

A rare cause of congenital portosystemic shunt: type 2 Abernethy malformation

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The Abernethy malformation is characterised by congenital extrahepatic portosystemic shunts and is divided into two groups according to the type of anastomosis. In type 1, all portal venous blood is discharged into the inferior vena cava and there is no intrahepatic portal vein. In type 2, the portal vein is partially discharged to the inferior vena cava via side-by-side anastomoses. Imaging has an important role in the diagnosis and follow-up of this malformation. Magnetic resonance imaging should be preferred to demonstrate both vessel anatomy and associated anomalies. The aim of this study was to present a 17-year-old male patient and to discuss the imaging findings of Abernethy malformation. (Folia Morphol 2020; 79, 1: 172–175)

Key words: Abernethy malformation, magnetic resonance imaging, radiology

INTRODUCTION

The Abernethy malformation was first reported by John Abernethy in 1793 and is characterised by the removal of portal venous blood from the liver by end-to-end and side-to-side shunts [1]. Approximately 80% of cases are children aged < 18 years. Complications such as hepatic encephalopathy and hepatopulmonary syndrome may develop in patients. It is divided into two classes according to the type of anastomosis between the portal vein (PV) and the inferior vena cava (IVC) and the presence of intrahepatic PV supply. In Abernethy malformation type 1, all of the portal venous supply is discharged into the IVC and there is no intrahepatic PV. In type 2, the PV is partially discharged into the IVC via side-by-side anastomoses [10].

Imaging has an important role in the diagnosis and follow-up of this malformation. Magnetic resonance imaging (MRI) should be preferred to demonstrate both vessel anatomy and associated anomalies [5].

The aim of this study was to present a 17-year-old male patient and to discuss the imaging findings of Abernethy malformation.

CASE REPORT

A 17-year-old male was admitted to our hospital with complaints of abdominal pain, vomiting and mental fog, which had been ongoing for 3 days. There was no history of fever, abdominal trauma, weight loss or jaundice, and there had been no similar episode in the past. The family history showed no gastrointestinal cancer. There was mild epigastric tenderness on physical examination. There was no abnormality in the complete blood count. The serum C-reactive protein, sedimentation, alanine aminotransferase, aspartate aminotransferase, gamma-glutamyl transpeptidase and alkaline phosphatase values were within normal limits.

Magnetic resonance imaging was obtained for further evaluation. A side-to-side portosystemic shunt

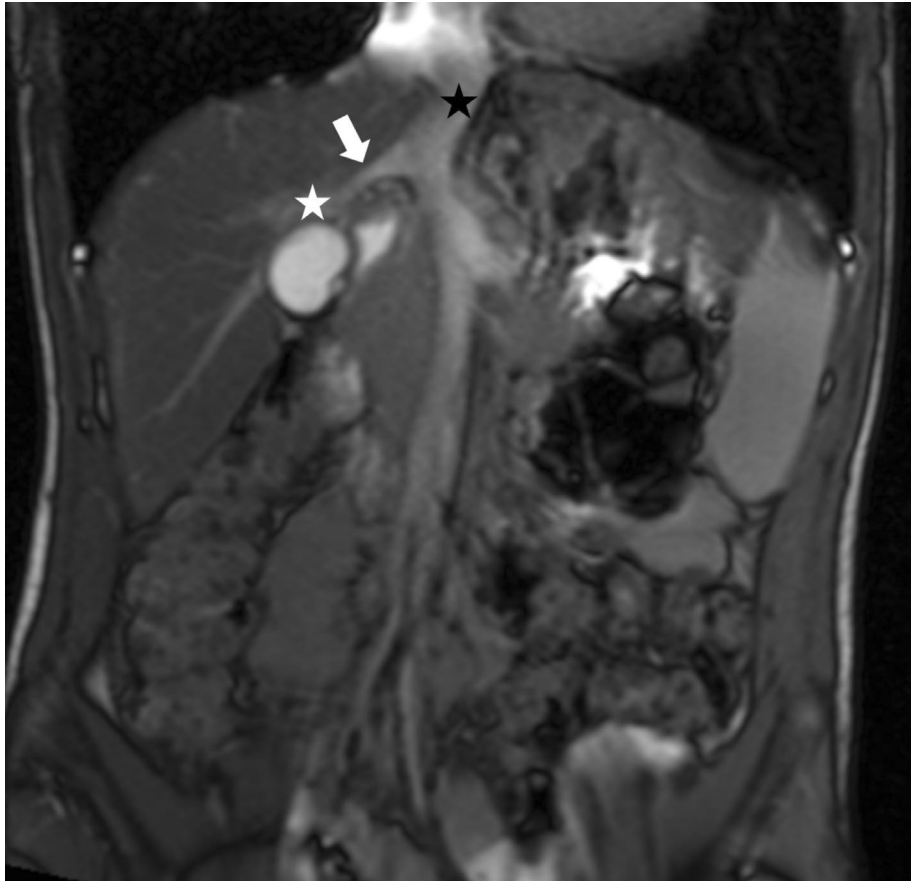


Figure 1. Coronal T2-weighted magnetic resonance images showing a side-to-side portosystemic shunt (arrow) between the inferior vena cava (black asterisk) and intrahepatic portal vein (white asterisk).

between the IVC and the intrahepatic PV was seen on serial post-contrast MRI (Figs. 1, 2). With these findings, the patient was diagnosed with type 2 Abernethy malformation, which was characterised by the absence of part of the PV with congenital portocaval shunt.

The patient was followed-up with conservative treatment. At the 6-month follow-up examination, the patient was asymptomatic. If hepatic encephalopathy develops in the future, it is planned to evaluate surgical closure of the shunt.

DISCUSSION

The PV system develops as a result of the selective apoptosis of a portion of the vitelline veins at 4–10 weeks of embryonic life. IVC development also coincides, so there is potential for congenital portosystemic shunt development [12]. There are two types of portosystemic shunt anomalies. In type 1 shunts, there is no intrahepatic PV and there is a complete end-to-side shunt. Type 1 shunts have two subtypes; type 1a shunts discharge separately into the superior mesenteric vein (SMV) and splenic vein (SV), IVC, iliac

veins or renal veins. In type 1b shunts, the SMV and SV converge to form a short extrahepatic PV. In type 2 shunts, there is a partial side-to-side shunt between the intrahepatic PV and PV. Portoportal collaterals develop when PV occlusion develops. Type 1 anomaly is more common [11]. The current patient was determined with type 2 anomaly.

Intrahepatic portosystemic shunts are classified by Park et al. [14] in four different types. In the first and most common type, the right PV is connected to the IVC through a large vessel. The second type has peripheral shunts in a single hepatic segment. In the third type, the shunt is provided with an aneurysm. The fourth type includes peripheral shunts in multiple hepatic segments. Persistent ductus venosus can also be evaluated as the fifth type.

Congenital extrahepatic portosystemic shunts are frequently seen with congenital heart disease, polysplenia, biliary atresia, malrotation, duodenal atresia, annular pancreas, situs inversus, urinary tract anomalies and skeletal anomalies [8]. However, no additional anomaly was present in the current patient. Congenital

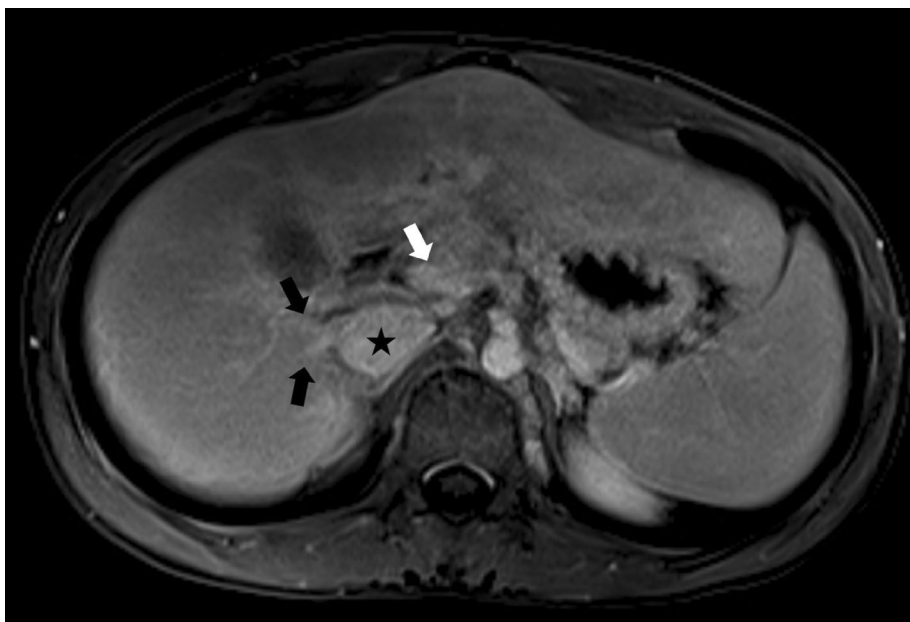


Figure 2. Axial postcontrast magnetic resonance images showing hepatic veins (black arrows) draining into inferior vena cava (asterisk) and a side-to-side portosystemic shunt (white arrow) between the inferior vena cava and intrahepatic portal vein.

extrahepatic portosystemic shunts are also associated with benign (focal nodular hyperplasia, hepatocellular adenoma, or nodular regenerative hyperplasia) or malignant (hepatocellular carcinoma or hepatoblastoma) liver neoplasms [4, 13]. It has been suggested that the presence of hepatotropic substances such as insulin and glucagon in the splanchnic venous blood outside the liver may cause changes in the liver's development, function and regeneration capacity. This deviation and associated increase in arterial hepatic flow can lead to neoplasm formation [4]. An imbalance between the hepatic artery and the PV is thought to pave the way for the development of neoplastic tumours [17]. In addition, beta-catenin gene mutations leading to tumour development have been demonstrated in a patient with Abernethy malformation [16]. In this context, it is important to monitor these patients for a long time because of the potential for benign formations to develop into malignant tumours [2].

Patients with Abernethy malformation may also have other symptoms, such as intravenous intrapulmonary dilatation and hepatopulmonary syndrome, which may occur due to hepatic encephalopathy or vasoactive mediators in systemic circulation as a result of the toxicity level of toxins produced in the intestines [19]. In the current patient, there was a mild mental fog at the time of admission.

The diagnosis of Abernethy malformation is currently usually made with imaging methods, such

as ultrasound, computed tomography (CT) or MRI showing shunt and intrahepatic PV branches. Doppler ultrasonography is a safe and non-invasive method for the diagnosis of intrahepatic vasculature, as the amount and direction of flow can be shown. However, it may not be able to detect associated anomalies, and the retroperitoneum cannot be well evaluated, especially in adult patients. Therefore, smaller shunts, in particular Type 1a, may not be clearly visible. Ultrasound may not be able to fully identify liver lesions seen in these patients. Associated anomalies and findings, especially lung and cardiac anomalies, will not be defined on ultrasound [6, 7]. CT is a rapid non-invasive method that demonstrates the anatomy and pathology in detail with spatial resolution. The greatest advantage of CT is that the portal anomaly and shunt type can be clearly visualised, which helps in treatment decision-making. CT evaluates the associated anomalies in patients with congenital heart disease who require the assessment of pulmonary vessels, or in patients with suspected hepatopulmonary syndrome, which require evaluation of the lungs. However, a very significant disadvantage is the radiation dose, which should be taken into consideration especially in paediatric patients [5, 15]. MRI, not only has the features of CT, but also helps detect and characterise the hepatic lesions in these patients. The use of liver-specific contrast agents in the characterisation of hepatic nodules is very help-

ful in the diagnosis. The most important superiority of MRI to CT is that it does not expose patients to ionising radiation. MRI is the method that should be used for serial monitoring of hepatic lesions [3, 18].

The prognosis depends on the location of congenital heart disease, liver disease, and portosystemic shunt. In patients with type 1 malformation, mesenteric venous blood is shunted in a single drainage pathway, and surgical closure is not performed in these patients. These patients should be followed up clinically and biochemically, and those with hepatic encephalopathy and malignant liver nodules should be evaluated for liver transplantation. If a serious complication such as hepatic encephalopathy develops in type 2 malformation, the shunt may be closed surgically or percutaneously [9].

CONCLUSIONS

In conclusion, although Abernethy malformation is quite rare, it can cause serious complications; therefore, early recognition is important for the implementation of proper follow-up and treatment to avoid complications.

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