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RESEARCH ARTICLE

ANNULAR PANCREAS DIVISUMTREATED WITH WHIPPLE PROCEDURE: A CASE REPORTAND UPDATE OF ARARE CONDITION

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Abstract

There are two distinct congenital anomalies of the pancreas—annular pancreas and pancreas divisum—both of which have the potential to manifest in infancy with symptoms that are taken to indicate duodenal blockage. However, symptoms such as stomach discomfort, postprandial fullness, and pancreatitis may not appear until maturity. An annular pancreatic divisum that required duodenopancreatectomyin a 32-year-old woman with a history of hyperthyroidism treated with carbimazole was admitted in our department. The MRCP was important in making the accurate diagnosis. This article describes the symptoms and potential treatments for annular pancreas divisum.

Introduction:-

Three out of every 20,000 autopsies are reported to show an annular pancreas [1]. There is either a partial or whole encirclement of the duodenum, making it impossible for digestive juices to pass through. According to Ravith and Woods [1], annular pancreas is the only real congenital defect of the gastrointestinal system that is seldom diagnosed in infants and young children.

Two percent to ten percent of the population suffer with pancreatic divisum [2-3]. This occurs when the ventral and dorsal pancreas fail to fuse between week 6 and 7 of embryogenesis [4], resulting in the pancreas draining into two separate ducts.

Only five occurrences of a patient with both diagnoses have been documented in the English literature [5-7]. We discuss the case of a patient with non-typical symptoms who was diagnosed with annular pancreas and treated with the Whipple technique, whereby the pancreas was divided.

Case Presentation

A 32-year-old female with history of hyperthyroidism treated with carbimazole presented to the emergency department of King Fahad General Hospital in Jeddah, complaining of persistent jaundice for 3 weeks, choluria and hitching. Associated symptoms of 15Kg of weight loss in 1 month and postprandial fullness also in the last month. No history of vomiting or fever. She described isolated episodes of mild abdominal pain in the epigastric area

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associated with meals since childhood. On physical examination, she was vitally normal, mild epigastric pain and clinically jaundice. Laboratory results are presented in table 1.

Table 1:- Laboratory results of the case.

Laboratory test	Value
Hemoglobin	11.3 g/dl
Lipase	45 U/L
Amylase	62 U/L
Total bilirubin	16.3 mg/dl
Direct bilirubin	13.2 mg/dl
Alkaline phosphatase (ALP)	227 U/L
Gamma-glutamyl transferase (GGT)	19 U/L
Aspartate Transferase (AST	59 U/L
Albumin	2.6 g/dl
CA19-9	24.8 U/ml
CA 125	17.4 U/ml
Alpha fetoprotein	4.2 Ng/ml
Hepatitis B serology	Negative

EGD evaluation showed D1, and D2 obstruction. ERCP couldn´t reach the ampulla because of a narrowing at the second part of the duodenum. CT showed appearance of annular pancreas encasing the second part of duodenum with moderate luminal narrowing. MRCP resulted with no biliary duct dilatation or filing defect, complete annular pancreas causing luminal narrowing of the second part of the duodenum and pancreas divisum showing two separated ducts (fig 1 and 2). Patient underwent for Whipple procedure (fig 3) to resolve duodenum and biliary obstruction; reconstruction was done with pancreatojejunostomy duct-to-mucosa, hepaticojejunostomy and gastrojejunostomy in single loop (fig 4), time of operation was 4,5 hours and blood loss was minimal. Histopathology of specimen showed pancreatic tissue surrounding the 2nd part of the duodenum, and there are two pancreatic ducts draining separately into the second part of the duodenum.

Outcome and follow up: close follow up after discharge showed no complications. Fife months after surgery patient presented with epigastric pain, nausea and vomiting, she was going through the first month of pregnancy, upper gastrointestinal endoscopy was done and showed gradeAesophagitis.

Discussion:-

Normal pancreatic duct development

At about 5 weeks of gestation, the dorsal and ventral anlages of the embryologic foregut fuse to produce the pancreas. The hepatobiliary system develops from the ventral system as well. The ventral pancreas moves behind the duodenum and settles below the dorsal pancreas's head at about 7 weeks into gestation. When fully developed, the dorsal bud serves as the backbone and the tail, while the ventral bud produces the underdeveloped head of the pancreas and the uncinate process. The major pancreatic duct develops when the ductular networks of the two pancreatic bud precursors fuse. The dorsal pancreatic duct gives rise to the auxiliary pancreatic duct (of Santorini) just distal to the point of fusion. When the ventral bud fails to rotate with the duodenum, the duodenum becomes encased in the pancreas, leading to the development of pancreas divisum, and when this fusion fails, annular pancreas occurs.

Therefor annular pancreas and pancreatic divisum comes under pancreatic congenital abnormality. Compared to annular pancreas, pancreatic divisum accounts for the vast majority of occurrences (about 10%). Previous research has shown a 30–38 percent overlap between the two disorders. However, results from a bigger research conducted recently revealed that the true rate may be as high as 50% [8-9]. As a result, the majority of instances are not discovered until imaging or autopsies are performed on deceased individuals, long after the patient has shown no signs of illness. Abdominal discomfort and gastric outlet blockage are the most prevalent symptoms, and they are most often experienced by individuals between the ages of 20 and 50 [10]. The estimated incidence of AP is at 1 in 1000, a number which has grown slightly with the advent of new imaging methods [11].

Clinical presentation:
Duodenal blockage, stenosis, and atresia may manifest in children, however they are usually linked to another congenital defect. Abdominal discomfort, fullness after eating, vomiting, acute or chronic pancreatitis, upper gastrointestinal hemorrhage due to peptic ulcer disease, and biliary obstruction are the most frequent first complaints in adults.

Radiology
In an annular pancreas, pancreatic tissue entirely or partially encircles the second portion of the duodenum, as shown on a CT scan. The proximal duodenum may also be dilated and narrowed in conjunction with this condition. This condition is often linked to pancreatitis in adults. The ductal structure of the pancreas, in addition to the annular characteristics of the pancreas, may be evaluated well using MR imaging thanks to MRCP. Typically, the annular duct connects to either the primary pancreatic duct or the auxiliary duct (duct of Santorini).

Diagnosing pancreas divisum has traditionally been done using endoscopic retrograde cholangiopancreatography (ERCP), although CT scan has the potential to help reveal changes in pancreatic ductal structure but has a poor sensitivity. According to studies, EUS has a high sensitivity for diagnosing pancreas divisum, anything from 87% to 95% [9]. Magnetic resonance imaging (MRI) is the standard of care today [12]. Imaging highlights include a direct connection between the dorsal pancreatic duct and the duct of Santorini, both of which empty into the minor papilla. In contrast to the dorsal duct, the ventral duct (duct of Wirsung) connects to the major papilla through the distal bile duct.

Management
The diagnosis and therapy of patients with both annular pancreas and pancreatic divisum lack clear standards. Symptoms, their length, their severity, and the occurrence of complications all play a role in determining the best course of therapy. Conservative treatment might range from medication to surgical intervention, such as duodenal bypass. In addition to its use in the treatment of malignant tumors, pancreaticoduodenectomy is also used to treat benign conditions. Imaging analysis led us to conclude that our patient was experiencing obstructive jaundice due to a case of annular pancreatic divisum. To reduce the severity of the patient's symptoms and increase their overall quality of life, pancreaticoduodenectomy is advised. Pre-procedure evaluation of perioperative treatment with the purpose of identifying potential morbidities and reducing hospital stay is recommended. Depending on the severity of the patient's disease, a surgeon may decide to perform by-pass techniques for annular pancreas or Whipple procedure for chronic pancreatitis in pancreas divisum.

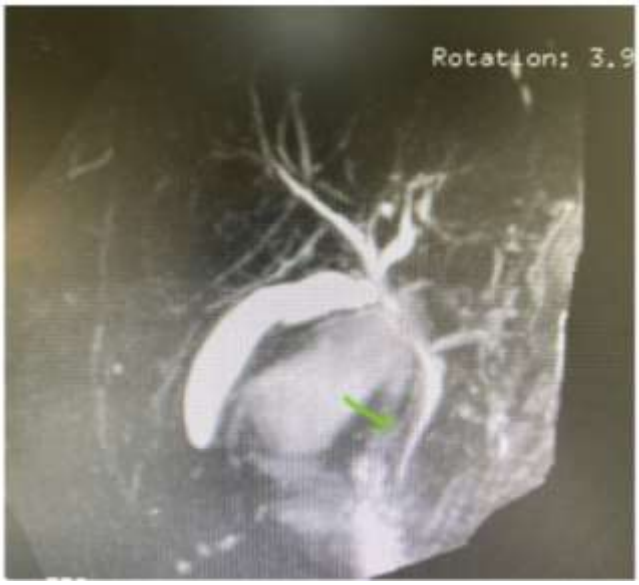

Figure 1:- MRCP showing pancreas divisum (arrow).

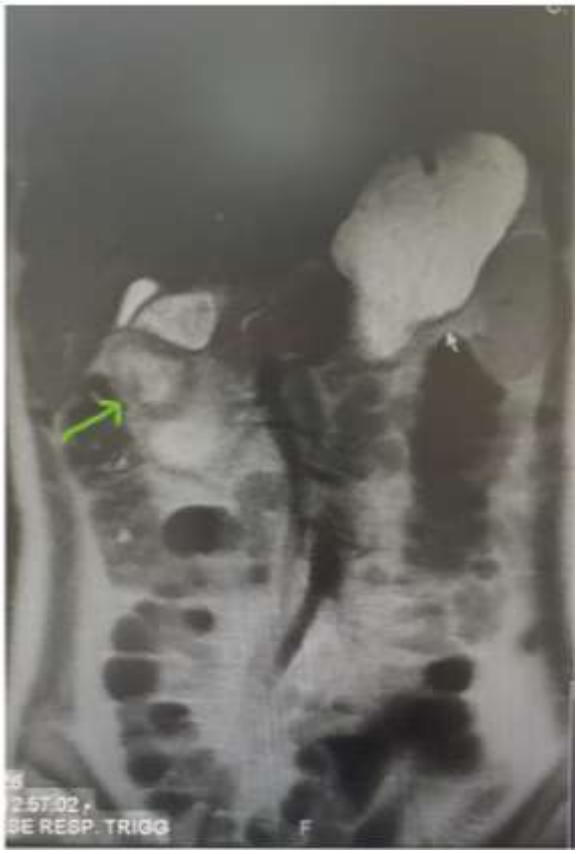

Figure 2:- MRI showing anular pancreas (arrow).

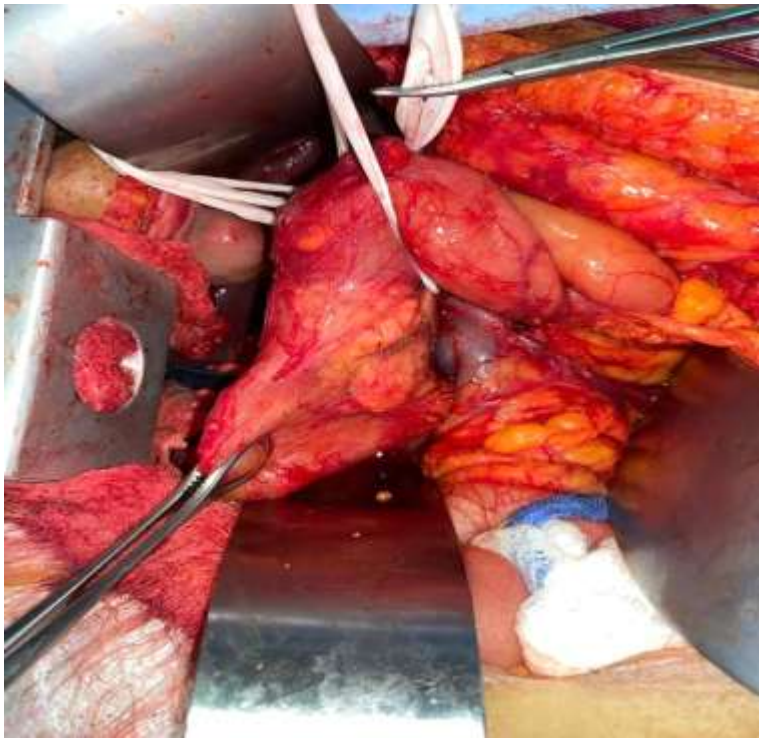

Figure 3:- Intraoperative finding.

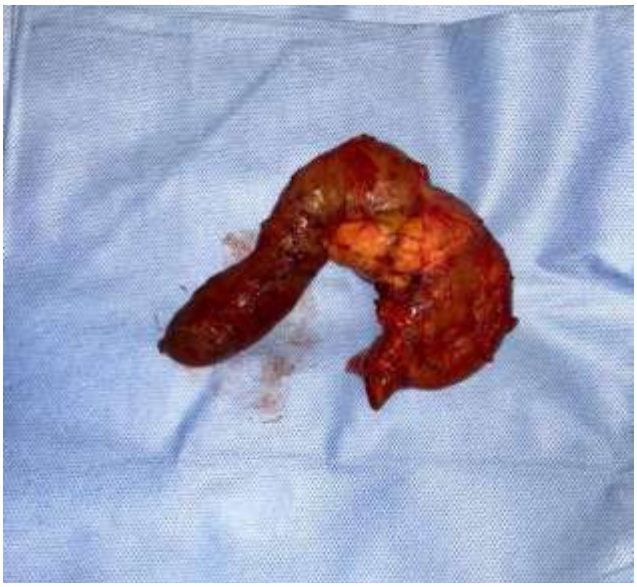

Figure 4:- Specim.

Conclusion:-

There are two separate congenital defects of the pancreas: annular pancreas and pancreasdivisum. Both have the potential to present in infancy with symptoms that are interpreted as duodenal obstruction. However, stomach pain, post-meal fullness, and pancreatitis may not manifest until adulthood. The MRCP was essential for establishing a correct diagnosis. In our case we performed Whipple procedure with successful outcome to treat the condition of annular and pancreas divisum. With developments in imaging modalities and a higher index of suspicion, more instances of symptomatic and asymptomatic annular pancreas in adults may be detected. Most studies of these lesions are single case reports or limited series, which do not let a single surgeon to gain substantial expertise; hence, the care of this congenital defect should be tailored to the accompanying comorbidities.

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