIP were discovered to have malignancies either at the time of AIP diagnosis or within one year [21]. The three most commonly diagnosed conditions were gastric, colorectal, and bladder cancer [22]. The underlying mechanisms are not known. It is hypothesized that there may be a component of paraneoplastic syndrome associated with AIP [23]. Moreover, a small study of 63 patients suggested that patients with Type 1 AIP have an elevated risk for pancreatic cancer similar to patients with chronic pancreatitis [24]. Further studies are needed to define the risk and relationship between AIP and malignancy. The effect of AIP on mortality also remains an area yet to be explored [25].

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