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A case of Hepatocellular Carcinoma in Abernethy syndrome

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Abstract

Abernethy malformation is a rare type of congenital malformation of the hepatic portal system. It can take one of three forms and is classified as an extrahepatic portosystemic shunt between the portal vein and a large systemic vein, usually the IVC.

Here, we present at 47-year-old male who was admitted with upper abdominal pain. His laboratory investigations showed a raised alkaline phosphatase, alanine aminotransferase and bilirubin but were otherwise within normal range. Ultrasound scan showed an indeterminate mass in the right lobe of the liver which was confirmed with a CT scan. An anomalous portal vein drainage system was also identified on CT scan. Following MDT discussion, the decision for surgical exploration was made to assess the resectability and potential functional liver remnant. At exploration, resection was precluded by the complete absence of portal veins with no viable reconstruction option due to the size of the tumour and the potential of inadequate functioning residual liver following resection. Both the splenic vein and SMV were visualised intraoperatively and seen to form a short convergence before draining directly into the IVC. This abnormality is in line with type 1b Abernethy malformation. Biopsies were also taken which confirmed a diagnosis of well differentiated HCC. Further treatment options including trans arterial chemoneminbolization (TACE) and trans arterial radioembolization (TARE) have been deemed inappropriate due to the lack of portal vein as well as the size of the tumour. Considering this the patient has been referred for systemic treatment.

Keywords: Abernethy syndrome; Liver resection; hepatocellular carcinoma

Case Presentation

A 47-year-old man presented to a district general hospital with upper abdominal pain. Ultrasound of the abdomen showed an indeterminate mass in the right lobe of the liver with no sign of intrahepatic biliary dilatation. His blood results revealed a raised alkaline phosphatase (286 U/L), raised alanine aminotransferase (158 IU/L) and a raised bilirubin (25 mg/dL). His urea was normal at 6.6 mg/dL. The patient has a past medical history of Type 2 Diabetes Mellitus, hypertension, and hypercholesterolemia. No history of excess alcohol consumption, risk factors for liver disease or family history of liver cancer. The patient underwent a CT scan of his chest, abdomen and pelvis which confirmed a mass lesion confined to the right lobe of the liver as well as an anomalous portal vein drainage system (**Figure 1**).

Oblique Maximum Intensity Projection (MIP) reconstruction of a portal venous phase CT scan further confirmed this (**Figure 2**). The patient was then referred to the regional HPB MDT. At MDT, surgical exploration to assess resectability and FLR was considered most appropriate. His Alphafetoprotein was 2 and a PET CT confirmed the lesion, which was non-avid and a presumptive diagnosis of a fibrolamellar hepatocellular carcinoma on the background of non-cirrhotic liver was made.

Following exploration, the tumour was deemed unresectable due to an absence of portal veins and aberrant anatomy of abnormal drainage of portal vein directly into the IVC. The SMV and splenic vein were seen intraoperatively to form a brief confluence before direct drainage into the IVC (**Figure 3**). This is characteristic of a type 1b Abernethy malformation (**Figure 4**). Normal hepatic artery supply was confirmed. Biopsies

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were also taken confirming the diagnosis of a well differentiated HCC.

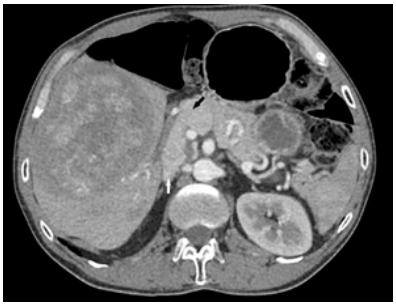

Figure 1: *Arterial phase CT scan shows areas of enhancement within a large right lobe liver lesion. Arterial phase enhancement and portal venous or delayed phase washout is a radiological hallmark of HCC. Note the ectatic SMV and splenic vein.*

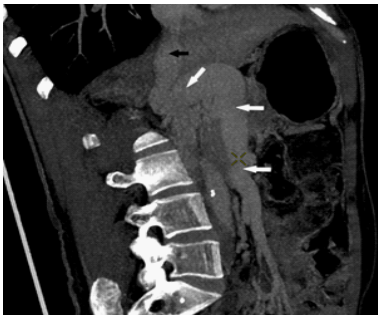

Figure 2: *Oblique MIP reconstruction of a portal venous phase CT scan shows a large calibre SMV (white arrows) with drainage into the IVC (black arrow).*

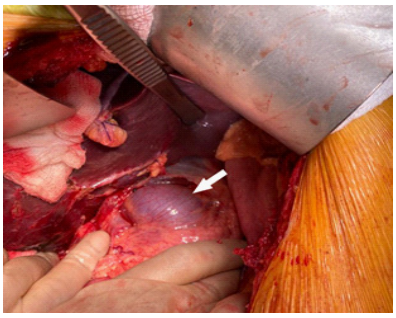

Figure 3: *Intraoperative image showing the SMV, splenic vein confluence draining into the IVC.*

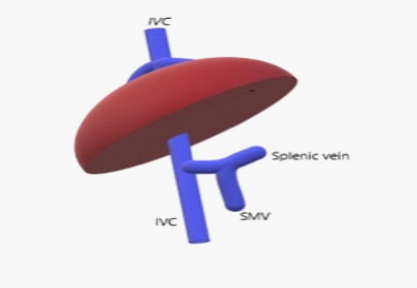

Figure 4: *Diagrammatic representation of an Abernethy type 1B malformation.*

Discussion

About 20-35% of the population have some congenital variation in the anatomy of their portal vein, most of which are not symptomatic but are incidental findings [1]. Abernethy malformation is a rare type of congenital malformation and is classified as an extrahepatic portosystemic shunt between the portal vein and a large systemic vein which is the inferior vena cava in most cases. Other areas the shunt may drain to include the right atrium, azygous or iliac veins [2]. How symptomatic these patients are depends mainly on the anatomy of their Abernethy malformation, some are completely asymptomatic, as with other congenital variations in portal vein anatomy or some present with features of cardiac failure, pulmonary hypertension and hepatic encephalopathy [1]. The most serious manifestation of Abernethy malformation is hepatopulmonary syndrome where there is severe hypoxaemia [3].

Abernethy malformation is divided into two subtypes consisting of type 1 where there is no intrahepatic branching of the portal vein meaning the entire venous drainage from the portal vein is emptied into a large systemic vein [1]. Type 1 comes in two types, type 1a has the splenic vein and superior mesenteric vein isolated, draining separately into the systemic circulation; type 1b has a confluence of the two to form the portal vein which then empties into the IVC [1]. A type 2 Abernethy has incomplete shunting between the portal vein and systemic circulation as there are intrahepatic portal vein branches present. Type 1 Abernethy is more commonly associated with other congenital abnormalities [1].

The malformation is rare. 310 cases have so far been reported though with more cases being found all the time as imaging and neonatal screening advances [4]. We are aware of only 16 other cases where the patient has an Abernethy malformation complicated by development of HCC [2]. However, it is known that patients with type 1 Abernethy malformations are more susceptible to developing HCC as well as other benign and malignant hepatic lesions [5].

Given the unresectable nature of the tumour, if we follow BCLC staging treatment guidelines, the next treatment option would be TACE [6]. However, in much the same way as TACE is contraindicated in portal vein thrombosis, the congenital lack of portal vein blood supply in this case means this is a treatment option would not be suitable as this patient's liver is solely dependent on their hepatic artery. Indeed, in two previous cases before the Abernethy malformation was identified use of TACE led to adverse outcomes [2,7].

Instead, success seems to have been found through use of TARE whereby the arterial supply is not compromised, rather, the microvasculature of the tumour itself is occluded [8]. However the size of the tumour and potential subsequent large area of necrosis may preclude this. Liver transplant with venous iliac graft to extend the donor portal vein has also been successful to treat a patient with Abernethy malformation [9]. Albeit this for an adenoma not HCC. Clearly the size of the tumour precludes transplant, even by extended criteria and so the patient has been referred for systemic treatment.

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