

Pyloric giant Brunner's gland hamartoma as a cause of both duodenojejunal intussusception and obscure gastrointestinal bleeding

Duodenojejunal intussepsiyon ve gizli gastrointestinal kanama nedeni olarak pilorik dev Brunner gland hamartoması

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Obscure gastrointestinal bleeding is an important dilemma. Brunner's gland hamartoma is an extremely rare tumor generally localized in the duodenal bulb. We present a 34-year-old woman who had suffered from several episodes of melena for the past three years. Endoscopic examinations were normal. Computed tomography showed a large target lesion over the right abdomen and an image representing intestinal malrotation, which was supported by enteroclysis. At exploratory laparotomy, ligamentum of Treitz was located in the mid-to-right side of the columna vertebralis, and the duodenal bulb was found to be invaginated into the proximal jejunum. After longitudinal duodenotomy, a pedunculated ring-shaped large polyp originating from the pyloric ring was seen and excised. Histology was consistent with Brunner's gland hamartoma. This case with obscure bleeding was original with respect to its rarity and being a huge, ring-shaped tumor with pyloric localization. Moreover, the patient had a rare clinical presentation of duodenojejunal intussusception with accompanying intestinal malrotation.

Obscure gastrointestinal kanama önemli bir sorundur. Brunner gland hamartoması oldukça nadir bir tümördür ve genellikle duodenal bulba yerleşimlidir. Burada üç yıldır birçok melena atağından yakınan 34 yaşında bir kadın hastayı sunmaktayız. Kompütürize tomografide batin sağ üst kadranda büyük bir hedef lezyon ve enteroklizis ile desteklenen intestinal malrotasyonu temsil eden görünüm izlendi. Eksploratif laparotomide Treitz ligamenti vertebral kolumnun orta-sağ tarafında lokalize idi ve duodenal bulbun proksimal jejunuma invajine olduğu gözlemlendi. Longitudinal duodenotomiden sonra pilorik halkadan köken alan saplı, halka şekilli büyük bir polip görüldü ve eksize edildi. Histoloji Brunner gland hamartoması ile uyumlu idi. Obscure kanamalı bu olgu, nadir ve pilorik yerleşimli büyük, halka şekilli tumor sergilemesi bakımından orjinaldi. Dahası, hastada intestinal malrotasyonun eşlik ettiği nadir bir sunum olan duodenojejunal intussepsiyon vardı.

Key words: Brunner's gland hamartoma, obscure bleeding

Anahtar kelimeler: Brunner gland hamartoması, gizli kanama

INTRODUCTION

The source of gastrointestinal bleeding is unidentified in about 5% of patients (1). As with endoscopies for the initial investigation of bleeding, repeat endoscopy is directed at the most likely site, and ideally, if performed during active bleeding, the specific diagnosis may be made. In patients with subacute or intermittent bleeding in whom repeat gastrointestinal endoscopy is negative, the focus of investigation should be broadened to include the small intestine. In patients less than 25 years of age, Meckel's diverticula are the most common source, whereas in patients between 30

and 50 years of age, tumors are the most common abnormalities in small bowel bleeding. Vascular ectasias are more common in older patients (2).

Intussusception is defined as the invagination of one segment of the intestine into another. It occurs mostly in the first year of life, generally between the ages of six months and two years. In some cases, the leading point of the intussusception is a recognizable lesion of the bowel wall such as Meckel's diverticulum, a nodule of ectopic pancreas, a polyp, intestine duplication, or a hypertrophied lymphoid patch (3). Duodenal polyps are ra-

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rely encountered masses in adults, with prevalence ranges between 0.6% - 4.6% (4, 5), and most of them that lead to duodenojejunal intussusception are benign (6, 7).

Intestinal malrotation is a congenital anomaly that results from abnormal or incomplete rotation of the midgut during embryonic development (8). This anomaly refers to either lack of or incomplete rotation of the fetal intestines around the axis of the superior mesenteric artery (SMA) (9). It is rare for this condition to present in adulthood, and may cause chronic but indistinct symptoms that are often difficult to diagnose in adult life. The rotation of intestinal development has been divided into three stages, and intestinal malrotation belongs to stage II anomalies (10). Malrotation can be diagnosed on computed tomography (CT) by the anatomic location of a right-sided small bowel, a left-sided colon, an abnormal relationship of the superior mesenteric vessels, and aplasia of the uncinate process (9). Abnormal positions of the superior mesenteric vein (SMV) in the setting of malrotation were described in patients, in whom CT examination demonstrated the SMV to the left or front of the SMA (11).

Brunner's gland hamartoma is an extremely rare duodenal tumor generally located in the duodenal bulb. This tumor is normally asymptomatic, but it might cause upper gastrointestinal bleeding or intestinal obstruction (12, 13). Herein, we introduce an exceptional Brunner's gland hamartoma with pyloric ring localization as a cause of both duodenojejunal intussusception and obscure gastrointestinal bleeding with accompanying intestinal malrotation.

CASE REPORT

A 34-year-old woman was hospitalized for gastrointestinal bleeding of unknown origin. She had suffered from postprandial abdominal pain, weight loss and several episodes of melena for three years. She also described intermittent sudden nausea and vomiting soon after the onset of the abdominal pain. Although she had been evaluated in different health centers repeatedly, the cause remained unresolved. There was no history of hematemesis attack. She had been taking iron supplementation for three months. No mucocutaneous pigmentation of the skin was evident and family history was negative for any malignancy. On physical examination, there was conjunctival paleness and one palpable mass with local tender-

ness over the right side of the mid-abdomen. On admission, laboratory data were as follows: hemoglobin, 5 g/dl; hematocrit, 17%; mean corpuscular volume, 78 fl; albumin, 3 g/dl; transferrin saturation rate, 6%; and ferritin, 4 ng/ml. Fecal occult blood test was positive. Other laboratory values including tumor markers were within normal ranges. Upper gastrointestinal endoscopic series were performed three times and revealed only indistinct erosive gastropathy and difficulty in passing from pylorus to duodenum. Colonoscopy was normal. Abdominal ultrasonography showed a large target lesion between the stomach and proximal jejunum. CT scan showed a large target lesion representing an intussusception over the right abdomen. It also showed the SMV lying anterior to the SMA, representing an intestinal malrotation. There was no significant enlargement of lymph nodes (Figure 1). Radiographic examination of the small bowel by enteroclysis showed malrotation signs, i.e., the duodenal-jejunal junction failed to cross

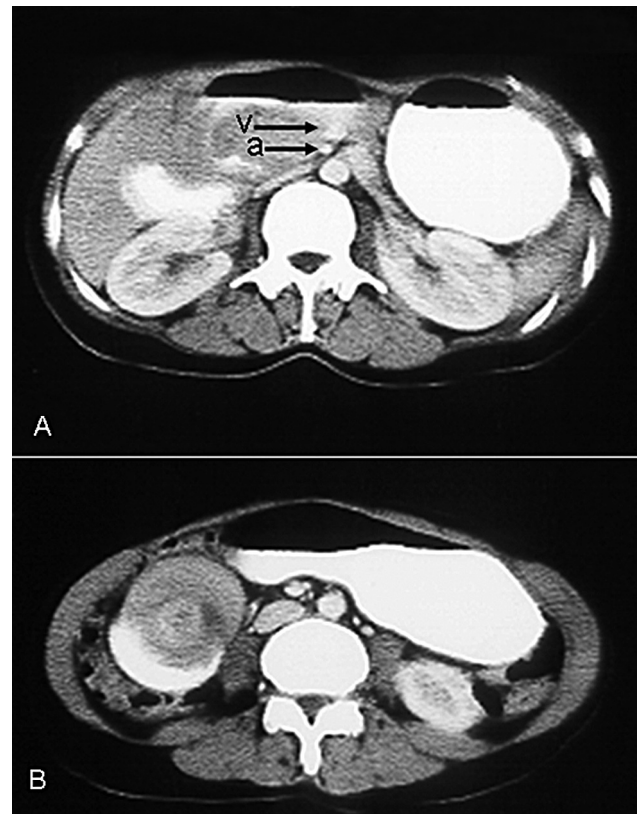


Figure 1. (A) Abdominal CT scan shows the superior mesenteric vein (v) lying anterior to the superior mesenteric artery (a). The SMV normally lies on the right side of the SMA. In malrotation, it lies either in front or on the left side. (B) Abdominal CT scan demonstrates a focal rounded mass representing an intussusception on the right side of the abdomen.

the midline and lay below the level of the duodenal bulb. The cecum was in normal localization but the jejunum was on the bottom right side of the abdomen and the ileum on the upper left side of the abdomen. The ileum also reached to the cecum, crossing the midline from left to right (Figure 2). Both superior mesenteric angiogram and Meckel's scan were negative. On exploratory laparotomy, ligamentum of Treitz was located in the mid-to-right side of the columna vertebralis, and the duodenal bulb was found to be invaginated into the proximal jejunum. The intussusception was reduced manually, and then a palpable lobular mass in the lumen of the duodenal bulb was noted. After longitudinal duodenotomy, a pedunculated ring-shaped large polyp (8 cm) originating from the pyloric ring was seen and excised with its base (Figure 3). The resected specimen was solid, irregular, ulcerative and brown-colored. Microscopically, the tumor was composed of Brunner's glands with the intervening fibrous bands. The connective tissue stroma contained a mixture of ducts, adipose tissues and smooth muscles without evidence of cellular atypia, findings consistent with a diagnosis of Brunner's gland hamartoma (Figure 4). There was no complication after operation, and she was discharged on the ninth hospital day in

stable condition. One month later, the patient was asymptomatic; the upper gastrointestinal endoscopy showed patent pylorus and duodenum without any mass.



Figure 3. A pedunculated ring-shaped large polyp (8 cm) originating from the pyloric ring is seen after longitudinal duodenotomy.

DISCUSSION

Brunner's glands are known as mucus-secreting glands found mainly in the duodenum, although they are occasionally found in the pylorus and distally as far as the jejunum (14). Polyps of these glands are rare benign lesions, accounting for less than 1% of small intestinal tumors (8). Brunner's gland polyps are mostly located in the proximal duodenum and are usually pedunculated, and a size greater than 2 cm is exceedingly rare (15, 16). Goldman (17), a well-known reviewer of the Brunner's gland "hyperplastic" polyps, pointed out that the histologic architecture of these lesions consists of a combination of ductal and acinar structures with fibromuscular and adipose elements. Furthermore, lack of an encapsulation fits the criteria for neither adenoma nor hyperplasia. Accordingly, he was the first to stress that these features strongly suggest a hamartomatous origin for Brunner's gland polyps (17). The distribution of Brunner's gland hamartomas is duodenal bulb (57%), the second (27%) and third (5%) portions of the duodenum, the pyloric channel (5%), jejunum (2%), and proximal ileum (2%) (18).

An entire gastrointestinal examination is mandatory since hamartomas are commonly associated

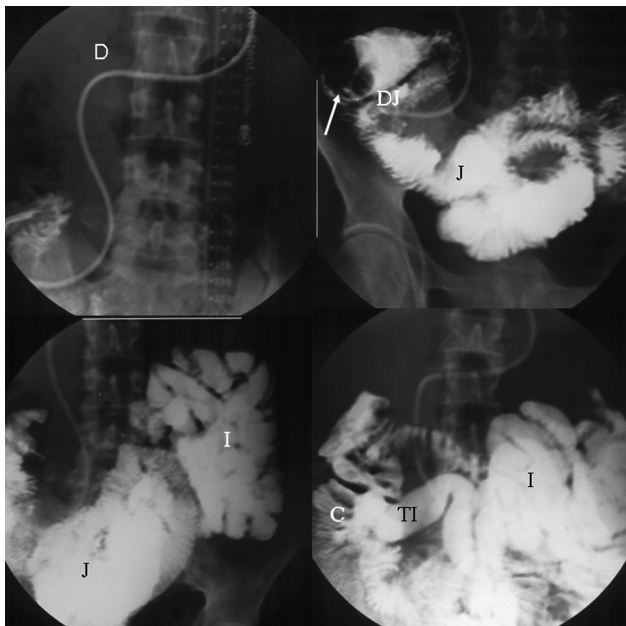


Figure 2. Radiographic images from a contrast material-enhanced gastrointestinal examination show the small intestine on the right side of the abdomen and the colon and cecum on the left side. The ileum is seen crossing the midline from right to left. Ligamentum of Treitz is located on the right side of the mid-abdomen.

with a generalized polyposis syndrome (19). Even though these tumors are dysembryoplastic lesions and exhibit a benign course without malignant predisposition (20, 21), the literature reports a proven case of cancer developing from this origin (22, 23). The differential diagnosis usually includes leiomyoma, adenoma of the mucosal glands, carcinoid tumor, lipoma, prolapsed pyloric mucosa, or antral polyp (24).

The majority of patients with Brunner's gland hamartomas are asymptomatic and the lesion is an incidental finding during endoscopy or other duodenal imaging procedures. The symptoms may be overt due to obstruction, motor disturbances, and ulceration or erosion of the tumor, which gives rise to upper gastrointestinal hemorrhage or anemia (21, 25). Duodenojejunal intussusception secondary to hamartomatous polyps is a scarce clinical

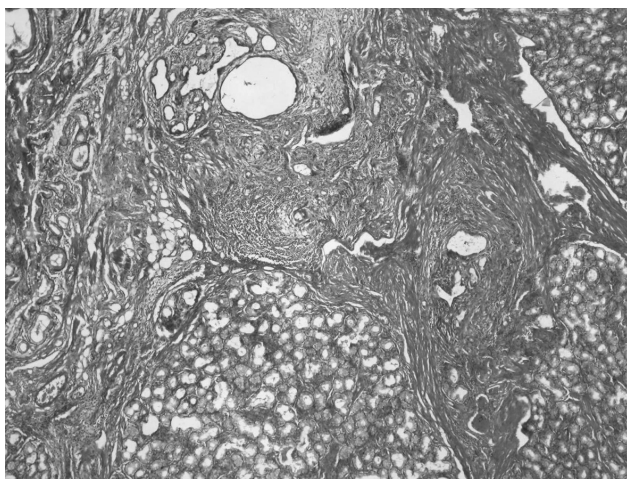


Figure 4. Histological findings of the polyp are consistent with Brunner's gland hamartoma: it is composed of Brunner's glands with the intervening fibrous bands. The connective tissue stroma contains a mixture of ducts, adipose tissues and smooth muscles without evidence of cellular atypia.

presentation reported in the literature (3). Ultrasonography may be the most useful initial screening examination for intussusception, which has been described as a "target" on transverse views and a "pseudokidney" on the longitudinal views (26). Our patient was investigated extensively after the ultrasonogram showed the "target" lesion. After the negative results of repeat endoscopies, we considered capsule endoscopy in order to determine the origin of the obscure bleeding. Since the patient also described intermittent sudden nausea and vomiting soon after the onset of the abdominal pain, which was compatible with ileus, this procedure was deemed too risky. Moreover, her clinical status was unstable. Finally, we consulted with surgeons and decided on an operation. We found this patient, who presented with recurrent episodes of melena and late postprandial abdominal pain, to have a polypoid Brunner's gland hamartoma with a long stalk in the pyloric ring. Interestingly, the tumor was reserved in the lateral pocket of the intussuscepted duodenojejunal area, which explains why it was invisible on upper endoscopic examinations. To our knowledge, this is only the second reported duodenojejunal intussusception secondary to hamartomatous polyp. Furthermore, we diagnosed intestinal malrotation utilizing enteroclysis, CT scan and exploratory laparotomy by the criteria mentioned in the features of the case.

In conclusion, our case was original in terms of the following features: i) a rare tumor: Brunner's gland hamartoma, ii) a rare localization: pyloric ring, iii) largeness and shape: 8 cm and ring-like, iv) rare clinical presentations: obscure bleeding and duodenojejunal intussusception, v) malrotation: predisposition to intussusception, vi) a rare instance: polyp out of the field of endoscopic view due to entrance into the intussuscepted area.

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