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Annular pancreas: A rare cause of duodenal obstruction

Anüler pankreas: Duodenal obstrüksiyonun nadir bir nedeni

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Abstract

Annular pancreas is a rare congenital anomaly. It occurs in 1 in 20,000 of the population. Annular pancreas clinically manifests as digestive stenosis which may be complicated with peptic ulcer. Digestive derivation surgery remains the essential treatment for annular pancreas. We report the case of an 18-year-old female patient with duodenal stenosis caused by an annular pancreas. **Keywords**: Annular pancreas, Duodenal obstruction, Congenital

Öz

Anüler pankreas nadir bir konjenital anomalidir. Nüfusun 20.000'inde 1'de görülür. Annüler pankreas klinik olarak peptik ülser ile komplike olabilen sindirim darlığı olarak kendini gösterir. Sindirim sistemi cerrahisi, anüler pankreas için temel tedavi olmayı sürdürmektedir. Bu yazıda, duodenal darlığı olan ve anüler pankreasın neden olduğu 18 yaşında bir kadın hasta sunuldu.

Anahtar kelimeler: Anüler pankreas, Duodenal tıkanıklık, Konjenital

Introduction

Annular pancreas occurs in 1 in 20 000 of the population. It develops during the fifth week of embryonic development when two lobes of a bilobed ventral pancreatic bud migrate in opposite directions around the duodenum [1]. The anatomy of annular pancreas was first described in 1818 by Tiedman but its name was first attributed in 1862 by Ecker [2]. About two-thirds of cases with annular pancreas remain asymptomatic for life, but several complications such as acute pancreatitis, duodenal stenosis, peptic ulceration, and chronic pancreatitis have been associated with this anomaly [3]. We report the case of an 18-year-old female patient with duodenal obstruction caused by an annular pancreas.

Case presentation

An 18 year old female patient was admitted for late postprandial vomiting associated with an altered level of consciousness. She had a history of being hospitalized a year ago for dehydration caused by chronic vomiting disorders in a context of apyrexia and general condition retention. Clinical examination found a confused (Glasgow score= 13), dehydrated, afebrile patient with stunted growth, tachycardia (110 beats per minute). Abdominal examination revealed a non-distended, supple and painless abdomen. There was no palpable mass. Laboratory investigation revealed the following: hyponatremia (Na=115 mEq/ l), hypokalemia (K=1.6 mEq / l) and renal insufficiency (Blood urea nitrogen=1.89 g / l and creatinine=36 mg / l). Upper gastrointestinal fibroscopy showed significant stasis with the presence of impassable post bulbar stenosis which was dilated with a balloon dilator revealing a normal duodenal mucosa. Abdominal computed tomography (CT) scan showed the head of the pancreas surrounding the pyloric region, D1 and part of D2 causing stenosis without suspected digestive thickening with a gastric distension (Figure 1).

The patient was consenting to her surgical management after being informed of her illness. The surgical procedure consisted of an omega loop gastric bypass (gastrojejunal anastomosis). The postoperative course was unremarkable.

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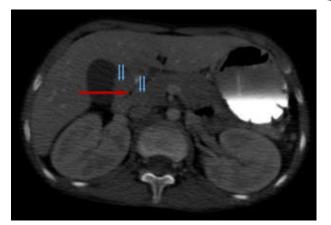


Figure 1: Blue arrow: CT image showing the head of the pancreas surrounding D2, Red arrow: CT image of the D2 $\,$

Discussion

Annular pancreas is a rare congenital anomaly. It is second the most common congenital anomaly of the pancreatic ducts after pancreas divisum. Its prevalence varies between 5-15 / 100,000 adults on an autopsy series, 1 in 250 on a retrograde endoscopic cholangiopancreatography study (ERCP). The prevalence rate between infants and adults is approximately 0.008% vs. 0.005%, respectively [4].

Many theories have sought to explain its embryological basis; however, no consensus has yet been reached regarding the exact mechanism of aberration. More recently, molecular investigations have shed light on some of these theories, confirming the origination of annular tissue from the ventral pancreatic bud. These studies highlight the role of the hedgehog signaling pathway in the development of this anomaly. Overexpression of the ventral-specific gene transmembrane 4 superfamily member 3 (tm4sf3) has also been associated with annular formation [5].

The ring of normal pancreatic tissue produces symptoms when it obstructs the duodenum. It has been estimated that only about 33% of the cases are symptomatic. 50% of patients present in the pediatric age group, 86% of these present in the neonatal period. In adults, annular pancreas usually presents between age 20 and 50 and is most commonly associated with abdominal pain and gastric outlet obstruction, secondary to duodenal stenosis. Additional presentations including pancreatitis, peptic ulcer disease and obstructive jaundice have been reported. Annular pancreas associated with a pancreatic tumor has also been reported. The diagnosis is usually made with computed tomography scanning and confirmed with upper gastrointestinal contrast fluoroscopy [6]. The imaging finding of pancreatic tissue with posterolateral extension to the duodenum in a patient with suspected chronic pancreatitis or gastric outlet obstruction or the finding of a crocodile jaw appearance of the pancreatic head should raise concern about the presence of annular pancreas [7].

Gastric bypass surgery remains the treatment of choice for annular pancreas: the duodenal ring must not be cut because it contains a pancreatic duct which complicated into a pancreatic fistula when ruptured. Duodenojejunostomy is most effective in treating duodenal stenosis which is sometimes associated peptic ulcer. Gastroenterostomy is more rarely proposed cephalic duodenopancreatectomy remains exceptional for this disease [8].

Conclusion

Annular pancreas is a rare congenital malformation. Its diagnosis is difficult to establish but it must be considered in adults with duodenal obstruction even in ulcer cases that do not respond to medical treatment, especially when it recurs despite the endoscopic dilation. The treatment of choice is surgery for annular pancreas.

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