Accessory Pancreatic Lobe with Gastric Duplication Cyst- A Rare Case of Recurrent Acute Pancreatitis

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ABSTRACT

Introduction: Hereditary anomalies of the pancreas and the pancreatic duct is not uncommon and can pose a diagnostic challenge. Accessory pancreatic lobe (APL) is a very rare and curable congenital anomaly of the pancreas which can present as recurrent acute pancreatitis (RAP) and association of APL with a gastric duplication cyst is even rarer. To the best of our knowledge, there are only 18 cases reported so far in the world literature and therefore the aim of this article is to highlight the significance of recognising these conditions in clinical practice for avoiding misdiagnosis.

Case Discussion We present an interesting case of 28-year old lady who presented with severe abdominal pain with similar episodes in the past but never evaluated before. A possibility of recurrent acute pancreatitis (RAP) was considered, further confirmed on serum amylase, lipase and imaging. CT panreatography (CTP) followed by MRCP showed an accessory lobe in the head region of pancreas with a separate pancreatic duct associated with a gastric duplication cyst. She was managed as recurrent acute pancreatitis with conservative measures and she improved.

Conclusion Prompt identification and understanding of the congenital anomalies related to pancreas and pancreatic duct is imperative as these represent correctable causes of RAP. Moreover, as in our case, the pancreas can have lobulated contours, simulating a pancreatic tumor/tumor deposit or lymphadenopathy. Therefore it is prudent to be aware of this condition to avoid misdiagnosis and to offer appropriate treatment.

Keywords: Accessory pancreatic lobe (APL), computed tomography of pancreas, panreatography (CTP), Recurrent Acute pancreatitis (RAP) MRCP (Magnetic resonance cholangiopancreatography)
INTRODUCTION

A 28-year old lady from Philippines presented with complaints of severe intensity epigastric pain followed by vomiting of 2 days duration. She had similar pain episodes in the past starting at the age of 14 years but was never evaluated before. At presentation, She was tachycardic (HR - 124 beats per minute), BP 90/70 mm of hg and had shallow respiration of 24/minute. Skin was warm and diaphoretic and abdominal examination revealed diffuse tenderness, guarding and hypoactive bowel sounds. Investigations revealed neutrophilic leucocytosis (TLC 12,400/mm³), serum Amylase levels > 1500 IU/l and serum Lipase levels > 45,000 IU/l (normal upto to 300 IU/l). Urgent USG abdomen done was noncontributory. Since she had pain abdomen starting in her childhood, further evaluation was done to establish and rule out the cause of recurrent acute pancreatitis. Lipid profile, calcium, phosphorus and ANA were all within normal range. Since she had no definite identifiable risk factors for RAP, (due to the absence of alcohol, gall stones, surgery, use of corticosteroids etc) possibility of congenital anomalies of pancreas/pancreatic duct was considered and was advised further evaluation. Imaging studies CT pancreatography followed by MRCP was done to confirm the diagnosis and to rule out congenital anomalies of the pancreas. Contrast enhanced CT abdomen confirmed features of acute pancreatitis with an accessory pancreatic lobe located anteroinferior to the head region of pancreas with a separate dilated and tortuous pancreatic duct and possibility of associated gastric duplication cyst. There were no signs of chronic pancreatitis on CT. (Fig 1,2 ). For further delineation and confirmation, MRCP (Figure 3) was done which confirmed the presence of accessory lobe of pancreas and irregular dilated duct with calculi within. The accessory pancreatic lobe was seen associated with a gastric duplication cyst with isodense contents within. (Fig 4) Therefore, the diagnosis of recurrent acute pancreatitis with accessory pancreatic lobe and gastric duplication cyst, an extremely rare anomaly was offered and she was managed with nil per oral status, Ryles tube aspiration, fluid resuscitation, proton pump inhibitors and supportive measures. She remarkably improved and was discharged on the 4th day after admission. She is advised surgery for further management.
Figure 1. Axial contrast enhanced CT (A and B) showing accessory pancreatic lobe with a dilated duct (white arrows, A) located anteroinferior to the main pancreatic tissue. Acute interstitial pancreatitis is evident by the increased bulk of the main pancreatic tissue (black arrows, B) and ascites (arrowhead, B).

Figure 2. Coronal (A and B) and sagittal (C) reformatted CT images depicting the accessory pancreatic lobe with dilated duct (white arrows). Note the accessory lobe is contiguous with the main pancreatic tissue (black arrow, A). Gastric duplication cyst with isodense contents (*, B) is seen associated with the accessory lobe (white arrows, B). Sagittal CT image showing the accessory pancreatic lobe and the main pancreatic tissue (black arrowhead, C).

Figure 3. Coronal T2W image (A) showing the dilated duct of the accessory lobe (white arrows, A) with irregular contour and intramural signal void representing ductal calculus (white arrowheads, A). Note the duct further extends into the main pancreatic duct (black arrows, A). Axial T2W MRI (B) showing accessory duct (white arrows, B) and the gastric duplication cyst (*, B).
DISCUSSION

Recurrent acute pancreatitis (RAP) is defined as more than two episodes of acute pancreatitis without any features of chronicity\(^1\). The estimated prevalence is approximately 1 in 10,000 and the disease predominantly affects males in their forties\(^2\). RAP has multiple causes, alcohol and cholelithiasis being the most common (followed by hypertriglyceridemia, biliary tract infection, congenital anomalies, duodenal obstruction, medications, and neoplasms). In 10% to 40% of patients with RAP, the cause is not identifiable and the diagnosis 'idiopathic' RAP (IRAP) is given\(^3\). Most of the patients of RAP are at heightened risk of having frequent episodes unless the offending cause/agent is eliminated and these patients often develop chronic pancreatitis in due course of time if not properly evaluated and treated. Frequent episodes of inflammation can progress and lead on to chronic pancreatitis based on "necrosis -fibrosis hypothesis" \(^3\). Very often the recurrence is predictable and there is a window of opportunity to identify and treat the cause to reduce recurrence. Therefore, if the common identifiable risk factors are not present (alcohol, gall stones, metabolic causes, infections, drugs, surgery etc), these patients must be further evaluated to find out the aetiology. With the advent of newer noninvasive imaging modalities like MRCP, the structural congenital anomalies are recognized more frequently with good specificity and sensitivity. Varied spectrum of anomalies of the pancreas and the ductal system are routinely encountered during radiologic procedures. These anomalies can simulate various neoplastic, inflammatory and posttraumatic conditions. Anatomic variants commonly encountered include developmental anomalies (e.g., pancreas divisum, annular pancreas, ectopic pancreas, pancreatic agenesis, congenital pancreatic cysts etc. Also not uncommon are potential imaging pitfalls (uneven fat distribution, "pseudo masses" etc). All these create a real diagnostic challenge for the treating physician and the radiologist. Therefore knowledge and recognition of the pancreas and pancreatic duct developmental anomalies is important as these anomalies may be a correctable cause of RAP. Congenital anomalies are usually not detected until adulthood and often detected as incidental findings. With the advent of
newer imaging modalities, MRCP, and CT pancreatography these anomalies are recognized more frequently. The common congenital variations of the pancreas are pancreatic divisum, annular pancreas, pancreatic agenesis, ectopic pancreas etc. Accessory pancreatic lobe (APL) is an extremely rare congenital anomaly and is defined as an accessory lobe of pancreatic tissue that originates from the main pancreatic gland and contains an aberrant separate duct \cite{1}. There is very limited literature on this entity and so far only 18 cases have been reported in the world literature. RAP is the commonest (66\%) clinical manifestation of this anomaly\cite{2}. The accessory lobe can vary in size, with a wide or narrow communication to the main pancreas. This anomaly may be associated with a gastric duplication cyst. The aberrant duct in such cases communicates with the main pancreatic duct and along with the duplication cyst. The cause of RAP is hypothesized to be due to the blockade of the pancreatic duct by viscous mucus plugs/biliary sludge. In our patient, she had an accessory pancreatic lobe with a separate duct with stones within associated with a gastric duplication cyst\cite{3}. Duplication cysts of the GI tract are very rare and usually occur in ileum and ileocecal region, gastric duplication cysts are the rarest (3.8\%). Majority of these cysts are associated with other congenital anomalies like ectopic pancreas, spina bifida etc\cite{4}. Abnormalities in the foregut development is postulated to be responsible for the development of this congenital anomaly. McLetchie et al\cite{5} has suggested a neuroenteric hypothesis where in traction on the pancreatic duct by a neuroenteric band causes traction diverticula and pancreatic abnormalities. Most of these developmental anomalies were diagnosed only preoperatively, nevertheless, with newer imaging modalities these are picked up preoperatively as in our case. Although ERCP is the gold standard for evaluating pancreatobiliary anomalies, this procedure is invasive and has increased complications in these patients. Though the presentation of the gastric duplication cyst has been with abdominal pain, nausea and vomiting, rare cases of complications in the form of peritonitis, malignancy is also reported. However in patients with accessory pancreatic lobe associated with a gastric duplication cyst, the cause of abdominal pain is usually from acute pancreatitis as in our case as confirmed on serum amylase and lipase levels. Management of gastric duplication cyst with accessory pancreatic lobe includes surgery and endoscopic drainage. However since there have been reports of malignant transformation of the duplication cyst, resection is preferred and surgery was performed in most of the cases reported so far. The surgery done was the ligation of the accessory pancreatic lobe with transection at its origin along with the local excision of the gastric duplication cyst\cite{6}.

In conclusion hereditary anomalies pertaining to the pancreas and the pancreatic duct are clinically of great importance as it usually creates a diagnostic dilemma. Physicians and radiologists ought to be aware of these varied presentations to distinguish them from other pancreatic conditions. Accessory pancreatic lobe with gastric duplication cyst is an extremely rare cause of recurrent acute pancreatitis and prompt identification of these developmental anomalies helps in surgical planning and prevention of inadvertent ductal injury.

REFERENCES

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