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Case Report

Pancreatic Insulinoma Associated with Stenosis of the Celiac Axis Due to Median Arcuate Ligament Compression

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ABSTRACT: Median arcuate ligament syndrome (MALS) is a rare stenotic vascular disorder caused by fibrous bands connecting the left and right crus of the diaphragm. A 42-year-old man was admitted to our hospital. Computed tomography revealed 20-mm mass at the tail of pancreas. He was diagnosed as having insulinoma. Angiography showed increased blood flow from dorsal pancreatic artery to insulinoma. Enhanced computed tomography revealed stenosis in the celiac artery. Thus, he was suspected to be suffering from MALS. MALS should be considered when angiography of area surrounding the pancreas reveals blood flow different from that observed in normal anatomy.

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KEYWORDS: median arcuate ligament syndrome, insulinoma

Median arcuate ligament syndrome (MALS) is a rare disorder caused by compression of celiac artery by the median arcuate ligament. MALS was first described in 1963.[1] MALS is a rare vascular disorder caused by fibrous bands connecting the left and right crus of the diaphragm. Clinical symptoms are postprandial abdominal pain, vomiting, and weight loss. We herein report a case of MALS in an insulinoma patient.

Case Report

A 42-year-old man complaining of hypoglycemia was admitted to our hospital. On examination, his height, body weight, and body mass index were 165 cm, 62.8 kg, and 23.1, respectively. There was no abdominal pain or vomiting. His abdomen was soft with no tenderness, palpable mass, or organomegaly.

Laboratory data were as follows: serum glucose level 41 mg/dl, HbA1c 4.0%, and plasma insulin 7.7 μU/mL. He had no significant medical history. He had never received insulin injection or medicine such as sulfonylurea compounds. Enhanced computed tomography revealed a 20-mm mass at the tail of pancreas (Fig. 1). He underwent a 72-h fast protocol that was terminated because of hypoglycemia. Finally, he was diagnosed as having insulinoma.

To determine the operability and location of the insulinoma, angiography and selective arterial calcium injection (SACI) test were performed. SACI test gave positive results for superior mesenteric artery (SMA). Angiography showed that blood flow in splenic artery was decreased. In addition, blood flow in SMA was increased. On the other hand, increase in blood flow in SMA was found from the dorsal pancreatic artery to insulinoma (Fig. 2A). In inferior

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pancreaticoduodenal artery (IPDA), retrograde blood flow was found from SMA to liver (Fig. 2B). Enhanced computed tomography revealed stenosis in the celiac artery (Fig. 3). Thus, he was diagnosed as suffering from MALS. Second SACI test gave positive results for dorsal pancreatic artery, but negative results for IPDA. Hence, the location of the insulinoma was determined. Laparoscopic spleen preserving distal pancreatectomy was successfully performed on the 97th day. Splenic artery and vein were divided and preserved (Fig. 4). Pathologic evaluation of the resected tissue revealed a well-differentiated tumor with a distinct border surrounding the exocrine pancreatic tissue. Immunohistochemical evaluation showed that the cells were positive for CD56, synaptophysin, chromogranin-A, and insulin.

On post-operative day 10, the patient was found to have recovered without any surgical complication and was discharged.

Discussion

Insulinoma is the most common cause of hyperinsulinemic hypoglycemia. They are insulin-secreting tumors of pancreatic origin that cause hypoglycemia. Insulinoma had been found in 2-4 subjects per million, per year.[2] There are clinically significant variations of insulinoma, i.e., 4.8% of them demonstrate celiac artery stenosis.[3] According to

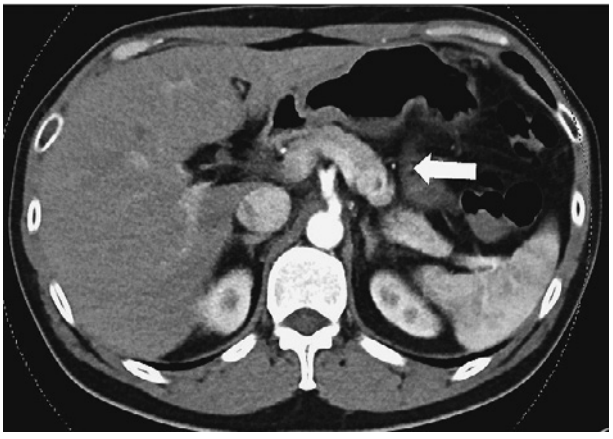

Fig. 1 Enhanced computed tomography revealed 20 mm of mass at the tail of pancreas.

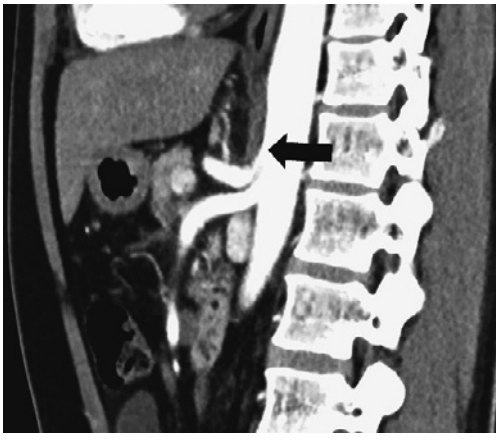

Fig. 3 Enhanced computed tomography revealed stenosis in celiac artery (arrow).

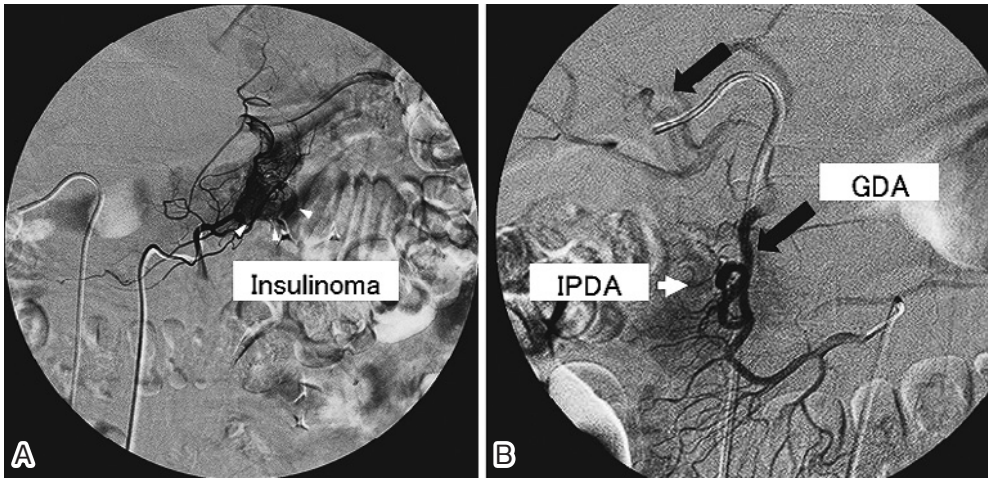

Fig. 2
A) Angiogram reveals the blood flow from dorsal pancreatic artery to insulinoma (arrow).
B) In inferior pancreaticoduodenal artery (IPDA), retrograde blood flow was found from SMA to liver by angiogram (black arrow).
IPDA, inferior pancreaticoduodenal artery (white arrow); GDA, gastroduodenal artery.

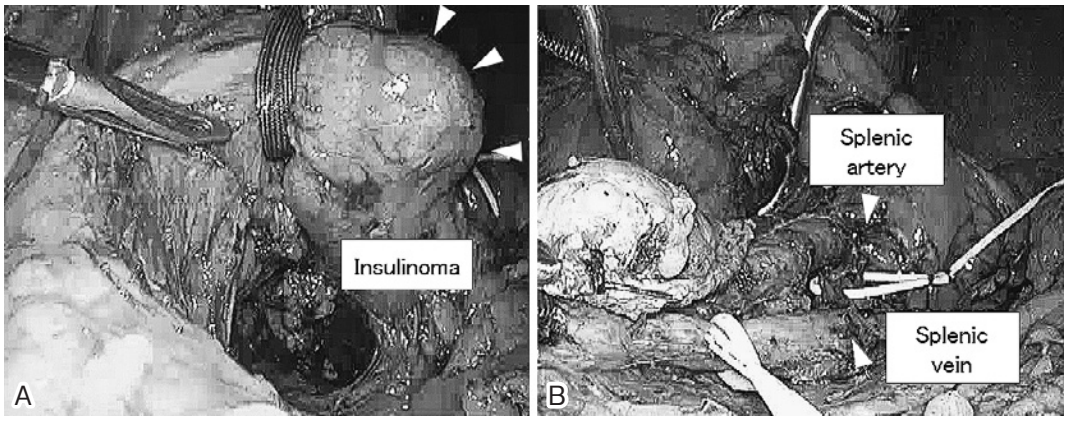

Fig. 4 Tumor imaging during surgery
A) Mass at the tail of pancreas was clearly identified (arrow).
B) Resection of the tumor was successfully performed. Splenic artery and vein were divided and preserved (arrow).

Scott et al., diagnostic arteriography in combination with biochemical, pancreatic arterial anatomy, and reginal perfusion data improved sensitivity of localization to 92.9%.[3] MALS typically occurs in young to middle-aged adults.[4] MALS has been reported in 4% of patients undergoing pancreaticoduodenectomy (PD).[5] In the present case, PD was not performed before Laparoscopic spleen preserving distal pancreatectomy. The incidence of celiac axis stenosis caused mainly by MALS is around 7.3% in asymptomatic individuals.[6] Decompression surgery, open surgery, laparoscopic surgery, and robotic surgery are hallmarks of the treatment.[7, 8]

To our knowledge, there were no reports of MALS in insulinoma patient. Because of MALS, celiac artery was compressed. Blood flow in splenic artery was decreased. In the present case, dorsal pancreatic artery was branched from SMA. On the other hand, blood flow in the dorsal pancreatic artery and SMA were increased. According to Woodburne, 21.5% of dorsal pancreatic artery arises close to the source of the SMA.[9] Therefore, blood flow from dorsal pancreatic artery to insulinoma was found. In addition, retrograde blood flow was found from SMA to liver in IPDA. MALS should be considered when angiography of the area surrounding the pancreas reveals blood flow different from that observed in normal anatomy.

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Consent: Informed consent was obtained from the patient and family for publication of this case report and the accompanying images.

Conflicts of interest: None declared.

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