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

Type 2 Abernethy malformation

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Abstract

The Abernethy malformation is characterized 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.

Key words: Abernethy malformation, magnetic resonance imaging, radiology

INTRODUCTION

The Abernethy malformation was first reported by John Abernethy in 1793 and is characterized 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 portal vein supply. In

Abernethy malformation type 1, all of the portal venous supply is discharged into the inferior vena cava and there is no intrahepatic PV. In type 2, the portal vein is partially discharged into the IVC via side-by-side anastomoses [2].

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 [3]. 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 CRP, sedimentation, ALT, AST, GGT and ALP values were within normal limits.

Magnetic resonance (MR) imaging was obtained for further evaluation. A side-to-side portosystemic shunt between the inferior vena cava (IVC) and the intrahepatic portal vein was seen on serial post-contrast MR images (Figure 1 and 2). With these findings, the patient was diagnosed with type 2 Abernethy malformation, which was characterized by the absence of part of the portal vein 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 portal vein 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 [4]. There are two types of portosystemic shunt anomalies. In type 1 shunts, there is no intrahepatic portal vein 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), inferior vena cava (IVC), iliac veins or renal veins. In type 1b shunts, the SMV and SV converge to form a short extrahepatic portal vein. In type 2 shunts, there is a partial side-to-side shunt between the intrahepatic portal vein and portal vein. Portoportal collaterals develop when portal vein

occlusion develops. Type 1 anomaly is more common [5]. The current patient was determined with type 2 anomaly.

Intrahepatic portosystemic shunts are classified by Park et al. [6] in 4 different types. In the first and most common type, the right portal vein 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 [7]. However, no additional anomaly was present in the current patient. Congenital 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 [8, 9]. 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 [8]. An imbalance between the hepatic artery and the portal vein is thought to pave the way for the development of neoplastic tumors [10]. In addition, β-catenin gene mutations leading to tumor development have been demonstrated in a patient with Abernethy malformation [11]. 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 tumors [12].

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 [13]. 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, CT or MRI showing shunt and intrahepatic portal vein 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 [14, 15]. Computed tomography (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 visualized, 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 pediatric patients [3, 16]. MRI, not only has the features of CT, but also helps detect and characterize the hepatic lesions in these patients. The use of liver-specific contrast agents in the characterization of hepatic nodules is very helpful in the diagnosis. The most important superiority of MRI to CT is that it does not expose patients to ionizing radiation. MRI is the method that should be used for serial monitoring of hepatic lesions [17, 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 [19].

CONCLUSIONS

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

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FIGURE LEGENDS

Figure 1. Coronal T2-weighted MR images showing a side-to-side portosystemic shunt (arrow) between the IVC (black asterix) and intrahepatic portal vein (white asterix).

Figure 2. Axial postcontrast MR images showing hepatic veins (black arrows) draining into IVC (asterix) and a side-to-side portosystemic shunt (white arrow) between the IVC and intrahepatic portal vein.