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Case Report

A rare case of autoimmune pancreatitis with pancreatic divisum

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ABSTRACT

Chronic pancreatitis has a fibro-inflammatory subtype called autoimmune pancreatitis. A combination of imaging tests, including a CT scan and pancreatography, lab tests that check for IgG4 and/or autoantibodies, histological analysis, and a favourable response to corticosteroid treatment are used for diagnosis. Hereby we present a case of a young boy who presented to our hospital with recurrent abdominal pain and on further investigation was found to have elevated levels of IgG4 as well as a developmental abnormality of pancreatic duct known as pancreatic divisum.

Keywords: Autoimmune pancreatitis, IgG4, Pancreatic divisum

INTRODUCTION

A fibro-inflammatory variant of chronic pancreatitis called autoimmune pancreatitis makes for 5-6% of all cases of chronic pancreatitis.

It is divided into two groups.[1] Although it affects both sexes, type I is more common in men and is linked to hyperimmunoglobulin G4, anti-nuclear, anti-smooth muscle, anti-lactoferrin, and anti-carbonic anhydrase antibodies.[2,3] Type I autoimmune pancreatitis frequently has extra-pancreatic symptoms and is linked to other autoimmune illnesses. In type II, no specific autoantibodies nor extra-pancreatic symptoms are linked.

Even though high IgG4 levels have been recorded in pancreatic cancer and chronic pancreatitis, type I autoimmune pancreatitis can be distinguished from other pancreatic illnesses by increased IgG4 to levels above 135 mg/dl.

However, pancreatic hypertrophy and pancreatic duct narrowing should be taken into account when diagnosing autoimmune pancreatitis.

According to histopathological analyses, the pancreatic ducts have lymphocytic and plasmacytic infiltration, lymphoid follicles, fibrosis, a rise in the thickness of the intralobular septate, and obliterans phlebitis.

The presence of IgG4+ plasma cell infiltration around the pancreatic ducts and an acceptable response to corticosteroid therapy helps distinguish this disorder from primary sclerosant cholangitis, despite the fact that it can cause extra-pancreatic lesions that are identical to that condition. The prognosis as a whole is vague.

The colon, duodenum, and stomach may all have localized infiltration of plasma cells, according to an endoscopy.[4] Other autoimmune conditions like ulcerative colitis or autoimmune hepatitis may coexist with autoimmune pancreatitis.

Images from a CT scan can reveal a variety of problems, such as a normal pancreas, a pancreatic tumour, or local or diffuse pancreatic enlargement.[5]

The presence of localized moderate lymphadenopathy is also typical. The head of the pancreas is where focal involvement most frequently occurs. The absence of extensive pancreatic duct stenosis in pancreatic carcinoma

cases should be used to distinguish autoimmune pancreatitis from pancreatic carcinoma.[6,7]

The existence of local or widespread pancreatic duct stenosis, which may be connected to biliary duct stenosis, is shown by cholangiopancreatography.[8]

Radiologic, analytical, and histopathologic results are all combined to make the diagnosis of autoimmune pancreatitis.

Here we present a case of a young boy who presented to our hospital with recurrent bouts of abdominal pain and vomiting and tenderness in the umbilical and the periumbilical region, and the unique finding of our patient having pancreatic divisum.

CASE REPORT

A 13-year-old child had repeated episodes of abdominal pain for the past one year. Two years back, he was admitted with severe abdominal pain on the umbilical and periumbilical region which was associated with vomiting and food intake and the pain used to get settled if the patient was kept NPO.

There was a history of re-current hospitalization with above mentioned complains. The patient kept getting bouts of abdominal pain 2 to 3 times a month, thus the patient was referred to our hospital for further Workup.

There was no significant personal, family and drug history. On work up in our hospital routine blood reports were all normal.

Serum amylase and lipase were checked and were found to be elevated: Amylase-960, lipase-2370.

Serial USG was done which was suggestive of acute necrotizing pancreatitis.

In the view of repeated exacerbation, CECT abdomen, gene study, IgG4 levels, MRCP was done.

Table 1: Additional investigations.

Variables	Findings
CECT abdomen (Figure 1)	Bulky edematous pancreas with multifocal areas of necrosis. Peripancreatic fluid present.
Gene study	CFTR, SPINK, PRCSI mutation negative
IgG4	2.74 gm/L
MRCP (Figure 1)	Bulky pancreas with peripancreatic inflammation. Pancreatic division present. Mildly dilated main pancreatic duct (3 mm) suggestive of acute edematous pancreatitis.

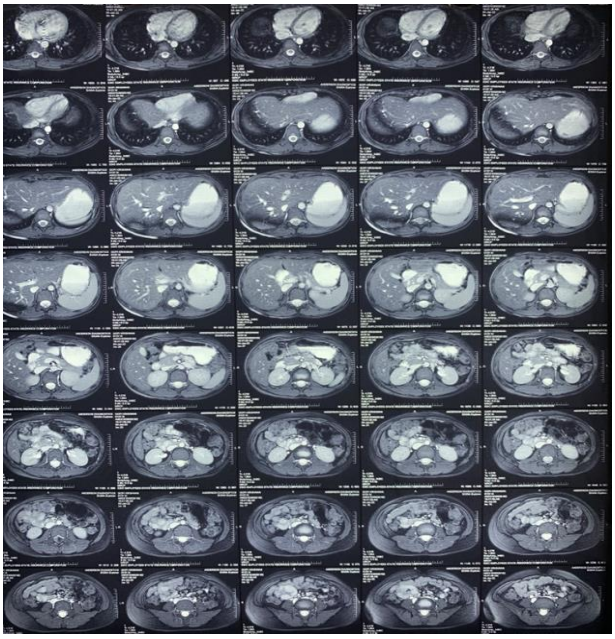

Figure 1: Bulky edematous pancreas with multifocal areas of necrosis. Peripancreatic fluid present.

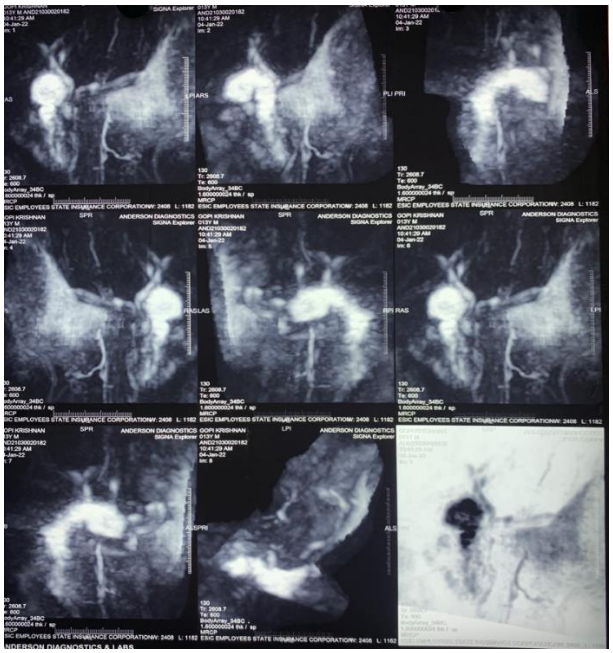

Figure 2: Bulky pancreas with peripancreatic inflammation. Pancreatic division present. Mildly dilated main pancreatic duct (3 mm) suggestive of acute edematous pancreatitis.

Therefore, keeping autoimmune pancreatitis in mind, our patient was started on prednisone 1 mg per kg, with tapering doses every 10th day. In the view of pancreatic division, the patient underwent ERCP and successful stent placement was done.

Currently our patient is on a stable dose of 5 mg prednisone and is on remission and regular follow up.

DISCUSSION

Males are more likely to have autoimmune pancreatitis, which frequently manifests as mild stomach pain without the usual pancreatitis pain and occasionally also includes obstructive jaundice.[1] Increased levels of hepatobiliary and pancreatic enzymes may also exist.[9] As a result, the pancreas' endocrine and exocrine functioning might be compromised.

Histopathological analysis of biopsies (gold standard) and autopsies reveals intralobular fibrosis, lymphocytic and IgG4+ plasma cell infiltration, and obliterans phlebitis. In alcoholics and those with Sjögren's syndrome who have chronic pancreatitis, none of these features are evident. 13-16 A close relationship with mutational fibrosclerosis is strongly suggested by specific inflammatory alterations in autoimmune pancreatitis patients.

The most common organ, second to the pancreas involved in people suffering from IgG4 disease is cholangitis, which is observed in 83% of the patients. IgG4 cholangitis is characterized mostly by jaundice, as well as cytolysis and cholestasis

IgG4-RD is a corticosteroid-sensitive disorder with a 97% to 100% response rate, and the same was observed in our patient. [10] Although the most common age of diagnosis is the 6th decade of life, our patient was a young boy, and also had a anatomical variance of the pancreatic duct which is rarely ever seen.

IgG4 pancreatitis is a recurring pathology, with a relapse incidence ranging from 26-70% after stopping corticosteroids.[10]

CONCLUSION

The IgG4-related disease is a newly discovered and poorly recognized condition. It was recognized as a unique systemic disease entity around 20 years ago.

The most frequent involvement is in the pancreas and biliary system. They primarily mimic cholangiocarcinoma and pancreatic cancer.

Numerous diagnostic criteria have been approved by international consensus in order to prevent diagnostic errors. To further define IgG4 disease and close gaps in our knowledge of pathogenesis and diagnosis, more prospective trials are required.

As our patient was a young male, autoimmune pancreatitis should be suspected and high clinical suspicion should be present even in young patient presenting with recurrent abdominal pain.

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