ABSTRACT Background: Pancreas divisum is one of the rare causes of recurrent pancreatitis. Diagnosis is challenging and diagnostic failure leads to recurrent episodes of pancreatitis and ultimately pancreatic failure. Case: Here we are presenting a case of a young female with a history of recurrent attacks of upper abdominal pain which on work up was diagnosed as pancreas divisum. This patient was successfully treated with ERCP with minor papillotomy followed by stenting in major papilla. Written informed consent was taken from the patient before writing this case report. Conclusion: This report emphasises the need for early suspicion of disease, in patients with recurrent idiopathic pancreatitis, by clinician and efficacy of minimally invasive procedures (ERCP) as a definitive treatment option.

KEYWORDS Pancreas Divisum, Recurrent pancreatitis, Endoscopic Papillotomy

Introduction

Pancreas divisum is one of the most commonly encountered congenital anomalies in the hepatobiliary system. The incidence is variable, but it ranges from 5% to 14% of the general population [1]. The disease occurs due to failure of fusion of the two embryonic parts of the pancreas i.e. non-fusion of dorsal and ventral ducts of pancreas resulting in openings at abnormal positions, This abnormal opening of primary pancreatic duct at minor papilla is not enough to completely drain the duct secretions resulting in stasis of enzymes and premature activation of these enzymes results in recurrent attacks of pancreatitis [2, 3].

Most of the patients are asymptomatic, and diagnosis is incidental. Abdominal ultrasound is not a very useful modality for diagnosis. Computed tomography (CT) of the abdomen may help in diagnosis, but imaging of choice is cholangiography (ERCP or MRCP). MRCP being better as it is noninvasive but ERCP has an advantage in its diagnostic as well as therapeutic capacity. Treatment is usually nonsurgical in the form of endoscopic sphincterotomy.

Here we are presenting a case of 24 years young female with a history of recurrent attacks of pancreatitis, which on work up is found to be secondary to pancreas divisum. She was successfully treated with endoscopic sphincterotomy with the pain-free post-procedure course. Report points out the need for early disease suspicion and efficacy of endoscopic sphincterotomy for the treatment of this disease.

Case Presentation:

Written informed consent was taken before reporting this case report. Our patient was a 24 years old lady, presented in the emergency department with a complaint of sudden onset severe continuous epigastric pain for last four days. This pain was not radiating, aggravated by food intake and relieved only with opioid analgesics. The pain was also associated with non-bilious vomiting. She had an episode of similar pain about one year back for which she was admitted and managed as acute pancreatitis. She had a history of the acid peptic disease, anorexia, and malaise and weight loss of 6 Kg in last three years. On examination, she was dehydrated and jaundiced. Her pulse was 96/minute; BP was 117/68, afebrile. Abdominal examination revealed tenderness in an epigastric area without any mass or palpable visera. Rest of systemic examination was unremarkable.

Her initial laboratory workup showed TLC of 10600/mm3, total bilirubin 1.2 mg% (0.2-1.2) with direct bilirubin 0.9 mg% (0.0 to 0.4), alanine aminotransferase of 71 IU/l (normal 0-31
IU/l), aspartate aminotransferase of 251 IU/l (normal 0-34 IU/l), alkaline phosphatase of 85 IU/l (12-38 IU/l), serum amylase was 99 IU/l (25-125 IU/l) and serum lipase was 167 IU/l (3-60 IU/l).

Ultrasonography (US) of abdomen surprisingly revealed a hypoechoic solid mass in the head of the pancreas resulting in atrophy of pancreatic body and tail, dilatation of pancreatic duct and prominent common bile duct suggestive of the neoplastic process.

To further delineate this situation, CT scan of the abdomen was done which showed a hypodense mass involving the uncinate process of the pancreas causing atrophy of body and tail of pancreas without any infiltration into surrounding structures. Pancreatic duct and common bile duct were prominent without dilatation of intrahepatic biliary channels appeared dilated. (Figure 1) This CT scan was discussed in length with radiologists as it is hard to differentiate between Pancreas Divisum and pancreatic malignancy. Finally, a provisional diagnosis of Pancreas Divisum was made as there was a convincing evidence of opening of dilated primary pancreatic duct at minor papilla.

ERCP was done as diagnostic as well as therapeutic measure, which confirmed the presence of minor papilla about 5 mm proximal to the major papilla, major pancreatic duct was opening at minor papilla while minor pancreatic duct was opening at major papilla (figure 2). Minor papillotomy was done and plastic stent placed in major pancreatic duct. The patient tolerated the procedure well, and no immediate or delayed complication was identified.

Post procedure, the patient had a smooth recovery with significant improvement in abdominal pain and appetite. She was discharged on 3rd post procedure day. On clinic follow-up of 6 months, she has remained pain-free and leading a normal healthy life.

Discussion:

Pancreatic Divisum is a congenital anatomical anomaly characterised by the lack of fusion of the ventral and dorsal parts of the pancreas during the eighth week of fetal development. This condition is found in 5% to 14% of the general population [1]. In a large retrospective study from India, pancreas divisum was more frequent in patients with pancreatitis than in those with biliary diseases or obscure abdominal pain (9 versus 2 percent).

The major pancreatic duct (Wirsung’s duct), drains the secretions from the head, body and tail of the exocrine pancreas, and ends at the major duodenal papilla (hepatopancreatic ampulla);
the accessory pancreatic duct (Santorini’s duct) extends through the head of the pancreas, crosses Wirsung’s duct and ends at the minor duodenal papilla; both pancreatic outlets are located on the medial wall of the second part of the duodenum at a distance of approximately 10 to 15 mm from each other; the minor papilla are above, the major duodenal papilla below.

In Pancreatic Divisum, the dorsal pancreatic section drains into the minor duodenal papilla through the major pancreatic duct; the ventral pancreatic duct, the smaller part of the pancreas, merges with the common bile duct at the hepatopancreatic ampulla. There are two types of Pancreatic Divisum: complete (most common) and incomplete (much less common), in which the ventral and dorsal systems remain connected through small-caliber branch ducts. Approximately 15 percent of cases of pancreas divisum are of the complete type. However, the implications of incomplete pancreas divisum are the same as for classic (or complete) pancreas divisum.

In PD, the increased incidence of acute and chronic pancreatitis is caused by inadequate drainage of secretions produced by the body, tail and part of the pancreatic head through an orifice which is too small [4, 5]. There is a group of patients with pancreas divisum who are subject to recurrent bouts of seemingly idiopathic pancreatitis [6]. In these patients, the minor papilla orifice is so small that excessively high intrapancreatic dorsal ductal pressure occurs during active secretion, which may result in inadequate drainage, ductal distension, pain, and, in some cases, pancreatitis. Although many times, patients with pancreas divisum remain clinically asymptomatic, other common forms of clinical presentations range from recurrent attacks of varying degree of pancreatitis, bowel obstruction, ascites, jaundice, shock and in its most severe form, can lead to shock. Alcohol seems to be the triggering factor for the attack of pancreatitis [6]. Our patient presented with upper abdominal pain secondary to pancreatitis.

Diagnostic workup ranges for laboratory tests to imaging modalities. Laboratory workup may show raised amylase or lipase levels indicating an episode of pancreatitis, but they are not accurate for diagnosing Pancreas Divisum. Imaging modalities which may help in diagnosis include ultrasound, CT scan abdomen but definitive diagnosis is made with some form of cholangiography either ERCP or MRCP. PD is most of the times diagnosed from MRCP but the important point to note here is that currently used 64 slicer CT scan is a good modality to diagnose PD like in our case, especially when the diagnosis is not being suspected [7].

The usual therapeutic solution for symptomatic Pancreatic Divisum is a sphincterotomy of the minor duodenal papilla, which decongests Wirsung’s duct [8, 9]. Clinical improvement with such treatment has been observed in up to 75 percent of patients. Rarely, in selected cases only, is surgical treatment indicated: surgical sphincterotomy, draining or even partial pancreatectomies and their results are comparable with those achieved by endoscopic procedure [6].

Conclusion

This report emphasizes the need of early suspicion of disease, in patients with recurrent idiopathic pancreatitis, by clinician and efficacy of minimally invasive procedures (ERCP) as definitive treatment option.

References