

Case Report

Dorsal agenesis of the pancreas associated with mixed carcinoma: a rare case report and review of literature

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ABSTRACT

Dorsal agenesis of the pancreas is a very rare congenital anomaly with less than 100 cases reported in the world literature till 2020. Dorsal agenesis associated with pancreatic tumors is extremely rare; only 17 cases have been reported to date. Here we reported a rare case of dorsal agenesis of the pancreas associated with mixed carcinoma of the pancreas.

Keywords: Dorsal agenesis of the pancreas, Mixed carcinoma, Pancreatic tumors, Congenital anomalies of pancreas

INTRODUCTION

Dorsal agenesis of the pancreas is a very rare congenital anomaly that occurs due to the failure of the dorsal pancreatic bud to form the body and tail of the pancreas. The exact prevalence is not known due to its rare condition. To date, only 100 cases of dorsal agenesis of the pancreas have been reported in the world literature.¹ Dorsal agenesis of the pancreas associated with pancreatic tumors is extremely rare; only 17 cases have been reported till now.² Here we report probably the first case of dorsal agenesis of the pancreas associated with mixed carcinoma of the head of the pancreas.

CASE REPORT

A 62-year-old woman with a history of recently detected uncontrolled diabetes presented with a history of jaundice, and abdominal pain for the one-month duration. She also had a history of loss of weight and appetite. She had no history of clinical features suggestive of exocrine pancreatic insufficiency. Apart from diabetes, she had no

previous medical or surgical history. She had a poor performance status of ECOG 3. On physical examination, she had icterus, pedal edema, and palpable gall bladder. Laboratory investigations showed anemia (Hb=8 g/dl), normal leukocyte and platelet count, elevated serum bilirubin levels of total bilirubin/direct: 10.2/9.3 mg/dl, serum alkaline phosphatase level of 449 IU/l, SGOT/SGPT are within normal range, blood urea 37 mg/dl and serum creatinine 1.7 mg/dl, serum album level of 1.8 g/dl, random blood glucose levels of 290 mg/dl with urine acetone positive and her glycated hemoglobin (HbA1c) was 11.2%. Serum amylase and lipase were within normal range. Ultrasound of the abdomen revealed dilated proximal biliary system with narrowing at the distal end of CBD.

After stabilizing her diabetes, hemoglobin, and renal status CECT abdomen was done which revealed non visualization of the body and tail of the pancreas with 1.9×1.3 cm hypodense lesion in the periaampullary region with gross upstream dilatation of CBD (13 mm), CHD (16 mm), and IHBR, MPD (3 mm) (Figure 1). She was

subjected to an MRI abdomen with MRCP which revealed a similar finding as CT along with non-visualization of the pancreatic duct in the body and tail (Figure 2). Her upper GI scopy was normal and serum CA 19.9 was 160 U/ml. Surgery in the form of a total pancreatectomy was planned and discussed in a multidisciplinary tumor board. Because of her poor performance status and failed endoscopic and percutaneous intervention for biliary drainage, it was decided to do a biliodigestive bypass followed by palliative chemotherapy. Intraoperatively there was a hard mass of approx. 1.5×1.5 cm was noted in the head of the pancreas with an absent body and tail of the pancreas (Figure 3). FNAC was taken from the mass lesion and it was positive for malignancy, mixed carcinoma (acinar cell and ductal carcinoma). We did cholecystojejunostomy with gastrojejunostomy and jejunojunctionostomy and postop course was uneventful.

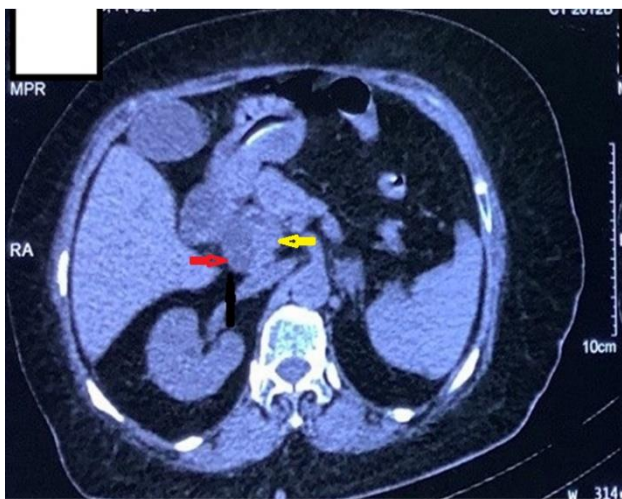


Figure 1: Computed tomography image showing-dilated common bile duct (red arrow), and mass in the head of the pancreas (yellow arrow).

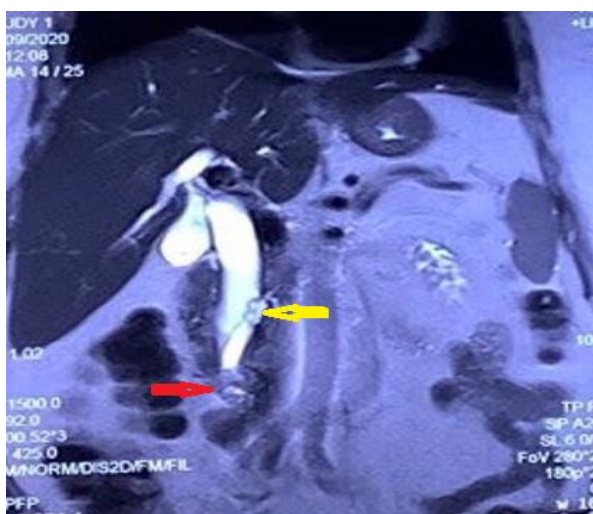


Figure 2: Magnetic resonance imaging showing-absent pancreatic duct in the body and tail of the pancreas (red arrow), and abrupt cutoff at the distal common bile duct.

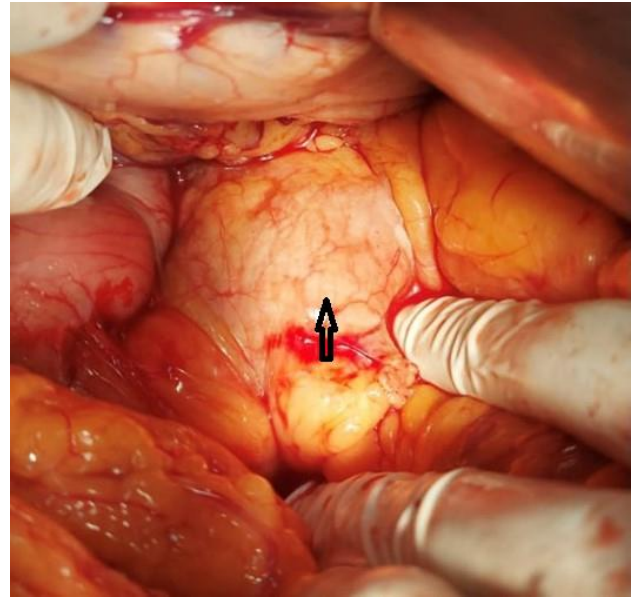


Figure 3: Intra-operative picture showing a mass in the head of the pancreas (black arrow).

DISCUSSION

The pancreas develops from ventral (caudal) and dorsal (cranial) buds originating from the endodermal lining of the duodenum at the junction of the foregut and midgut during the fourth week of gestation.³ During the second month of development, the ventral pancreatic bud migrates dorsally, coming to lie posteroinferior to the dorsal bud and fusing with it.⁴

The ventral bud forms the inferior head and uncinate process of the pancreas and the dorsal bud forms the superior part of the head, body, and tail.⁵ Failure of the dorsal pancreatic bud to form the body and tail of the pancreas results in dorsal agenesis of the pancreas.⁵ This rare entity was first described in 1911 by Heiberg during an autopsy in a patient with diabetes mellitus.⁶ So far around 100 cases of dorsal agenesis of the pancreas and 17 cases of pancreatic tumors associated with this anomaly have been reported in the literature till the year 2020.⁷ The majority of patients with dorsal agenesis of the pancreas are asymptomatic and discovered incidentally, some patients may develop associated clinical manifestations like abdominal pain, pancreatitis, diabetes, bile duct obstruction, compensatory hypertrophy of the ventral part of the pancreas, and rarely tumours.^{8,9} Because islet cells are predominantly located in the tail of the pancreas approximately 50% of the patients with this anomaly may have diabetes as in our patient.^{9,10}

A combined imaging modality with ultrasound, CT, and MRCP helps in the diagnosis of dorsal agenesis but confirmation is by demonstration of the absence of pancreatic duct in the body and tail by MRCP/ERCP.^{9,10} The dependent stomach and intestine signs are hallmarks of dorsal pancreatic agenesis.¹¹ In our case CT helped in the diagnosis of the pancreatic tumor with dorsal pancreas

ventral to the splenic vessels is not seen and stomach and small bowel loops occupying the distal pancreatic bed. We confirmed by MRCP of the absent dorsal duct. The association between dorsal agenesis and the pancreatic tumor is not well studied and not clear, some hypothesize dorsal agenesis increases the risk of chronic pancreatitis, which in itself is a risk factor for pancreatic tumors.¹²

In the literature, till the year 2020, only 17 cases of pancreatic tumors associated with dorsal agenesis of the pancreas have been reported.¹² Among them, the most common is ductal adenocarcinoma in 12 cases, two neuroendocrine tumors, and three of them precancerous.¹²⁻¹⁴ The most common type of pancreatic malignancy is pancreatic ductal adenocarcinoma (PDAC) accounting for about 90% of pancreatic neoplasm.¹⁵ Acinar cell carcinoma of the pancreas (ACC) is rare accounting for 1-2% of all pancreatic cancer and mixed acinar and ductal carcinoma is even rare as only a few case reports have been reported in the literature.¹⁶ We reported probably the 18th case of dorsal agenesis associated with a tumor and it is the first case report of mixed carcinoma (acinar cell and ductal carcinoma) in the literature.

The treatment of pancreatic malignancy associated with dorsal agenesis is total pancreatectomy with lifelong pancreatic exocrine enzyme and insulin replacement therapy. In our case since the patient's poor general condition, we did a biliodigestive bypass and subjected her to palliative chemotherapy. The limitation in this case report is pathological diagnosis is based on FNAC and not the complete surgical specimen as pathological diagnosis of mixed carcinoma requires at least 25% of ductal morphology in the tumor with a predominant acinar cell component.

CONCLUSION

Here we reported a probable 18th case of dorsal agenesis of the pancreas associated with a pancreatic tumor which is the first case with mixed carcinoma (acinar cell and ductal components). The association between dorsal agenesis of the pancreas and pancreatic tumor is not clear but should have a high index of suspicion when patients with dorsal agenesis present with symptoms and total pancreatectomy is the treatment of choice with lifelong pancreatic enzyme and insulin replacement therapy.

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