

Recurrent acute pancreatitis due to a santorinicele in a young patient

Khan S A, Chawla T, Azami R

ABSTRACT

A cystic dilatation of the terminal portion of the minor pancreatic duct (duct of Santorini) is referred to as a santorinicele. It is usually associated with pancreas divisum and has been suggested to be a cause of relative stenosis of the minor papilla, often leading to recurrent pancreatitis. While this anomaly has been reported in the paediatric population, it is more commonly found in the elderly. We present a 27-year-old woman with recurrent acute pancreatitis attributed to a santorinicele with a dorsal duct-exclusive pancreatic drainage.

Keywords: cystic dilatation of dorsal pancreatic duct, minor pancreatic duct, pancreas divisum, recurrent acute pancreatitis, santorinicele

Singapore Med J 2009;50(5):e163-e165

INTRODUCTION

Santorinicele is a term used to describe a focal cystic dilatation of the dorsal pancreatic duct (duct of Santorini) in its terminal portion at the minor papilla.⁽¹⁾ While there is some controversy as to the origin of this condition, it is often considered to be a cause of the relative stenosis of the minor papilla, which, in the presence of a dorsal duct predominant drainage (as in pancreas divisum), likely contributes to episodes of recurrent pancreatitis associated with this condition.⁽¹⁻⁴⁾ Most cases reported in the literature are associated with pancreas divisum, and there are only two reports of santorinicele without the latter anomaly.^(2,5) In both of these cases, however, the ventral duct, in addition to its connection with a complete anomalous dorsal duct, was found to be draining normally at the major papilla. In this report, a young patient with recurrent acute pancreatitis associated with santorinicele in the presence of a “dorsal-duct-exclusive” drainage is described.

CASE REPORT

A 27-year-old woman presented with a history of epigastric pain and vomiting for one day. She was a known case of recurrent (non-gallstone-induced) pancreatitis. She was first admitted to our hospital at

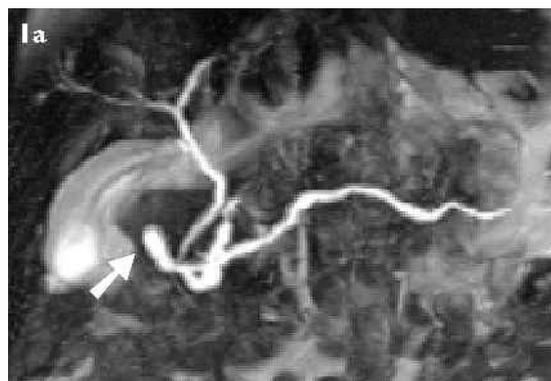


Fig. 1a Coronal MRCP image shows the santorinicele towards the extreme left. Also note the dilated ventral and dorsal pancreatic duct, and the exclusive drainage of the pancreas through the dorsal duct into the santorinicele at the minor papilla. The arrow indicates the distal common bile duct that circumvents the entire pancreatic system to drain at its usual location at the major duodenal papilla.

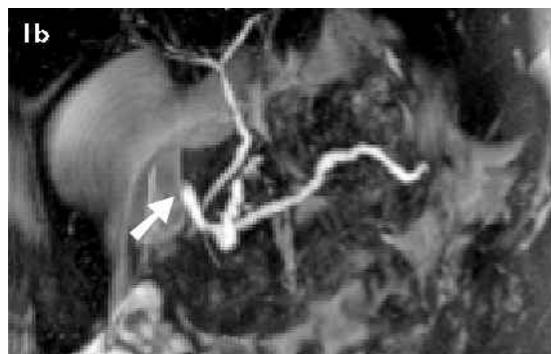


Fig. 1b Coronal MRCP image. The distal extension of the common bile duct beyond the pancreatic duct drainage (white arrow) is well visualised.

the age of 16 years, with severe acute pancreatitis with a cumulative Ranson's score of 3. Upper abdominal ultrasonography done on that admission showed no evidence of cholelithiasis. Other investigations, including a fasting lipid profile and liver function tests, were also within normal limits. She was conservatively managed (including a stay in the high-dependency unit for four days) and then discharged. Over the next five years, she was admitted twice to the hospital for recurrent episodes of mild acute pancreatitis. During each of these admissions, she was investigated with ultrasonography, but no evidence of gallstones was found. At the time of her third admission, she was scheduled for elective

Department of
Surgery,
Aga Khan University,
Stadium Road,
Karachi,
Pakistan

Khan SA, MBBS
Resident Surgeon

Chawla T, MBBS,
FRCS
Assistant Professor
and Consultant

Azami R, MBBS,
FCPS
Associate Professor
and Consultant

Correspondence to:
Dr Tabish Chawla
Tel: (92) 21 486 4751
Fax: (92) 21 493 4294
Email: tabish.chawla
@aku.edu

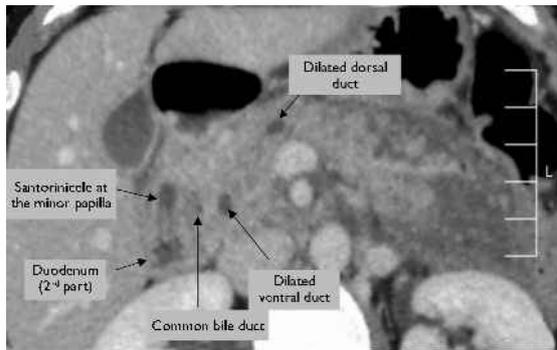


Fig. 2 Axial contrast-enhanced CT image taken through the pancreas shows the santorinicele at the minor duodenal papilla as well as the dilated ventral and dorsal pancreatic ducts in relation to the common bile duct. Also note extensive inflammation and oedema in the region of the tail and body of the pancreas.

endoscopic retrograde cholangiopancreatography (ERCP). The ERCP demonstrated a normal common bile duct, but repeated attempts to cannulate (and demonstrate) the pancreatic ducts were unsuccessful. She tolerated the procedure well without any complications.

For the next six years, the patient remained symptom-free until the most recent episode brought her to the hospital. At presentation, she had tachycardia (106/min) and was otherwise stable. The rest of her physical examination was unremarkable, except for moderate tenderness in the epigastrium. Significant among her laboratory investigations was a raised amylase (3,933 IU/L), lipase (3,070 IU/L), lactate dehydrogenase (601 IU/L) and leucocyte count of 21,000 (94% neutrophils). Ultrasonography on this occasion revealed an oedematous pancreas, and normal common bile duct and gallbladder, with no evidence of gallstones or sludge. She was admitted to the general ward with a Ranson's score of 2 (same as her cumulative Ranson's score after 48 hours), and managed conservatively with an initial 36-hour period of intravenous (IV) fluids and absolute restriction of oral fluids/diet, followed by the gradual introduction of an oral diet. No antibiotics were administered.

While in the hospital, she was investigated with magnetic resonance cholangiopancreatography (MRCP) using 200 ml of pineapple juice as T2-negative contrast (without secretin stimulation). MRCP showed dilated dorsal and ventral duct (approximately 5.5 mm) with the dorsal duct exclusive drainage at the minor papilla (Fig. 1). In addition, a cystic dilatation (santorinicele) of the dorsal duct was demonstrated. The common bile duct was found to be of normal calibre and draining to the site of the major papilla, isolated from any of the pancreatic ducts. The ventral duct was seen draining exclusively into the terminal dorsal pancreatic duct. Inflammation

of the pancreatic parenchyma was demonstrated on T1-weighted images. The patient subsequently underwent multidetector computed tomography of the pancreas, enhanced with 150 ml of IV contrast (iopromide) and oral administration of 1,200 ml of water (pancreatic protocol). Portal venous phase images were obtained 70 minutes after the injection of IV contrast via a mechanical injector. Computed tomography (Fig. 2) confirmed the findings, demonstrating the santorinicele and the resultant dilatation of both pancreatic ducts with evidence of inflammation of the pancreatic parenchyma (more marked in the tail and body regions).

The patient improved on conservative management. The various available options for further management were considered and discussed with the patient. As the episodes were infrequent and her symptoms did not significantly affect her lifestyle, an approach of watchful surveillance was adopted and any possible intervention(s) deferred, pending future evolution of her condition.

DISCUSSION

Santorinicele was first described as a focal cystic dilatation of the terminal pancreatic duct in association with pancreas divisum and recurrent pancreatitis in 1994.⁽¹⁾ Several more cases have been reported ever since.⁽¹⁻⁶⁾ The issue of aetiopathogenesis of this anomaly is a matter of some controversy. The relatively more common occurrence in the elderly⁽¹⁻⁵⁾ and its association with duodenal diverticula^(1,3-5) point towards an acquired aetiology. Reports of its existence in the paediatric population, however, indicate that in at least some instances, a santorinicele may be congenital.⁽⁶⁾ It is postulated to contribute to relative stenosis at the minor papilla, leading to inadequate drainage of the dorsal pancreatic duct which results in high intraductal pressure, pain and recurrent episodes of pancreatitis, a sequence known as the "dominant dorsal duct syndrome".⁽⁷⁾ There is, however, still a debate about whether a santorinicele is an incidental finding or a cause of minor papilla stenosis and subsequent pancreatitis.⁽⁵⁾

In uncontrolled trials and case series, dominant dorsal duct syndrome responds variably to dilatation of the minor papilla with endoscopic sphincterotomy (and/or stenting), with the greatest benefit demonstrated in patients with acute relapsing pancreatitis, and the presence of santorinicele predicting a favourable outcome.^(1,7,8) Restenosis of the minor papilla is relatively common with extended follow-up after endoscopic sphincterotomy,⁽⁹⁾ but this may respond to repeat intervention or open surgical sphincteroplasty.⁽⁷⁾

To the best of our knowledge, this is the third case of santorinicele without pancreas divisum reported in the English literature,^(2,5) and the only one with a “dorsal-duct-exclusive” pancreatic drainage.

REFERENCES

1. Eisen G, Schutz S, Metzler D, Baillie J, Cotton PB. Santorinicele: new evidence for obstruction in pancreas divisum. *Gastrointest Endosc* 1994; 40:73-6.
2. Nam KD, Joo KR, Jang JY, et al. A case of santorinicele without pancreas divisum: diagnosis with multi-detector row computed tomography. *J Korean Med Sci* 2006; 21:358-60.
3. Costamagna G, Ingrosso M, Tringali A, Mutignani M, Manfredi R. Santorinicele and recurrent acute pancreatitis in pancreas divisum: diagnosis with dynamic secretin-stimulated magnetic resonance pancreatography and endoscopic treatment. *Gastrointest Endosc* 2000; 52:262-7.
4. Seibert DG, Matulis SR. Santorinicele as a cause of chronic pancreatic pain. *Am J Gastroenterol* 1995; 90:121-3.
5. Byeon JS, Kim MH, Lee SK, et al. Santorinicele without pancreas divisum. *Gastrointest Endosc* 2003; 58:800-3.
6. Tang H, Kay CL, Devonshire DA, Tagge E, Cotton PB. Recurrent pancreatitis in a child with pancreas divisum. Endoscopic therapy of a Santorinicele. *Surg Endosc* 1999; 13:1040-3.
7. Madura JA 2nd, Madura JA. Diagnosis and management of sphincter of Oddi dysfunction and pancreas divisum. *Surg Clin North Am* 2007; 87:1417-29, ix.
8. Delhaye M, Matos C, Devière J. Endoscopic technique for the management of pancreatitis and its complications. *Best Pract Res Clin Gastroenterol* 2004; 18:155-81.
9. Kamisawa T. Clinical significance of the minor duodenal papilla and accessory pancreatic duct. *J Gastroenterol* 2004; 39:605-15.