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CASE REPORT

Heterotopic Ileal Pancreas with Lipoma and Coexisting Fibromatosis Associated with a Rare Case of Gastrointestinal Bleeding. A Case Report and Review of the Literature

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ABSTRACT

Context The development of pancreatic tissue outside the confines of the main gland represents a congenital abnormality referred to as heterotopic pancreas. This is a rare pathological and surgical entity which remains mostly asymptomatic.

Case report We present the case of a 28-year-old male, who was admitted to hospital because of a history of blood in bowel movements. After a normal gastroscopy and colonoscopy, Tc^{99m}-tagged red blood cells scintigraphy showed enrichment in the right lower abdomen. At double-balloon endoscopy, a intraluminal polypoid mass 8 cm in diameter was revealed 120 cm from the ileocecal valve. The initial macroscopic diagnosis was a gastrointestinal stromal tumor. During surgery, the diagnosis of heterotopic pancreas with lipoma and fibromatosis was made. To our knowledge this is the first case of ileal heterotopic pancreatic tissue and lipoma described to date in the literature.

Conclusion Ileal heterotopic pancreas is a rare entity with potentially life-threatening complications, local excision being the appropriate indicated treatment.

INTRODUCTION

The development of pancreatic tissue outside the confines of the main gland represents a congenital abnormality referred to as heterotopic pancreas. This is a rare pathological and surgical entity which remains mostly asymptomatic. However, depending on its location, certain complications may occur which define its clinical significance. We present the case of an ileal heterotopic pancreas with lower gastrointestinal bleeding emphasizing the main diagnostic and therapeutic problems this rare abnormality poses.

CASE REPORT

A 28-year-old male patient was first admitted to an external department for internal medicine because of a history of blood in bowel movement, without fever or pain for 2 days. The color of the blood ranged from light to dark red with decreasing bleeding. There was no history of NSAID intake or any other drugs. The patient's weight had remained constant over the previous 6 months.

Physical examination revealed good a general condition, stable hemodynamics (blood pressure: 120/80 mmHg) and regular heart rhythm of 72 bpm. The abdomen was soft and

non-distended, with bowel sounds present and without palpable tumor. Digital examination of the rectum was without pathological findings, other than remnants of coagulated blood. In the laboratory blood samples, hemoglobin was 12.8 g/dL (reference range: 14-18 g/dL). Due to a relevant drop of the patient's hemoglobin, a total of four units of red blood cells were given.

Besides slight edema at the pyloric area, nothing suspicious was found during gastroscopy. During colonoscopy, coagulated blood was revealed in the proximal ileum, but the source of the bleeding could not be located. Thereafter, conventional catheter mesentericography was performed. The examination did not detect any intestinal bleeding.

An ultrasound scan was carried out, without suspicious findings. Tc^{99m}-tagged red blood cell scintigraphy showed enrichment in the right lower abdomen. Therefore, a double-balloon endoscopy was performed. This examination revealed a bleeding ulceration 30 cm from the ileocecal valve. After submucosal adrenalin injection, the bleeding was successfully terminated.

A follow up examination two weeks later revealed an additional drop of hemoglobin from 10.7 g/dL to 9.3 g/dL and a weight loss of 4 kg. Blood in the bowel movement was not observed.

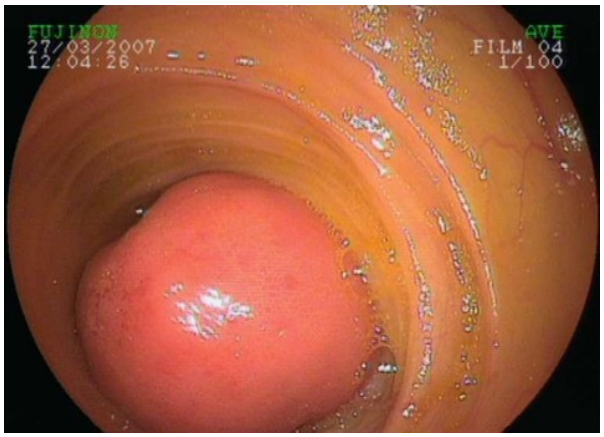

Figure 1. Endoscopic view. Suspicious formation found 120 cm from ileocecal valve. Its macroscopical appearance was consistent with a gastrointestinal stromal tumor (GIST).

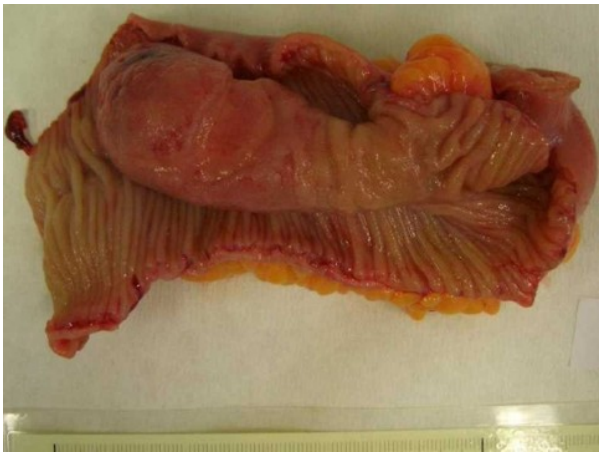

Figure 2. Specimen. A 15 cm long ileum with the polypoid mass. Note: mucosa lesion.

An ultrasound scan of the abdomen showed no abnormalities. Cervical, axillary and inguinal lymph nodes were not suspicious. During double-balloon endoscopy, the former ulceration could not be identified. However, a polypoid lesion 8 cm in diameter was revealed 120 cm from the ileocecal valve. The formation subtotally occluded the ileum and its macroscopic appearance was consistent with a gastrointestinal stromal tumor (GIST) (Figure 1). Multiple biopsies were taken.

Computed tomography of the abdomen revealed multiple moderately enlarged lymph nodes (approximately 9 mm in diameter) surrounding Bauhin's valve. No other abnormal formations were seen. Due to ongoing gastrointestinal bleeding with respect to endoscopic findings consistent with a GIST, we decided to perform a diagnostic laparoscopy. Inspection of the intraperitoneal cavity showed no abnormal macroscopic findings. However, careful palpation of the suspicious ileum region as described by the previously performed endoscopy, indicated an intraluminal mass. A partial ileum resection (length 150 mm) and an end to end ileum anastomosis were performed. A suspicious peritoneal lesion at the ileocecal area was also resected. The patient's postoperative course was uneventful and he was discharged on the fifth postoperative day. At the moment, follow-up is on an outpatient basis without any relevant problems.

Pathologic Findings

The specimen consisted of 150 mm of ileum with a 90 mm long and 35 mm wide polyp. (Figure 2). Macroscopically, the mucosa showed a 30x12 mm laminar lesion. Histological examination revealed a lipoma of the ileal mesenterium. The prominent ileal mucosa showed an ulceration and a nodule of exocrine heterotopic pancreas tissue (Figure 3). The suspicious peritoneal lesion at the ileocecal area was identified as fibromatosis.

DISCUSSION

By definition, heterotopic pancreas is pancreatic tissue lacking anatomic and vascular continuity with the main body of the pancreas [1]. It is a rare entity with an estimated incidence of 1 in 500 surgical procedures in the upper abdomen or 0.5-13.7% in autopsy cases [2, 3, 4].

The most common locations of a heterotopic pancreas are the stomach (24-38%), duodenum (9-36%), jejunum (15-21.7%), and occasionally the esophagus, gallbladder, bile duct, spleen, mesentery, fallopian tubes and Meckel diverticulum [3, 4, 5, 6].

Depending upon the anatomical location and the tumor size, a heterotopic pancreas might be asymptomatic or may otherwise manifest with atypical abdominal pain, weight loss, nausea, vomiting and bleeding with melena or anemia, as in the case presented [1, 3, 4, 5]. Due to the exocrine and endocrine function of the heterotopic pancreatic tissue, complications from the main body of the pancreas, such as acute or chronic pancreatitis, may occur [7, 8]. The incidence of malignant transformation is not clear because only 15 cases have been reported to date [3]. An ileoileal intussusception, due to the quantity of heterotopic tissue, has also been described [9].

Heterotopic pancreas in the ileum is rare. Barbosa *et al*. reported the first case of ileal heterotopic pancreas complicated with intussusception in 1946 [10] and only 16 cases of ileal heterotopic pancreas and gastrointestinal bleeding have been reported to date in the literature [4, 11].

The pathogenetic mechanism of pancreatic ectopia remains unclear. Armstrong *et al*. suggested separation of the pancreatic tissue occurring during embryonic rotation and fusion of the ventral and dorsal pancreatic buds [1]. Skandalakis *et al*. suggested a metaplasia of pluripotential endodermal cells of the embryonic foregut as an origin for heterotopic tissue, which might explain occasional reports on unusual sites such as the fallopian tube [12].

CT findings of heterotopic pancreas tissue appear to be nonspecific for diagnosis, except for location. Despite modern techniques, such as multislice spiral CT and portovenous i.v. contrast, it remains difficult to distinguish heterotopic pancreatic tissue from other submucosal tumors [13].

Diagnosis is usually made after histological examination of the suspected tissue in symptomatic patients. In a series of 32 ectopic pancreas cases, Pang *et al*. reported an incidental diagnosis in 18 (56%) asymptomatic patients while 14 patients had symptoms. Intraoperative diagnosis by frozen section was made in 28 (87%) cases. Preoperative diagnosis was not made in any of these cases [5]. The exact diagnosis was also histologically confirmed in our case.

Depending on its location, conventional or laparoscopic ileum resection with restoration of the continuity by means of an anastomosis has been shown to be an adequate treatment

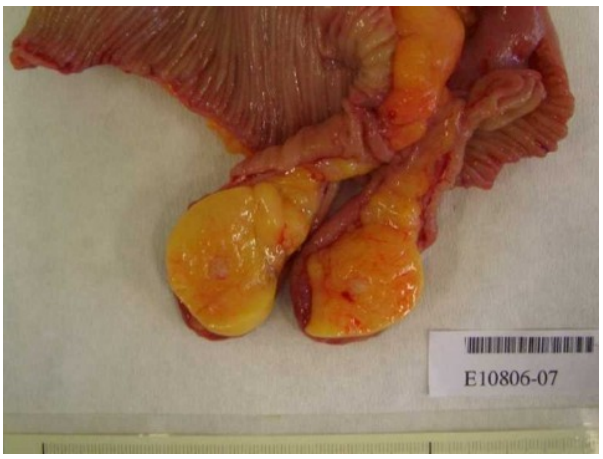

Figure 3. Specimen. Pathologic examination revealed a nodule of exocrine heterotopic pancreas tissue in the mucosal ulceration.

for ileal pancreas [9, 14]. Subsequent histologic examination to exclude the presence of malignant disease is warranted.
In our case, histology revealed no malignancy; it confirmed the presence of pancreatic tissue along with a lipoma and coexisting fibromatosis. To our knowledge, this is the first case of ileal heterotopic pancreatic tissue and lipoma described to date in the literature.
In conclusion, ileal heterotopic pancreas is a rare entity with potentially life threatening complications. Despite modern diagnostic methods, preoperative diagnosis remains difficult. Local resection of the heterotopic tissue is the appropriate indicated treatment.

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Keywords Gastrointestinal Hemorrhage; Lipoma; Pancreas

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