Idiopathic chronic calcific pancreatitis in a child: An uncommon entity

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Abstract

Inflammatory disease of pancreas can be acute or chronic. Acute pancreatitis is a reversible process whereas chronic pancreatitis produces irreversible changes in the architecture and function of pancreas. Although pancreatitis is less common in children than in adults it still occurs with regularity and should be considered in any child with acute or chronic abdominal pain. The main difference between chronic pancreatitis in children and adults is in the etiology. We present a case of idiopathic chronic calcific pancreatitis in a child thereby signifying the importance of this entity at this age.

Key words: *Chronic pancreatitis, child, pancreatitis*

INTRODUCTION

Pancreatitis in children is uncommon and represents a diagnostic challenge for the clinicians. Importantly, the clinicians evaluating pediatric patients with abdominal pain should have a high index of suspicion for pancreatitis, as it is associated with significant morbidity and mortality. Chronic pancreatitis in children can be due to hereditary pancreatitis, idiopathic chronic pancreatitis, cystic fibrosis, tropical chronic pancreatitis, hypertriglyceridemia or hyperparathyroidism. All these types of pancreatitis can present with repeated acute attacks progressing to chronic calcific pancreatitis.

Here is such a case of a seven-year-old male child diagnosed as chronic calcific pancreatitis with no etiological cause thereby pointing towards the hereditary or idiopathic variety.

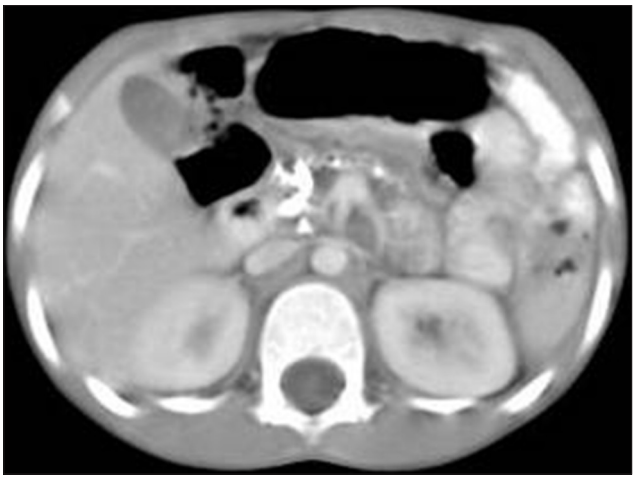

Figure 1: Pancreas showing multiple coarse calcific foci with dilated and tortuous main pancreatic duct (modified computed tomography score 4)

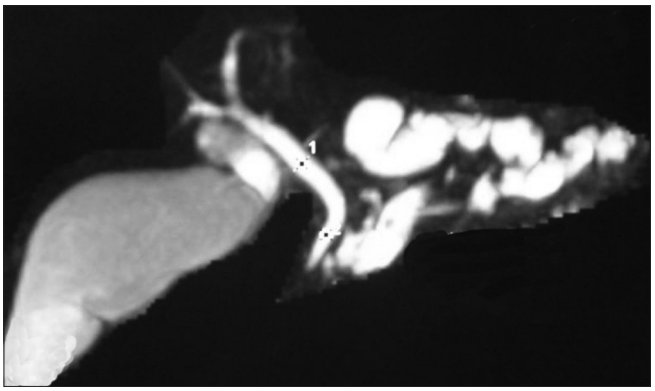

Figure 2: Magnetic resonance cholangio pancreatography showing grossly dilated and tortuous main pancreatic duct hypodense filling defects s/o calculi

CASE REPORT

A seven-year-old boy was examined in the outpatient department with pain in abdomen. Pain was in the epigastric region, radiating to the back and episodic since one year. There was no history of associated vomiting. These episodes of pain were managed at home. There was no history of drug intake or trauma. There was no family history of chronic abdominal problem. On examination the child was average built without any epigastric tenderness or abdominal distension. Vital signs were within normal limits. There was mild pallor. Results of laboratory studies were within normal limits; the exception was raised amylase and lipase concentrations. Serum amylase level was raised (205 U/L)(N 28 - 100U/L). Serum lipase value was also raised 113U/L (N 5 - 31U/L). Serum calcium, glucose, triglyceride levels were normal (9.8 mg/dl, 86 mg/dl and 150 mg/dl respectively). An abdominal sonogram revealed dilatation of the main pancreatic duct (MPD) with calcifications and multiple cystic areas. Computed tomography (CT) scan of the patient showed heterogeneous attenuation of pancreas with multiple coarse calcific foci throughout the pancreatic parenchyma. MPD was dilated and tortuous and measured 9 mm in the body and tail (modified CT score 4) [Figure 1]. Magnetic Resonance Cholangio Pancreatography revealed atrophic pancreas with grossly dilated and tortuous main pancreatic duct. Hypointense filling defects suggestive of calculi were also seen in the body and tail. The side branches were also dilated [Figure 2].

DISCUSSION

Chronic pancreatitis is a longstanding inflammation of the pancreas that alters its normal structure and functions. It can present as episodes of acute inflammation in a previously injured pancreas or as a chronic damage with persistent pain or malabsorption. In hereditary pancreatitis, the inheritance pattern is strongly suggestive of an autosomal dominant trait. Mutations in the cationic trypsinogen (PRSS1) gene play a causative role in chronic pancreatitis. Other genes, such as the anionic trypsinogen (PRSS2), the serine protease inhibitor kazal Type 1 (SPINK1), and the cystic fibrosis transmembrane conductance regulator (CFTR) have been found to be associated with chronic pancreatitis (idiopathic and hereditary) as well.[1,2] Chymotrypsin C (CTRC) variants play a significant role in the pathogenesis of tropical calcific pancreatitis (TCP).[3] N34S SPINK1 represents the major genetic risk factor in TCP.[4] Strong genetic susceptibility due to SPINK1 and CFTR gene mutations and comparative phenotype of idiopathic chronic pancreatitis in India suggest that the term TCP is a misnomer.[5] Variations in Glycoprotein 2 gene are unlikely to play a role in the etiology of chronic pancreatitis.[6] There are a number of reports on hereditary pancreatitis from various countries including India.[7,8] Clinical features of hereditary pancreatitis include recurrent episodes of acute pancreatitis in 80% of the family members and eventual chronic pancreatitis in about 20 to 30% of family members.[9,10] Tropical calcific pancreatitis though initially described from Southern India, is seen in all parts of our country.[8,9,11] This type of pancreatitis is seen exclusively in the tropics. It is also characterized by recurrent episodes of abdominal pain, protracted malnutrition and ketosis-resistant diabetes. Ten per cent patients develop pancreatic cancer. Almost one-third of patients with tropical calcific pancreatitis get recurrent episodes of pain in childhood and can develop diabetes in childhood. Dense calcification is seen in pancreatic parenchyma. Though the exact etiology of tropical chronic pancreatitis is unknown, malnutrition with protein deficiency, cassava toxicity, impaired immune response, viral infections and genetic susceptibility have been considered as various factors in aetiopathogenesis.[8] In idiopathic pancreatitis the clinical presentation is the

same as hereditary pancreatitis but complications like pancreatic ascites, pseudo cyst and portal vein thrombosis are uncommon.[12] In North India, idiopathic chronic pancreatitis differs from the classical tropical pancreatitis described in the literature as it is associated with a higher prevalence of pain and lower frequency of diabetes, calcification and intraductal calculi.[13] As with other forms of chronic pancreatitis, treatment of chronic calcific pancreatitis includes control of diabetes, relief of pain with analgesics, pancreatic enzyme replacement, endoscopic and surgical decompression of dilated pancreatic ducts and removal of calculi. Immediate and long-term results from the surgical procedures have shown encouraging results and confirmed the beneficial effect of surgery in well-selected cases with a goal of avoiding total pancreatectomy.[14]

CONCLUSION

The present case stresses the need to diagnose chronic pancreatitis in the pediatric population and not to underestimate the importance of this relatively uncommon entity in this age group. It also stresses the importance of various etiologies of chronic pancreatitis in children and timely management so as to avoid long-term complications and/or pancreatectomy.

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