CASE REPORT

Accessory pancreatic lobe with gastric duplication cyst: diagnostic challenges of a rare congenital anomaly

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SUMMARY

Reports of combined congenital abnormalities of gastric duplication cysts and accessory pancreatic lobes are rarely reported. Patients with such anomalies may require appropriate surgical intervention tailored to the individual patient for complete cure. A multimodality diagnostic approach using ultrasonography, CT and MRI is useful for appreciation of the relevant anatomy of the congenital abnormality and for proper surgical planning. We present a case of a gastric duplication cyst with accessory pancreatic lobe and ectopic pancreatic rest in a 5-year-old child presenting with symptoms of recurrent pancreatitis.

BACKGROUND

An enteric duplication cyst is an uncommon congenital abnormality that may occur anywhere between the mouth and rectum.^{1 2} Among these cases, pancreatic duplication or accessory pancreatic lobe is an extremely rare congenital anomaly.³ Further, reports of coexistence of gastric duplication cysts with pancreatic anomalies are even rarer.⁴ The coexistence of gastric and pancreatic duplications is not easy to diagnose. Patients may present with a variety of symptoms, including recurrent abdominal pain, vomiting, or pancreatitis, which warrant appropriate management.² In this context, we report a case of a 5-year-old child diagnosed with pancreatic and gastric duplication cysts, who also had ectopic pancreatic rest. We emphasise that a multimodality approach using ultrasonography, CT scan, MRI and MR cholangiopancreatography (MRCP) is essential for appreciation of the relevant

anatomy and for appropriate surgical planning, as demonstrated in our case.

CASE PRESENTATION

A 5-year-old girl presented with recurrent upper abdominal pain for the past 4 months. Physical examination revealed abdominal guarding. Laboratory investigations exhibited elevated levels of serum amylase and lipase. Other laboratory examinations were within normal range. The patient was diagnosed as a case of pancreatitis. She was referred to the Department of Radio Diagnosis and Imaging the hospital for further investigations.

INVESTIGATIONS

Abdominal sonography revealed a bulky pancreas with coarse echotexture. A dilated pancreatic duct with a few small cysts was identified in the body and tail region of the pancreas (figure 1). The gallbladder and common bile duct were normal. Contrast-enhanced CT scan of the abdomen revealed an enlarged pancreas with duplication of pancreatic gland in the body and tail regions, which were attached to the main pancreatic gland by a well-defined stalk (figures 2 and 3). The accessory pancreatic lobe was atrophic and displayed multifocal dilations and strictures of its duct. Further, the accessory pancreatic lobe was closely associated with a soft-tissue density lesion along the greater curvature of the stomach, with enhancement similar to the pancreas, raising the suspicion of ectopic pancreatic rest (figures 2 and 3). This lesion was further evaluated by sonography, which revealed wall thickening with a layered appearance and finger-like

Figure 1 Abdominal sonography revealed an enlarged pancreas with coarse echotexture and dilated pancreatic duct with a few small cysts in the body and tail regions.

Figure 2 Contrast-enhanced CT scan of the abdomen revealed an accessory pancreatic lobe with signs of pancreatitis and its close association with a gastric duplication cyst.

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Figure 3 Contrast-enhanced sagittal CT scan reconstruction revealed an accessory pancreatic lobe between a gastric duplication cyst and main pancreatic lobe; mucosal fold thickening was noted in the adjacent gastric wall.

projections into the lumen (figure 4). On dynamic scan after distending the stomach with water, the lesion, which had a collapsed lumen, now exhibited certain peristaltic movements and had moving internal echoes, leading to the diagnosis of a gastric duplication cyst with gastric communication (figure 5). The main pancreatic duct was normal in course and calibre, and was opening normally with the common bile duct at the major duodenal papilla. The CT scan revealed a small, submucosal lesion, with enhancement similar to that seen in the pancreas, having a duct-like structure within, which was projecting into the lumen of the first part of the duodenum, raising suspicion of ectopic pancreatic rest (figure 6). Subsequently, MRI was performed for evaluation of the ductal anatomy. MRI with MRCP confirmed the presence of a gastric duplication cyst and duplication of pancreatic duct in the body and tail regions, with multiple strictures and cystic dilation of the duplicated duct (figure 7). The duplicated duct of the accessory pancreatic lobe was connected to the main pancreatic duct in the body of the pancreas. It also demonstrated the normal course of the main pancreatic duct and common bile duct, and their common opening at the major papilla of the duodenum. The T2-weighted image also confirmed the presence of an accessory pancreatic lobe with gastric duplication cyst (figure 8). It also revealed ectopic pancreatic rest in the first part of the duodenum.

Figure 4 Sonography revealed thick-walled structures with a layer appearance and finger-like projections into the lumen, attached to the stomach wall, suggesting a gastric duplication cyst.

Figure 5 Diagnosis of gastric duplication cyst with gastric communication by dynamic scan.

OUTCOME

Surgical excision of the gastric duplication cyst and accessory pancreatic lobe was planned based on the diagnostic findings. The surgery was successful. At 6-month follow-up, the patient was asymptomatic and had not experienced any episodes of acute pancreatitis.

DISCUSSION

Duplications of gastrointestinal organs are very rare congenital abnormalities with an occurrence rate of 1 in 4500–10 000 live-births.¹ Gastric duplications account for 3.8% of all duplications of the gastrointestinal tract.² Further, this gastric anomaly in association with accessory pancreas and pancreatic duplications is rarely reported.⁵ In fact, to the best of our knowledge, this is the 24th reported case of a combined gastric duplication cyst and duplicate pancreas,⁵ and the first reported case from Gujarat, India.

The underlying cause of combined duplications of pancreas and gastric cysts during embryological development is hypothesised to be traction along a neurenteric band between the stomach and the pancreas.⁶ Such patients usually present with symptoms of recurrent acute pancreatitis as a result of obstruction of the pancreatic duct by viscous mucus secretions, ulcer bleeding or biliary sludge.⁴ The duplicate pancreatic lobe is presented as an accessory lobe of pancreatic tissue connected with the main pancreatic gland and containing an aberrant duct.⁴ On the other hand, gastric duplication cysts result from abnormal

Figure 6 CT scan revealed a small, submucosal lesion having a duct-like structure within, raising the suspicion of ectopic pancreatic rest.

Figure 7 MR cholangiopancreatography revealed duplication of pancreatic duct in the body and tail region with a few cystic dilations in the duct of an accessory pancreatic lobe and its close association with a gastric duplication cyst.

foregut development and usually occur in the greater curvature.² Pathologically, most enteric duplications present as cysts in relation to the organ, with a thickened muscular wall that usually shows some evidence of inflammation, and which is the cause of recurrent pain.²⁻⁵ However, in some cases, the inflammatory response may destroy the lining and may lead to diagnostic confusion with idiopathic pseudocysts.⁵ Therefore, it is important not only to order the appropriate diagnostic modality, but also to interpret the data correctly.² It should be noted that not all cystic structures near the stomach and pancreas are pancreatic pseudocysts.² In our case, we successfully identified combined duplication of the pancreatic lobe and gastric cyst. Additionally, our patient also exhibited ectopic pancreatic rest, presenting as pancreatic tissue that lacked anatomic or vascular continuity with the normal pancreas. Ectopic pancreas is reported to have an incidence rate of 1–15%,⁴ and is usually confused with pancreatic submucosal tumours.⁷ Nevertheless, contrast-enhanced CT images helped in differential diagnosis in our case, as the

structure appeared as a homogeneously enhancing intramural subepithelial mass with irregular border and intraluminal growth.

The analysis of 23 previously reported cases of combined gastric and pancreatic duplication revealed that the majority of such cases were reported among females (>65%). Further, the congenital abnormality was identified before 12 years of age in more than 50% of the patients. The common finding in all these patients was presentation of recurrent abdominal pain. Data of anatomical evaluation revealed that gastric duplication was connected to the main pancreatic duct in 78% of patients. 10 patients (43.5%) had undergone multiple operations before achieving complete symptom resolution.² Here, we emphasise that diagnostic accuracy of congenital anomalies of the pancreas and pancreatic duct is imperative because these anomalies can be corrected surgically, resulting in complete cure for recurrent pancreatitis or gastric outlet obstruction, and can avoid multiple operations and years of morbidity. Therefore, we recommend that pancreatitis in young patients should be thoroughly investigated. Our case reflects the role of a multimodality approach in providing critical diagnostic information. Each modality has its specific role. In our case, CT scan revealed the diagnosis of accessory pancreatic lobe, ultrasonography was helpful for diagnosis of gastric duplication cyst, while MRI and MRCP were valuable for depicting the ductal anatomy and ectopic pancreatic rest.

Although rarely reported, the combined duplication of pancreatic lobe and gastric cyst is curable in the majority of patients.² The optimal treatment includes local excision of the gastric duplication and duplicate pancreas as well as closure of the pancreatic duct.² We believe that improved awareness of the condition and proper diagnosis with accurate interpretation would help practitioners in appropriate surgical planning with a tailored approach for each patient.

Learning points

- ▶ Congenital abnormality reports of coexistence of gastric duplication cysts and accessory pancreatic lobes are very rare.
- ▶ Acute recurrent pancreatitis in young patients should be investigated thoroughly.
- ▶ Besides ordering the appropriate diagnostics, it is also important to interpret the data correctly.
- ▶ A multimodality approach using ultrasonography, CT scan and MR cholangiopancreatography is essential for appreciation of the relevant anatomy and for appropriate surgical planning.

Figure 8 The T2-weighted image confirmed the presence of an accessory pancreatic lobe in close association with a gastric duplication cyst.

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