Annulling An Annulled Annular Pancreas: A Case Report

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ABSTRACT

Annular Pancreas (AP) is a rare congenital developmental anomaly which results from failure of the ventral bud to rotate to fuse with the dorsal bud, resulting in a ring of pancreatic parenchyma that surrounds the second portion of the duodenum. This rare entity can be partial or complete and the partial is even rarer. Most cases manifest in the infancy but may remain asymptomatic and various nonspecific clinical manifestations are seen in adults. We report a rare case of partial annular pancreas with atrophy of body and tail of pancreas which presented with upper gastrointestinal bleeding and obstruction. The challenge in this case was the annular pancreas was the only functioning pancreatic tissue.

KEYWORDS

Annular; Atrophy; Bleeding; Pancreas.

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INTRODUCTION

Annular Pancreas (AP) is a rare congenital developmental anomaly which results from failure of the ventral bud to rotate during the 7th week of gestation to fuse with the dorsal bud, which results in a ring of pancreatic parenchyma that surrounds the second portion of the duodenum.1 Tiedeman first reported this anomaly in 1818 and the term annular pancreas was coined by Ecker in 1862.2,3 It usually affects approximately one in 20000 live births.3 The reported incidence in adults is 0.050-0.015%, but the exact prevalence is unknown as the majority of patients are asymptomatic.4 At times it is associated with other congenital anomalies like Down's syndrome, tracheoesophageal fistula, intestinal atresia, pancreas divisum and pancreaticobiliary malrotation.5

Incomplete or partial AP is a very rare entity with unknown exact prevalence.6 Only few cases of incomplete AP have been reported and in contrast to complete AP, the patients with incomplete or partial AP can be totally asymptomatic.6 We report a case of a rare partial AP with atrophy of body and tail of pancreas in an adult who developed severe symptoms and ultimately required surgical management.

CASE REPORT

A 26 years gentleman presented to outpatient clinic with complaints of melena for four days, vomiting and pain abdomen for two days. He was in his usual condition of health four days back when he noticed black colored foul-smelling stool and this melena was followed by multiple episodes of bilious vomiting. He also had food particles consumed a couple of days back in his vomitus. This bilious vomiting was associated with dull aching epigastric pain without radiation and his pain was relieved after vomiting. He didn't give history of weight loss, anorexia, jaundice, palpitation, shortness of breath, altered sensorium or abnormal body movements. He doesn't have history of chronic comorbid conditions. He is a social alcohol consumer and has never smoked in his life. He is married and sexually promiscuous individual with healthy sleeping habit. He gives similar episode of pain abdomen and vomiting around six months back and was diagnosed as having AP with stricture of his second part of his duodenum for which he underwent balloon dilatation for the stricture of the duodenum at another center. He was symptomatically better after the procedure.

At presentation he was tachycardiac while his blood pressure was normal. He was pale and his abdomen was soft, non-tender and normal bowel sounds were heard. He had melena in digital rectal examination. He was resuscitated with two wide bore cannulas, Ryle's tube aspiration, foley catheterization and various pertinent diagnostic investigations were done. He had hemoglobin of 8.9gm/dl with normal serum amylase and lipase and contrast enhanced Computed Tomography of abdomen revealed that second part of duodenum was partially encased by pancreatic tissue (more than 180-degree circumference) compressing and narrowing the its lumen, resulting in dilatation of stomach (Figure 1) while pancreatic body and tail were absent (Figure 2). Hence the diagnosis Partial annular pancreas with atrophy of body and tail of pancreas causing stricture of D2 leading to GOO was made. Furthermore, the upper GI Endoscopy revealed that the pylorus was deformed with scarred are in first part of duodenum and the mucosa of the second part of duodenum was friable with a small ulcer with stigmata of recent bleeding (the cause of melena) and the scope couldn't be negotiated further.

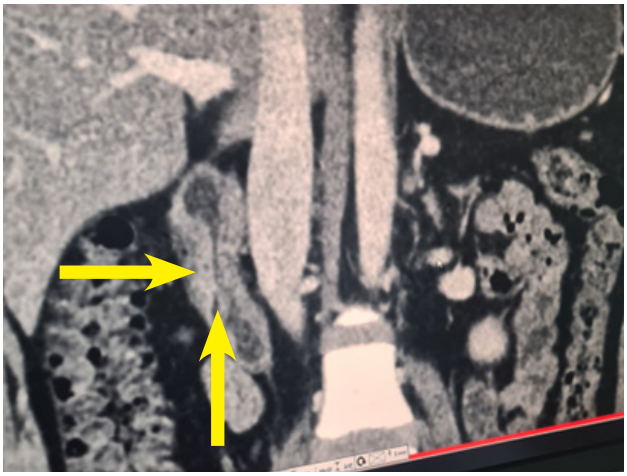
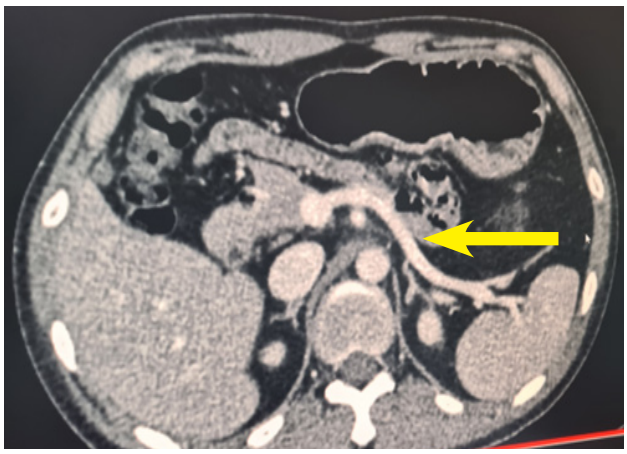


Figure 1 Arrow shows that the second part of duodenum is partially encased by pancreatic tissue compressing and narrowing the lumen.



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After stabilization the patient underwent Exploratory Laparotomy which confirmed the radiological findings. The patient had partial AP with stricture of second part of duodenum with atrophy of body and tail of pancreas. The exploration was followed by D2 stricturoplasty and Loop Gastrojejunostomy with Braun's Jejuno-jejunostomy. The gentleman recovered in a week and is asymptomatic nine months after the surgery.

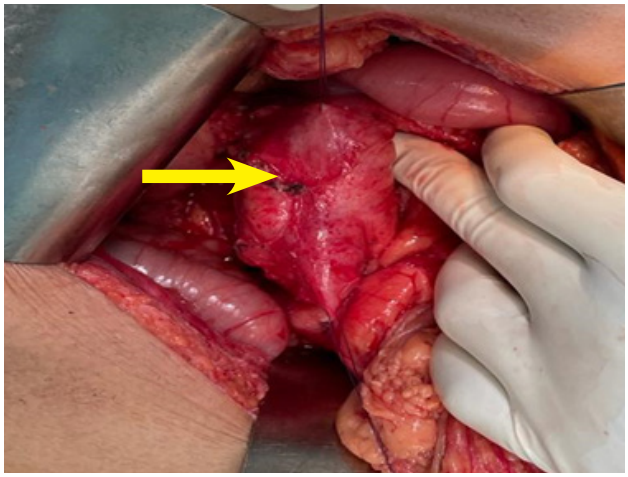

Figure 3 Partial annular pancreas with stricture of second part of pancreas

DISCUSSION

Annular pancreas (AP) in adults is a rare embryologic abnormality detected after development of complications or as incidental finding.4 Most cases are diagnosed in neonatal period or infancy but in a few cases the condition may be found later in life where the symptoms are directly related to the degree of duodenal obstruction.3,7 In some cases, the obstruction is not significant until inflammation of the annulus narrows the duodenum, while in other cases the diagnosis is incidentally made when the patient is being evaluated for symptoms of pancreatitis or peptic ulcer disease.4 Drey has classified it according to age of presentation into neonatal, pediatric, adult asymptomatic and adult symptomatic annular pancreas.8

Most of the symptomatic individuals aged 20 to 60 years show symptoms of upper GI obstruction.9 The associated complications of AP include peptic ulceration with bleeding, acute or chronic pancreatitis, biliary obstruction and rarely malignancy.9

The initial diagnostic modality for AP is ultrasonography or plain abdominal x ray which reveals the classic double bubble sign in case of duodenal atresia associated with AP.4 Computed tomography (CT) or Magnetic resonance (MR) imaging both are used for diagnosis of AP and both reveal pancreatic tissue encircling the duodenum.10 Sandrasegaran et al. have described different configurations of pancreatic tissue around the second part of the duodenum.11 A crocodile jaw configuration, where the pancreatic head is found anterior and posterior to the second part of the duodenum, or the presence of pancreatic tissue posterolateral to the duodenum, is thought to be highly suggestive of annular pancreas.11 MR is a better imaging modality compared to CT due to its high signal intensity on T1 fat-suppressed imaging showing pancreatic tissue surrounding the duodenum with similar findings on T2-weighted imaging including magnetic resonance cholangiopancreatography.12 For preoperative diagnosis of AP endoscopic retrograde cholangiopancreatography(ERCP) is considered the gold standard despite the risk of acute pancreatitis or inaccessibility due to some sort of duodenal stenosis.13

The management of AP depends upon the clinical spectrum in which the patient presents to us. While acute pancreatitis is managed conservatively; various by pass techniques like duodeno-duodenostomy, duodenojejunostomy and gastrojejunostomy are required in cases of outlet obstruction.1,4,10 The resection of the annulus is not recommended as it has been associated with various complications like pancreatic fistula, pancreatitis, and incomplete relief of duodenal obstruction.14 Pancreatic resection is reserved for cases where suspicion of malignancy is unavoidable due to its adverse effects like pancreatitis, pancreatic fistula formation, or incomplete obstruction relief.4,15

CONCLUSION

Annular pancreas is a rare entity and even rare is partial annular pancreas. The annular pancreas associate with atrophy of body and tail is itself even more rare and to our knowledge is the first reported case. The management of this entity should be carefully planned as the annular pancreas is the only functioning pancreatic tissue in the patient and the procedure planned should incur minimal insult to the pancreas. The AP should be suspected in patients presenting with unusual epigastric pain with features of upper GI obstruction after ruling out common entities and should be dealt with accordingly.

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