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PANCREATICO-PLEURAL FISTULA, AN UNUSUAL CAUSE OF RECURRENT PLEURAL EFFUSION IN CHILDREN: A CASE REPORT

Pediatrics

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ABSTRACT

Pancreatico pleural fistula (PPF) represents an uncommon but significant complication of chronic pancreatitis (CP), necessitating a heightened level of clinical suspicion in patients who exhibit pleural effusion. The confirmation of the presence of a fistula tract, extending from the pancreatic duct to the pleural space, can be established through the use of radiological imaging techniques. A 10-year-old girl presented with a history of abdominal pain dating back to one year and difficulty in breathing for the past 10 days. USG scan of the chest and abdomen revealed right pleural effusion. Magnetic resonance cholangio pancreatography (MRCP) was suggestive of chronic pancreatitis with pancreatic pleural fistula. In the current case, the child received urokinase treatment for three days through the catheter in a dose of 100,000 IU diluted in 100 ml of normal saline resulting in significant expansion of the child's lungs. The child had shown clinical improvement with octreotide infusion (2µg/Kg IV bolus followed by 2µg/Kg/hr IV continuous infusion for one day and then 4µg/Kg/hr IV for one day and thus child was managed conservatively.

KEYWORDS

Chronic pancreatitis, pleural effusion, pediatric pancreatitis, pancreatic pleural fistula (PPF), urokinase, octreotide infusion.

INTRODUCTION

Pancreatitis, a condition characterized by the inflammation of the pancreas is relatively uncommon ailment in children. It is defined by the presence of clinical manifestations, including epigastric abdominal pain and elevated levels of digestive enzymes in the blood. This condition is associated with considerable morbidity and mortality rates [1]. In recent years, there has been a notable increase in the prevalence of pediatric pancreatitis [2]. Among the identifiable etiologies, biliary causes, trauma, infections and medications are found to be the most prevalent, even though the precise underlying cause for pediatric pancreatitis cannot be determined in most cases [3]. Due to its infrequency, the diagnosis is often postponed in a considerable number of cases, necessitating a high degree of suspicion for the timely diagnosis and management.

The relationship between pancreatic diseases and pleural effusions (accumulation of fluid in the pleural space) is a subject of considerable recognition [4]. Acute pancreatitis frequently manifests with the presence of minor, transient effusions predominantly on the left side. It is hypothesized that these effusions result from a combination of lymphatic or sympathetic mechanisms. Conversely, chronic pancreatic disease whether it is accompanied or not by pseudocysts often leads to persistent pleural effusions of an increased size [5]. Furthermore, over the past decade, chronic ascites along with chronic pleural effusion have been identified as additional complications associated with chronic pancreatitis. Chronic or recurrent inflammatory conditions may lead to both exocrine insufficiencies, characterized by steatorrhea, and endocrine insufficiency, which can culminate in the development of diabetes [6]. Generally, individuals suffering from pleural effusion present with primary pancreatic disease frequently does not manifest any noticeable symptoms. Present case focus on the management of recurrent episodes of pancreatitis and pleural effusion and in a girl child.

Case Presentation

A 10-year-old girl child presented to us with abdominal pain for 1 year and difficulty in breathing for 10 days. The child underwent a comprehensive evaluation for abdominal pain, receiving treatment with Proton Pump Inhibitors (PPIs) and supportive measures on several outpatient visits. The child had a history of accidental ingestion of a foreign body (a hairpin), which was uneventful and there was no history of blunt abdominal trauma. On her admission to our hospital, child had dyspnea grade 3 with room air saturation being normal.

Respiratory system examination showed decreased chest movements on the right hemithorax and trachea is shifted to left side, decreased tactile fremitus and vocal resonance on right side with stony dull percussion note on the right side. The examination of the cardiovascular system was found to be within normal parameters, with the sole exception being the apex beat, which shifted outward and downwards. The child experienced considerable weight reduction with loss of appetite. Her anthropometric measurements indicated acute malnutrition characterized by a weight > 3rd centile for her age (5 years), height > 10th centile for her age (9 years) and a BMI > 3rd centile for her age. Initial differential diagnoses included tuberculous pleural effusion and chronic pancreatitis. Further, CBP evaluation was suggestive of microcytic hypochromic anemia and total leucocyte count was found to be normal with few toxic granules and thrombocytosis. The chest X-ray of the child indicated a right pleural effusion, accompanied by a mediastinal shift (Figure 1). Subsequently, the child underwent a ultrasonography (USG) of the chest, followed by therapeutic and diagnostic pleural fluid aspiration, achieved through the insertion of a pigtail catheter (Figure 2). The intercostal pig tail catheter drains initially collected pleural fluid around 1400ml, but the amount of fluid drained significantly reduced despite clear signs of pleural effusion. Pleural fluid was grossly hemorrhagic and serum analysis showed WBC count of 510 cell/cumm with neutrophils 65%, lymphocyte 35%, RBC were plenty, sugar 78mg/dl and proteins 4g/dl. The pleural fluid analysis revealed a suggestive exudative type of effusion without any indication of malignant etiology or bacteria (the pleural fluid culture was sterile) and the Cartridge based nucleic acid amplification test (CBNAAT) tested negative for Mycobacterium tuberculosis (MTB).

Figure 1: Chest X-ray of the child showing right pleural effusion with a mediastinal shift.

Figure 2: Fluid aspired through the insertion of a pigtail catheter.

Pleural fluid levels of amylase and lipase were significantly increased (amylase 33950 u/l, lipase 69800 u/l). However, the USG scan of the chest and abdomen revealed multiple septation in the right pleural space and pancreatic atrophy with dilated main pancreatic duct.

In view of chronic pancreatitis with right pleural effusion, pancreatico pleural fistula (PPF) was considered and child was further evaluated with Magnetic resonance cholangiopancreatography (MRCP) and Multi-Detector Computed Tomography (MDCT) to look for pancreatic structural anomalies. MRCP was suggestive of chronic pancreatitis with peripancreatic fluid collection with extension in to the mediastinum across the diaphragmatic hiatus with massive right pleural effusion. The primary pancreatic duct was observed to interact with peripancreatic fluid results indicating a pancreatico pleural fistula (PPF) (Figure 3).

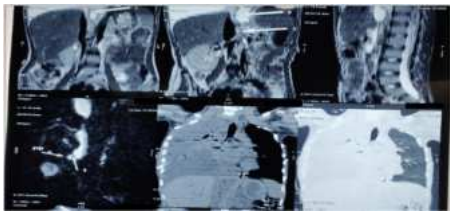

Figure 3: Pancreatitis with peripancreatic fluid collection and extension of peripancreatic collection in to the mediastinum with massive right pleural effusion. Main pancreatic duct appears to communicate with peripancreatic fluid findings suggestive of pancreatic pleural fistula (PPF)

Given the significant accumulation of right pleural effusion, characterized by numerous septations and inadequate drainage via the pigtail catheter, urokinase was administered intrapleural. Urokinase was given for three days through the catheter in a dose of 100,000 IU diluted in 100 ml of normal saline. The pleural effusion showed a favorable response after urokinase treatment, resulting in significant expansion of the child's lungs. The child was started on partial parenteral feeding and diet that is high in calories, proteins and low in fat. Octreotide infusion has been commenced in response to the presence of pancreatico pleural effusion (PPF). Throughout the treatment, the child's blood sugar (8.0mmol/L) and blood pressure (115/75) were meticulously monitored, and observed that these parameters remained within the normal range. Pediatric gastroenterologist opinion was sought for clinical improvement with octreotide infusion (2µg/Kg IV bolus followed by 2µg/Kg/hr IV continuous infusion for one day and then 4µg/Kg/hr IV for one day) and thus the child was managed conservatively.

DISCUSSION

Chronic pancreatitis (CP) is a progressive fibroinflammatory disorder marked by the irreversible destruction of pancreatic parenchyma, ducts and the loss of exocrine function [7]. The two primary etiologies identified in children include trauma to the pancreas and hereditary pancreatitis. Additionally, children afflicted with cystic fibrosis, a progressive, debilitating, and incurable lung disease are at an increased risk of developing pancreatitis [8].

In a significant proportion of cases, the underlying cause for developing pancreatitis remains unidentified. In the present case, the probable cause might not be attributed to the accidental ingestion of foreign body (hair pin) however it was uneventful while there was no history of blunt abdominal trauma as well. Subsequently, following the persistent inflammation of the pancreas, the primary pancreatic duct becomes obstructed and ruptured during the subacute phase. The disruption of the pancreatic duct is a consequence of alcoholic pancreatitis or trauma in adults, and the development of pseudocysts throughout the progression of chronic pancreatitis in children [9]. In the case of ductal leakage occurring in an anterior direction, pancreatic secretions are able to freely enter the abdominal cavity, leading to the development of pancreatic ascites [10]. Conversely, when the ductal leakage is posterior, the retroperitoneal pancreatic fluid is more likely to migrate towards the mediastinum, influenced by the transdiaphragmatic pressure gradient that exists between the abdominal and pleural cavities [11]. Here the child demonstrated pancreatic atrophy with dilated main pancreatic duct. Owing to the posterior rupture of the pancreatic duct and decompression of enzymes into the mediastinum, this child has few abdominal signs and symptoms.

Pancreatico pleural fistula (PPF) represents a rare complication associated with chronic pancreatitis. It occurs when there is a leakage of pancreatic fluid, resulting from the disruption of a pancreatic pseudocyst into the pleural cavity [12]. PPF along with pleural effusion from various other causes, often exhibits similar clinical symptoms, including shortness of breath, dyspnea, or orthopnea [13]. The diagnosis of PPF can be substantiated through the analysis of pleural effusion and the identification of a pseudocyst and/or fistula in imaging studies [14]. In the current study, the child reported dyspnea of grade 3, while the saturation of room air was found to be within the normal range. Individuals suffering from pancreatic bronchial fistulas may additionally experience recurrent lobar lung consolidations.

When a Pancreatico pleural fistula (PPF) is suspected, it is imperative to obtain detailed radiographic imaging to definitively confirm the diagnosis. MRCP is a non-invasive imaging modality for the identification of specific abnormalities associated with Chronic Pancreatitis (CP), including the dilation and narrowing of the main pancreatic duct and its branches, as well as the presence of pancreatic duct stones [15]. The diagnostic sensitivity of CT, endoscopic retrograde cholangio pancreatography (ERCP) and magnetic retrograde cholangio pancreatography (MRCP) in identifying a PPF respectively stands at 47%, 78%, and 80% [16]. Currently, it is advisable to utilize MRCP as the imaging modality of choice for confirming a PPF. This preference is based on the superior diagnostic capability of MRCP in delineating the presence of a fistula, in conjunction with its non-invasiveness, as opposed to that of ERCP. In the current instance, the MRCP findings were indicative of chronic pancreatitis, characterized by the presence of peripancreatic fluid collections that extended into the mediastinum, traversing the diaphragmatic hiatus, and accompanied by a significant accumulation of fluid in the right pleural space.

Conservative treatment is initially recommended as the primary therapeutic approach, which includes gastrointestinal rest, total parenteral nutrition (TPN), thoracic drainage, somatostatin analogues and has demonstrated success rates ranging from 33 to 60% in adult patients [17]. Endoscopic pancreatic stenting has been proven to effectively alleviate pancreatico pleural fistula (PPF) [18]. Endoscopic retrograde cholangiopancreatography (ERCP) is considered a safe procedure with a minimal risk of bleeding, estimated at less than 1% in pediatric patients [19]. In a series of minor research endeavors conducted with pediatric patients suffering from pancreatico ductal disruption and PPF, the success rates of both ERCP and stenting procedures were found to be 80% [20]. ERCP is considered an invasive procedure with a relatively low but significant complication rate. These complications include acute pancreatitis, along with sepsis and bleeding. ERCP may not always be accessible, and its performance can be challenging in pediatric patients. In such cases, initial conservative management strategies including tube thoracostomy and pancreatic enzyme suppression should be implemented. Invasive interventions, such as pancreatic duct stenting or surgical treatment, are warranted when conservative management fails. In the management of pleural effusion, the utilization of intrapleural fibrinolytics presents itself as a potentially safe, straightforward and cost-effective approach. Furthermore, the use of urokinase and tissue plasminogen, have demonstrated successful outcomes [21]. In the present case, the child was given Urokinase treatment, which gave favorable response. Octreotide provides a valuable addition to the therapeutic armament of the pediatric gastroenterologist for a wide variety of disorders [22]. In

this case, the child received conservative treatment, including octreotide infusion and the result was favorable.

CONCLUSION

In conclusion, while the occurrence of pancreatico pleural fistula (PPF) is secondary to chronic pancreatitis, it merits consideration in the differential diagnosis of pleural effusion in pediatric patients. The elevated concentrations of pleural fluid and the utilization of advanced imaging modalities, especially magnetic resonance cholangiopancreatography (MRCP) are crucial for the diagnosis of PPF. Consequently, an initial approach involving conservative treatment and endoscopic management is advisable.

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