CASE REPORT

Disconnected Pancreatic Duct Syndrome: A Case Series

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Abstract

Disconnected pancreatic duct syndrome (DPDS) is a condition where there is a ductal disconnection between viable secreting distal pancreatic tissues and the gastrointestinal tract. It may follow acute or chronic pancreatitis, abdominal trauma, and pancreatic surgery, leading to necrosis or structural disintegration of the pancreatic duct.

Aim: The aim of our study is to describe the imaging features of DPDS on ultrasound, computed tomography, and magnetic resonance cholangiopancreatography (MRCP) that helps in diagnosis.

We present a case series of DPDS with their imaging features in two settings, one in the patient with acute necrotizing pancreatitis and the other with blunt abdominal trauma.

Conclusion: Imaging plays a significant role in preoperative diagnosis. Contrast-enhanced computed tomography provides a comprehensive assessment of pancreatic duct integrity, and it shows its type and site of ductal disruption. It is a simple, effective noninvasive imaging modality in diagnosing pancreatic duct disruption.

Keywords: Acute necrotizing pancreatitis, Contrast-enhanced computed tomography, Disconnected pancreatic duct syndrome, Magnetic resonance cholangiopancreatography, Main pancreatic duct.

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Background

Disconnected pancreatic duct syndrome (DPDS) is a circumferential disruption of the continuity of the pancreatic duct between viable secreting distal pancreatic tissues and the gastrointestinal tract.[1] Acute pancreatitis, chronic pancreatitis, pancreatic surgery, or trauma are possible causes. The disconnected segment can occur anywhere throughout the duct; however, it usually occurs in the pancreatic head or body. This compromised ductal integrity leads to extra ductal leakage of pancreatic secretions and the destruction of viable pancreatic tissue surrounding the duct.[1,2] There can be partial or complete duct disruption.

The diagnosis is usually made by ultrasound, contrast-enhanced computed tomography (CECT), and MRCP. The diagnosis is confirmed by endoscopic retrograde cholangiopancreatography (ERCP).

We report a case series of DPDS following acute necrotizing pancreatitis and blunt abdominal trauma with their imaging features and concluding CECT as a noninvasive primary modality in diagnosing pancreatic ductal anatomy and its injury.

Case Description

Case 1 (Disconnected Pancreatic Duct Syndrome—Following Acute Necrotizing Pancreatitis)

A 70-year-old man presented to the emergency department with acute epigastric pain that radiated to his back. The patient had generalized abdominal tenderness, abdominal rigidity, and rebound tenderness in the epigastrium. He also complained of severe nausea and vomiting. At the time of presentation, the patient was conscious, oriented, and had a low-grade fever (38.4°C), and his heart rate was elevated (121 bpm).

All the blood parameters were normal except for a borderline white cell count of 11×10^9/L with elevated lipase levels 2401 U/L, amylase levels 250 U/L, and hemoglobin was 8.8 g/dL.

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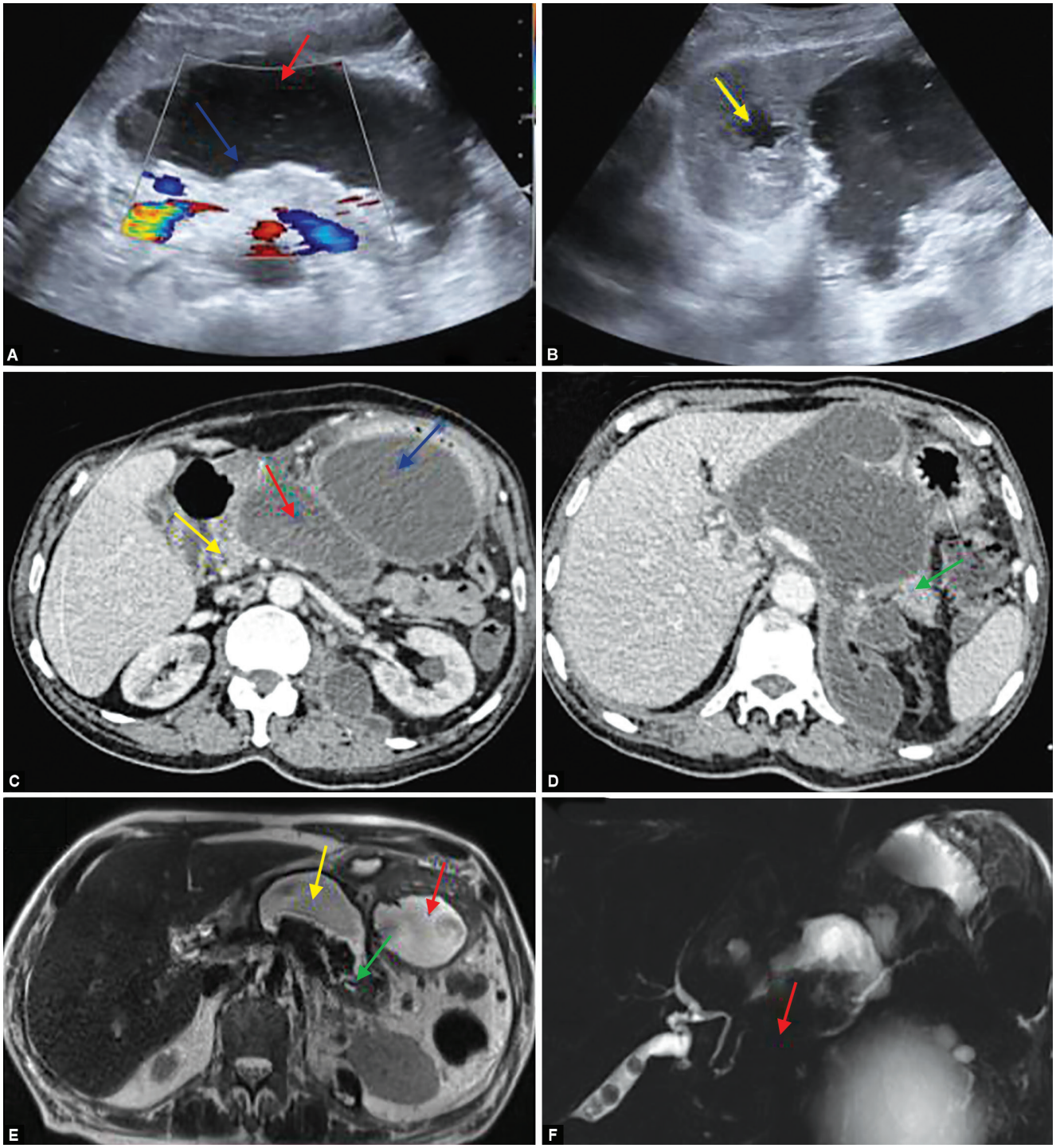
Ultrasonography (USG) of the whole abdomen with splenoportal axis Doppler reveals extensive collection extending from lesser sac to hypogastrium with internal echoes resembling pancreatic necrotic collection, which was communicating with a collection in a left lobe of the liver (Figs 1A and B). He also had cholelithiasis and mild free fluid in the abdomen.

A CECT of the abdomen reveals intrapancreatic collections measuring $4.3 \times 5.4 \times 4.8$ cm, completely replacing parenchyma in the neck and body region of the pancreas with nonvisualization of the main pancreatic duct in that region. The head, uncinate process, and tail region appear normal with prominent MPD. There was evidence of walled necrosis in the peripancreatic region and ascites (Figs 1C and D). There was also hepatomegaly with heterogeneously enhancing multiloculated cystic lesion in the left lobe of the liver and features of portal hypertension. As the collection was nonresolving along with other imaging features, a possibility of DPDS was considered.

Further evaluation by MRCP was advised to see the biliary and ductal anatomy.

Magnetic resonance cholangiopancreatography confirmed the nonvisualization of the main pancreatic duct in the neck and proximal body region. However, it was normal in the head and rest of the body region. Multiple intrapancreatic and peripancreatic



Figs 1A to F: A 20-year-old patient of acute necrotizing pancreatitis presenting with complication of disrupted pancreatic duct syndrome: Ultrasound abdomen image (A) Shows a large well-defined hypoechoic collection in suprapancreatic region at neck and body region (red arrow) compressing them inferiorly with few hyperechoic debris within (blue arrow) and showing no vascularity on color Doppler; (B) Reveals extension of collection superomedially segment III (yellow arrow) of the left lobe liver; CECT axial image of abdomen; (C) Reveals a large well-defined intrapancreatic hypodense lesion (red arrow) replacing pancreatic parenchyma in neck and body region s/o walled-off necrosis and another well-defined hypodense lesion in the left lumbar region (blue arrow) closely abutting above collection s/o extrapancreatic walled-off necrosis with normal proximal MPD (yellow arrow); (D) Reveals normal tail of pancreas in morphology and enhancement pattern and distal main pancreatic duct is opening into collection with duct making an angle of 90° with collection (green arrow); T2 HASTE axial section of MRCP; (E) Reveals a large hyperintense lesion in the pancreatic neck and body region with multiple T2-hypointense–dependent non liquefied debris within s/o walled-off necrosis (yellow arrows). Another well-defined heterogeneously hyperintense lesion in the left lumbar region is extrapancreatic collection (red arrows). The distal MPD at tail region making an angle of 90° with the collection and also opening into it suggestive of disconnected pancreatic duct syndrome (green arrow); T2 HASTE coronal thick multislice sequence of MRCP; (F) Reveals nonvisualization of MPD (red arrow) in neck and body region with normal caliber MPD at head and tail region

cystic lesions replaced pancreatic parenchyma in the neck and body region, which was hyperintense on T2WI with few dependent hypointense nonliquefied debris within s/o walled-off necrosis with cholelithiasis. There was communication between residual MPD with the pancreatic cystic collection, suggesting DPDS (Figs 1E and F).

The patient had been advised iv tramadol for pain with an antibiotic. Ultrasound-guided percutaneous drainage was done for the extensive collection, and the patient was kept on follow-up with percutaneous drainage output monitoring. The output reduced significantly after 1 week of treatment.

A follow-up CECT revealed multiple walled-off necrosis in pancreatic parenchyma with features of portal hypertension and an evolving abscess in the left lobe of the liver. Collection in the different pancreatic regions reduced drastically after percutaneous drain.

After 4 months, a follow-up ultrasound of the abdomen showed cholelithiasis and fatty liver; however, there was no evidence of intra-/extrapancreatic collection.

Case 2 (Disconnected Pancreatic Duct Syndrome—Following Blunt Abdominal Trauma)

A 20-year-old boy presented with abdominal pain and distension in the emergency department following blunt trauma to the abdomen by a steering wheel of the tractor. The patient had generalized abdominal tenderness, abdominal rigidity, and generalized abdominal distension. The patient was conscious, oriented, and his blood pressure was 100/60 mm Hg. All the blood parameters were normal except for raised amylase (137), lipase (1769), and low hemoglobin was 8.8 g/dL.

Ultrasonography of the whole abdomen with spleno-portal axis Doppler reveals complete transection of the head with loss of communication between head and body of pancreas with a hypoechoic collection in the neck region and free fluid in the abdomen.

Contrast-enhanced computed tomography abdomen revealed complete transection of pancreatic parenchyma in the neck and proximal body region (grade IV) with main pancreatic ductal disconnection and pancreatic and peripancreatic collection (Figs 2A and B). Immediate exploratory laparotomy with peritoneal lavage and bilateral drain placement was done.

Magnetic resonance cholangiopancreatography (MRCP) was done after exploratory laparotomy. It showed complete transection of the neck and proximal pancreatic body with discontinuity of main pancreatic duct (MPD) with biloma formation in segment V of the liver. Endoscopic retrograde cholangiopancreatography (ERCP) with pancreatic duct stenting was done, but unfortunately, the stent could not connect the two ends of the disrupted pancreatic duct.

The patient still had pus drainage from the initial drainage site even after 3 months; he was advised for follow-up CECT. A follow-up CECT abdomen was done and showed discontinuity of pancreatic parenchyma in the neck region with evidence of collection measuring 2.2 × 1.7 × 2.5 cm showing air-fluid levels within. Laterally, the collection was seen communicating with the dilated main pancreatic duct, and the pancreatic duct stent seemed to reach along with the medial wall of the collection. The body and pancreatic tail region appeared normal. There was also walled-off necrosis in the peripancreatic region with ascites.

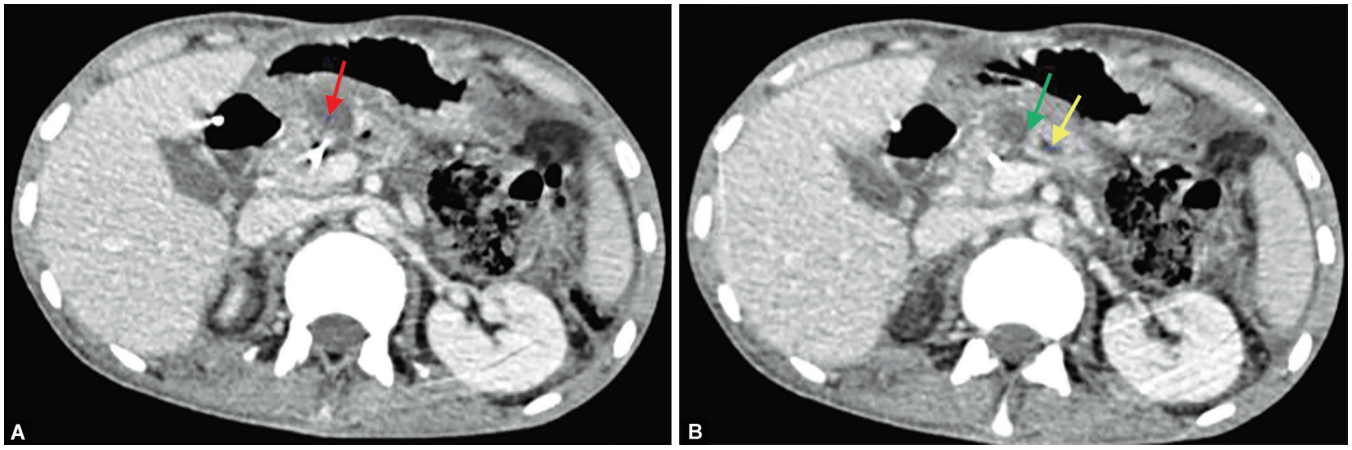
The final diagnosis of DPDS was made based on CECT features leading to pancreatic ascites and walled-off necrosis. The patient had been planned for distal pancreatectomy.

Discussion

Disconnected pancreatic duct syndrome occurs when the pancreatic duct's continuity is disrupted. This disease usually develops secondary to acute necrotizing pancreatitis, chronic relapsing pancreatitis, pancreatic surgery, or trauma.[1,2] The prevalence of DPDS is unknown, but studies have shown that DPDS can complicate acute necrotizing pancreatitis in 16–23% of cases.[1,3] Disrupted pancreatic duct leak into the peritoneal cavity can present as pancreatic ascites or communicate with the pseudocyst.

Significant pancreatic necrosis, the presence of intrapancreatic fluid adjacent to the main pancreatic duct, and viable pancreatic tissue distal to the disconnected duct segment are usually required to make a diagnosis.[4,5]

Contrast-enhanced computed tomography is a noninvasive procedure and can be used during acute pancreatitis without the risk of aggravation of pancreatitis. According to one study, CT scans had high accuracy for detecting pancreatic duct damage and discontinuity.[6] To avoid complications, such as creating a



Figs 2A and B: Axial CECT image of the abdomen in a patient with a history of blunt trauma abdomen reveals discontinuity in pancreatic parenchyma in its neck region replaced by an ill-defined collection (red arrow). Distal main pancreatic duct is dilated (yellow arrow) and opens into the collection with duct making an angle of 90° with collection (green arrow). Disconnected pancreatic duct syndrome

fistula, early detection and diagnosis of this disease are critical.[2] To confidently diagnose disconnected pancreatic duct, all of the following features must be present: (a) necrosis of at least 2 cm of the pancreas, (b) viable pancreatic tissue upstream (i.e., toward the pancreatic tail) from the site of necrosis, (c) extravasation of contrast material injected into the main pancreatic duct at pancreaticography, and (d) duct in the pancreatic tail segment should make the angle of 90° with the collection.[4]

Endoscopic retrograde cholangiopancreatography or secretin-stimulated MRCP is usually used to confirm the diagnosis. ERCP provides dynamic information about pancreatic duct disruptions, and it shows a contrast leak from the pancreatic duct. However, it has a few limitations. It is an invasive procedure and requires a skilled endoscopist. It is associated with complications such as pancreatitis and infection.

Secretin MRCP should not be used in an acute setting as secretin aggravates the inflammatory process. So their usefulness is weighed by their complications.

The first-line treatment for DPDS has been CT-guided percutaneous drainage of fluid surrounding the detached segment due to the low efficacy of conservative management. The installation of an indwelling stent between the two detached ends of the main pancreatic duct via ERCP appears to be a viable alternative for DPDS patients, as this procedure minimizes the occurrence of pancreatic fluid collection.[7,8] Roux-en-Y internal drainage (by pancreaticogastrostomy, pancreaticojejunostomy, or fistulotomy) and distal pancreatic splenectomy are the two main types of surgery recommended for patients with DPDS for whom other nonsurgical measures have failed.[9]

Conclusion

Imaging plays a significant role in the diagnosis and follow-up of DPDS patients. CECT can provide a comprehensive and noninvasive assessment of pancreatic ductal injury and acute pancreatitis, and it helps diagnose pancreatic duct disruption and intervention guidance.

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