Symptomatic Annular Pancreas in an Elderly Patient with Situs Inversus Totalis: Case Report

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Abstract

Annular pancreas is a rare congenital abnormality that is identified either incidentally or when individuals present symptomatically. It is known to be associated with other congenital anomalies. In this case report, we discuss a very rare case of an annular pancreas in an elderly male patient with known Situs Inversus Totalis (SIT). After presenting to the general surgical team with acute pancreatitis, an annular pancreas was identified incidentally on Computed Tomography (CT) imaging. He was managed conservatively and discharged with outpatient follow up. Patients with annular pancreas should always be considered as an important differential in adult patients with SIT. It remains integral for surgeons and radiologists to be aware of this rare association. We hereby discuss this patient, followed by a brief literature review about annular pancreas and its association to SIT.

Keywords: Annular pancreas; SIT; Computed tomography; Radiology

Introduction

SIT, also known as situs inversus with dextrocardia, refers to the complete inversion of the thoracic and abdominal viscera [1]. It is a rare congenital disorder, demonstrating an autosomal recessive pattern of inheritance [2]. Annular pancreas, also a rare congenital abnormality, occurs in less than 1% of the population [3]. It results as a consequence of an error during human embryogenesis, when there is abnormal or failed migration/rotation of the ventral pancreatic bud [3]. This results in a ring of pancreatic tissue either partially or completely encircling the second part of the duodenum [4]. The clinical manifestation of this abnormality may present at any age, however it is more common to ensue in early years of life [4]. Conditions associated with annular pancreas include duodenal atresia, and tracheoesophageal fistula, acute pancreatitis and peptic ulcer disease [3,4]. In this case report, we present an incredibly rare case of an adult patient with known SIT to have an annular pancreas and present with acute pancreatitis.

Case Presentation

A 70 year old gentleman presented into the emergency department with a two day history of sudden onset epigastria pain, radiating into the back. This pain was associated with nausea and vomiting. He also reported loose stools, and recent weight loss. He had a background of SIT, hypertension, type two diabetes mellitus and alcohol abuse. He was also known to have undergone an open cholecystectomy and splenectomy in 1997. His regular medications included metformin, furosemide and ramipril. His observations were stable on admission. On examination, he had clinical evidence of icterus. His abdomen was soft, and non-distended, however, there was tenderness of his epigastrium and left iliac fossa region. The remainder of his systems enquiry was unremarkable. The significant findings of his laboratory blood results were as follows: White cell count 13.5 × 10^9 cells/L (RR 4.3 to 11.2 cells/L), neutrophils 11.03 × 10^9 cells/L (RR 2.1 to 7.4 cells/L), CRP 2.8 mg/L (RR 0 to 5 mg/L). His LFT results: AST unavailable (hemolysed sample), alanine transaminase 213 U/L (RR 0 to 41 U/L), alkaline phosphatase 117 U/L (RR 30 to 130 U/L), gamma-glutamyl transferase 1008 U/L (RR 10 to 71 U/L), bilirubin 31 umol/L (RR 0 to 21 umol/L). The patient’s serum amylase was 195 U/L (RR 28 to 100 U/L).

Thoracic and abdominal CT scans were done, which demonstrated situs inversus (Figure 1). The inferior vena cava was noted to be on the left side, with the apex of the heart pointing to the right,
Annular pancreas was first identified by Tiedemann in 1818, and later given the name "pancreas annulare" in 1862 by Ecker [5,6]. It occurs as a result of impaired embryological migration of the ventral pancreatic bud, subsequently causing duodenal envelopment [7]. There have been many theories which attempt to postulate the origin of annular pancreas, with Leeco’s and Baldwin’s theories being more recognized. Leeco states that failed migration of the ventral bud is as a result of its adherence to the duodenal wall. Baldwin, however, postulates it occurs due to impaired movement of the bud [8]. 74% of cases involve D2 [8]. Although rare, its exact prevalence is difficult to ascertain as many adults may not present symptomatically. It is estimated to be 15 to 400 cases in 100,000 adults [9]. There is also no clear gender predilection for this anomaly that can be identified from the literature reviews [9]. More often, cases of annular pancreas are identified in neonates and infants in their first year of life [5]. In infancy, it presents predominantly with symptoms of gastric outlet obstruction such as intractable vomiting, bloating and feeding intolerance [4,5,9]. In adults however, approximately two-thirds are asymptomatic [8]. Symptoms are related to the degree of narrowing, and may not necessarily present with obstruction until there is significant annular inflammation to cause duodenal narrowing [4]. It can also present with epigastric pain, postprandial fullness and vomiting [5,10]. Annular pancreas has been associated with conditions such as peptic ulcer disease, pancreatitis, biliary obstruction with jaundice, pancreatic head carcinoma [5]. Due to annular pancreas being a risk factor for pancreas to biliary malignancy, our patient was discussed at the MDT to exclude this. This is integral as there have been eight cases of ampullary region carcinomas in patients with annular pancreas that have required resection from English literature [8]. In our case, the patient presented with acute pancreatitis, and an annular pancreas was incidentally discovered. A constant peristaltic wave of the proximal duodenum causes reflux of these contents into the pancreatic ducts, resulting in pancreatitis [10]. In this patient, he has a known background of SIT. The association between annular pancreases with acute pancreatitis is more established than that with SIT. There is very little published literature on the presentation of complications of annular pancreas in adult patients with SIT. There are however reports of congenital duodenal obstruction secondary to annular pancreas in young patients with situs inversus [11,12]. Therefore, this does suggest a link exists between SIT and annular pancreas, albeit a rare association. Annular pancreas is a radiological finding the various diagnostic imaging modalities: CT, MRCP, Endoscopic Retrograde Cholangiopancreatography (ERCP), may show pancreatic tissue enveloping the duodenum, in keeping with annular pancreas [4]. With these modern imaging devices, 1 in 250 cases are reported to have been found to have an annular pancreas [8]. No such statistical figures exist for patients with annular pancreas on a background of SIT, which remains of particular interest. The treatments for these patients are individualized as no guidelines currently exist for its management [9]. As with our patient, symptomatic acute pancreatitis should be managed with supportive care. Duodenal bypass surgery is reserved for those presenting with refractory duodenal or gastric obstruction, by performing a duodenoduodenostomy or gastrojejunostomy [9]. More complex procedures entail pancreaticoduodenectomy or hepaticojejunostomy; however this is less common [9].
Conclusion

Annular pancreas is a rare congenital abnormality, which is often associated with other congenital abnormalities. It more often presents during infancy however may also manifest during later life as a result of its complications. In individuals with known SIT presenting with signs and symptoms indicative of acute pancreatitis, annular pancreas should also be considered as a part of the differentials. Radiological investigations will identify this pancreatic abnormality, therefore it is important for radiologists to also be familiar with this association when reporting imaging findings. These patients are managed on an individual basis as no guidelines exist presently. As there is minimal literature on adults with SIT identified to have an annular pancreas, we emphasize the importance of surgeons and radiologists being aware of this rare association.

References