Acute relapsing pancreatitis in children due to congenital anomalies - Case Series

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ABSTRACT

Congenital variants of Pancreas such as Pancreas divisum, annular pancreas, and partial agenesis are seen in 10% of general population. Most of these variants are of no clinical significance and are found incidentally at endoscopy, surgery or autopsy. Magnetic resonance Cholangio Pancreaticography (MRCP) constitutes a valuable tool in the evaluation of Pancreatico Biliary Anomalies. When there is acute pancreatitis in children one should suspect these congenital anomalies and necessary investigations have to be done. Recently we had come across two children diagnosed as recurrent acute pancreatitis and were found to have congenital anomalies on MRCP & CECT. Various diagnostic methods and possible methods of correction of these anomalies in a symptomatic child were discussed.

Keywords: Congenital anomalies of the pancreas, Biliary Pancreatico ductal system, Pancreas divisum, ERCP & MRCP

Introduction:

Congenital variants of the pancreas are seen in approximately 10% of the general population and include both malformation variants such as pancreas divisum and malrotation variants such as annular pancreas and partial agenesis [1] (Figure 2). Most of these congenital variants are of limited clinical importance and are found incidentally at endoscopy, surgery, or autopsy. Pancreatitis is associated with several morphological anomalies such as anomalous pancreatico biliary junction[2] pancreatic divisum.[3-5] Approximately 20-30% cases of acute pancreatitis remain idiopathic.

Acute pancreatitis (AP) is an emerging problem in pediatrics. Although the cause is unclear, it may be explained by a heightened awareness of AP in children. Although magnetic resonance cholangio-pancreatography (MRCP) is seldom required for first attack of AP, it constitutes a valuable tool in the evaluation of pancreatico-biliary abnormalities. Congenital anomalies and normal variants of the pancreatic duct are often detected as incidental findings in asymptomatic patients. Occasionally it produces symptoms in
childhood and here is a patient presented to us with recurrent episodes of abdominal pain and MRCP helped us in diagnosing the condition and directed us for therapy.

We present here two cases of recurrent acute pancreatitis due to pancreatic duct anomalies

Case 1:

A 13 year old male presented to us with complaints of upper abdominal pain since 5 days, vomiting since 4 days. His abdominal pain is radiating to back. He had similar episodes for three times in the past 2 years but were treated conservatively at a local hospital and not been investigated. No history of haemetemesis or melena.

On examination , patient was conscious and oriented. Pulse rate -96bpm, blood pressure - 100/70mmHg, afebrile. No pallor, no icterus. Abdominal examination was normal.

Hb - 11g/dl, WBC -10,200cells/Cmm, serum creatinine - 0.8mg/dl, serum bilirubin - 0.98mg/dl, serum amylase- 1400IU/L, serum lipase - 526IU/L.

In view of history of recurrent acute pancreatitis, MRCP (figure 1) was done. It showed bifid duct of Wirsung with duct of Santorini opening into its anterior division with bulky body and tail of pancreas.

Patient was conservatively treated and he recovered well

CASE 2

A 12 year old female presented with complaints of upper abdominal pain for 2 days, vomiting for 2 days. Her abdominal pain is radiating to back. She had similar episodes 5 times since she was 4 year old. No history of haemetemesis or melena.

On examination, patient was conscious and oriented. Pulse rate- 100bpm,blood pressure - 100/70mmHg,Afebrile.No pallor, No icterus. Abdominal examination was normal.

Hb- 10g/dl,WBC- 10,000cells/Cmm, Serum creatinine -0.9mg/dl, Serum bilirubin - 0.9mg/dl, Alkaline phosphatase - 246IU/L, Serum amylase- 2100IU/L, Serum lipase - 1200IU/L.

CECT abdomen showed incomplete pancreas divisum with MPD 4mm.patient was conservatively treated and recovered well.

Discussion

There are many congenital anomalies of pancreatic duct which are described. Many of these are asymptomatic and does not require any treatment. However, MRCP is useful to identify these anomalies if patient presents with symptoms.

Some of the congenital variants which are clinically significant are the downstream ductal configuration most commonly manifests as a bifid configuration formed by the ducts of Wirsung and Santorini (60% of cases). Less common configurations include a rudimentary duct of Santorini (30% of cases), a dominant duct of Santorini (1%), and "ansapancreatica," in which the duct of Santorini forms a sigmoid curve as it courses to the duct of Wirsung (6). Narrowing of the caliber of the duct at the "knee" of the MPD
can be seen, a finding that represents the site of fusion of the dorsal and ventral ducts. The absence of dilatation of the proximal, or upstream, ductal system allows differentiation of this normal variant from a true stricture.

In case 1 scenario, ERCP may be helpful if symptoms recur but none of the existing data has shown statistically significant treatment available for the congenital variants causing pancreatitis as in pancreas divisum.

Pancreas divisum is the most common congenital pancreatic ductal anatomic variant, occurring in approximately 4-14% of the population at autopsy series, 3-8% at ERCP, and 9% at MRCP [7]. This anomaly results when the ventral and dorsal pancreatic ducts fail to fuse. The ventral duct (duct of Wirsung) drains only the ventral pancreatic anlage, whereas the majority of the gland empties into the minor papilla through the dorsal duct (duct of Santorini) [8].

Three variants have been described: type 1 or classical divisum in which there is total failure of fusion; type 2 in which dorsal drainage is
dominant in the absence of the duct of Wirsung; and type 3 or incomplete divisum where a small communicating branch is present (9). Case 2 fits to type 3 criteria.

The clinical relevance of pancreas divisum remains controversial. Most patients with pancreas divisum are asymptomatic. However, in some patients, this anomaly is associated with recurrent episodes of pancreatitis. Of those with idiopathic recurrent pancreatitis (10), 12-26% of patients have pancreas divisum, as opposed to 3-9% of the general population. Focal dilatation of the terminal portion of the dorsal pancreatic duct, a condition known as santorinicele, is described in association with pancreas divisum and relative obstruction at the minor papilla (11).

The definitive diagnosis of pancreas divisum is made with endoscopic retrograde pancreatography. Multi-detector row CT may also depict pancreas divisum, but only when the pancreatic duct is visualized (12). MRCP provides a noninvasive means of diagnosing pancreas divisum without the use of contrast material and avoids the risk of ERCP-induced pancreatitis. The main features of pancreas divisum when using MRCP include the dorsal pancreatic duct in direct continuity with the duct of Santorini, which drains into the minor ampulla, and a ventral duct, which does not communicate with the dorsal duct but joins with the distal bile duct to enter the major ampulla.

Recent research shows that the administration of secretin improves the sensitivity of MRCP in diagnosing pancreatic divisum. It has been suggested that PD predisposes to an obstructive pancreatopathy as the major part of the pancreatic secretion must flow through the minor papilla [13]. However, most people with PD will never develop pancreatitis since PD occurs in about 5 to 10% of the population while the incidence of pancreatitis from all causes is only 0.1% [14].

**Treatment Options**

Most authors acknowledge the fact that results are better when the indication for treatment used is that of recurrent acute pancreatitis as compared to that used for patients with pain alone or chronic pancreatitis. Surgical treatment consists of transduodenal sphincteroplasty of the minor papilla sometimes associated with cholecystectomy and major papilla sphincteroplasty making interpretation of the results somewhat confusing [15]. Endotherapy options include endoscopic sphincterotomy of the minor papilla using a wire-guided standard sphincterotome or a needle-knife cutting along a previously inserted 5F or 7F guiding stent which is removed after a few days. Stenting of the pancreatic dorsal duct has also been proposed either as a therapeutic trial for patients who are having daily pain or as a long-term treatment (16).

**Conclusion**

Congenital variants of the biliopancreatic ductal system provide interesting challenges when discovered during the diagnostic workup of idiopathic acute recurrent pancreatitis. However, most of these variants are clinically irrelevant. Methods for the selection of patients most likely to benefit from invasive therapy need to be refined.

**References:**

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