

◆ CASO CLÍNICO

Different clinical aspects of Wirsungocele: case series of three patients and review of literature

Djalma Ernesto Coelho,¹ José Celso Ardengh,² Eder Rios de Lima-Filho,³ José Flávio Coelho⁴

¹ Department of Surgery, Universidade Estácio de Sá - UNESA, Rio de Janeiro, Brasil.

² Department of Anatomy and Surgery, Faculdade de Medicina de Ribeirão Preto - Universidade de São Paulo- USP.

³ Department of Surgery, Hospital dos Servidores do Estado - HSE, Rio de Janeiro, Brasil.

⁴ Department of Surgery, Universidade Federal do Rio de Janeiro - UFRJ, Brasil.

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Summary

The association of Santorinicele with pancreas divisum has been described. This anatomic condition creates ideal conditions for acute pancreatitis episodes and chronic abdominal pain. Saccular dilation of main pancreatic duct has also been described as incidental finding and causing episodes of acute pancreatitis. However, there is no description of associated chronic abdominal pain. Three detailed cases of Wirsungocele demonstrated by endoscopic retrograde cholangiopancreatography are presented. Two of them had episodes of acute pancreatitis and one had chronic abdominal pain. All patients were treated by endoscopic biliopancreatic sphincterotomy. After a follow-up for more than two years, none presents clinical recurrence. Endoscopic biliopancreatic sphincterotomy for symptomatic patients with this anatomic condition seems safe and effective.

Key words. Endoscopic retrograde cholangiopancreatography, pancreatic ducts, pancreatitis, endoscopic sphincterotomy.

Diferentes aspectos clínicos del Wirsungocele: serie de tres pacientes y revisión de la literatura

Resumen

Se ha descrito la asociación de Santorinicele con páncreas divisum. Esta característica anatómica crea con-

diciones ideales para los episodios de pancreatitis aguda y dolor abdominal crónico. La dilatación sacular del conducto pancreático principal también ha sido descrita como hallazgo incidental y como causa de episodios de pancreatitis aguda. Sin embargo, no hay una descripción del dolor abdominal crónico asociado. Se presentan tres casos detallados de Wirsungocele demostrados por colangiopancreatografía retrógrada endoscópica. Dos pacientes tuvieron episodios de pancreatitis aguda y uno de dolor abdominal crónico. Fueron tratados con esfinterotomía endoscópica biliopancreática. Después de más de dos años de seguimiento ningún paciente presenta recurrencia clínica. La esfinterotomía endoscópica biliopancreática parece segura y efectiva para los pacientes sintomáticos con esta condición anatómica.

Palabras claves. Colangiopancreatografía retrógrada endoscópica, conductos pancreáticos, pancreatitis, esfinterotomía endoscópica.

Intramural cystic dilation (intraduodenal) of distal common bile duct is known as choledocele, and cystic dilation of intramural dorsal main pancreatic duct (MPD) is known as Santorinicele.¹⁻⁵ All these anatomical changes are known and have been previously described.¹⁻⁴ The later, when associated with pancreas divisum, has an increased risk for acute pancreatitis.⁶ Nevertheless, because it is rare, epidemiology and evolution are not defined. Cystic dilation of terminal ventral MPD (Wirsung's duct) is known as Wirsungocele. This anatomical abnormality was first described in 2004⁷ as an incidental finding. Gupta et al showed a Wirsungocele in a patient with recurrent episodes of acute pancreatitis.⁸

Correspondence: José Celso Ardengh
Alameda dos Arapanés, 881 - cj 111
CEP 04524-001 - Moema - São Paulo - SP
Phone: (11) 50557134
E-mail: jcelso@uol.com.br

To date, bouts of chronic abdominal pain were only related to Santorinicele. We found no reports of abdominal pain caused by Wirsungocele, such as those described by Seibert et al.⁴

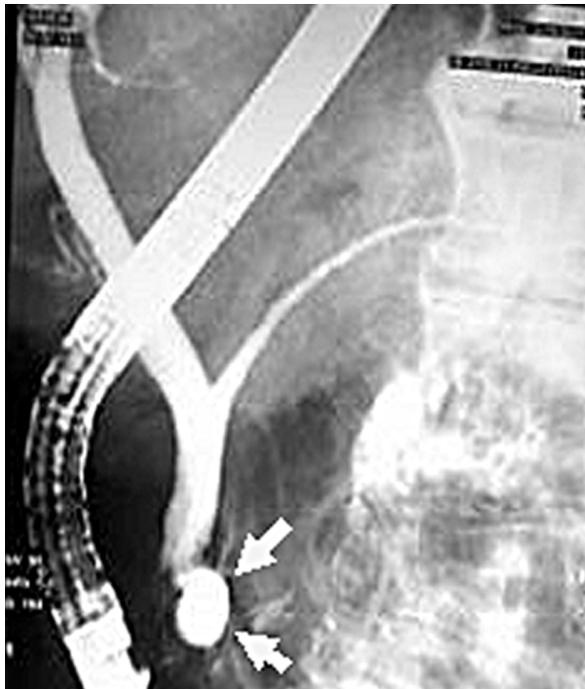
We present three cases of Wirsungocele demonstrated by endoscopic cholangiopancreatography (ERCP) and treated by pancreatic sphincteroplasty and biliary sphincterotomy. Two had recurrent episodes of acute pancreatitis and one had chronic abdominal pain. All had a good outcome and after a prolonged follow-up had not the symptoms presented before endoscopic treatment. Besides, we make a review of literature, comparing it with our patients.

Case reports

Case 1

A 80-year-old man presented jaundice, abdominal pain, nausea and vomiting presented several bouts of acute pancreatitis with no apparent cause. Blood tests showed total bilirubin 11.0 mg/dL (conjugated 7 mg/dL), gamma glutamyltransferase 327 IU/L, amylase 5,518 IU/L, and white blood count 17.000 per microliter. Transabdominal ultrasonography (US) revealed mild dilation of intrahepatic bile duct, thickened wall of gallbladder and

Figure 1. ERCP shows main pancreatic duct, Wirsungocele in distal portion (arrows) and opacification of a dilated common bile duct.



microlithiasis. A computerized tomography (CT) confirmed US findings. ERCP was performed based on clinical, laboratory and radiological examinations. Duodenoscopy showed normal Vater's ampulla. Selective cannulation showed a dilated main bile duct and normal caliber of intrahepatic bile duct. Selective catheterization of MPD showed a 2.0 cm cystic dilatation of its terminal portion (Figure 1). MPD above cystic dilatation showed mild dilation. Biliary sphincterotomy and pancreatic sphincteroplasty were performed. Patient is asymptomatic 2 years after treatment with no further episodes of acute pancreatitis.

Case 2

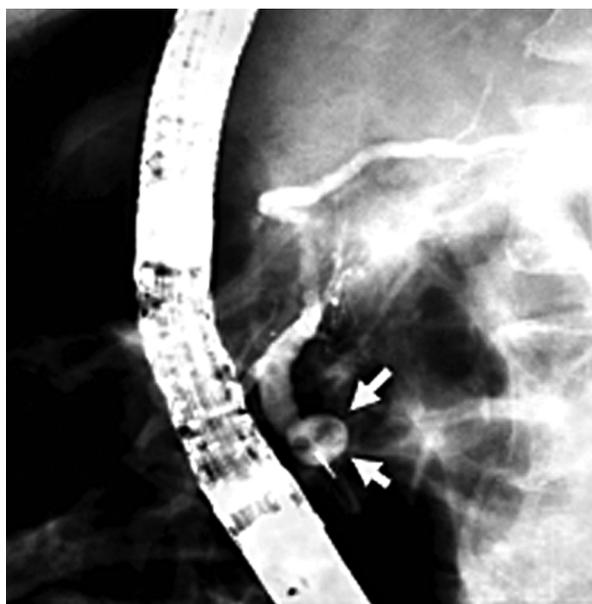
A 78-year-old man presented chronic abdominal pain and one episode of upper gastrointestinal bleeding due to gastritis caused by NSAIDs. He denied choloria, jaundice, fecal acholia, cholangitis, and alcohol intake. Clinical investigation was performed during hospital stay. US revealed a normal gallbladder, with small calculi and dilated intrahepatic bile duct. CT showed mild dilatation of intrahepatic bile duct and common bile duct (1.0 cm). CT confirmed US findings, but did not define the source of biliary obstruction. Biochemical tests were normal except for gamma glutamyltransferase (885 IU/L) and alkaline phosphatase (1,219 IU/L). ERCP was performed based on clinical, laboratory and radiological

Figure 2. Duodenoscopy shows bulging of Vater's ampulla (cholechocele?).



gical examinations. Duodenoscopy showed a prominent Vater's ampulla suggesting choledochocoele (Figure 2). Selective catheterization showed mild dilation of common bile duct and suspected choledochocoele could not be assured. Intrahepatic bile duct had normal caliber. Selective catheterization of MPD showed a 2.5 cm cystic dilation of terminal MPD. MPD above cystic dilatation was slightly dilated with irregular wall, as described by Kasugai et al (Figure 3).⁹ Biliary and pancreatic sphincterotomy were performed. Patient is asymptomatic after more than 48 months.

Figure 3. ERCP shows partial opacification of dilated terminal main pancreatic duct (Wirsungocele).

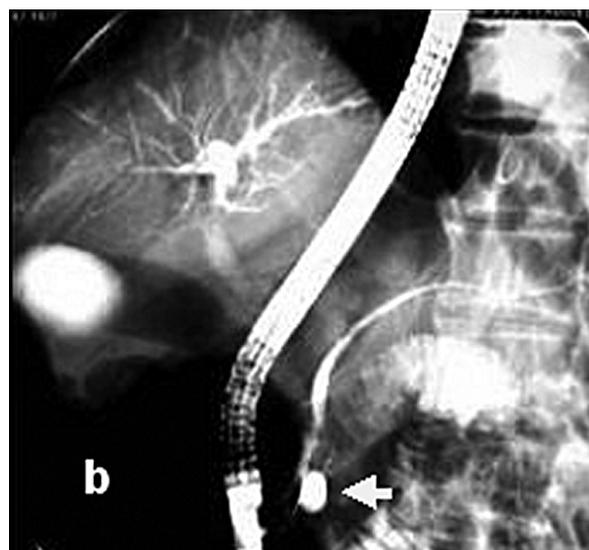
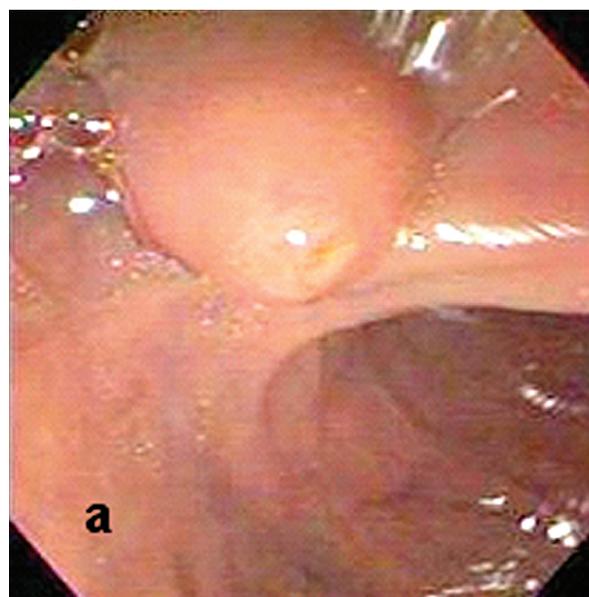


Case 3

A 76-year-old woman presented acute pancreatitis associated with jaundice, abdominal pain and fever. Amylase (three times the normal value) and lipase (five times the normal value) were high and associated with leukocytosis (18,000 per mcL). US showed normal bile duct and gallbladder containing no gallstones. CT confirmed US findings and added no important data for the management. The duodenoscopy during ERCP showed ampullary bulge suggesting choledochocoele (Figure 4a). Selective catheterization showed main and intrahepatic bile duct of normal appearance. MPD catheterization showed a 2.0cm terminal cystic dilatation. MPD above cystic dilatation was slightly dilated with a small stenosis in the boundary of head / body and poor opacification of branch duct (Figure

3b). Biliary and pancreatic sphincterotomy were performed. Patient is asymptomatic after 3 years of follow-up and had no episodes of acute pancreatitis.

Figure 4. a) Duodenoscopy shows suggestive appearance of choledochocoele b) ERCP: opacification of dilated terminal main pancreatic duct (Wirsungocele, arrow) and opacification of common bile duct.



Discussion

Santorinicele, a cystic dilation of Santorin's duct into papilla minor, has been well described in patients with recurrent attacks of acute pancreatitis associated to pancreas divisum.^{3-4,10} It has been postulated that stenosis of papilla minor in patients

with pancreas divisum could lead to high intraductal pressure.⁸ This high pressure (congenital or acquired), along with weakness of ductal wall, leads to Santorinicele which, in turn, predisposes to papilla minor obstruction causing recurrent attacks of pancreatitis.^{6,11} Moreover, episodes of recurrent acute pancreatitis related to Wirsungocele have been recently reported.⁸

Wirsungocele was reported as an incidental finding by Baron et al.⁷ In this series, endoscopic US (EUS) showed no dilation, while magnetic resonance colangiopancreatography (MRCP) and ERCP clearly demonstrated a Wirsungocele accompanied by episodes of acute pancreatitis.⁷ In our case series, we had no available EUS and diagnosis was performed by ERCP. Several theories have been proposed to explain etiology and pathophysiology of terminal duct cystic dilatation. It has been postulated that a decreasing in autonomic innervation of Oddi's sphincter leads to uncoordinated sphincter and functional obstruction of papillary orifice.¹² A defect in common channel with anomalous pancreatobiliary junction, greater than 5 mm, has also been proposed as an important factor for dilated pancreatic and choledochal ducts.¹³

Another study shows a focal MPD dilatation in the head of pancreas with increasing age.¹⁴ None of our patients had juxtapapillary diverticula or any abnormality at the junction to explain predisposition for MPD saccular dilation. In our opinion the age and weakness of ductal wall relative to Oddi's sphincter low pressures could explain this phenomenon. However, functional obstruction of papillary orifice can not be ruled out. In our opinion, pathophysiological mechanism for Wirsungocele is unclear. Another important factor so far from being explained is whether association of recurrent acute pancreatitis and Wirsungocele is causal or accidental. Likewise, several authors believe that pancreatic sphincterotomy has not background.⁸ However, in our series, after pancreatic and biliary sphincterotomy, patients were free of the symptoms that led to perform ERCP

In conclusion, our case series show definite Wirsungocele in two patients with recurrent acute pancreatitis and in one with chronic abdominal pain who underwent endoscopic treatment. This association between chronic abdominal pain and Wirsungocele has been never published.

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