Case 13694

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Disconnected pancreatic duct syndrome: imaging findings and endoscopic treatment

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DOI: 10.1594/EURORAD/CASE.13694 ISSN: 1563-4086 Section: Abdominal imaging Area of Interest: Pancreas Procedure: Catheters Procedure: Diagnostic procedure Imaging Technique: Fluoroscopy Imaging Technique: MR Imaging Technique: CT Special Focus: Acute Case Type: Clinical Cases Authors: Tonolini Massimo, M.D.1; Vella Adriana, M.D.1; Bareggi Emilia, M.D.2; Gambitta Pietro, M.D.2 Patient: 56 years, male

Clinical History:

Middle-aged male patient with history of prolonged hospitalization and medical treatment at another institution because of severe necrotic-haemorrhagic acute pancreatitis (AP) from biliary sludge, and allergy to iodinated contrast medium.

Currently suffering from persistent abdominal pain and biochemical abnormalities ten months after the initial AP episode.

Imaging Findings:

Previous CT reports described severe AP with pleural effusions and 8 cm dominant peripancreatic post-necrotic fluid collection (PPNFC), which was initially treated with endoscopic ultrasound-guided cystogastrostomy with metallic stent (Fig. 1), achieving incomplete cyst resolution.

Transferred to our hospital, the patient underwent pancreatic necrosectomy and positioning of pigtail stent through the cystogastrostomy (Fig. 2a-b); after initial clinical improvement both stents were endoscopically removed (Fig. 2c). A month later, multidetector CT (Fig. 3) and MRI (Fig. 3a-g) showed the reappearance of sizeable, elongated PPNFC located above the pancreatic body-tail, which showed preserved anatomic continuity and enhancement without appreciable ductal dilatation. Additional MR-cholangiopancreatography (MRCP, Fig. 3h-i) showed mild Wirsung duct dilatation at the pancreatic head, plus segmental discontinuity of the main pancreatic duct (MPD) at the body.

Endoscopic retrograde cholangiopancreatography (Fig. 4a) confirmed MRCP findings and allowed definitive treatment by positioning of long stent through the disrupted MPD, ultimately resulting in relieved complaints and laboratory changes.

Discussion:

The disconnected pancreatic duct syndrome (DPDS) is an anatomic condition characterised by absent continuity between viable secreting pancreatic tissue and the gastrointestinal tract. This increasingly recognized condition occurs as a complication of acute necrotizing pancreatitis (ANP), occasionally following pancreatic traumatic injury with segmental necrosis [1-3].

Mostly seen in adults, DPDS generally presents late (2-9 months) after alcohol-related, gallstone or idiopathic ANP. The commonest presentation is persistent illness and non-resolving collection despite ineffectual percutaneous, endoscopic or surgical procedures. Meanwhile, patients remain at risk of delayed bleeding or infection. Alternatively, DPDS may occur in the setting of chronic pancreatitis (CP). Long-term complications include persistent symptoms, development of CP, pancreatic atrophy, diabetes (16-53% of patients), rarely left-sided portal hypertension. The mortality is non-negligible (up to 2%) [1-3].

Traditionally, the DPDS hallmark was endoscopic retrograde cholangiography (ERCP) demonstration of main pancreatic duct (MPD) discontinuity at the neck (58%), body-tail (23%) or mid-body (19%). Contrast extravasation at the site of ductal disruption at ERCP was reported in up to half of patients, particularly in the distal MPD [2-4]. Contrast-enhanced multidetector CT represents the mainstay technique to assess severity, complications and prognosis of acute pancreatitis [5, 6]. DPDS is present in up to 16% of patients with walled-off pancreatic necrosis (WOPN). Making the diagnosis of DPDS is important since—unlike other post-inflammatory peripancreatic fluid collections—conservative management is generally unsuccessful. Unfortunately, despite most patients receiving serial CT studies, DPDS is missed at primary CT interpretation in the majority of cases, thus contributing to diagnostic delay: therefore, the radiologists' awareness is advisable. DPDS is suggested by viable, enhanced pancreatic parenchyma after ANP plus a non-resolving collection of greatly variable size (4-74 cm2). As this case exemplifies, collections may or may not be located along the expected course of the MPD. Although false positives may result from partial duct disruption, additional investigation with MR-cholangiopancreatography (MRCP) may reveal ductal discontinuity which requires ERCP for diagnosis confirmation and treatment [2-4].

Managing DPDS requires either redirection of secretions into the digestive tract, or resection of the viable disconnected pancreatic segment. Repeated surgery is reported in 6% of patients. Surgery may perform internal drainage into a Roux-en-Y jejunal limb, or distal pancreatic resection. Endoscopic treatment includes ultrasound-guided transgastric drainage of WOPN with pigtail stents and ERCP-assisted stenting of the MPD, is increasingly recognized as the best therapeutic option and is successful in 75% of patients [1, 2, 7-11].

Differential Diagnosis List: Disconnected pancreatic duct syndrome following acute necrotizing pancreatitis, Pancreatic pseudocyst, Walled-off pancreatic necrosis, Peripancreatic abscess

Final Diagnosis: Disconnected pancreatic duct syndrome following acute necrotizing pancreatitis

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Description: Cysto-gastrostomy with metallic stent (thin arrows) and nasal-cystic tube (arrows) was previously positioned to drain the major peripancreatic collection (*). **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



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Description: Maximum-intensity projection (MIP) images showed the metallic stent (thin arrows) and nasal-cystic tube (arrows) previously positioned at the cysto-gastrostomy. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



Description: Pancreatic necrosectomy was performed through the cysto-gastrostomy with nasal-cystic tube (arrows). **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



Description: The nasal-cystic tube was then removed and replaced with a pigtail stent (arrow) positioned through the metallic stent already in place. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



Description: After clinical improvement, both stents were endoscopically removed. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



Description: Unenhanced (a) and post-contrast (b,c) images showed reappearance of the major postnecrotic fluid collection (*) measuring approximately 9x2.5 cm, located above the pancreatic body-tail (+). **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



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Description: The pancreatic body and tail (+) were recognizable with preserved anatomic continuity and enhancement. The main pancreatic duct was not perceptible. Note major post-necrotic fluid collection (*). **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



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Description: Axial T1-(a), T2-(b), diffusion-weighted (c) and apparent diffusion coefficient (ADC) map (d) showed stable major post-necrotic fluid collection (*) measuring approximately 9x2.5 cm, located above the pancreatic body-tail. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



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Description: Axial T1- (e) and T2-(f) weighted images confirmed pancreatic body and tail (+) with preserved anatomic continuity and thickness within normal range. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



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Description: MRCP showed mild dilatation of the Wirsung duct at the pancreatic head (arrow), and segmental discontinuity (thick arrow) of the main pancreatic duct in the body. Normal appearance of the biliary tract. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)

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Description: Repeated ERCP confirmed MRCP findings of mild dilatation of the Wirsung duct in the pancreatic head (arrow), and focal discontinuity (thick arrow) of the main pancreatic duct in the body. **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)



Description: Definitive endoscopic treatment required positioning of a long pancreatic stent (arrowhead) through the main pancreatic duct discontinuity. Note mild dilatation of the Wirsung duct in the pancreatic head (arrow), **Origin:** Tonolini M, Radiology Department, "Luigi Sacco" University Hospital – Milan (Italy)