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Case Report

Acute recurrent pancreatitis in a child with pancreatic divisum– A case report

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ABSTRACT

Introduction and importance: In children, acute recurrent pancreatitis is attributed to pancreato-biliary anomalies, hereditary pancreatitis and cystic fibrosis. Pancreatic divisum is a common congenital ductal anomaly that leads to recurrence of pancreatitis.

Case presentation: A 13 years old female presented with clinical features of acute recurrent pancreatitis. After ruling out common causes, magnetic resonance cholangiopancreatography was done which showed pancreatic divisum. Her symptoms resolved following duodenum preserving pancreatic head resection.

Discussion: Acute recurrent pancreatitis is attributed to raised intrapancreatic dorsal ductal pressure due to ductal anomalies especially pancreatic divisum (PD). It is the embryological failure in the fusion of the dorsal and ventral ductal system. PD is further classified into a classical subtype where there is complete failure of ductal fusion and an incomplete subtype where there is partial fusion of the ductal system. The diagnosis is commonly done through abdominal imaging with secretin enhanced magnetic resonance cholangiopancreatography being the choice of imaging modality. The initial approach is endoscopic intervention unless patients present with signs of pancreatic fibrosis where a duodenum preserving pancreatic head resection can be carried out.

Conclusion: A keen suspicion should be given towards anatomical or structural variants in absence of common etiologies. Early identification and management of pancreatic divisum prevents the recurrence of pancreatitis.

1. Introduction

Acute recurrent pancreatitis (ARP) refers to a clinical entity characterized by distinct episodes of acute pancreatitis (AP) which occurs on more than one occasion. AP is an inflammatory disease of the pancreas that requires two out of the three features of characteristic abdominal pain, raised serum lipase/amylase which is greater than three times the upper limit and computed tomography scan suggestive of AP [1]. Similarly, chronic pancreatitis (CP) is considered to be a sequelae of untreated AP and is a progressive fibroinflammatory disease that exists in large-ducts (often with intraductal calculi) or the small-ducts [2].

Unlike in adults, the etiology of AP in children is quite different. AP in children is mostly associated with abdominal trauma, infections, drugs and systemic or metabolic disorders while ARP is related to pancreato-biliary anomalies, hereditary pancreatitis and cystic fibrosis [3]. Likewise, it is believed that any factor capable of causing an initial episode of acute pancreatitis has the potential to incite recurrent episodes [4].

ARP and CP have been diagnosed in children at increasing rates over the past decade. However, as pediatric ARP and CP are still relatively rare conditions, little quality evidence is available on which to base the diagnosis and determination of etiology [3].

The most common etiology in ARP is attributed to alcohol and gallstone but other conditions like pancreatic divisum and genetic factors are also represented in patients with ARP [5,6].

Pancreatic divisum (PD) is a common congenital anomaly and is seen

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in up to 12% of the population [7]. This embryological abnormality occurs due to the failure of fusion in the dorsal and ventral ductal system in utero.

There have been very few studies indicating the association of ARP in individuals with pancreatic divisum [5,6,8,13]. We report a 13 years old female who presented with recurrent episodes of pancreatitis which on further evaluation was attributed to pancreatic divisum.

2. Methods

We reported this case following the updated consensus based Surgical Case Report (SCARE) guidelines [9].

3. Case Presentation

A 13 years old female with a past medical history of recurrent pancreatitis presented to the emergency department with severe abdominal pain that was radiating to the back and the left inferior angle of the scapula. The pain was acute in onset and was not associated with fever, nausea and reduced appetite. She did not have jaundice and hematemesis. She described her pain as sharp, stabbing in nature. Her mother mentioned that she had experienced this type of pain before, but it had been one year since her last episode. The patient mentioned that her first episode of pancreatitis was at the age of eight. There was no history of trauma or use of any medications. There was no history of such events in her family members. On her physical examination, she was anxious and malnourished. She was afebrile, tachycardic and tachypneic and was in acute distress due to pain. On examination of the abdomen, the abdomen was soft and non-distended but presented with guarding and tenderness, which was especially tender in the epigastric region. On the rectal examination, there was a normal tone and finger stained with stool. Laboratory investigation revealed normal white blood cell count of 8000/mm^3 (normal range 4000–11000/mm^3) with neutrophilia, hemoglobin was 13 gm/dL (normal range 12–16 gm/dL) and albumin 3.9 (normal range 3.4–4.8 g/dL). Her lab values showed amylase and lipase values as 415 U/L and 705 U/L respectively. She was admitted in the High Care Unit and further workup was done to rule out hereditary pancreatitis. Her alpha 1 antitrypsin (AAT) serum was 137 mg/dL (normal range 90–200 mg/dL). Her ENA (Extractable Nuclear Antigens) qualitative profile was found to be negative ruling out hereditary pancreatitis.

On her radiological examination, her ultrasonography of abdomen and pelvis showed a bulky head of pancreas and tail measuring approximately 2.5 cm (anteroposteriorly) at the body with small hypoechoic collection seen in the anterior part of pancreas. Peripancreatic fat thickening was also noted along with mild free fluid in the pelvis. Her contrast enhanced computed tomography (CECT) examination showed evidence of pancreatitis. We suggested magnetic resonance cholangiopancreatography (MRCP) radiological analysis for the patient. The MRCP findings demonstrated the dorsal main pancreatic duct entering the minor papilla as shown in Fig. 1 . Common bile duct was seen entering the ampulla with evidence of pancreatic divisum.

She was then referred to the pediatric surgery department where she was planned for a duodenum preserving pancreatic head resection (DPPHR). The child was followed up after a month of surgery. During her scheduled follow ups, she was free of any abdominal symptoms.

4. Clinical discussion

Acute recurrent pancreatitis (ARP) and chronic pancreatitis (CP) are considered to be separate entities but surveys have shown that patients with CP have had a history of ARP suggesting that ARP and CP are disease continuum [10]. Though the current knowledge regarding AP and CP are based on studies of adults, recent reports from the International Study Group for Pediatric Pancreatitis: In Search of a Cure (INSPPIRE) have provided new quality evidence regarding pediatric risk factors, diagnosis, and management [3]. Single-center studies estimate that 9%–35% of children with AP have recurrent episodes and the incidence of CP is approximately 0.5 per 100 000 persons per year in young adults [11].

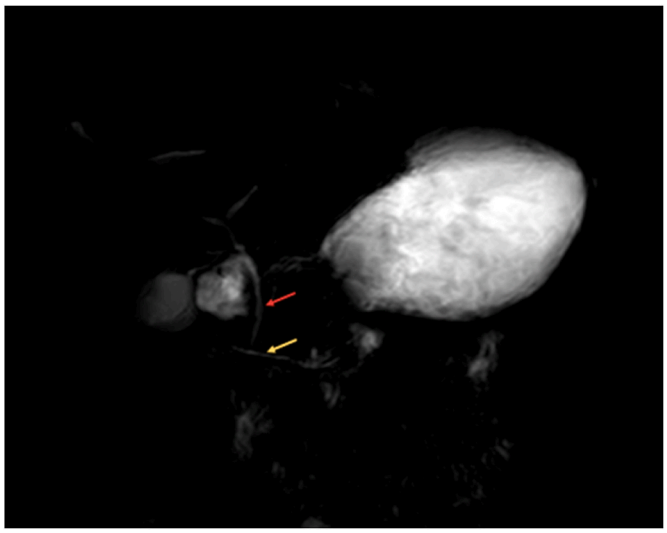

Fig. 1. Magnetic resonance cholangiopancreatography (MRCP) of the abdomen showing pancreas divisum
(A) Common Bile Duct (CBD) which drains into duodenum via major papilla (red arrow). (B) dorsal pancreatic duct which drains directly into duodenum via minor papilla (which is the most prominent duct in pancreatic divisum) (yellow arrow). . (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

A study conducted in China showed trauma to be a major cause for pancreatitis in pediatric age groups followed by the presence of any systemic diseases or structural abnormalities, use of toxins or drugs, autoimmunity or any idiopathic etiology [12]. Certain studies suggest the association of structural anomaly (pancreatic divisum) with pancreatitis is more prominent in cases of recurrent pancreatitis [8,13]. About 20% of patients with ARP have associated pancreatic duct anomalies [7].

Pancreatic divisum (PD) is the most common variant of pancreatic ductal anatomy. The abnormality lies in the embryological failure in the fusion of the dorsal and ventral ductal system. PD is further classified into a classical subtype where there is complete failure of ductal fusion and an incomplete subtype where there is partial fusion of the ductal system. The pathogenesis of developing pancreatitis in PD is attributed to raised intrapancreatic dorsal ductal pressure which thereafter results in inadequate drainage and distention of the ductal system. A study suggests that intrapancreatic ductal hypertension makes the pancreas more susceptible to injury from alcohol, toxins or trauma [14].

In most instances, patients with PD are asymptomatic. In symptomatic cases, PD has been associated with recurrent pancreatitis [15]. The diagnosis is commonly established following abdominal imaging. The use of abdominal computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) helps identify this anatomical aberrant. The imaging modality of choice for identifying PD is secretin enhanced MRCP [16].

The clinical history in our patient was inconclusive for any evidence of trauma, underlying systemic diseases or use of drugs. Likewise, autoimmune causes for pancreatitis were also ruled out by the negative result of the qualitative profile of Extractable Nuclear Antigens (ENA). In order to assess for any anatomical defects, MRCP was suggested which demonstrated the presence of PD.

In patients of PD with infrequent symptoms, conservative management can be indicated with analgesics, anticholinergics and pancreatic enzyme supplementation. In recurrence of pancreaticobiliary pain and

acute or chronic pancreatitis, treatment aimed on dilation of the dorsal ductal system can be used prior to surgery. Procedural options aim at endoscopic and surgical sphincterotomy which can be performed depending on the condition of the patient. According to a systematic review on endotherapy for PD, ARP due to PD showed a better response rate than in cases with CP [17].

Only symptomatic patients are treated for ARP. Endoscopic intervention is considered as the initial approach for ARP. The standardized approaches include ERCP with subsequent endoscopic minor papilla sphincterotomy and sphincteroplasty or endoscopic dilatation and

stenting. However, endoscopic approaches are associated with significant complications when done repeatedly and can fail too. The patients who fail to respond to endoscopic treatment benefit by surgical modalities. For patients who fail at endoscopic sphincterotomy, surgical resection and reimplantation of the pancreatic dorsal duct creates a greater orifice than simple endoscopic sphincterotomy or stenting and presents with a better outcome. Likewise, in patients with signs of pancreatic fibrosis and/or inflammatory mass, a duodenum preserving pancreatic head resection (DPPHR) can be carried out. In patients where there are no such signs or suturing the pancreas is not difficult, a pylorus preserving whipple resection is recommended [18].

5. Conclusion

Our aim in this case report is to highlight the various pathological and structural causes of Acute Recurrent Pancreatitis in a pediatric population. A keen suspicion should be given towards anatomical or structural variants (pancreatic divisum) in absence of common etiologies. Early identification and management of pancreatic divisum prevents the recurrence of pancreatitis.

Ethical approval

Not required in our case.

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None.

Author contributions

Sunil Basukala (SB) = Conceptualization, Supervision.

Sunil Basukala (SB), Ojas Thapa (OT) = Writing - original draft.

Ojas Thapa (SP), Sunil Basukala (SB), Yugant Khand (YK), Soumya Pahari (SP) = Writing - review & editing.

All the authors read and approved the final manuscript.

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Consent

As the patient is a minor, written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

No conflict of interest.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104642.

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