CASE OF THE MONTH

An unusual case of duodenal beaking

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Radiology

A 35-year-old female presented with complaints of several episodes of malaena and frank blood in the stools for the past 1 year. The frequency of these episodes had increased in the past 2 months. The patient complained of extreme weakness and palpitations. There was no history of abdominal pain, nausea, vomiting or haematemesis. There was no history of fever, jaundice, weight loss or constipation. Physical examination was unremarkable except for the presence of severe pallor. Digital rectal examination revealed black tarry stools staining the glove. There was no palpable growth; first degree haemorrhoids were visible at the 3 o'clock position; no active bleeding was discernible. Her haemoglobin was 4 g dl^{-1} , packed cell volume 12.5%, total leukocyte count $7900\text{ cells mm}^{-3}$ and platelet count $5.34 \times 10^5\text{ mm}^{-3}$. Peripheral smear showed features of

anaemia. Total serum bilirubin was 2.5 mg dl^{-1} (direct bilirubin 1.2 mg dl^{-1}). The rest of the liver function tests were within normal limits. HBsAg (the surface antigen of the hepatitis B virus) was negative and human immunodeficiency virus was non-reactive. A chest radiograph demonstrated mild cardiomegaly and the lung fields were clear. Colonoscopy was within normal limits.

An upper gastrointestinal endoscopy was performed, which showed a normal stomach and first part of duodenum. The endoscope could not be negotiated into the third part of the duodenum.

A barium meal examination was done for further evaluation of the upper gastrointestinal tract. Two radiographs of an upper gastrointestinal barium series are shown (Figure 1).

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(a)

(b)

Figure 1. (a) Barium meal upper gastrointestinal tract: smooth narrowing or “beaking” at the junction of the second and third parts of the duodenum, with mucosal folds running parallel to the lumen and a small C-loop of the duodenum. (b) Barium follow-through study showing dilated proximal jejunum with a lobulated filling defect (arrow), and two parallel lines of barium extending from the duodenum into the jejunum, suggesting intussusception. This appearance is not seen in extrinsic compression of the duodenum.

What are the findings?

The upper gastrointestinal barium series demonstrates a distended stomach and proximal duodenum with a small C-loop. A smooth tapering or “beaking” is seen at the junction of the second and third parts of duodenum, showing mucosal folds running parallel to the luminal axis (Figure 1a). However, the barium passed easily into the proximal jejunum, which is dilated, and shows thickened valvulae and a lobulated filling defect. Two parallel lines of barium are seen extending from the duodenum into the proximal jejunum (Figure 1b).

Differential diagnosis

The possibility of a benign duodenal stricture with a polypoid mass or tumour in the proximal jejunum was entertained.

The distal duodenal stricture could occur because of intrinsic causes (e.g. duodenal tuberculosis, Crohn’s disease, Zollinger–Ellison syndrome, chronic pancreatitis, strongyloidiasis [1], intramural duodenal hematoma, an intraluminal duodenal diverticulum that has not filled with barium or, rarely, an intussusception [2]). Duodenal tuberculosis is usually associated with diffuse ulceration, mucosal irregularity and thickening of folds, which were

not seen in this case. Also, the patient had no fever or abdominal pain, and her chest radiograph did not show any evidence of tuberculosis. Tuberculosis of the duodenum may be indistinguishable from Crohn’s disease and duodenal stenosis due to strongyloidosis. There was no history of chronic pain in the abdomen, so Zollinger–Ellison syndrome and chronic pancreatitis were not considered. Intramural duodenal haematoma may be seen as a concentric obstructive lesion in the second or third part of duodenum, sometimes giving a coiled-spring appearance. But there was no history of trauma, anticoagulant therapy, bleeding diathesis or endoscopic biopsy in our patient. Extrinsic causes of duodenal compression or narrowing (e.g. duodenal bands, preduodenal portal veins, annular pancreas and duodenal duplications [1]) usually present children and do not show mucosal folds running in line with the duodenal lumen; this appearance can be seen in narrowing secondary to intussusception [3].

Sonography

Sonography was undertaken to evaluate the cause of the duodenal stricture. It demonstrated a normal gut signature (with alternate hypoechoic and hyperechoic

layers in the third part of duodenum). There was no wall thickening to suggest the possibility of a duodenal stricture or any mass in this region. The pancreas appeared normal and the pancreatic duct was not dilated. The common bile duct was dilated (8 mm), but tapering at its distal end. A target-shaped lesion suggestive of intussusception was seen in the left lumbar region, containing a lobulated hypoechoic mass measuring 3.8×2.6 cm (Figure 2).

What is the diagnosis and how will it be confirmed?

The diagnosis is duodenojejunal intussusception due to a duodenal tumour. A contrast-enhanced CT scan of abdomen confirmed the diagnosis of duodenojejunal intussusception. It showed "beaking" of the duodenum overlying the spine. A smoothly marginated mass showing intense, homogeneous enhancement, seen within the intussusception, appeared to be the lead point (Figure 3a). No lymph nodes were identified. The pancreatic duct was dilated while the common bile duct appeared normal in calibre. The head and uncinate process of the pancreas appeared to be displaced and extending towards the left of the spine (Figure 3b).

Intra-operatively, the duodenum was very mobile. The third and fourth parts of the duodenum were found telescoping into the jejunum, just beyond the duodenojejunal junction. The jejunum was dilated but visibly healthy. The intussusception was reduced back easily. There was a highly vascular, ulceroproliferative growth in the anterolateral wall of the third part of duodenum, 2 cm beyond the region of the ampulla of Vater. The common bile duct was slightly dilated. The pancreas was normal. Multiple enlarged mesenteric and retroperitoneal nodes were found. A standard Whipple's procedure was performed.

Figure 2. Sonography: a lobulated, well-margined, hypoechoic mass seen within the intussusception.

The post-operative period was uneventful and the patient was well on follow-up after 6 months. Histopathological examination showed the lesion to be a benign gastrointestinal stromal tumour (GIST) of the duodenum, while the lymph nodes showed reactive hyperplasia.

Discussion

Duodenojejunal intussusception is a rare entity owing to the relatively fixed position of the duodenum in the retroperitoneum. In adults it occurs secondary to tumours of the duodenum such as lipoma, adenoma, hamartomatous polyps (Peutz-Jeghers syndrome), Brunner's gland hyperplasia and hamartoma [4], and malignant duodenal ulcer [5]. It can also occur because of intramural duodenal diverticula and duodenal hematoma. Rarely, a duodenal GIST may present with duodenojejunal intussusception [6].

The clinical symptoms of duodenojejunal intussusception are not specific. Epigastric pain, abdominal mass, gastrointestinal bleeding and enteral obstruction may be present. Malaena of short duration and anaemia have also been reported [5, 7]. Uggowitz et al [7] reported hypochromic microcytic anaemia that was resistant to therapy for several years, as was the case in our patient complaining of extreme weakness and palpitations due to malaena for 1 year.

Generally, the biliary and pancreatic system are not involved. Intravenous cholangiography documented the absence of biliary dilatation in a case with duodenojejunal intussusception [8]. However, this is not always the case. Intussuscepting Brunner's gland hamartoma may present with acute jaundice and appears as a homogeneously or heterogeneously enhancing mass at the lead point on CT scan [4]. Uggowitz et al [7] reported another case of duodenojejunal intussusception detected because of obstructive jaundice, due to dislocation and compression of the involved duodenum. McGrath et al [9] and Chalmers et al [10] described the occurrence of dilated extrahepatic bile ducts and pancreatic duct, but it was not specified whether the biliary obstruction was caused by the duodenal tumour or by the intussusception. Our patient showed a high serum bilirubin level, and transient dilatation of the common duct (on sonography) and pancreatic duct (on CT scan). We postulate that the intermittent dilatation of the common bile duct and pancreatic duct in our case may have been caused by the intussuscepted duodenal segment, which was found to be easily reducible pre-operatively.

In a case of duodenojejunal intussusception reported by Neogi et al [2], gastroduodenoscopy revealed a smooth stricture with pin-hole opening in the third part of the duodenum, and a pre-operative diagnosis of a duodenal stricture was made. At laparotomy, a tubulovillous adenoma of 4 cm found inside the third part of the duodenum was resected and a duodenojejunal anastomosis performed. In the present case also, the endoscope could not be negotiated into the third part of duodenum.

In duodenojejunal intussusception, the classic coiled-spring appearance is not apparent in the duodenum on barium studies [5]. As a result of the fixity of the

Figure 3. Contrast-enhanced CT scan. (a) Intussusception showing a homogeneously enhancing mass, which proved to be a benign gastrointestinal stromal tumour. (b) "Beaking" in the third part of duodenum and displacement of the uncinate process of pancreas (arrow) to the left of the spine.

duodenum, while part of the intussusception remains above the junction of the first and second part of the duodenum (the genu inferius), the leading edge of the intussusception is found at the duodenojejunal junction, often resulting in the appearance of two separated masses. This appearance of intussusception on upper gastrointestinal series, reported by van Beers et al [8] in three patients, should not be mistaken as two separate masses. Narrow, tapered barium streaks representing the intussusceptum have been reported in a case of gastroduodenal intussusception [5]. Streaking of barium on the intussusceptum, which mimics the lumen within a lumen of a complete invagination, has been reported in a case of duodenal intussusception [3]. While radiolucent areas suggestive of Brunner's gland adenomas may be seen in the proximal duodenum [3], polypoid lesions may be identified in the gastric antrum and jejunum, on barium study in a case of duodenojejunal intussusception due to Peutz-Jeghers syndrome [5]. Duodenal lipoma causing intussusception may be seen as a multilobulated filling defect in the duodenum on barium study or as a mass showing fat density on CT.

Neogi et al [2] reported a barium study revealing an abrupt, smooth narrowing in the third part of duodenum in duodenojejunal intussusception. In addition, the duodenojejunal junction was not opacified; there was distension of the stomach and the first and second parts of the duodenum, and there was delayed emptying of the stomach. They concluded that clinicians should be suspicious of duodenojejunal intussusception when they encounter a duodenal stricture [2]. Benign strictures of the distal duodenum are rare [1]. Beaking has been reported in ileal intussusception. Duodenal intussusception should be specifically considered if there is evidence

of duodenal malrotations such as an abnormally configured C-loop [11].

Sonography can make the diagnosis of an intussusception when a target-like or bull's eye lesion is seen. A pathognomonic bowel-within-bowel configuration, with or without contained fat and mesenteric vessels, may be visible on CT scan. CT can directly show the elongated duodenum with or without the characteristic target-like lesion in the proximal jejunum, accompanied by dislocation of the ampulla of Vater, in duodenojejunal intussusception [5].

In patients with malaena or anaemia of long duration, inability to negotiate the distal duodenum on endoscopy and presence of an abrupt, smooth narrowing in the third part of duodenum, with mucosal folds running parallel to its luminal axis on barium study, should alert one to the possibility of a duodenojejunal intussusception. Associated findings of a malrotated or small C-loop, distension of the stomach and proximal duodenum, and a coiled-spring appearance in the proximal jejunum may be present. Intermittent dilatation of the bile duct or pancreatic duct, and displacement of the uncinate process of pancreas to the left of the spine, can also be found on sonography and CT scan, respectively. Thus we conclude that "beaking" of the third part of the duodenum is a useful new sign of duodenojejunal intussusception on upper gastrointestinal barium series.

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