CASE REPORT

Percutaneous drainage of a bleeding pancreatic duplication cyst: a case report

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SUMMARY

Pancreatic duplication cyst is rare in children. Diagnosis may be difficult as it usually presents with abdominal pain and other varying non-specific symptoms. Cross-sectional imaging such as computed tomography (CT) scan is usually needed to diagnose the condition and provide more information for surgical planning. Definitive treatment remains excision of the cyst. We report a case of pancreatic tail duplication cyst in a 2-year-old girl who presented with abdominal pain and anaemia due to a bleeding and perforated cyst diagnosed by ultrasound and CT scan. The patient was managed initially with percutaneous drain insertion followed by spleen-preserving distal pancreatectomy and excision of the cyst. The diagnosis of pancreatic duplication cyst was confirmed on histopathology. The child remained well after one year of follow-up.

INTRODUCTION

Pancreatic duplication cysts are the rarest among duplication cysts.[1] Diagnosis is difficult and presentations vary, most often presenting as abdominal pain.[2-6] Excision of the cyst with or without excision of parts of the pancreas is the management of choice.[2-4] We report a pancreatic duplication cyst presenting with abdominal pain and anaemia that was initially managed with percutaneous drainage before definitive surgery.

CASE PRESENTATION

A 2-year-old girl presented with fever, abdominal pain, non-bilious vomiting and diarrhoea for 1 week. She had a similar episode of pain at 1-year-old with abdominal fullness in the left abdomen but did not seek further treatment or investigation for the symptoms. She otherwise did not have any other significant medical history or history of trauma. The child was pale, but active on examination with a pulse rate of 134/min, blood pressure 118/58 mmHg, and temperature of 38°C. There was a tender left abdominal mass. Full blood count revealed haemoglobin of 6.3 g/dL, total white blood count of 9.7 × 10^9/L, platelet 440 × 10^9/L. Renal profile and liver function tests were normal. Serum amylase was 77 units/L. A provisional diagnosis of intussusception was made.

Abdominal ultrasound revealed a large left paracolic collection measuring 5 × 5 × 10 cm (Figure 1A). No evidence of intussusception was seen. The initial diagnosis of intraabdominal abscess was made. The child was transfused with 240 mL of packed cells followed by ultrasound-guided percutaneous drain insertion that drained up to 500mL of stale blood. Fluids sent for cultures and acid-fast bacilli were negative, while fluid for amylase was 138 units/L.

The child was treated with intravenous cefotaxime and metronidazole for 7 days. Her pain and fever resolved, and she regained her appetite. However, she continued to drain about 200 mL per day, at first hemoserous, then serous after 6 days. The colour of the drain output turned to light green with blackish sediments after 7 days. The persistent and unusual drain output prompted further investigations.

Repeated ultrasound revealed a gut signature sign of the cystic lesion which is relatively specific for duplication cyst (Figure 1B). Contrast-enhanced computed tomography (CT) abdomen performed showed rim-enhancing collection in the left retroperitoneal region. A bifid pancreatic tail was seen with a subjacent cystic mass measuring 1.6 × 2.6 × 2.1 cm and a claw sign at the ventral tail of the pancreas (Figure 2). The wall of the cystic lesion has a corrugated appearance resembling a gastric wall. A separate collection was seen anterior to the left Gerrota's fascia measuring 3.0 × 1.7 × 2.8 cm with the tip of the percutaneous catheter in situ. A tiny defect in the cystic wall that communicated with the collection suggested perforation (Figure 3). The distal pancreatic duct was dilated but did not demonstrate any connection to the cystic lesion. There was no radiological evidence of pancreatitis.

A laparotomy was performed with the operative findings of a distal pancreatic cyst measuring 6 × 7 cm. Spleen preserving distal pancreatectomy and excision of the cyst was performed. The histopathology report described the cyst as composed of full-thickness gastric tissues with an abnormal dilated pancreatic duct structure. The surrounding pancreas showed changes of chronic pancreatitis with areas of mucosal ulcerations along the pancreatic duct.

Post-operatively the child recovered uneventfully. She was followed up in the clinic 1 year after the surgery without further complications.

DISCUSSION

The incidence of alimentary tract duplications that may occur from the mouth to the anus is 1:4500, with duplication of the pancreas the rarest among them.[1] Pancreatic

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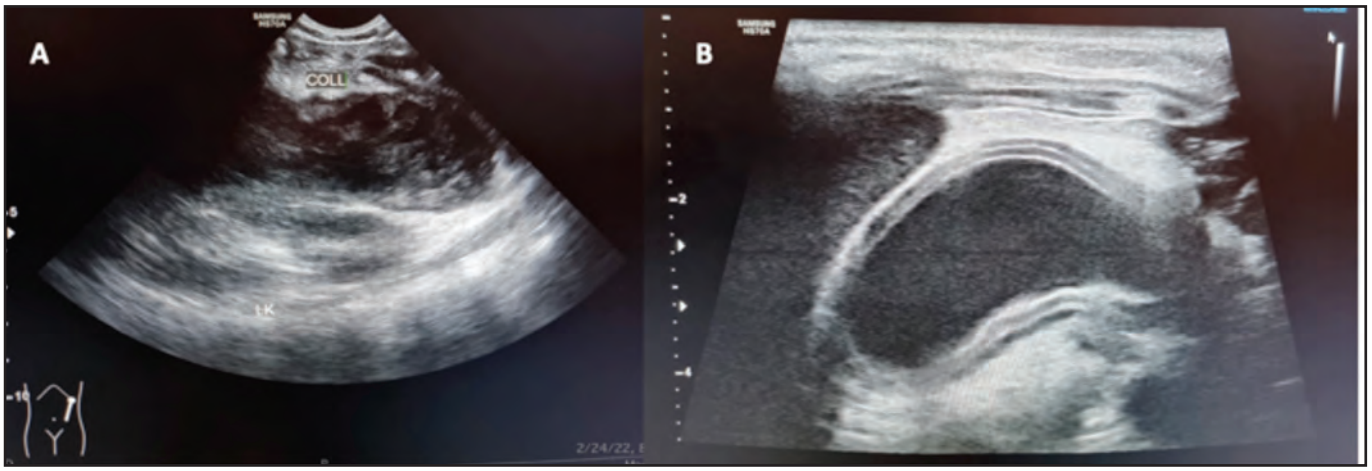

Fig. 1: Initial ultrasound showed heterogenous hypoechoic collection in the left retroperitoneal region (A). Targeted relook ultrasound showed a cystic lesion with the gut signature sign, with echogenic mucosa and hypoechoic muscularis propria, relatively specific for duplication cyst (B).

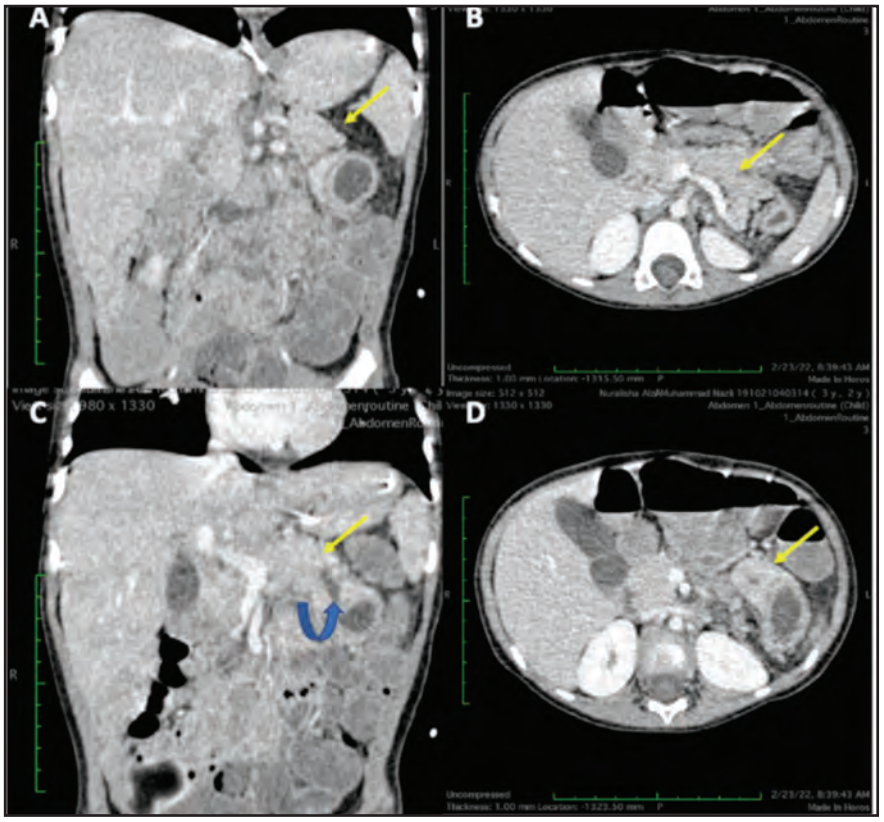

Fig. 2: Contrast-enhanced CT abdomen on coronal reconstruction and axial view showing a bifid pancreatic tail with dorsal tail (A, B) and ventral tail (C, D). Note that the thick-walled cystic lesion is subjacent to the ventral tail of the pancreas, with a dilated distal pancreatic duct as denoted by curved arrow (C). Note that the thick wall cystic lesion has a claw sign with an adjacent ventral tail of the pancreas (D).

duplication cyst with gastric-type mucosa lining is rarer still.[5,6] In the literature, the nomenclature of duplication cysts may be defined based on anatomical origin or histological features.[4,7] In our report, we prefer to classify duplication cysts based on the anatomical origin to avoid confusion. The majority (50%) of the cysts originate from the head of the pancreas, while origin from the body and the tail represent about a quarter each.[4]

Several reports described pancreatic duplications associated with bifid pancreas similar to our case.[2,3,7,8] The cyst may or may not communicate with the main or accessory pancreatic buds.[3,7] These various associations and communication with the pancreatic ducts suggest embryological malformation.[3,7,8]

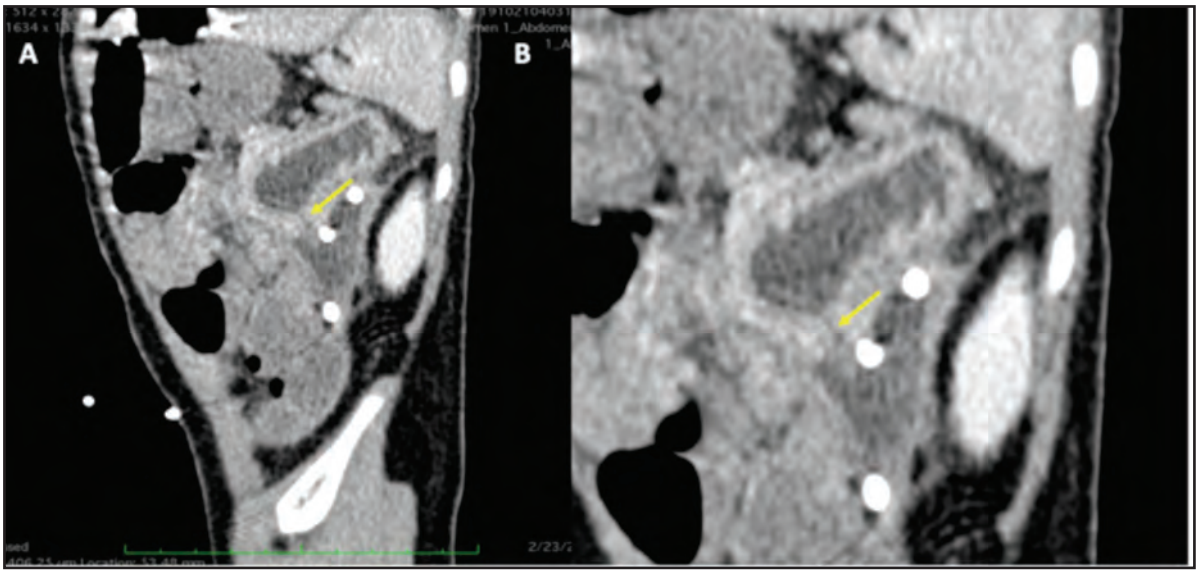

Fig. 3: Contrast-enhanced CT abdomen on sagittal reconstruction showed a focal wall defect at the inferior aspect of the cystic lesion which communicates with the left retroperitoneal collection (A, B). Note the corrugated appearance of the wall of the cystic lesion resembling the gastric wall (B).

There are a few postulations regarding the formation of duplication cysts including partial twinning, split notochord theory, diverticula and canalization defects, and environmental factors.[1] In pancreatic duplication cysts, the most popular theory to its formation is due to a traction diverticula formation along the neuroenteric band between the stomach and the pancreas due to impaired separation of the notochord and endodermal layers.[7,8] This theory may also explain the common association of gastric duplication or pancreatic duplication containing gastric-type mucosa with the formation of a bifid pancreas as the malformation may develop on either end.[8]

The majority of pancreatic duplication cysts present with abdominal pain (67–72%).[2-4] The pain may be due to inflammation and ulceration of the cyst, pancreatitis or perforation leading to peritonitis.[2-4] Repeated infections, obstruction of the pancreatic duct from viscous mucus secretions, or biliary sludge may exacerbate the pancreatitis and ulceration of the cyst.[3] This may be associated with vomiting and the presence of an abdominal mass.[4] Anorexia and weight loss have been reported mimicking presentations of pancreaticoblastoma.5 Rarely the cyst may bleed resulting in anaemia and ulceration into a bowel manifesting as a gastrointestinal (GI) bleed due to hydrochloric acid secretion from gastric mucosa lining the cyst which most likely occurred in our patient.[6,8]

Serum amylase and lipase levels are usually raised but may be normal.[2] Ultrasonography is the first-line imaging for diagnosis and the appearance of the 'gut signature sign' is specific for duplication cysts.[9] However, this feature may not be seen due to the chronic inflammation and perforation of the cyst that has disrupted the cyst wall and was only seen later after drainage of the collection in our patient. Hence CT scan or MRI would be required to identify the location, extent, anatomic relations, and characteristics of the lesion to differentiate it from other cystic pancreatic lesions such as pancreatic neoplasms and pancreatic pseudocyst.[10] MRCP, ERCP, endoscopic ultrasound, and intra-operative pancreaticography are useful adjuncts to further delineate the relationship of the cyst with pancreatic ductal anatomy although all these procedures require general anaesthesia if performed in small children. Ultimately operative findings and histology are required for the final diagnosis.[3,10]

Excision of the cyst is curative.[4] The type of surgical management depends on the location of the pancreatic cyst. For lesions located at the tail of the pancreas, spleen-preserving distal pancreatectomy and cyst excision are recommended to prevent recurrence of the cyst and pancreatitis as well as to prevent potential malignant transformation.[3] Excision of the cyst without damaging the surrounding pancreatic tissue is possible although some patients with lesions at the head of the pancreas may require pancreaticoduodenectomy or Roux-en-Y cystojejunostomy.[4] The overall outcome of pancreatic duplication cyst is good with very low complications.[4]

Although surgical excision is the definitive treatment for the lesion, initial drainage of the cyst should be considered in selected patients. Placement of a drain offers initial symptomatic relief, aid in the diagnosis, and treat infective collections that may potentially progress to sepsis while planning for surgical management of the patient. The presence of stale blood and later greenish fluids that may likely be pancreatic fluids or bile in this case prompted a reassessment of the patient with further imaging that led to the final diagnosis of pancreatic duplication cyst. Percutaneous drain insertion should only be considered if there is an adequate window for puncture and drainage of the collection without injuring vital surrounding structures. Endoscopic transgastric cystogastrostomy has been reported for initial symptomatic relief for pancreatic duplication but this technique is more invasive and would require general anesthesia.[7] Percutaneously inserted drain is less invasive

and can be performed with sedation. This was performed for our patient without any complications and with marked improvement in symptoms before spleen-preserving distal pancreatectomy and cyst excision were performed.

CONCLUSION

Pancreatic duplication cyst is a rare anomaly that usually presents with abdominal pain but rarely with anaemia due to bleeding and perforated cyst. Diagnosis is difficult, a computed tomography (CT) scan is usually needed for diagnosis and surgical planning. In selected patients, an initial percutaneous drain insertion to evacuate the collection from the perforation or the cyst is feasible to provide early symptomatic relief and allow time for further work-up and surgical planning. Treatment of the lesion remains excision of the cyst, preferably with the surrounding pancreatic parenchyma to prevent recurrence and risk of malignant change.

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CONFLICT OF INTERESTS

There was no conflict of interest to be declared.

ETHICS BOARD APPROVAL

For the publishing of this article, approval was sought and exempted by the Medical Research and Ethics Committee MREC on 27 March 2023.

CONSENT

Permission was obtained from the parents for this case report.

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