Case report

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Painless Silent Chronic Fibrosing Pancreatitis Causing Complete Biliary Obstruction In A Toddler: A Case Report

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All authors searched medical literature, databases, conceptualized, conducted the case review and reviewed the final manuscript. All authors have read and agreed to the published version of the manuscript.

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1. Abstract

Chronic pancreatitis in toddlers presents with epigastric pain radiating to the back, steatorrhea, or recurrent acute pancreatitis, chronic weight loss, even when eating habits and amounts are normal, nausea, vomiting, and/or diarrhea. Among adults, almost 10% with chronic pancreatitis do not report pain. We report a 15-month-old boy who presented with jaundice, clay-colored stool, hepatomegaly and elevated liver enzymes. The mother reported that the child had no complaints otherwise. Hepatitis A virus IgM was positive. He was advised conservative treatment. Two months later he presented with olive green jaundice, pruritus, and clay stools, and no other complaints at all. The COVID-19 anti-spike IgM was positive. Abdominal ultrasound revealed dilatation of intrahepatic biliary radicals, right, left and common hepatic ducts with abrupt no dilation at common bile duct. MRCP and dynamic MRI of pancreas confirmed the findings and detected bulky head of the pancreas with atrophic body and tail compressing the common bile duct. He underwent percutaneous trans hepatic drainage with T tube insertion for drainage of the biliary obstruction followed 10 days later by Frey procedure with coring of the pancreatic head to allow biliary drainage. The post-operative coursewas uneventful and within 3 weeks the jaundice resolved completely, and the amylase and lipase dropped to normal levels within 8 weeks. Chronic fibrosing pancreatitis associated with acute hepatitis or COVID-19 might be painless and get masked by obstructive jaundice. Chronic pancreatitis presenting with biliary obstruction might be mistaken for prolonged cholestatic hepatitis A infection. Chronic pancreatitis should be excluded in children with suspected prolonged cholestatic hepatitis A infection. Percutaneous transhepatic drainage might be a necessary step before Frey's procedure. Chronic pancreatitis in children is amenable to successful surgical intervention.

Keywords:
Chronic pancreatitis; COVID-19; Frey procedure; painless; toddler.
Abbreviations: COVID-19: corona virus disease of 2019

2. Introduction

Pancreatitis in children presents with symptoms such as abdominal pain, which may be severe and persistent, nausea, vomiting, fever, and a distended abdomen. Other possible signs and symptoms include poor appetite, weight loss, jaundice, and diarrhea. In severe cases, children with pancreatitis may also experience dehydration, low blood pressure, and organ failure. It is crucial for children exhibiting these symptoms to seek medical attention promptly for proper evaluation and management of pancreatitis (1). In the young age, pancreatitis is rare and it's etiology is poorly understood. Few reports describe hepatitis A virus-induced pancreatitis. It occurs during the third week of infection. (2) Others report that COVID-19 is an underestimated cause of pancreatitis as it might occur silently in almost 17% of cases with COVID-19 pneumonia (3). Autoimmune pancreatitis is responsible for 2-6% of cases in adult population (4,5). Pancreatic fibrosis is the prevailing reversible pathology and diagnostic factor in early chronic pancreatitis (6). Pancreatic fibrosis was reported in scarce literature to cause obstructive jaundice in adults (7). We aim to describe our experience with a toddler who suffered from painless chronic fibrosing pancreatitis that caused complete biliary obstruction.

3. Case Presentation

A 21-month-old boy presented to New Children Hospital, Faculty of Medicine, Cairo University, Egypt by jaundice, dark urine, and pale stool for 6 months. The parents reported that the jaundice had a gradual onset and progressive course without abdominal pain, fever, vomiting or diarrhea. There was no history of trauma or previous operations. There was no history of bleeding, rash, drug intake, gastrointestinal tract symptoms, renal symptoms or other system affection. Initially, HAV IgM was positive so patient was advised conservative home treatment in the form of diet regimen. Two months later he was still jaundiced with clay

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colored stools and no other complaint. He was diagnosed with prolonged cholestatic hepatitis A viral infection. There was no family history of a similar condition in the family or previous hospital admissions. He was born by cesarian section to non-consanguineous parents. By examination, the child's skin color was deep olive green, and his weight and height percentiles were at the 3rd percentile for his age (9 kg and 76cm respectively). Abdominal examination revealed hepatomegaly with no splenomegaly. Otherwise, the examination was unremarkable. His total bilirubin was 14.2mg%, direct bilirubin was 6.7mg/dL, alanine aminotransferase (ALT) was 108IU/L (expected upper level of normal was 36IU/L), aspartate aminotransferase (AST) was 142IU/L (expected upper level of normal was 36IU/L), gamma glutamyl transferase (GGT) was 837 IU/L (expected upper level of normal was 60 IU/L), serum cholesterol 283 mg/dL (expected upper level of normal is less than 170 mg/dL), serum triglycerides was 185 mg/dL (expected upper level of normal is less than 175 mg/dL), low density lipoproteins and high density lipoproteins were 203 mg/dL and 15.19 mg/dL (expected upper level of normal is less than 100 mg/dL and 15-25mg/dL) respectively. His serum amylase and lipase levels were elevated, 125 U/L and 174 U/L (expected upper level of normal is less than 115U/L and 60U/L respectively). We investigated the child for an underlying cause of this pancreatitis. His antinuclear antibodies (ANA), anti-liver microsomal antibodies (LKM), anti-tissue transglutaminase TTG IgA, anti-tissue transglutaminase TTG IgM, anti endomysial Ab (IgA- IgG), and calprotectin in stools were negative. His IgA level in serum was 261.4 (n 20-100) and IgG level was 1242 (n 258- 1393). His COVID-19 anti-Spike antibodies were positive 11.4 U/ml (positive >10U/ml) (COVID 19 IgM was 10.39 U/ml (reactive >10 U/ml) COVID 19 IgG: 0.44 U/ml (non-reactive <10 U/ml).

The pelviabdominal ultrasound revealed dilated intrahepatic biliary radicals, right and left hepatic and common hepatic ducts with abrupt change at the common bile duct. No biliary stones were detected. Magnetic resonance cholangiopancreatography (MRCP) and dynamic magnetic resonance imaging (MRI) revealed non visualized distal part of the common bile duct and adjoining part of pancreatic duct with associated dilatation of the proximal part of pancreatic duct, proximal part of common bile duct, hepatic ducts and intrahepatic biliary radicals. Bulky head of pancreas with atrophic body and tail. He underwent percutaneous transhepatic drainage (PTD) with T-tube insertion for drainage of the biliary obstruction. (Figure 1). Jaundice resolved immediately after the PTD along with normalization of urine and stool colour. Following the drop in bilirubin the child underwent Frey's operation of non-anatomical excision of the fibrotic pancreatic head with longitudinal pancreatico jejunostomy and hepatico jeujenostomy (Roux en y). (Figure 2). The biopsy of the excised tissue of head of pancreas showed preservation of lobular architecture with fibrous thickening of interlobular septae with mild chronic inflammatory cells. No lymphoplasmacytic sclerosing pancreatitis, no granuloma or evidence of neoplasia were detected. Excised gall bladder showed focal hyperplastic mucosa, mild transmural chronic inflammatory cellular infiltrate, and fibrosis. The post-operative ultrasound revealed echogenic fat planes surrounding the pancreas likely due to inflammatory changes. Two weeks post-operatively the child's total bilirubin was 0.29 mg/dL (normal value: 0-1.2 mg/dL), direct bilirubin was 0.1 mg/dL (normal value: 0-0.3 mg/dL), ALT was 18 IU/L (normal value is up to 36 IU/L), AST was 41 IU/L (normal value is up to 36 IU/L), GGT was 611 IU/L (normal value: 8-60), serum amylase and lipase was 68 U/L and 104 U/L (expected upper level of normal is less than 115U/L and 60U/L respectively). Four weeks post-operatively all lab tests normalized. Blood sugar and glycated hemoglobin A1c were normal all through. Patient followed -up 4 months post operative with no interval symptoms of pancreatitis and laboratory studies showed no evidence of endocrine or exocrine function loss.

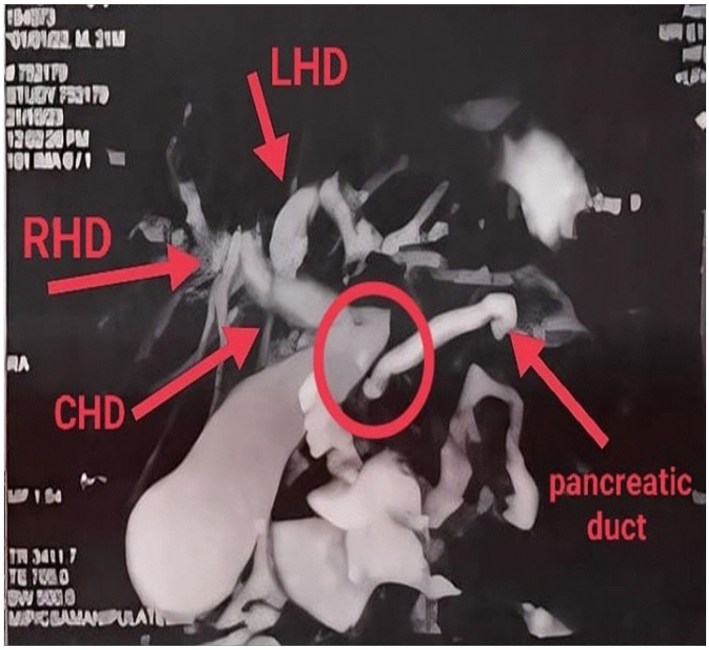

Figure 1: Dynamic MRCP revealed Dilated CBD, right and left hepatic ducts and intrahepatic biliary radicals, and distended gall bladder so as the cystic and pancreatic ducts.

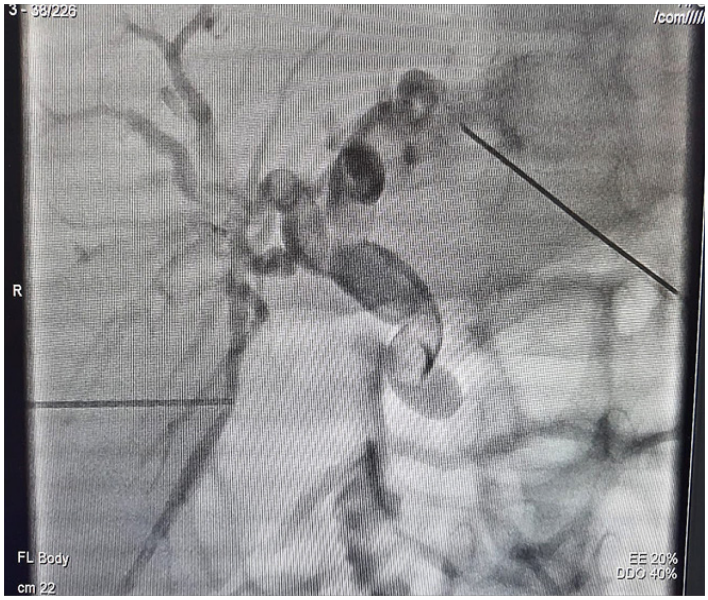

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Figure 2: Percutaneous trans hepatic drainage with T tube insertion for drainage of the biliary obstruction of the studied case.

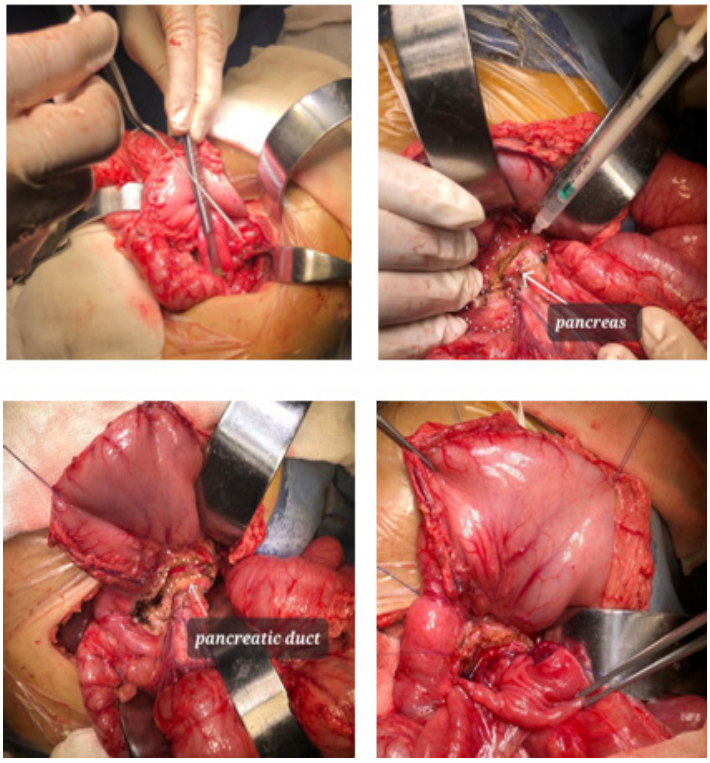

Figure 3: Non-anatomical excision of the fibrotic pancreatic head with longitudinal pancreatico-jejunostomy and hepatico-jejunostomy.
a) Opening of the lesser sac and full exposure of the pancreas. b) Longitudinal incision of the dilated pancreatic duct and aspiration of the retained secretion. c) Non- anatomical resection of pancreatic head and longitudinal incision of the main pancreatic duct. d) Pancreatico-jejunostomy.

4. Discussion

Common bile duct stenosis/stricture has long been known to complicate 3-46% of chronic advanced pancreatitis among adults (8), but not toddlers. Painless silent pancreatitis is being more recognized among children, reaching up to 13% (9). Our studied child sustained chronic fibrosing pancreatitis, complete biliary obstruction, and clay stools for almost 4 months, without any other symptoms. There was no abdominal pain, fever, poor activity, no diarrhea, no symptoms of inflammatory bowel syndrome. The painless silent pancreatitis in our studied toddler masqueraded as prolonged cholestatic hepatitis A viral infection. Yet, prolonged cholestatic hepatitis A viral infection is an uncommon debilitating condition with failure to thrive, malabsorption, pruritus, and not a clinically silent condition. Prolonged cholestatic hepatitis A viral infection is not a painless condition. Hence, it seems necessary to revise the diagnosis of prolonged cholestatic hepatitis A viral infection among children. Other diseases should be ruled out, as the hepatitis A IgM might falsely test positive among those with autoimmune disease, acute and chronic infections as typhoid fever that trigger polyclonal gammopathy (10). Among adults, pancreatitis was reported to complicate hepatitis A viral infection due to edema of the ampulla of Vater with obstruction to the outflow of pancreatic secretions (11), but exceptionally rare among children (12), let alone toddlers. Yet, pancreatitis associated with hepatitis A infection is mostly self-limiting and not chronic fibrosing pancreatitis. Our child had no history of inflammatory bowel disease, gall stones, hypertriglyceridemia . He had no history of recurrent chest infections, diarrhea, malabsorption that might be suggestive of cystic fibrosis. There was no reported history of trauma. Drug induced pancreatitis constitutes almost 25% of pancreatitis was a remote possibility in our studied child owing to the lack of history of drug intake. Also the very rare possible etiology of autoimmune pancreatitis and malignancy of head of pancreas were excluded by the biopsy histopathology findings.

Underlying anatomic pancreatic divisum was excluded in our studied case by the pre-operative MRCP, that did not demonstrate anatomic abnormality of the pancreatic duct (13). Pancreatitis complicates almost 1% of cystic fibrosis patients. Yet, cystic fibrosis is very unlikely in our studied child, given the immediate catch up and lack of history incriminating cystic fibrosis (14). A genetic susceptibility cannot be ruled out as he did not undergo whole exome sequencing for mutanations known to be associated with pancreatitis as SPINK1. PRSS1, CFTR, CRTC. (15) The recent subclinical infection by COVID 19 infection diagnosed by the positive COVID 19 anti-spike IgM antibodies might be responsible for the painless silent pancreatitis in our studied child. It is not known if COVID 19 infection might cause a false-positive hepatitis A virus IgM, yet it causes false positive hepatitis E virus IgM (16). COVID 19 might be the inciting factor for the development of pancreatitis in our studied child. Yet, almost 95% of adults with COVID 19 induced chronic pancreatitis have a benign self-limiting course (17, 18). Idiopathic fibrosing pancreatitis might be another possible cause, as it presents by biliary obstruction and has an excellent prognosis up to 25 years after intervention (19, 20). PTD proved to be an exceptionally valuable intervention in our studied case. It resulted in adequate draining of bile and was an essential step before the final Frey's procedure. It is intriguing however that the liver did not sustain injury by the 6 months of biliary obstruction, and once the obstruction was relieved the liver size, and functions were restored.

5. Conclusion

Chronic fibrosing pancreatitis associated with acute hepatitis or COVID-19 might be painless and get masked by obstructive jaundice. Chronic pancreatitis presenting by biliary obstruction might be mistaken for prolonged cholestatic hepatitis A infection. Chronic pancreatitis should be excluded in children with suspected prolonged cholestatic hepatitis A infection. Percutaneous trans hepatic drainage might be a necessary step before Frey's procedure. Chronic pancreatitis in children is amenable to successful surgical intervention.

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6. Declarations

Consent to participate: Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient/ parent/guardian/ relative of the patient. A copy of the consent form is available for review by the Editor of this journal

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Competing interests The authors declare no conflict of interest in connection with the reported study. Authors declare veracity of information.

Ethics approval :
Ethical clearance was obtained for the research project

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