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Pyloric atresia associated with Dieulafoy lesion and gastric dysmotility in a neonate

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Abstract In this case study, we report a male infant with pyloric atresia, extreme gastric distension, and a caliber-persistent gastric artery (Dieulafoy lesion) with massive gastric bleeding. After a transverse pyloroplasty and endoscopic hemoclip application to the caliber-persistent gastric artery, very slow gastric emptying developed, which required repeated surgical interventions. Gastroduodenostomy failed to promote gastric emptying. The intraoperative and postmortem histologic examinations of the gastric wall revealed a loss of interstitial cells of Cajal, which possibly explains the extreme motility disorder.

Dieulafoy lesion refers to a protuberant submucosal ectatic artery in the gastrointestinal tract larger than the vessels usually in that area [1]. It occurs most frequently in the stomach. The protuberant artery causes brisk bleeding with little or no surrounding ulceration. The etiology is unknown. Dieulafoy lesion (also known as *caliber-persistent artery* or *cirsoid aneurysm*) in the stomach can be a source of massive, potentially life-threatening, and often recurrent upper gastrointestinal bleeding [1]. In most of the reported cases, the median age of the patients is in the mid 50s; the youngest patients described to date were 2 days and 8 weeks old, respectively [2,3]. Manifestation of this malformation immediately after birth has not been previously reported in the English-language literature.

We describe a male neonate with severe gastric bleeding caused by Dieulafoy vascular malformation of the stomach, manifested in the delivery room after birth in association with pyloric atresia and extreme gastric enlargement. This unusual combination resulted in delayed gastric emptying even after surgical and endoscopic treatment.

1. Case report

A small-for-date male infant weighing 1990 g was born at 36 weeks of gestation by vaginal delivery as a twin A; the Apgar scores were 8 at 1 minute and 9 at 5 minutes. Although the prenatal ultrasound study at 32 gestational weeks was unremarkable, a repeat sonogram at 35 weeks of gestation demonstrated significant gastric enlargement. At birth, during insertion of the postnatal feeding tube in the delivery room, 40 mL of fresh blood was aspirated from the

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stomach, and the first postnatal ultrasound examination, performed within 3 hours after birth, demonstrated a large stomach filled with blood clots. Massive hematemesis commenced at approximately 10 hours postnatally and did not stop after the transfusion of fresh-frozen plasma and phytomenadione (Konakion MM paediatric, Roche, Switzerland) administration. A plain abdominal radiograph at 11 hours was indicative of pyloric atresia, with no gas observed in the intestines (Fig. 1A). A laparotomy was performed at 13 hours postnatally. At operation, a membranous diaphragm was found in the pylorus; fresh bleeding was not seen. The web was excised, a huge gastric hematoma was evacuated, and a transverse pyloroplasty was performed with insertion of a transpyloric catheter. However, the cause of bleeding could not be identified intraoperatively. On the fourth postoperative day, massive bleeding recurred. Endoscopy revealed a caliber-persistent artery (Dieulafoy lesion) on the greater gastric curvature (Fig. 1B), and a hemoclip was applied (Figs. 1C and D). Enteral alimentation was initiated through the transpyloric catheter, and the infant passed a stool. After removal of the feeding tube on day 9 postoperatively, the gastric emptying became extremely slow. During intraoperative revision of the pyloric region on postoperative day 12, an intraluminal obstruction caused by severe postoperative edema was noted, and a gastroduodenostomy was performed, with placement of a transanastomotic catheter. Histologic examination of the specimen from the stomach at the anastomotic site showed normal smooth muscle and neural structures (Fig. 2). Immunohistochemistry against C-kit and peripherin revealed a normal distribution pattern of interstitial cells of Cajal (ICCs) and neuronal networks (Fig. 2). Enteral feeding via the transanastomotic catheter was successful, and 2 weeks after the second operation, the catheter was removed. However, intolerance of the oral feeding developed, the gastric residuals increased, and vomiting occurred. Repeated examination with a plain abdominal radiograph revealed a massively distended stomach without emptying (Fig. 1D). The infant underwent endoscopy, which demonstrated stenosis at the gastroduodenal anastomosis, and a repeat laparotomy was carried out 3 weeks after the second operation. Intraoperatively, only mild intestinal adhesions were found, but the external appearance and size of the anastomosis appeared adequate, and the stenosis was readily dilated through a gastric incision. A jejunostomy feeding tube was inserted to permit enteral feeding and avoid possible hepatic complications of long-term parenteral nutrition. Further surgical intervention was considered to deal with the abnormal gastric emptying, but the infant died of respiratory complications after an methicillin-resistant Staphylococcus aureus (MRSA) infection 51 days after birth. C-kit and peripherin immunohistochemistry was carried out on necropsy stomach specimens. Histologic examination revealed a reduced number of ICCs at necropsy as compared with the biopsy specimen taken during the second surgical exploration. The number of peripherin-immunostained cells was also moderately decreased (Fig. 2).

2. Discussion

The most frequent cause of delayed emptying in children is gastroesophageal reflux, but anatomical variations (eg, pyloric atresia or a gastric smooth muscle defect) and a motility disorder of the antrum likewise can result in prolonged emptying [4]. Postoperative stomach conditions

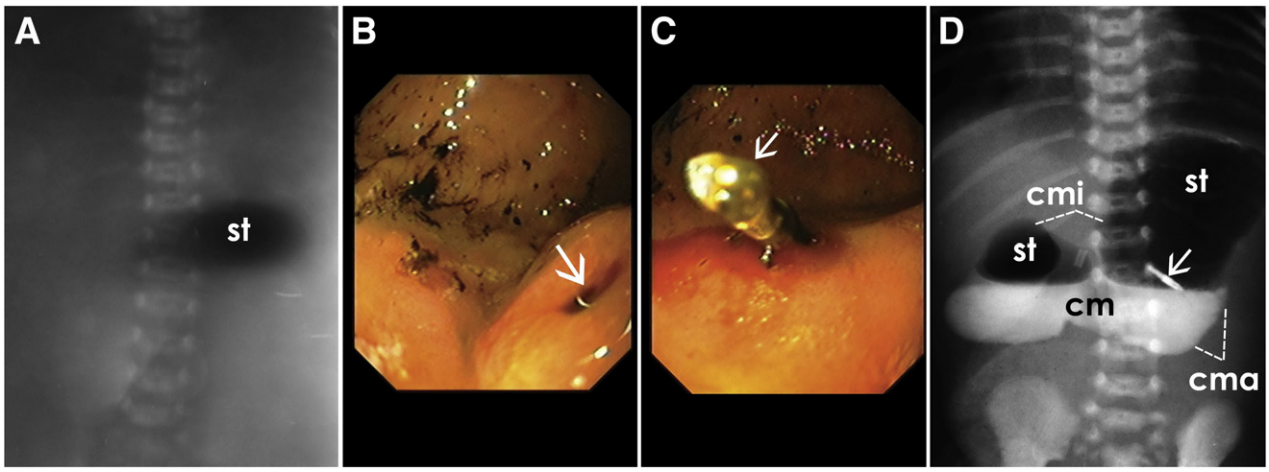

Fig. 1 Radiographic (x-ray) and endoscopic appearance of a caliber-persistent artery on the greater curvature in the infant. A, The abdominal x-ray study of the pyloric atresia. Gas-filled stomach (st) and lack of gas in the intestines. B, Endoscopic image of a caliber-persistent artery (arrow) in the cardiac stomach wall taken on the fourth postnatal day. C, Endoscopic image on the closed artery. Arrow indicates the hemoclip. cmi indicates minor curvature; cma, major curvature. D, Upper gastrointestinal contrast study taken on the 28th postnatal day showing the accumulation of contrast material (cm) in the stomach (st). Arrow indicates a hemoclip, which has been introduced to stop bleeding from the caliber-persistant artery shown on panel B.