Agenesis of Dorsal Pancreas Associated with Periampullary Pancreaticobiliary Type Adenocarcinoma

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ABSTRACT

Context Agenesis of the dorsal pancreas is one of the rare congenital malformations of pancreas. The association of agenesis of the dorsal pancreas with pancreatic tumors is extremely rare and only around 9 cases being reported till date. Case report We report a case of a fifty one year old woman with an agenesis of the dorsal pancreas with periampullary pancreaticobiliary adenocarcinoma. She presented with features of obstructive jaundice without pain abdomen or fever. Laboratory data showed hyperbilirubinemia, raised alkaline phosphatase and impaired glucose tolerance. Ultrasound abdomen showed periampullary mass. MRI abdomen and MRCP demonstrated dorsal agenesis of the pancreas, dilated intra and extra hepatic bile ducts with narrowing of distal CBD with periampullary mass. Pancreatic tumor was considered as preoperative diagnosis, and pancreaticoduodenectomy was performed. Histopathology confirmed pancreaticobiliary type of adenocarcinoma. Conclusion A rare case of dorsal agenesis of the pancreas with periampullary pancreaticobiliary type of adenocarcinoma was presented. Therefore this case therefore merits reference as a rare clinical presentation.

CASE REPORT

Agenesis of Dorsal Pancreas

INTRODUCTION

Agenesis of dorsal pancreas is a rare congenital malformation, resulting from failure of development of the dorsal pancreatic bud in the fetus [1]. This leads to absence of body and tail of the pancreas. The pancreas development begins during fourth week of gestation from the two endoderm buds of the foregut, ventral and dorsal bud. Ventral bud forms the head and uncinate process of pancreas. Dorsal bud forms the upper part of head, isthmus, body and tail of the pancreas. Both the ventral and dorsal bud fuse to form the complete pancreas by sixth to seventh week of gestation [1]. The duct of ventral bud fuses with the duct of dorsal pancreatic bud to form duct of Wirsung (main pancreatic duct) and the remaining proximal duct of the dorsal bud forms the duct of Santorini (accessory duct). Agenesis of dorsal pancreas is a very rare condition; only around 55 cases have been reported in the literature from 1913 to 2013 [2]. Because of its rarity, the exact incidence of ADP is not known [2]. The existence of ADP was first described in 1911 in an autopsy study and was associated with diabetes mellitus [3]. The association of ADP with pancreatic tumors is extremely rare and only around 9 cases being reported till date. With this background we report a case of agenesis of dorsal pancreas with periampullary carcinoma.

CASE SUMMARY

A fifty-one-year-old female, presented with a history of jaundice, generalized itching, and weight loss of one month duration, without any history of pain abdomen, fever, nausea, vomiting or steatorrhea. She was a known hypertensive for last 2 years on medication, without any significant past history of diabetes mellitus, pancreatitis or malignancies. There was no history of smoking or alcohol intake. Her family history was not significant. Physical examination revealed icterus, pallor and blood pressure of 140/84 mmHg. Abdominal examination did not reveal any significant findings. Laboratory investigation showed: total bilirubin 13.4 mg/dL (reference range: 0.2-1.3 mg/dL), conjugated bilirubin 10.77 mg/dL (0-0.3 mg/dL), unconjugated bilirubin 2.33 mg/dL (0-1.1 mg/dL), alkaline phosphatase 396 IU/L (38-126 U/L), AST 85 IU/L (17-59 U/L), ALT 74 IU/L (21-72 U/L), serum albumin 3.6 g/dL (3.5-5 g/dL), prothrombin time 15.9 sec (12-16 sec), INR 1.3 (1-1.5) and haemoglobin 10.2 g/dL (11-14 g/dL). She had impaired glucose tolerance, with HbA1c of 6.3%.

An USG abdomen showed mild hepatomegaly with extrahaepatic biliary dilatation and periampullary mass. MRI and MRCP abdomen revealed dilated intra and extra hepatic bile ducts with narrowing of distal CBD with periampullary mass (Figures 1 and 2). MRI abdomen also showed hypoplasia of body and tail of the pancreas suggestive of agenesis of dorsal pancreas (Figure 2). ERCP was done and put biliary stent.

Patient underwent surgery, where a large mass of size 3 cm was found in the periampullary region with absence of body and tail of the pancreas confirming the diagnosis of agenesis of dorsal pancreas. A total pancreatectomy, duodenectomy, Roux-en-Y gastrojejunostomy, hepaticojejunostomy, and...
cholecystectomy were performed. On gross examination, a large ulceroinfiltrative lesion (3.0 × 2.5 × 2.0 cm) involving mostly periampullary region was found which was firm in consistency, pale greyish white in appearance with ill defined edges appearing to invade almost full thickness of the duodenal wall extending onto subserosa (Figure 3). Histopathological examination revealed invasive poorly differentiated adenocarcinoma with diffuse deep infiltration involving almost full thickness of the duodenal wall and invading subserosa (Figure 4a). Immunohistochemistry was done and the tumor cells were positive for immunohistochemical markers CA 19-9 (Figure 4b), cytokeratin 7, KI-67 (Figure 4c) and negative for CEA, cytokeratin-20. No lymph node metastasis was found.

Post operatively patient developed hyperglycemia and she was managed with insulin and advised adjuvant chemotherapy.

DISCUSSION

Agenesis of dorsal pancreas is a very rare congenital pancreatic malformation [4]. Complete agenesis of pancreas is not compatible with life. In the literature only around 55 cases have been reported in the last 100
ADP is due to developmental failure of dorsal pancreatic bud during embryogenesis. Exact cause of agenesis of the dorsal pancreas is unknown, but it may be due to primary dysgenesis of the dorsal pancreatic bud and lack of blood supply during pancreas development are the possible mechanisms [6]. Familial occurrence has been reported and this is suggestive of genetic predisposition. Exact genetic factors in humans have not been detected. However in mice studies, mice deficient in homeobox gene H1xb9 [7] and mutation in Raldh-2 (retinaldehyde dehydrogenase 2) or deficiency of retinoic acid leads to gene

Patients with ADP are mostly asymptomatic and detected incidentally during an evaluation for an unrelated cause [5] ADP presents with various clinical manifestations like abdominal pain, pancreatitis, diabetes mellitus and exocrine pancreatic body, accessory papilla, terminal end of the duct of Santorini is present and only the tail of the pancreas is absent [5].

ADP may be complete or partial, partial ADP is more frequent. In complete ADP, body, tail of the pancreas and duct of Santorini are absent whereas in partial ADP pancreatic body, accessory papilla, terminal end of the duct of Santorini is present and only the tail of the pancreas is absent [5].

CONCLUSION

Agenesis of dorsal pancreas is a very rare pancreatic congenital anomaly detected incidentally or with various clinical manifestations. With the advent of newer imaging techniques this rare congenital anomaly is being reported increasingly. Association of ADP with periampullary carcinoma and pancreatic cancer is very, but it is a serious condition. Therefore when a case of ADP is detected, we should also keep in mind the possibility of coexistence of pancreatic tumors, because these patients needs complete pancreatectomy and will develop diabetes mellitus requiring lifelong insulin therapy.

Conflict of Interest

The authors did not report any potential conflicts of interest.
References


