CASE REPORT

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Ectopic pancreas presenting with intractable diarrhea: Case report

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Ectopic pancreas is an uncommon congenital anomaly, which is usually found incidentally in clinical practice. It presents with non-specific gastrointestinal symptoms like epigastric pain and dyspepsia and rarely with the clinical findings of obstructive jaundice or intestinal obstruction, or it may mimic gastrointestinal system cancer. Herein, we describe a case of ectopic pancreas in the duodenum, which was the cause of the intractable diarrhea. In our patient, upper gastrointestinal endoscopy and endoscopic ultrasonography revealed a 1.5 cm submucosal lesion, which was umbilicated centrally with a normal in appearance overlying mucosa. Endoscopic biopsy of the lesion was normal. Pathological examination of the lesion after surgical excision was compatible with ectopic pancreas. After total excision of the lesion, the clinical findings of the patient normalized. Ectopic pancreas presenting with diarrhea has not been reported previously in the literature.

Key words: Ectopic pancreas, diarrhea, duodenum

Ektopik pankreasa bağlı inatçı ishal: Olgu sunumu

Ektopik pankreas genellikle tesadüfen saptanan nadir bir konjenital anomalidir. Epigastrik ağrı, dispepsi gibi spesifik olmayan gastrointestinal semptomlarla kendisini gösterir. Nadiren tıkanma sarılığı, intestinal obstrüksiyon kliniği verebildiği gibi gastrointestinal sistem kanserini de taklit edebilir. Biz bu çalışmada, inatçı ishale neden olan duodenumda lokalize bir ektopik pankreas olgusu sunuyoruz. Hastamızda üst gastrointestinal sistem endoskopisinde ve endoskopik ultrasonda, ortasında göbeklenme olan, yüzeyel mukozanın normal olduğu, 1.5 cm çapında submukozal bir lezyon izlendi. Lezyondan endoskopik yolla alınan biyopsiler normal idi. Cerrahi yolla eksize edilen lezyonun patoloji sonucu ektopik pankreas ile uyumlu bulundu. Lezyonun total eksizyonu sonrasında hastanın kliniği normale döndü. İshal ile birliktelik gösteren ektopik pankreas olgusu literatürde daha önce yayınlanmamıştır.

Anahtar kelimeler: Ektopik pankreas, ishal, duodenum

INTRODUCTION

Ectopic pancreas is a congenital anomaly, which can be defined as the presence of pancreatic tissue with no vascular or anatomic connection with the normal pancreas. The frequency of ectopic pancreatic tissue in autopsy material has been reported to range from 0.55% to 13.7% (1). The most common sites are the stomach, duodenum, proximal jejunum, and ileum. Less common sites are the umbilicus, common bile duct, gallbladder, Meckel's diverticulum, and the hilus of the spleen, as well as perigastric and paraduodenal locations (2). Ectopic pancreas is usually asymptomatic and discovered incidentally, or presents with non-specific gastrointestinal symptoms (e.g., epigastric pain, dyspepsia). However, some rare complications have been described in the literature, including obstructive jaundice, intestinal obstruction, gastric outlet obstruction, and development of car-

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cinoma in the ectopic pancreas tissue (3-6). This report describes a case of ectopic pancreas in the duodenum, which was the cause of the intractable severe diarrhea.

CASE REPORT

A 26-year-old female was admitted to the hospital with the symptom of diarrhea without blood or mucus, with a frequency of 20 times a day, with accompanying abdominal pain beginning from the epigastric area and radiating to the umbilical area, regressing with defecation. The amount of diarrhea was about 3 liters per day. She reported no nausea or vomiting, but weight loss of 7 kg within five months and her body mass index (BMI) was 17. Her physical examination was normal. Vital signs, temperature, pulse, and blood pressure were normal. The stool culture and parasite examination were negative. Upper gastrointestinal endoscopy revealed a 1.5 cm submucosal lesion, which was umbilicated centrally with a normal in appearance overlying mucosa (Figure 1). Endoscopic biopsy of the lesion was normal. Work-up included colonoscopy, abdominal ultrasonography, barium studies, double balloon enteroscopy, abdominal tomography, and capsule endoscopy, but they showed no pathological signs. The blood values of thyroid function, anti-endomysial and antigliadin antibodies, serum cortisol level, parathyroid hormone (PTH), calcitonin, vasoactive intestinal polypeptide (VIP) and 5-hydroxyindoleacetic acid (5-HIAA) in 24-hour urine were all within normal limits. Positron emission tomography-computed tomography (PET-CT) and octreotide scintigraphy in order to rule out any kind of malignancy revealed normal results. During the follow-up period, the number of defecations increased, and a new upper gastrointestinal endoscopy and endoscopic ultrasonography (EUS) were carried out to further evaluate the lesion in the bulbus. EUS revealed that the lesion was originating from the submucosa and muscularis propria (3rd and 4th layers) with normal in appearance overlying mucosa. The echogenicity of the lesion was similar to that of the pancreas (Figure 2). Partial resection of the lesion was applied endoscopically and the number of stools per day decreased about 50% after resection, so total excision of the duodenal mass was performed by surgery (Figure 3). Subsequent pathology revealed ectopic pancreas (Figure 4a and 4b). The number of stools per day decreased dramatically to normal after excision of the ec-

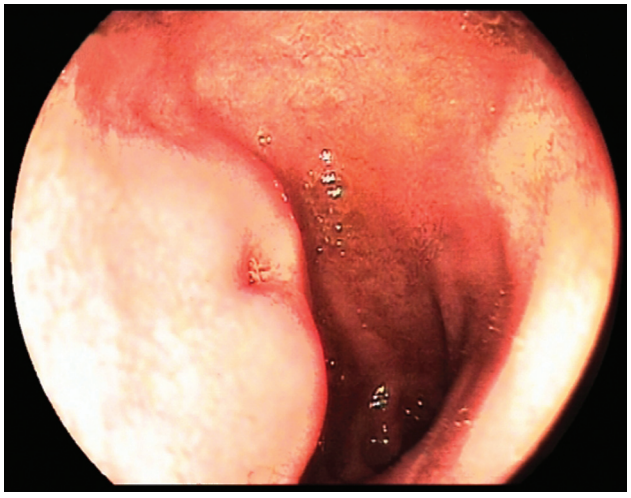

Figure 1. Endoscopic view of the ectopic pancreas in the duodenum showing a submucosal lesion with central umbilication and normal overlying mucosa.

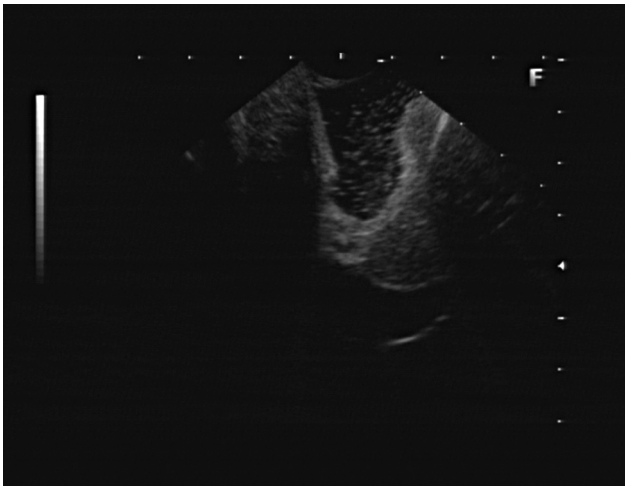

Figure 2. Endosonographic view of the submucosal lesion involving the 3rd and 4th sonographic layers of the duodenum. The echogenicity of the lesion was similar to that of the pancreas.

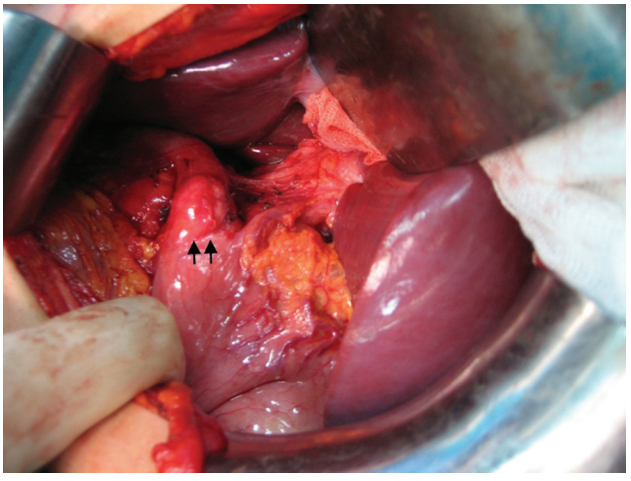

Figure 3. Intraoperative view of the ectopic pancreas in the duodenal wall.

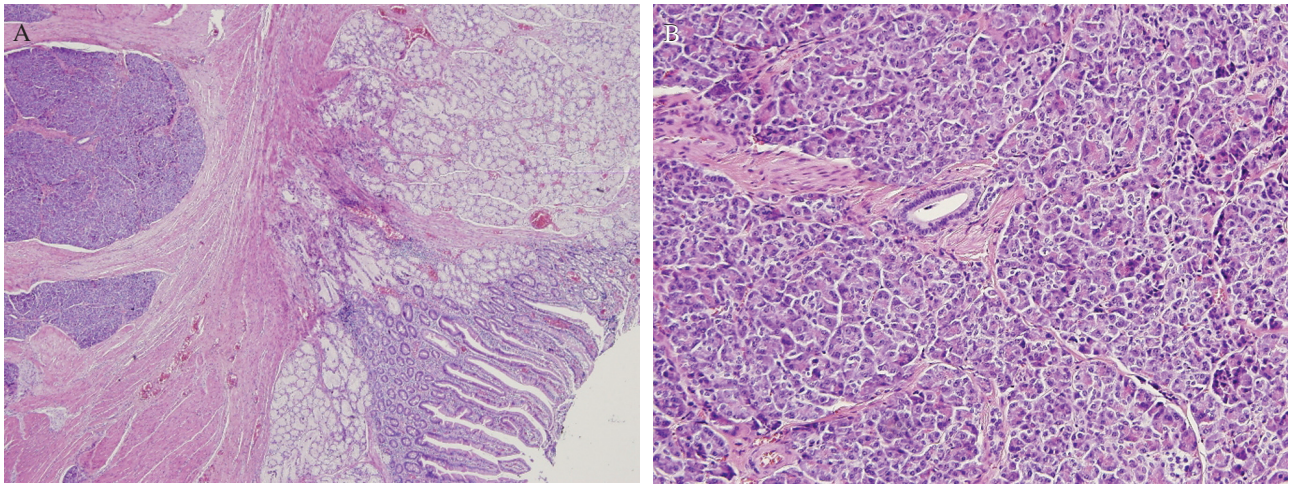

Figure 4. A: Ectopic pancreatic tissue located in the muscularis propria of the duodenal wall (Hematoxylin and eosin, X20). **B:** Histopathological sections of the ectopic pancreas with ductus and asinus (Hematoxylin and eosin, X40).

topic pancreas. During the one- year follow-up of the patient, the frequency of stools per day remains normal.

DISCUSSION

Ectopic pancreas was first described in 1727, when it was found in an ileal diverticulum (7). It can be defined as pancreatic tissue that is outside its normal location, having no connection with the main pancreatic tissue. The etiology of ectopic pancreas is unknown. It is thought that early in fetal life, during rotation of the foregut and fusion of the dorsal and ventral parts of the pancreas, small fragments become separated from the main gland and continue their development in different locations, the so-called misplacement theory (8). The other theory is pancreatic metaplasia of endodermal tissues, which can be explained as the presence of an ectopic pancreas away from the main gland (9).

Ectopic pancreas is mostly localized in the submucosa, and rarely can be found in muscularis mucosa, subserosa and serosa (8). It has its own exocrine tissue (acini) and excretory ducts, and islets of Langerhans can be seen. The ducts drain separately into the lumen (9).

Ectopic pancreas is most often detected incidentally during routine upper endoscopy. Grossly, ectopic pancreas in the stomach and duodenum is a round or oval subepithelial nodule with central umbilication (6). However, pathological diagnosis of ectopic pancreas is usually very difficult because pinch mucosal biopsies during endoscopy do not provide enough tissue for diagnosis, since they are mostly submucosal lesions (10). Excision of the tissue, either endoscopically or laparoscopically, is the best means of diagnosis.

Most cases of ectopic pancreas are asymptomatic, but symptoms may occur because of the hormones and enzymes secreted by the ectopic pancreatic tissue (9). Histologically, ectopic pancreas may resemble a normal pancreas with acini, ducts and islets of Langerhans, or may contain widely separated ducts with a muscular stroma (10). Our case is the only case presenting with diarrhea.

In the literature, there are a limited number of case reports about ectopic pancreas. Obermaier (11) reported a case of ectopic pancreatic tissue localized in the major duodenal papilla, causing biliary obstruction and mimicking pancreatic head tumor. In that case, pancreatic head resection was performed for diagnosis and treatment. Hsu (12) also reported a patient presenting with symptoms of ampullary tumor with obstructive jaundice. They had performed a Whipple procedure on the suspicion of a tumor of the ampulla of Vater, but the pathological result was ectopic pancreas. Papaziogas (6) described a case of endoepithelial carcinoma arising in a gastric heterotopic pancreas, in which partial gastrectomy was performed. Khashab (10) performed successful band ligation-assisted endoscopic mucosal resection of a gastric heterotopic pancreas in two cases. Erkan (4) reported a case of ectopic pancreas in the jejunum, with the findings of intestinal obstruction. In that case, segmental resection of the jejunum was performed successfully.

The management of ectopic pancreas changes according to the presence of symptoms. When the patient is asymptomatic, regular follow-up is preferred. If the patient is symptomatic, surgical management is indicated. The type of the procedure should be performed according to the type and size of the pancreatic tissue (3).

In conclusion, our case is the first case of ectopic pancreas in the literature presenting with intractable severe diarrhea. Since ectopic pancreatic tissue is generally submucosal, we suggest excision of the lesion rather than mucosal biopsies for the correct diagnosis in symptomatic patients.

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