Pancreatic pseudocyst: a rare presentation
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Pseudocyst formation is defined as a localised collection of pancreatic juice enclosed by a wall of fibrous or granulation tissue arising as a result of acute and chronic pancreatitis or pancreatic trauma. They are more commonly found in men than women and are solitary in most cases. Mostly occurs in lesser sac and peripancreatic region. Thoracic / mediastinal the extension is rare. About 100 cases have been reported in literature. The pathogenicity includes isomism involved in the formation of pleural effusion include direct contact of pancreatic enzymes with the diaphragm, haematogenous transfer of pancreatic enzymes into pleura, transfer of pancreatic secretions through trans diaphragmatic lymphatics and results in the formation of pancreatico pleural fistula which results in direct communication of pancreatic pseudocyst within the pleural cavity. The clinicopathologic features of the cyst include a cystic wall without an epithelial cell lining and a black cystic fluid with high amylase concentration. Pseudocyst persisting for longer than 6 weeks have been associated with three folds increases in complication rates. The complications of pseudocyst include infection, haemorrhage, obstruction, rupture and erosion into neighbouring structures including major blood vessels. Pancreatic fistula and ascitis result when there is leakage or rupture of pseudocyst into the abdominal cavity. Repeated pleural effusion presumably resulting from rupture and fistulation from pancreatic duct initial account the pleural cavity. Such presentation account for less than 3 % of cases and even rarer to be an asymptomatic event. Resection is the optimal treatment for persistent leakage from the tail of pancreas.

Keywords: Thoraco-abdominal cyst, No ductal communication, MRCP
Introduction

Pseudocyst of the pancreas is localised fluid collection rich in amylase and other pancreatic enzymes. A mediastinal extension is rare. The frequency of pancreaticopleural fistulas in patients with pancreatitis is estimated to be 0.4 to 4.5%. 80% with fistulas are associated with mediastinal pseudocyst. A posterior mediastinal cyst can be foregut duplication cyst, pancreatic pseudocyst, lymphatic malformation, cystic teratoma and cystic neuroblastoma. Differentiating all these needs prompt clinical examination and diagnostic modalities [1-6].

We report this rare case of mediastinal pseudocyst with the unusual presentation without any ductal communication. The maturation period of pancreatic pseudocyst is reported to be 2 to 6 weeks, during this time 33% of the cyst are expected to spontaneously resolve [7,8,9,10]. Only a few cases were reported in the literature. In our case, a large abdominal thoracic thin-walled pseudocyst with a major thoracic component behind the heart and extending along esophageal hiatus with a smaller component in the lesser sac is seen. Cyst measures were 14*4.1*6.5cm with the volume of 186cc. The pseudocyst has no fistulous communication with pleura / mediastinal structures. Paediatric surgeons proceeded with Bypass pancreas procedure with thoracoscopic drainage of pseudocyst with ICD placement on both sides. Intra and post OP period was uneventful. Cystic fluid showed elevated levels of amylase 9230 IU/L (1 in 10 dilutions) and lipase 45201 (1 in 100 dilutions). The histopathology showed thick cyst wall with no definite lining epithelium. It has a collection of chronic inflammatory cells with granulation tissue which was also consistent with a pancreatic pseudocyst. ICD clamped and removed on POD5. The child was discharged on incentive spirometry and pancreatic enzyme supplementation and on regular follow up.

Case Report

A 15-year-old boy presented with progressive worsening of breathlessness and cough for the past 1 month. He had been admitted 5 months ago for similar complaints where chest x-ray did show bilateral pleural effusion for which left side pleural taping was done. Pleural fluid analyses were negative for tuberculosis workup. But, Child was started on empirical ATT, but compliance was poor. CECT thorax done showed bilateral moderate pleural effusion, chronic calcific pancreatitis with pseudocyst extending to the posterior mediastinum. The child was referred to a higher center for further management. On examination, air entry was reduced on bilateral lung field particularly in mid and lower zone regions. Per abdomen examination was normal.

Routine blood investigations were normal. FBS, PPBS was normal. Serum amylase and lipase were elevated. MGE advised MRCP which showed atrophied pancreas, main (4mm) and accessory pancreatic duct dilated and multiple foci of strictures are seen. No peripancreatic inflammation. A large abdominal thoracic thin-walled pseudocyst with a major thoracic component behind the heart and extending along esophageal hiatus with a smaller component in the lesser sac. Cyst measures were 14*4.1*6.5cm with the volume of 186cc. The pseudocyst has no fistulous communication with pleura / mediastinal structures. Paediatric surgeons proceeded with Bypass pancreas procedure with thoracoscopic drainage of pseudocyst with ICD placement on both sides. Intra and post OP period was uneventful. Cystic fluid showed elevated levels of amylase 9230 IU/L (1 in 10 dilutions) and lipase 45201 (1 in 100 dilutions). The histopathology showed thick cyst wall with no definite lining epithelium. It has a collection of chronic inflammatory cells with granulation tissue which was also consistent with a pancreatic pseudocyst. ICD clamped and removed on POD5. The child was discharged on incentive spirometry and pancreatic enzyme supplementation and on regular follow up.

Discussion

Pancreatic pseudocyst in children is rare, although it is well documented as a primary consequence of pancreatitis needing in most of the cases surgical or endoscopic intervention. Regardless of the etiology of the pseudocyst, the incidence is low, 1.6% - 4.5%, or 0.5 – 1 per 1000 individuals per year [11].Pancreatic pseudocyst is a well recognized and common complication of acute and chronic pancreatitis. In acute pancreatitis, fluid gets collected in the peripancreatic region due to the inflammatory process. After 4 weeks, it gets walled up with granulation and connective tissue. It is called pseudocyst because it has no epithelial lining. Pancreatic fluid in retroperitoneal space tracks into mediastinal space through aortic, oesophageal hiatus and forms mediastinal, thoracoabdominal pseudocyst. In our patient, it was thoracoabdominal. Most of such patients have pleural effusion. Effusion can be either due to reactionary changes or fistulous connection.

It is recognised with increasing frequency in the paediatric population as a result of trauma, biliary tract disease, viral illness, state of intracranial hypertension and steroids.
Trauma is the most common cause of pseudocyst in children. Other causes are congenital anomalies, drug-induced. The anomalous pancreatic duct, anomalous pancreaticobiliary junction, pancreatic divisum, annular pancreas causes recurrent pancreatitis. Ectopic pancreatic tissue with the formation of pseudocyst is also reported in the literature.

The usual symptom of pseudocyst will be tense cystic mass in epigastric, umbilical, or in the left hypochondrium. Misleading presentation in case of mediastinal pseudocyst is mainly due to compression or invasion of adjacent structures by pseudocyst i.e. dyspnoea, cough, chest pain, dysphagia, hemoptysis, cardiogenic shock, acute respiratory failure, pseudo achalasia, weight loss [12]. Abdominal symptoms will be rare in the case of the mediastinal cyst.

In the absence of any peripancreatic collection/communication, these cysts are difficult to say as a pseudocyst of the pancreas. A chest X-ray can show pleural effusion.USG/ CT will detect the size, location, and wall thickness of the cyst. MRCP and ERCP will establish fistulous communication between the pancreatic duct and pleura. ERCP leads to diagnosis in 80% of cases, demonstrates a fistulous tract in 59%, and allows stenting. Pleural fluid will high amylase and albumin content (In pancreaticpleural fistula it may exceed 10,000IU/L).Endoscopic ultrasound gives more information about pancreas and duct if any communication. Endoscopic USG guided aspiration of fluid and analysis of fluid will be the definitive diagnostic measure.

Most pseudocyst will resolve spontaneously with supportive care. Spontaneous resolution occurs in about 50% of cases and complications occur in 5 to 40% of cases. Spontaneous regression of pseudocyst is likely with conservative management with bowel rest, total parenteral nutrition and non-operative management are successful, particularly when the pseudocyst is less than 6 cm. If any ductal obstruction or communication with the cyst is there, it should be managed first with ERCP stenting. In cases where the size of pseudocyst exceeds 6cm, conservative management is ineffective thus the choice of treatment includes surgical internal drainage (Cystogastrostomy/Marsupialization, Roux-en-Y cystojejunostomy,), endoscopic drainage procedures and percutaneous catheter drainage methods. For mediastinalpseudocyst, these options are not available.

Endoscopic / Thoracoscopic aspiration of pseudocyst is also reported.

In our patient, since major part eas intrathoracic and lower part of cyst is not accessible through the abdomen, thoracoscopic drainage of pseudocyst was done with bilateral ICD placement. Possible complications associated with pseudocyst include pancreatic abscess, rupture, haemorrhage or gastric outlet obstruction.

**Conclusion**

Pseudocyst of the pancreas in children is a very rare but potentially troublesome clinical entity, whose management depends upon the size, location and symptoms of such a pseudocyst. This is a case of pseudocyst presenting as Pseudo Meig's syndrome, an abdomen case presenting with respiratory symptoms, where a stepwise approach helped in making the diagnosis and appropriate management.

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