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Ectopic Pancreatic Tissue in The Small Bowel

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1. Abstract

The ectopic pancreas is a relatively rare submucosal tumor in the gastrointestinal tract. It is defined as pancreatic tissue without vascular or anatomical continuity with the main body of the pancreas, usually distantly implanted. Ectopic pancreas in the ileum is a rare examination finding. We report a case of ectopic pancreas in the ileum causing recurrent abdominal pain treated by laparoscopic surgery.

2. Introduction

The ectopic pancreas represents a pancreatic tissue without continuity and is isolated from the orthotopic pancreas, lacking anatomical and vascular connections [1,2]. Male involvement is predominant, but its incidence is difficult to determine since most affected patients are asymptomatic [1,3]. Clinical suspicion arises during surgeries for other diseases or autopsies [4,5]. Its true etiology is unknown, but some theories try to explain this phenomenon. The gold standard diagnosis is histological confirmation. The treatment of choice is surgical resection. However, despite the absence of symptoms in most patients, the ectopic pancreas can lead to complications, such as bleeding and tumor formation. Thus, early diagnosis and adequate treatment of ectopic pancreas are essential in the clinic. We report a case in which the patient had an ectopic pancreas, confirmed by histology, to update and add knowledge about this rare pathology to health professionals since its incidence has increased.

3. Case Report

A 62-year-old woman attended our hospital with intermittent abdominal pain and abdominal distension. The patient denied any relevant medical history. Physical examination only revealed epigastric tenderness. Laboratory tests results were normal, and the computed tomography (CT) did not show significant changes. A Magnetic Resonance (MR) showed a 0.4 cm nodular lesion on the antimesenteric wall of the small bowel at 60 cm from the duodenojejunal flexure, with no signs of invasion of adjacent structures. After careful evaluation of the MR, a diagnostic laparoscopy was performed. The lesion described by the RM was identified at 45 cm from the duodenojejunal flexure, with a 0,5 cm nodular lesion in the antimesenteric wall of the small bowel (Figure 1). The patient underwent segmental enterectomy with exiguous margins and primary enteroenterostomy with a laparoscopic stapler. The intestinal segment proceeds to anatomopathological frozen analysis that confirmed the heterotopic pancreas with necrosis and

inflammatory process inside the pancreatic tissue with punctual malignant degeneration, with free surgical margins.

The histopathological examination later confirmed the findings described by the pathologist in the operating room. The patient recovered satisfactorily, accepted the diet after the second day of surgery, and was discharged from the hospital on the third day of hospitalization. The patient is being followed up as an outpatient without recurrence for four months, without presenting any current abdominal symptoms.

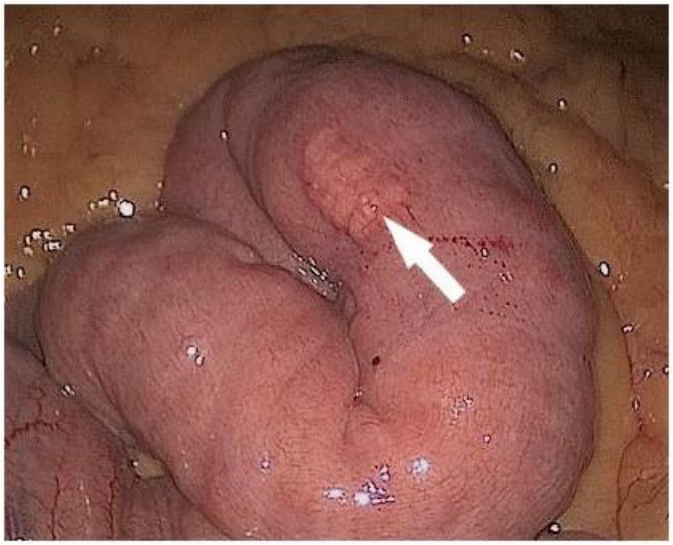

Figure 1: laparoscopic finding of a small bowel segment with a 0.5 cm diameter nodular lesion at 45 cm from the ligament of Treitz.

4. Discussion and Conclusion

Jean Schultz, in 1727, described a presumed case of tissue similar to the pancreatic gland at the base of the ileal diverticulum, which is likely the first case of ectopic pancreas recorded in the literature [6]. More than 100 years later, in 1859, Klob described the histological features of this condition [7,8]. Ectopic pancreas (EP) is considered a rare benign congenital disease, with its formation during embryonic development, characterized by the appearance of pancreatic tissue in areas other than its anatomical location and without any ductal or vascular continuity [1,6]. Although its true incidence remains unknown, as most patients are asymptomatic, some studies report incidental findings of the ectopic pancreas during abdominal surgeries for other indications, with a percentage ranging from 0.2% to 0.3%, or in autopsy series, with a higher value ranging from 0.5% to 14% [5,9]. The ectopic pancreas can occur throughout the gastrointestinal system but is found primarily in the upper gastrointestinal tract, less frequently in the small intestine [1]. The stomach and duodenum are the two most common sites of the ectopic pancreas, accounting for 24% to 43% and 9% to 36% of all cases, respectively [1,10]. The etiology of EP is unknown, but there are two predominant theories about the origin of pancreatic ectopic, the migration or displacement theory and the metaplasia theory [6]. Within the migration/displacement theory, during embryonic rotation of the dorsal and ventral intestinal buds, pancreatic tissue deposits migrate from the main body of the pancreas. As a result, they occur at various ectopic sites [6,7]. Most cases of pancreatic heterotopia are asymptomatic and discovered by chance, as it is a silent malformation [8]. Only a few patients with EP have clinical symptoms, often nonspecific. When symptoms are present, upper abdominal pain is most common. On the other hand, jaundice, vomiting, nausea, and weight loss are few reported complaints [1,10]. In the Hancheu study, it was revealed that 8 of the 10 symptomatic cases in the study were associated with abdominal pain and gastrointestinal bleeding or melena, as well as syncope and abdominal distention in two cases, vomiting, and diarrhea in one case, concluding that most lesions found in clinical practice are associated with abdominal pain, gastrointestinal bleeding, and anemia [5]. They may become clinically evident when complicated by chronic inflammation or lesion growth. It has been noted that the severity of symptoms depends on the size of the ectopic lesion; therefore, lesions larger than 1.5 cm have been associated with more severe symptoms. The most reasonable explanation is that the pain results from the secretion of hormones and enzymes that trigger the onset of spasms, chemical irritation, and inflammation of the surrounding tissue [8, 9,10]. The preoperative diagnosis of EP requires high clinical suspicion. It can be divided into stages that complement each other and help in the conclusion of the pathology since, usually, the finding of the ectopic pancreas is accidental, characterized by clinical picture, exploration, and diagnostic confirmation [1,5,9]. Since only the histological examination from a biopsy can confirm the presence of an ectopic pancreas. There are no markers that can verify the existence of the pancreatic mass, so laboratory tests provide information about the patient's condition. Amylase and lipase levels may increase in the presence of EP and, combined with other symptoms and pathological findings, may serve as an indication for diagnosis [6].

The investigation of ectopic pancreatic tissue is complex because

1. The symptoms are nonspecific and can be confused with a gastrointestinal stromal tumor, carcinoid tumor or gastrointestinal carcinoma;

2. Generally, the size of the ectopic pancreas is small and may go unnoticed by imaging tests;

3. EP frequently stays in the deepest layer of the submucosa of the intestine, which makes it hard for biopsy.

Consequently, the mucosa is removed from the intestine and not from the ectopic pancreas, causing an erroneous negative result [1]. Thus, making a diagnosis of EP is more accessible in the upper gastrointestinal tract; on the other hand, visualization in the distal portion of the small intestine is deceptive, so the diagnosis in the ileum is rare and accidental [9]. This disease lacks specific tests for diagnostic confirmation, so it is easy for patients with the

ectopic pancreas to be misdiagnosed [1,7]. Imaging studies, laboratory tests, and endoscopic views are neither specific nor sensitive enough to analyze ectopic pancreas. Thus, the definitive diagnosis and gold standard to date can be made only in the histopathological examination [1,6,9]. Multiple complications occur secondarily in the ectopic pancreas, including ulceration or inflammation, leading to ectopic pancreatitis, pseudocyst formation, malignant degeneration, gastrointestinal bleeding, intestinal obstruction, perforation and intussusception [7,9]. Rarely, malignancy such as adenocarcinoma, mucinous papillary neoplasm, and solid pseudopapillary tumor may develop in EP. Malignant transformation of topical pancreatic tissue ranges from 0.7 to 1.8% among all cases of ectopic pancreatic tissue. These tumors are usually located in the submucosal layer and only occasionally expand into the muscularis propria [11,12].

The ectopic pancreas is a rare pathology that is challenging to recognize. Its nonspecific and often asymptomatic symptoms make diagnosis even more difficult. Treatment, if not carried out early, increases the chances of complications. Therefore, this study aimed to update the information about this pathology so that clinical and surgical measures are recognized early to avoid a worse prognosis for patients, especially men aged between 30 and 50 years, from the description of a case report.

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