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Acute pancreatitis associated with periampullary hamartomatous polyp in Peutz–Jeghers syndrome

Edson Kenzo Mizushima[1], Augusto Ricken Siqueira[1], Luis Carlos Anflor Jr[1,2], Luiz Felipe Peres Giesta[1,3]

[1]Department of Radiology. Hospital de Clínicas de Porto Alegre. Porto Alegre, Rio Grande do Sul, Brazil. [2]Universidade Federal de Ciências da Saúde de Porto Alegre (UFCSPA). Porto Alegre, Rio Grande do Sul, Brazil. [3]Grupo Hospitalar Conceição. Porto Alegre, Rio Grande do Sul, Brazil.

Correspondence: Augusto Ricken Siqueira

E-mail: augustoricken@live.com

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Case report

An 11-year-old girl with Peutz–Jeghers syndrome (PJS) and a history of prior bilateral oophorectomy for a sex cord–stromal tumor, currently receiving vitamin D (7000 IU weekly) and estradiol/norethisterone (Natifa®, 0.5 mg/0.1 mg, three times per week) and under multidisciplinary follow-up with pediatric gastroenterology, pediatric oncology, endocrinology, and medical genetics, was admitted with acute abdominal pain, nausea, and vomiting. Laboratory tests revealed elevated pancreatic enzyme levels. Contrast-enhanced abdominal computed tomography demonstrated pancreatic enlargement with peripancreatic fat stranding, consistent with acute interstitial pancreatitis.

Abdominal ultrasound, performed on two separate occasions, showed no gallstones or biliary sludge, with mild extrahepatic bile duct ectasia (common bile duct up to 0.9 cm). Magnetic resonance imaging demonstrated two contiguous polypoid lesions in the distal second portion of the duodenum extending into the third portion, appearing as a bilobulated mass (largest component 6.2 × 3.8 cm on MRI), located in the periampullary region. The lesion was associated with mild biliary dilatation and upstream dilatation of the

main pancreatic duct (up to 0.3 cm), without evidence of gallstones on MRCP (Figures 1 and 2). Diffusion-weighted imaging showed pancreatic diffusion restriction, supporting acute pancreatitis (Figure 2).

A structured etiologic assessment for pediatric acute pancreatitis was performed. Biliary pancreatitis was considered unlikely given the absence of gallstones or biliary sludge on two separate ultrasound examinations, the absence of choledocholithiasis on MRCP, and the lack of gallstones on macroscopic examination of the resected gallbladder. There was no history of abdominal trauma, systemic infection, or prior recurrent pancreatitis. Metabolic causes were not supported, as serum triglycerides (37–112 mg/dL on serial measurements) and serum calcium (8.7–9.6 mg/dL on repeated determinations) were within normal limits. Drug-induced pancreatitis was also considered unlikely, as the patient was receiving only vitamin D and low-dose estradiol/norethisterone, which are not established causes of pediatric pancreatitis. Cross-sectional imaging showed no congenital pancreaticobiliary abnormalities, including no evidence of pancreas divisum. In this context, the presence of a large periampullary duodenal polypoid mass associated with upstream dilatation of the main pancreatic duct supported a mechanical obstructive mechanism at the ampullary region as the most plausible etiology.

Upper gastrointestinal endoscopy confirmed a large periampullary polypoid lesion, markedly friable on minimal contact, with the papillary orifice not clearly identifiable because it was obscured by the lesion (Figure 3). Given the large size, broad-based morphology, and close relationship with the ampullary region, endoscopic resection was considered high-risk for bleeding, perforation, and incomplete excision. Therefore, surgical resection of the duodenal polyps with cholecystectomy was performed, achieving complete removal of the obstructing periampullary lesions.

Histopathological analysis demonstrated a distinctive papillary villous architecture with tree-like arborization of smooth muscle and no evidence of dysplasia, consistent with a hamartomatous polyp of Peutz–Jeghers syndrome (Figure 3). The patient recovered uneventfully and remains asymptomatic, with no recurrence of pancreatitis after at least 6 months of follow-up.

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Discussion

Peutz–Jeghers syndrome is an autosomal dominant disorder characterized by mucocutaneous pigmentation and hamartomatous polyps throughout the gastrointestinal tract (1). Duodenal involvement is less common, and periampullary localization is rare (2). Large periampullary lesions may impair pancreatic duct outflow through extrinsic compression and inflammatory edema involving the ampullary complex, leading to acute pancreatitis (3,4).

Acute pancreatitis secondary to a large periampullary hamartomatous polyp in the setting of PJS is uncommon. In this patient, the absence of biliary lithiasis on repeated ultrasound examinations, MRCP, and gallbladder macroscopic analysis, the lack of congenital pancreaticobiliary anomalies, and the demonstration of a large lesion adjacent to the ampullary region with upstream pancreatic duct dilatation supported an obstructive mechanism as the most plausible cause. Surgical resection achieved complete symptom resolution, with sustained clinical stability during follow-up.

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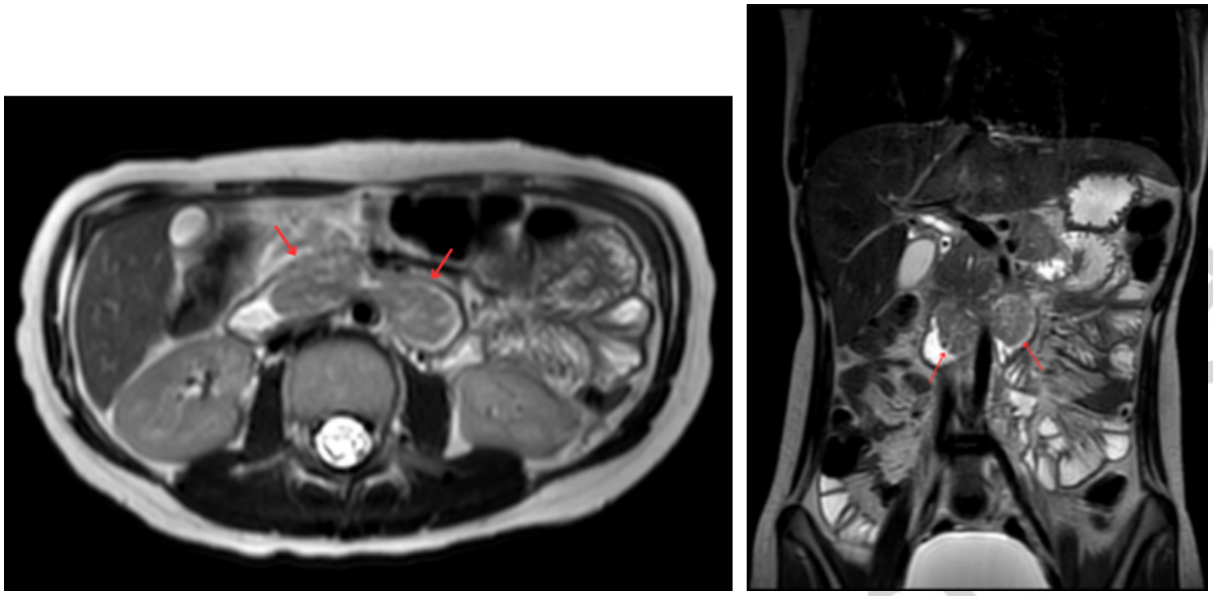

Fig 1. Axial (left) and coronal (right) T2-weighted images demonstrating a bilobed polypoid lesion within the duodenum.

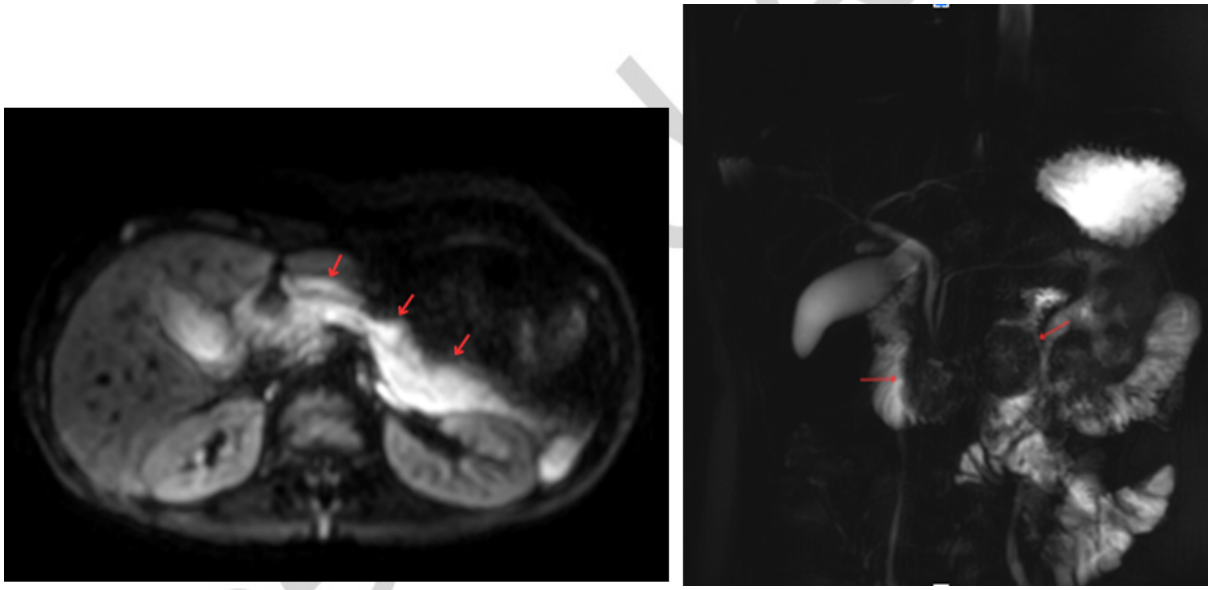

Fig 2. Pancreatic parenchyma showing diffusion restriction (left), consistent with acute pancreatitis. Cholangiographic section (right) demonstrating periampullary involvement by the large duodenal polypoid lesion.

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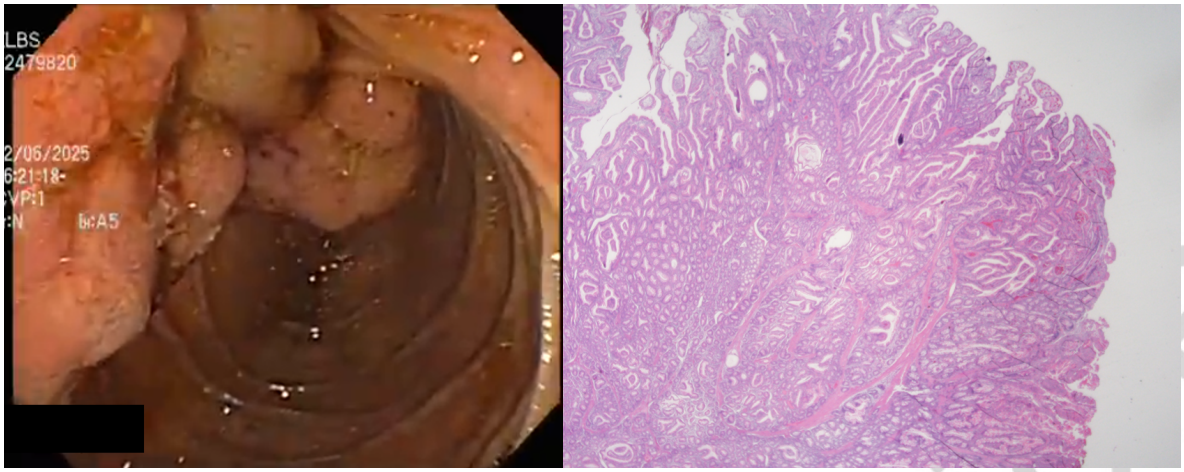

Fig 3. Upper digestive endoscopy (left) demonstrating a large periampullary polypoid lesion making it difficult to visualize the papillary orifice. Histopathological analysis (right) demonstrates distinctive papillary villous architecture in polypoid lesion, with tree-like arborization of smooth muscle, consistent with hamartomatous polyp (H&E, 12,5x).