ORIGINAL RESEARCH PAPER

General Surgery

AUTOIMMUNE PANCREATITIS - A DOPPLEGANGER

KEY WORDS: autoimmune pancreatitis, pancreatic cancer, IgG4,steroids

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ABSTRACT

Evaluating mass in the head of pancreas is clinically challenging. We herein report a case of 46 year old male patient ,chronic alcoholic ,smoker, with history of upper abdominal pain ,significant loss of weight and epigastric mass for six months .On further evaluation ,he was provisionally diagnosed with carcinoma head of pancreas and proceeded with pancreaticoduodenectomy. Mass lesion in the head of pancreas can be due pancreatic carcinoma or secondary to inflammation. Recognising this is crucial to avoid unnecessary surgery.

INTRODUCTION:

Autoimmune pancreatitis is a rare pancreatic manifestation of IgG4 related disease. It is characterized by irregular narrowing of the main pancreatic duct ,lympho plasmacytic infiltration of the pancreas .The presence of focal mass in the pancreas due to hypermetabolic activity in the autoimmune pancreatitis clinically mimics pancreatic cancer. Differentiation between autoimmune pancreatitis and pancreatic cancer is crucial because the clinical course, treatment and prognosis of these two disease entities are quite different. AP responds well to steroid therapy, while pancreatic cancer may require surgical intervention with or without adjuvant chemotherapy. Despite having diagnostic criteria for AP ,clinical and radiological differentiation between AP and pancreatic cancer remains challenging. Hereby reporting a case of AP who presented with pancreatic head mass that mimicked pancreatic cancer.

Case Report:

A 46 year old, thin built, male, chronic alcoholic, smoker, presented with history of upper abdominal pain which was not associated with food intake , vomiting, loss of appetite and significant loss of weight for six months. On examination 10×6 cm non tender ,firm, epigastric mass extending into right hypochondrium and umbilicus was felt. Laboratory investigation revealed elevated total bilirubin 10.7 mg/dl and direct bilirubin 7.6mg/dl , other parameters were within normal limits. CECT abdomen showed heterogeneous enhancing mass lesion in the head of pancreas with prominent vascularity in the periphery of lesion with coarse calcification with MPD dilatation and CBD dilation of 1.3cm, with multiple enlarged peripancreatic para,aortocaval lymph nodes. A malignant pancreatic tumor was suspected.

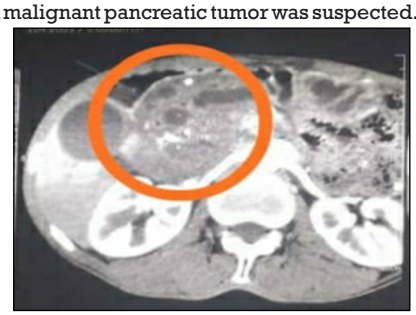

Fig.1: CECT Abdomen Showing Heterogeneous Enhancing Mass Lesion In The Head Of Pancreas.

Tumor markers CA 19 – 9 : 262u/ml & chromogranin 622ng/ml, elevated. Image guided biopsy attempted turned out to be inconclusive Esophagogastroduodenoscopy revealed pangastritis and extraneous compression along the lesser curvature,which raised suspicion of pancreatic cancer. Hence patient was clinically diagnosed with carcinoma head of pancreas and proceeded with pancreaticoduodenectomy.

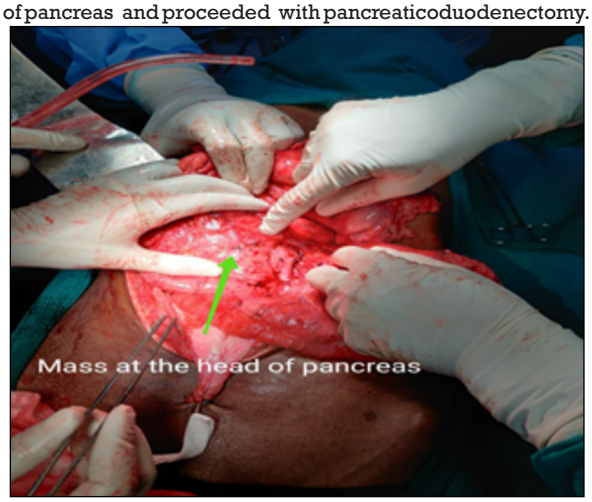

Figure.2: Intra Op Images Showing Mass At The Head Of Pancreas

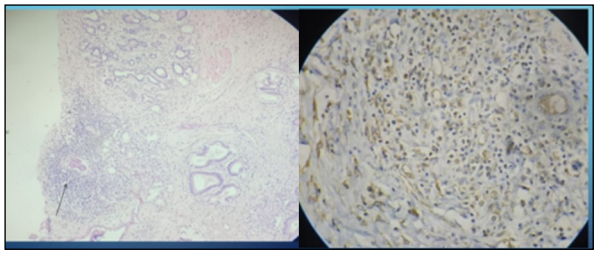

Figure 3: 3a - HPE image shows pancreatic acini replaced by neutrophils and plasma cells and 3b- IHC showing IgG4 positivity in plasma cells.

Intra operatively diffusely enlarged pancreas with altered texture with firm mass of 10x 10 cm in the head of pancreas with extensive peripancreatic edema and multiple enlarged peripancreatic and para aortic nodes identified. Resected

specimen sent for histopathological examination showed most of the pancreatic acini being replaced by sheets of neutrophils and plasma cells ,surrounding stroma with myxoid degeneration. On IHC, IgG4 was positive in more than 50 plasma cells in high power field and turned out to be type 1 IgG4 related autoimmune pancreatitis Post operatively patient was started on steroids and doing well on follow up .

DISCUSSION :

Autoimmune pancreatitis, also known as lymphoplasmacytic sclerosing pancreatitis, is a rare cause (1%) of chronic pancreatitis. It occurs in both sexes, however, it is predominant in men (male: female ratio of 3:1). With respect to age distribution, AP occurs mostly in elderly patients. Clinically, patients with AP may present with obstructive jaundice and abdominal pain, and imaging examination may reveal a mass. The clinical symptoms of localized AP, mimic those of pancreatic cancer. However, the clinical course, treatment, and prognosis are quite different between AP and pancreatic cancer. AP is classified into type 1 or type 2 depending on its histopathological findings. Type 1 AP, characterized by dense infiltration of T lymphocytes and IgG4-postive plasma cells, storiform fibrosis, and obliterative phlebitis. Serum IgG4 is elevated in type 1 AP and responds to steroids. The main histological feature of type 2 AP (idiopathic duct-centric pancreatitis) is neutrophilic infiltration in the epithelium of the pancreatic ducts. Absence of IgG4 elevation is typical of type 2 AP. In 2008, Korean and Japanese pancreatologists revised the diagnostic consensus on AP .Five cardinal features are used to diagnose AP and they are imaging findings of the pancreatic ducts and parenchyma, serology, involvement of other organs, pancreatic histology, and clinical responsiveness to steroid therapy. The typical imaging finding of the pancreatic ducts and parenchyma is diffuse enlargement with delayed enhancement, sometimes associated with rim-like enhancement.

According to established guidelines,steroid treatment is the standard therapy for AP. Initially, oral prednisolone (30–40 mg/day or 0.6 mg/kg per day) is administered for 2 to 4 weeks; thereafter, the dose is tapered by 5 mg every 1 to 2 weeks. This treatment may be continued for 3 to 6 months while carefully monitoring the patient's symptoms and biochemical, serological, and imaging ndings until a maintenance dose is reached. Morphological and serological evaluation should be performed to assess the disease responsiveness after the initial steroid therapy.

CONCLUSION:

The prevalence of autoimmune pancreatitis is relatively low compared to pancreatic cancer . Focal mass forming AP mimics pancreatic cancer as in our case ,challenging to diagnose. Serum IgG4 should be considered in the evaluation of doubtful cases of pancreatitis with mass lesion .In patients with high suspicion of AP ,trial therapy with steroids and immunosuppresants can be considered .

Abbreviations:

AP – autoimmune pancreatitis
CECT – contrast enhanced computed tomography
HPE – histopathological examination
IHC – Immune HistoChemistry
CBD – common bile duct
MPD – main pancreatic duct
Conflict of interest – nil
Funding – nil
Ethical committee approval – not required

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