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Case Report

Sphincter of Oddi incompetence associated with duodenal stenosis in a newborn: A case report ☆

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ABSTRACT

Newborn incompetence of the sphincter of Oddi is rare. While there are many causes of reflux of air or ingested contrast material into the biliary tree in adults, in the newborn, it is usually due to incompetence of the sphincter of Oddi associated with partial or complete duodenal obstruction. This paper presents upper gastrointestinal series findings of incompetence of the sphincter of Oddi associated with duodenal stenosis in a 3-day-old newborn. If pneumobilia is identified in the newborn, although the possibility is low, clinicians should consider incompetence of the sphincter of Oddi with duodenal obstruction as well as portal vein gas.

Introduction

Congenital duodenal stenosis causes upper gastrointestinal tract obstruction in neonates. Duodenal stenosis is less common than atresia; however, the etiology is the same [1]. Both duodenal atresia and stenosis are thought to be due to a failure of recanalization following epithelialization, proliferation and subsequent vacuolization of the duodenum [2]. Although approximately 30% of these patients have Down syndrome, it may also be associated with other congenital anomalies such as: other intestinal atresias, biliary tract anomalies, congenital heart disease, renal anomalies, and vertebral anomalies [3,4]. Duodenal stenosis commonly occurs near the ampulla of Vater; hence, it is frequently accompanied by abnormalities of the bile duct and pancreas. While upper gastrointestinal series is performed in infant with duodenal stenosis, we experienced a reflux of ingested contrast material into the biliary tree. There are many published reports on duodenal stenosis [5,6]; nevertheless, herein we present an additional case of sphincter of Oddi incompetence associated with duodenal stenosis in a newborn and discuss the possible causes.

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Case report

A 36-year-old female delivered a 3500 g male newborn by normal vaginal delivery with a cephalic presentation, at 39 weeks of gestation. Three days after birth, the infant was admitted to the hospital due to vomiting after every feed. Physical examination on admission revealed mild dehydration. The patient had a small chin, slanted eyes, a protruding tongue, and a single crease on his palm. Laboratory studies were unremarkable. Transthoracic echocardiography revealed a ventricular septal defect. Plain abdominal radiography showed a mildly dilated stomach and proximal duodenum (Fig. 1). Upper gastrointestinal series was performed and it showed partial obstruction of the second part of duodenum. The stomach and duodenal bulb were distended and stasis of contrast material at the stenotic duodenum was noted. After a while, at a point distal to the stenotic duodenum, reflux of the ingested contrast material into the common bile and pancreatic duct was noted (Fig. 2A and B). The common bile and pancreatic duct were opacified without resistance by the contrast material and the distal part of the duodenum, including parts connected to the common bile and pancreatic duct, were still narrowed. Subsequently, the gallbladder and intrahepatic bile duct were opacified by contrast material and pneumobilia was noted (Fig. 2C and D). The third and fourth parts of the duodenum were unremarkable. Subsequently, duodenoplasty was performed the next day. During surgery, dilatation of the stomach and proximal duodenum was noted and there was stenosis of the second part of the duodenum without annular pancreatic or peritoneal bands. We were unable to check sphincteric tone; however, the ampulla of Vater was not found during surgery. Subsequent chromosomal analysis showed a karyotype of 47, XY +21. Postoperatively, the infant had no problems during 2-months of follow-up, after which he did not revisit the hospital.

Discussion

Reflux of air or ingested contrast material into the biliary tree of newborns is rare [5,6]. It is usually due to sphincter of Oddi incompetence associated with partial or complete duodenal obstruction [5–7]. In adults, sphincter of Oddi incompetence may be due to the repeated passage of stones through the sphincter or distal duodenal obstruction with the resulting elevated intraluminal pressure [7]. In newborns, elevated duodenal intraluminal pressure may be the cause of sphincter of Oddi incompetence; however, structural abnormalities near the sphincter of Oddi should also be considered [5,6]. Atkinson et al. [5] reported a case of sphincter of Oddi incompetence with a duodenal web; they failed to find sphincteric tone at the orifice of the common bile duct and a distinct ampulla of Vater during surgery suggesting that the incompetence was due to anomalous development. One of the limitations of our study was our inability to check sphincteric tone; however, a structural abnormality near the sphincter of Oddi may be considered the cause of incompetence rather than elevated intraluminal pressure in the duodenum as the reflux of ingested contrast material into the biliary tree occurred in the distal part of the stenosis and a distinct ampulla of Vater was not found. In previously reported upper gastrointestinal series of sphincter of Oddi incompetence with duodenal obstruction [5,6],

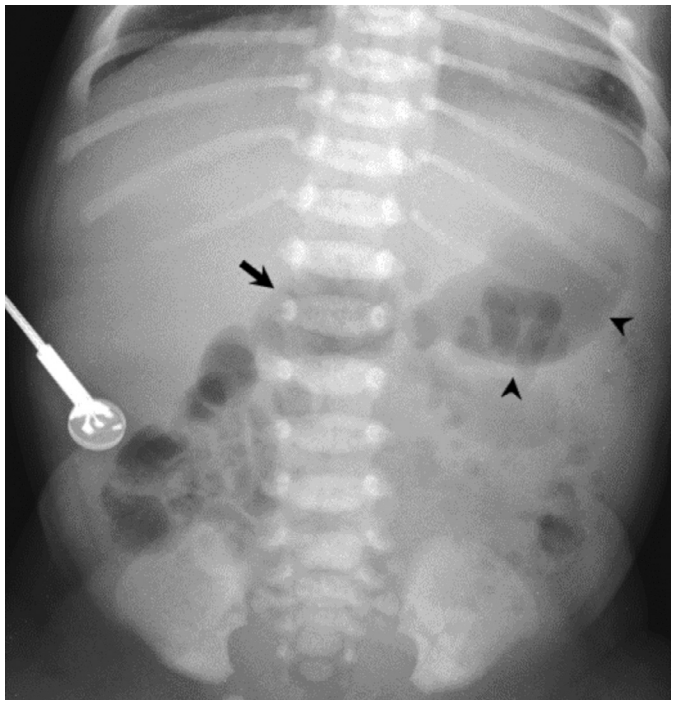

Fig. 1 – Sphincter of Oddi incompetence associated with duodenal stenosis in a 3-day-old boy who presented with vomiting. Plain abdominal radiography shows a mildly dilated stomach (arrow heads) and proximal duodenum (arrow).

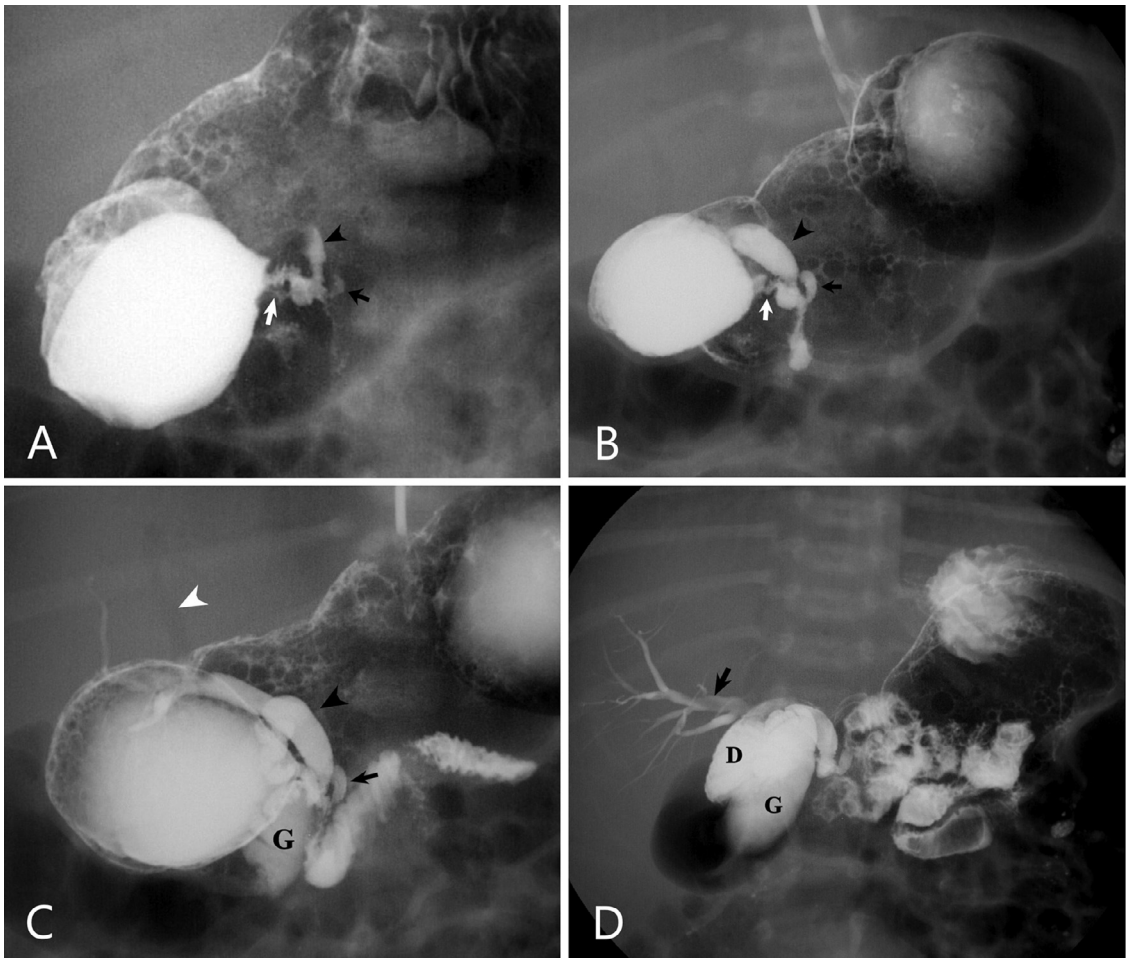

Fig. 2(A–C) – Spot images of the upper GI study shows distension of stomachm and duodenal bulb and partial obstruction (white arrow) of the second part of duodenum. At a point distal to the stenotic duodenum, reflux of ingested contrast material into common bile (arrow head) and pancreatic ducts (black arrow) is noted. The lumen of the duodenum, including parts connected to common bile and pancreatic ducts, is narrowed. Subsequently, the gallbladder (G) and intrahepatic bile duct are opacified by contrast material. Gas (white arrow head) in the intrahepatic bile duct is also noted. D. 15-minute delayed image of the upper GI study showing a distended stomach and duodenal bulb (D) with contrast material in the intrahepatic bile duct (arrow) and gallbladder (G).

since the biliary tree and the dilated stomach and duodenum were filled with contrast material, the relationship among the stenotic duodenum, common bile duct, and pancreatic duct was not apparent. In our case, the early spot images helped reveal the relationship between the duodenum and biliary tree.

There are some reports that biliary tree anomalies are related to congenital duodenal obstruction [8–10]. Most of them are related to an alteration of the normal branching pattern of the hepatopancreatic duct but there are also some regarding sphincter of Oddi incompetence [5,6]. Duodenal stenosis, like duodenal atresia, is thought to result from failure of recanalization of the duodenum between about the ninth and 11th weeks of gestation. During fetal development there is a temporary interruption of the duodenal lumen that occurs as the result of epithelial proliferation. Vacuoles form within the interrupted lumen and coalesce. Initially the duodenal lumen is recanalized into 2 parallel canals and the hepatopancreatic duct and accessory pancreatic duct end into each of the 2 lumens. Early on during development, 4 duct openings enter a short segment of the duodenum. Subsequently 1 terminal branch of the hepatopancreatic duct and 1 terminal branch of the accessory pancreatic duct regress. The region in which these complex events occur measures $\frac{1}{8}$ mm [2,8]. Boyden et al. [2] suggested that delayed coalescence of the vacuoles to form the 2 orifices of the hepatopancreatic and accessory pancreatic ducts has precedence over the vacuolization of adjacent duodenal portions creating a “traffic jam,” thereby suggesting the hepatopancreatic and accessory pancreatic ducts entry into the duodenum inhibits the recanalization of the main cavity. An epithelial barrier developing at the level of the ducts fails to be recanalized leaving a blind cavity above and below it. Ultimately this epithelial septum degenerates and is replaced by mesenchyme which then differentiates into connective tissue or muscle.

Sphincter of Oddi incompetence can cause confusion in making a proper preoperative diagnosis. On plain abdominal

radiography of newborns, linear radiolucent streaks overlying the liver shadow are usually gas within the portal vein which is usually seen in peripheral radicles and gas within the biliary tree is usually seen in central ducts. However, it is difficult to distinguish between gas in the portal vein and gas in the biliary system on plain radiography. If gas in the extrahepatic bile ducts or gallbladder is identified, gas in the portal vein can be excluded. Some authors [2,8–10] have reported cases of the paradoxical presence of small bowel gas in duodenal atresia. In these cases, the terminal portion of the hepatopancreatic duct divided into 2 branches before entering the duodenum. The proximal branch entered the duodenum above the atresia and the distal one below the atresia. Air or ingested contrast material entered the proximal branch and passed into the distal branch to bypass the atretic segment so that duodenal atresia was diagnosed as incomplete duodenal obstruction.

Sphincter of Oddi incompetence in the newborn is usually associated with complete or incomplete duodenal obstruction. It may be simply due to elevated intraluminal pressure in the duodenum or due to a structural abnormality in the region of the sphincter occurring during fetal development. In the neonatal period, sphincter of Oddi incompetence may be the main cause of reflux of air or ingested contrast material into the biliary tree. If pneumobilia is identified in the newborn, although the possibility is low, clinicians should consider sphincter of Oddi incompetence with duodenal obstruction as well as portal vein gas.

Patient consent

Consent for publication has been obtained.

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