CASE REPORT

Disconnected pancreatic duct syndrome: a multidisciplinary management dilemma

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SUMMARY

Disconnected pancreatic duct syndrome is a circumferential interruption of the pancreatic duct. It usually occurs secondary to pancreatitis and carries significant diagnostic and management challenges. We present a case of disconnected pancreatic duct syndrome that represented a diagnostic and management dilemma for both medical and surgical teams. The aim of this article is to share a successful management experience of disconnected pancreatic duct syndrome with other physicians and to perform a brief but focused literature review on this challenging condition.

BACKGROUND

Disconnected pancreatic duct syndrome (DPDS) is a circumferential disruption of the continuity of the pancreatic duct resulting from an area of cellular necrosis.¹ This compromised ductal integrity leads to extraductal leakage of pancreatic secretions and the destruction of viable pancreatic tissue surrounding the duct. In extreme cases, DPDS can cause leakage of pancreatic fluid into the peritoneal cavity, causing chemical pancreatic ascites.^{1 2} The disconnected segment can occur at any part of the duct, but it most commonly occurs within the head or the body of the pancreas. Trauma and necrotising pancreatitis (acute or chronic) are the two most common non-iatrogenic causes of DPDS.³ The diagnosis is usually made by MR cholangiopancreatography or endoscopic retrograde cholangiopancreatography (ERCP) with or without endoscopic ultrasound. The majority of cases do not respond to conservative management and therefore require more invasive therapies, such as ERCP stenting or surgical resection of the necrotised pancreatic segment. Also, DPDS represents a challenge for the hospital team because of the associated persistent pain and prolonged hospitalisation.^{2 3} We present a case of DPDS and illustrate therapeutic strategies with a focused literature review.

CASE PRESENTATION

A 35-year-old man with a history of chronic abdominal pain secondary to chronic alcoholic pancreatitis, complicated by pseudocyst formation, presented to the emergency department with acute epigastric pain that radiated to his back. He rated the pain as 9/10 on the pain severity score (where 0 represents no pain and 10 represent the maximum pain severity). The patient's new pain was associated with severe nausea and vomiting. He had a percutaneous drain that was placed to evacuate the pancreatic pseudocyst 20 days before presentation.

At the time of presentation, the patient had a low-grade fever (38.4°C), and his heart rate was elevated (121 bpm). The patient had generalised abdominal tenderness, abdominal rigidity and rebound tenderness in the epigastric and left lower quadrant. A scanty amount of clear fluid was draining from the drainage catheter in the pancreatic pseudocyst.

INVESTIGATIONS

The blood work-up was significant for elevated white cell count of $14 \times 10^9/L$ (normal $4\text{--}11 \times 10^9/L$) and elevated lipase levels 120 U/L (normal 7–60 U/L). Cultures from samples of the pancreatic fluid failed to grow any microorganisms. A contrast-enhanced CT scan of the abdomen showed pancreatic neck and body necrosis, peripancreatic fluid collection (figure 1) and a pocket of retroperitoneal fluid collection adjacent to the sigmoid colon.

The patient was treated with bowel rest, intravenous hydration and scheduled doses of hydromorphone for pain control. No improvement in the patient's condition was noted 48 hours later. The patient's symptoms were highly suspicious for pancreatic fistula or DPDS causing chemical peritonitis; therefore, ERCP and antegrade and retrograde endoscopic ultrasonography were performed to confirm the diagnosis.

During the retrograde endoscopic ultrasonography, 25 mL of clear, yellowish fluid was aspirated from the retroperitoneal pocket seen on the CT scan. The analysis of the fluid revealed a high amylase level, suggesting pancreatic fluid leakage. ERCP revealed opacification of the pancreatic duct and disruption of the duct in the pancreatic body at the genu (figure 2). The ERCP wire was unable to be passed distally into the pancreatic tail, confirming the diagnosis of DPDS (figure 2). Stent placement using ERCP was also unsuccessful, despite multiple attempts, leaving surgical intervention as the last available intervention to treat this patient's condition.

TREATMENT

Pain control was achieved by intravenous hydromorphone with the guidance of the pain management team. The location of the necrosis in the pancreas head, the relatively proximal site of the DPDS and the intact pancreatic body and tail made the resection of the disconnected segment by a distal pancreatectomy a non-preferred option for this patient. Internal drainage was then considered. The walled-off pancreatic necrosis, including the disconnected segment, was successfully drained into the stomach, using Roux-en-Y internal drainage pancreaticogastrostomy.

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Figure 1 (A) CT scan with contrast of the abdomen showing the area of pancreatic necrosis at the pancreatic head-neck segment (red arrow) with the tip of the drainage catheter at the exact same area. (B) Showing the rest of the pancreas (green arrow) is intact with no obvious damage or necrosis.

OUTCOME AND FOLLOW-UP

The postoperative course was complicated by healthcare-associated pneumonia that was treated successfully with a prolonged course of antibiotics. The patient recovered from the surgical procedure slowly but progressively and was discharged to an inpatient rehabilitation facility at postoperative day 23. Follow-up was coordinated between gastroenterology, pain management and general surgery.

Follow-up at 1 and 6 months revealed an improvement in the general condition of the patient. The patient described an interval improvement in the chronic abdominal pain and described a successful weaning process from opioid pain medications.

DISCUSSION

DPDS occurs when the pancreatic duct's continuity becomes interrupted. This disease usually develops secondary to acute

necrotising pancreatitis, chronic relapsing pancreatitis or pancreatic fistula.^{1 2} The prevalence of DPDS is unknown but studies have shown that acute necrotising pancreatitis can be complicated by DPDS in 16–23% of cases.^{1 3} Substantial pancreatic necrosis and viable pancreatic tissue distal to the disconnected duct segment are usually critical to establish the diagnosis.^{4 5} Findings of an intrapancreatic fluid adjacent to the main pancreatic duct and radiographically viable distal pancreatic tissue on contrast-enhanced CT scan are characteristic of DPDS diagnosis.¹ However, diagnosis is usually confirmed by ERCP or MR cholangiopancreatography. One study has shown that the accuracy of CT scans for detecting pancreatic duct trauma and discontinuity is high in the portal phase of contrast enhancement.⁶ However, the clinical application of this concept is still controversial due to the resemblance of multiple pancreatic pathology findings on imaging techniques.⁵ Fluid accumulation and necrosis near the pancreatic neck and genu also raise the suspicion for DPDS.⁵ Early identification and diagnosis of this syndrome is crucial to prevent complications, particularly fistula formation.²

With the limited success of conservative management for the resolution of DPDS, CT-guided percutaneous drainage of fluid surrounding the disconnected segment has classically been the first-line treatment. Unfortunately, those drainage procedures are amenable to be complicated by fistula formation and often require endoscopic repair procedures.^{5 7}

ERCP insertion of an indwelling stent between the two disconnected ends of the main pancreatic duct appears to be a useful option for this group of patients. Some studies have shown that this procedure reduces the recurrence of pancreatic fluid collection in DPDS patients; however, the rate of stent migration is high.^{7 8} Another challenge with ERCP stenting in DPDS is difficulty of passing the stent between the two disconnected ends of the pancreatic duct, which is similar to our experience. Those two factors make surgery the last resort for those in whom ERCP stenting was unsuccessful.

Roux-en-Y internal drainage (by pancreaticogastrostomy, pancreaticojejunostomy or fistulotomy) and distal pancreaticosplenectomy are the two main types of surgery that are generally recommended for patients with DPDS and pancreatic necrosis for whom other non-surgical measures have failed.⁹ Many of those surgical procedures are associated with a high level of mortality and morbidity represented by infection, chronic pancreatic insufficiency, necrosis, multiorgan failure and death.^{10 11} Our patient underwent Roux-en-Y internal drainage pancreaticogastrostomy and not distal pancreaticosplenectomy for two main reasons. First, the involved segment of the duct was near the head of the pancreas, mandating a complete resection of the head of the pancreas proximal to the disconnected segment and

Figure 2 Endoscopic retrograde cholangiopancreatography with fluoroscopy. (A) The stent is advanced in the early part of the pancreatic duct (arrow). (B) Endoscopic retrograde cholangiopancreatography wire is incapable of passing distal to the disconnected segment and near the drainage catheter (arrow). (C) Disrupted continuity of the duct and disconnected pancreatic duct syndrome as the contrast did not fill the distal part of the pancreatic duct (arrow).

an immediate drainage to the stomach. Second, the distal portion of the pancreas was completely intact, making its removal along with the spleen an excessively invasive procedure to a functional part of the pancreas which may carry procedural complications and ultimately, poor outcome.^{10 11} Splenectomy is usually performed when the distal pancreatic segments (body and tail) are surgically removed, which was not the case in our patient.^{9 10} Limited therapeutic options seem to be the main driver for the surgical intervention in this group of patients.

The high recurrence and failure rates of the DPDS complications pose a management challenge to the primary care team in the hospital as well. Challenges encountered by the primary care team in the hospital include coordination of prolonged hospitalisation and pain management. The recurrence rate of DPDS generally is estimated to be 25–30%.^{2 5} This fact usually leads to recurrent readmissions for pain and symptoms management before the next intervention. In our experience, it is advisable to involve pain management services for the pain control strategies in this group of patients. Our patient experienced improved pain control, decreased dosing of narcotics and easier weaning from opioids, which was facilitated by the pain management team.

Learning points

- ▶ Disconnected pancreatic duct syndrome is a management challenge in many patients who develop this disease secondary to pancreatitis.
- ▶ Multidisciplinary team effort is mandatory to manage patients with this morbid illness.
- ▶ Surgical intervention is inevitable in some cases; however, it carries high morbidity and mortality profile.

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