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Case Report

Intrahepatic inferior vena cava interruption with transhepatic venous continuation initially misdiagnosed as a congenital portosystemic shunt ☆☆☆☆

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ABSTRACT

We report a case of intrahepatic inferior vena cava interruption with azygos and transhepatic venous continuation discovered incidentally on CT angiography for acute aortic syndrome. The lesion was initially misdiagnosed as a congenital portosystemic shunt on multiphase CT of the liver but subsequent fluoroscopic venogram revealed no evidence of portosystemic shunting. While intrahepatic IVC interruption with azygos continuation is an uncommon but well-known anatomical variant, transhepatic venous continuation is extremely rare and only a few cases have been published. Excluding portosystemic shunting is important for determining management as persistent congenital portosystemic shunts can be associated with significant morbidity.

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Introduction

Intrahepatic inferior vena cava (IVC) interruption with azygos continuation is a rare but well-known anatomical variant with an overall prevalence of 0.6% [1]. Although originally thought to be largely associated with severe congenital heart disease

and polysplenia syndrome, this finding is now increasingly discovered incidentally in otherwise healthy individuals during cross-sectional imaging [1]. In contrast, transhepatic venous continuation is extremely rare and only a few cases of this phenomenon have been published in the literature [2]. We present a case of intrahepatic IVC interruption with transhepatic venous continuation discovered incidentally in a 65-

Abbreviations: CT, Computed tomography; CTA, CT angiography; ECG, electrocardiogram; HPB, hepato-pancreato-biliary; IVC, inferior vena cava; MDCT, Multidetector CT; MIP, Maximum intensity projection; MRI, Magnetic resonance imaging; VIBE, volumetric interpolated breath-hold examination.

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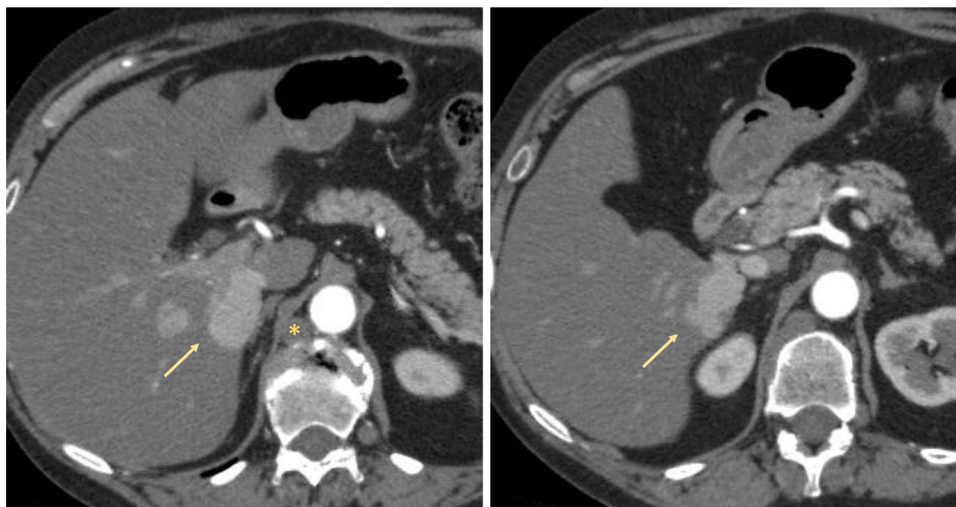


Fig. 1 – Axial MDCT images in arterial phase and soft tissue windows (left: superior; right: inferior) shows a lobulated lesion in hepatic segments 6/7 which enhances homogenously following hepatic venous blood pool (arrows). The intrahepatic IVC was not seen. Note the dilated azygos vein (*)

year-old woman and initially misdiagnosed as a congenital portosystemic shunt on cross-sectional imaging.

Case report

A 65-year-old woman presented to urgent care with complaints of vague chest and subscapular pain. She was well-appearing and in no distress but was noted to have a systolic blood pressure in the 190s. The patient's chest radiograph and ECG showed no concerning findings, and routine bloodwork was similarly unremarkable. The primary physician opted for a CT angiography (CTA) of the thorax, abdomen, and pelvis to rule out possible acute aortic syndrome given the patient's hypertension and subscapular pain.

CTA did not demonstrate acute aortic syndrome but incidentally found a lobulated lesion in hepatic segments 6 and 7 (Fig. 1). The lesion enhanced following venous blood pool and appeared indistinct from the right hepatic vein and main portal vein. The intrahepatic IVC was not seen, and the azygos vein was dilated. An intrahepatic venous malformation or vascular shunt was suspected, and outpatient multiphase CT of the liver was recommended for further evaluation.

The multiphase MDCT was performed on a 128-detector CT with non-enhanced, late arterial, and portal venous phase sequences (Fig. 2A and C). It confirmed that the lesion was vascular in origin and had no distinct soft tissue component. No arterial components were found, and there was no early venous filling. There was absence of the intrahepatic IVC, and the lesion communicated directly with both the inferior right hepatic vein, a normal anatomical variant that this patient possessed, and the infrahepatic IVC. Several tortuous venous collateral vessels appeared to anastomose the inferior right hepatic vein with the main right hepatic vein, with the largest calibre vessel containing an aneurysmal outpouching measuring 3.4×1.6 cm. There were no filling defects in the hepatic or portal veins and no other vascular malformations were

seen in the abdomen or pelvis. There were no findings to suggested cirrhosis, portal hypertension, or polysplenia syndrome. The lesion was indistinct from the main portal vein and while communication of the lesion with the portal system could not be determined with confidence, it was suspected. The diagnosis made was a complex congenital portosystemic shunt and recommendations were made for referral to hepatology and an ultrasound and MRI for additional evaluation.

The MR of the abdomen obtained with a 1.5 T MR system (Fig. 2B) however found no clear evidence of communication with the portal venous system. Instead, the findings were reported to represent intrahepatic IVC interruption with azygos and transhepatic venous continuation. Along with the 3.4 cm venous aneurysm arising from the main intrahepatic collateral vessel, an additional 1.1 cm aneurysm was suspected downstream. No other new or discordant findings were seen. Given the inconclusive findings on cross-sectional imaging, a fluoroscopic venogram was ordered for further anatomical characterization and to rule out portosystemic shunting. The ultrasound examination was not obtained.

Venogram (Fig. 3) was performed with a 5-French pigtail catheter placed in the infrarenal IVC and again confirmed both the absence of the intrahepatic IVC and an enlarged azygos vein serving as collateral drainage into the superior vena cava at the right paratracheal space. In addition, contrast flowed from a small calibre infrahepatic IVC into the dilated inferior right hepatic vein, then into several intrahepatic bridging venous collateral vessels, then into the main right hepatic vein, and finally directly into the right atrium. The two venous aneurysms arising from the dominant collateral vessel seen on cross-sectional imaging measured 3.3 cm craniocaudad by 2.5 cm medial – lateral and 2.6 cm craniocaudad by 1.3 cm medial – lateral. The catheter was then placed within the dominant venous aneurysm. Pressure measurements within the dominant venous aneurysm measured 10 mmHg and 11 mmHg in the infrarenal IVC. There was no evidence of portosystemic shunting. The venographic findings confirmed in-



Fig. 2 – Selected consecutive axial images of the upper abdomen with A) contrast-enhanced MDCT in soft tissue windows, and B) 1.5T gadolinium-infused T1-weighted fat suppressed VIBE MRI both in portal venous phase (left to right: superior to inferior). The lobulated hepatic lesion is confirmed to be vascular in origin and enhances following venous blood pool. It is part of a series of collateral venous structures that appear to connect the right inferior hepatic vein and the right main hepatic vein. The lesion appears intimate with the main portal vein (arrow) and appears indistinct on CT. C) Coronal MDCT MIP reconstructions (top: anterior, bottom: posterior) show the overall anatomy of the collateral network. The dominant saccular aneurysm is also more apparent



Fig. 3 – Fluoroscopic abdominal venogram. (A) There is interruption of the intrahepatic IVC with several collateral vessels between the inferior and main right hepatic veins. The dominant collateral network (white arrowheads) contains two saccular aneurysms (black and white arrows). The azygos vein is dilated (*). (B) Venogram with the 5-French catheter advanced into larger and more proximal aneurysm better highlights the anatomy of the collateral network. There is no evidence of portosystemic shunting

trahepatic IVC interruption with azygos and transhepatic venous continuation

This finding was novel to our institution, and the patient's case and future management options were discussed during interdisciplinary rounds between hepato-pancreato-biliary (HPB) surgery, hepatology, and radiology. It was decided that no treatment was necessary as the patient was asymptomatic and had no clinical or metabolic evidence of hepatic dysfunction. She remains under clinical follow-up with HPB

surgery, and a repeat CT was recommended in a year as a precaution to reassess the venous aneurysms.

Discussion

During embryogenesis, a right-sided pre renal (hepatic and suprarenal) IVC is normally formed following fusion of the

hepatic vein and the right subcardinal vein by means of the subcardinal-hepatic anastomosis between the sixth to eighth weeks [1,2]. Interruption of the intrahepatic IVC is suspected to occur following failure of this fusion. Blood is subsequently shunted into the supracardinal vessels, which give rise to the azygos venous system, or to collateral venous vessels [1–3]. This classically results in azygos continuation, but additional prominent venous collaterals may also form. Compared with azygos continuation, an already uncommon finding, transhepatic venous continuation is extremely rare and only a few cases have been described in the literature [2]. Along with a congenital etiology, there have also been reports of intrahepatic venous collateral vessels forming secondary to chronic occlusion of the IVC [4,5]. Both Takayasu et al. 1985 and Chevallier et al. 1999 postulated that the presence of an inferior right hepatic vein, a variant vessel found in ~10% of the population, is necessary for these intrahepatic venous collaterals to form [2,4].

This developmental anomaly appears to be a relatively benign finding; nearly all cases were discovered incidentally in asymptomatic individuals, and to our knowledge there has been no reported association with significant comorbidity [2,6,7]. One case described by Sahin et al. 2017 was associated with pelvic congestion syndrome, but the patient had several additional venous anomalies including a duplicated IVC, which is known to be associated with an increased incidence of venous congestion and thrombosis [8].

Our case was initially misdiagnosed as a complex congenital portosystemic shunt. Congenital portosystemic shunt persisting into adulthood is an additional extremely uncommon finding [9]. However, unlike intrahepatic IVC interruption with transhepatic venous continuation, congenital portosystemic shunts persisting into adulthood can be associated with significant morbidity such as pulmonary hypertension, heart failure and hepatic encephalopathy [9]. They are also associated with a higher risk of developing both benign and malignant hepatic tumors [9]. The discovery of a portosystemic shunt in our case would have likely prompted a different treatment approach.

The presence of venous aneurysms involving the intrahepatic collateral vessels presented additional consideration for possible treatment. Prior case reports also encountered dilated, tortuous, and aneurysmal collateral vessels in transhepatic venous continuation [2,6,7,8]. To our knowledge, there has not been a published case of any complications arising from these aneurysmal vessels, such as enlargement, thrombosis, or rupture. In general, venous aneurysms arising from the hepatic veins are extremely rare and only five cases have been

reported [10]. Given the overall rarity of this condition and subsequent lack of data, the temporal course for these anomalies are unclear and treatment options are non-standardized. In our case, the primary care team opted for no treatment given the lack of rationale and opted for surveillance with repeat CT as a precaution.

In summary, we present an additional case of an extremely rare developmental anomaly discovered incidentally during cross-sectional imaging. Fluoroscopic venogram confirmed no portosystemic shunting and no treatment was attempted.

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