Congenital extrahepatic portosystemic shunt in pancreatic cancer

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Introduction

Portosystemic shunts are generally related with portal hypertension but in few cases, they are associated with genetic abnormalities called congenital portosystemic shunts (CPSS). The CPSS is characterized by a large communication between the portal venous system and the systemic venous circulation in the absence of parenchymal or biliary disease. The incidence of CPSS is not well known and estimated to be 1/30,000 live births. Different types of CPSS have been described by abdominal imaging: extrahepatic and intrahepatic. The extrahepatic are classified as type I-a (splenic and the SMV drain separately into the IVC), I-b (portal vein drains into IVC), both without portal perfusion to the liver and II has normal vein draining to the liver and communicating with IVC. Herein, we present an unusual case of CPSS in an asymptomatic patient diagnosed with pancreatic cancer.

Case presentation

An 85-years-old female with a past medical history of a carotid endarterectomy, hypertension and no other comorbidities presented with an episode of decreased appetitive and obstructive jaundice. Imaging studies were performed showing the presence of a 3cm mass in the head of the pancreas.

Incidentally, there was a large portosystemic shunt extending from the left renal vein to the portal vein as well as a replaced right hepatic artery arising from the SMA noted. Both were asymptomatic to the patient.

An upper endoscopic ultrasound was carried out and showed the mass previously described and FNA biopsy confirmed the presence of an adenocarcinoma. The case was presented in a pancreatic board and was decided to go on neoadjuvant chemotherapy for 5 cycles. After that, a Whipple procedure was performed with no injury to the large CPSS.

The patient did well and was discharged on POD 5.

Conclusion

Few cases of congenital portosystemic shunts (CPSS) are described in the literature, especially with pancreatic cancer. Some reports show that these patients may have a higher rate of other genetic diseases but no associate with malignancy has been noted.
Patients with CPSS may develop complications just like encephalitis, portal hypertension, hepato-pulmonary syndrome and shunt closure during surgery must be considered. However, this patient was asymptomatic 85 therefore did not require taking down the shunt.
Care should be taken to identify these vascular abnormalities prior to surgery to avoid intraoperative injury of an anomalous vascular structure.

Figure 1:
Axial CT plane with a red arrow pointing to the shunt

Figure 2:
Coronal CT plane a red arrow pointing to the shunt

Figure 3:
Intraoperative shot with a yellow circle on the shunt

Figure 4:
Illustration Image with a red circle on the shunt

Figure 1

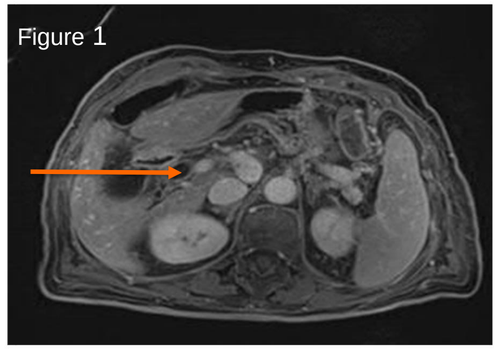


Figure 2

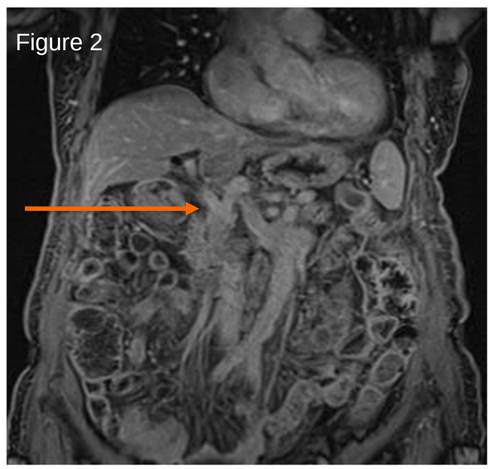
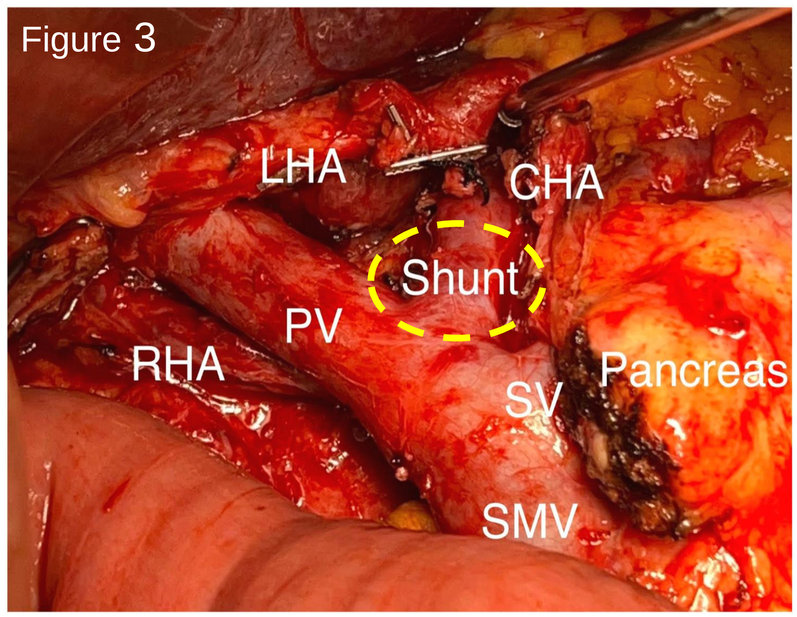


Figure 3



PV: portal vein	LHA: left hepatic artery
SV: splenic vein	RHA: right hepatic artery
SMV: superior mesenteric vein	CHA: common hepatic artery

Figure 4