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Case Report

Congenital Extrahepatic Portosystemic Shunt: Abernethy Malformation Type 2

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ABSTRACT

Abernethy malformation is a vascular congenital anomaly in which extrahepatic portocaval shunts develop. The patient was admitted to the gastroenterology department with complaints of abdominal pain and nausea. Ammonia and bilirubin levels were increased in the laboratory values and other liver function values were normal. The shunt was detected between the inferior vena cava and portal vein by ultrasonography and computed tomography, and the portal vein was hypoplastic. In this case report, we present a male patient diagnosed with Abernethy malformation type 2.

Keywords: Abdominal pain, abernethy malformation, congenital anomaly

INTRODUCTION

Abernethy malformation (AM) with congenital anomaly was defined by John Abernethy in 1973. This malformation is characterized by shunting between the portal vein (PV) and systemic circulation (1, 2). AM is frequently associated with other rare congenital anomalies, including the extrahepatic portocaval shunt, heterotaxy, biliary atresia, and liver nodules (3, 4). AM has been classified into two types on the basis of the pattern of anastomosis between the systemic circulation and PV and the presence of intrahepatic portal venous supply. AM type 1 portosystemic shunt is characterized by complete shunting and absence of a PV. AM type 2 is characterized by partial shunting with a small grade of PV flow to the liver (5). Assessment of the vascular anatomy and liver using new abdominal imaging modalities aids in treatment planning so that patients with AM can receive appropriate treatment. An alternative treatment mode is non-surgical endovascular treatment; if this treatment fails, liver transplant may be considered (5). In this study, we present the case of a male patient who was incidentally diagnosed with AM type 2.

CASE PRESENTATION

A 68-year-old male patient presented to the gastroenterology department with the complaints of abdominal pain and nausea. The patient had no significant personal or family medical history. No pathology except the right upper quadrant sensitivity was found on physical examination. The patient had undergone total gastrectomy for gastric adenocarcinoma 3 months ago. The laboratory test findings were as follows: hemoglobin, 13 g/dL; direct bilirubin, 1.24 µmol/L; indirect bilirubin, 3.70 µmol/L; lactate dehydrogenase, 322 U/L; alanine aminotransferase, 21 U/L; aspartate aminotransferase, 40 U/L; gamma-glutamyl transpeptidase, 32 U/L; alkaline phosphatase, 116 U/L; and prothrombin time, 39.0 s. Serologic test results for hepatitis B and C viruses were negative. Abdominal ultrasonography (USG) showed the presence of an anechoic tubular structure approximately 13 mm in diameter in the liver with no current Doppler signal. Computed tomography (CT) axial sections showed superior contrast enhancement in the right lobe of the liver, suggesting a hemangioma 5 mm in diameter. PV diameter was measured as 5 mm. PV superior mesenteric vein (SMV) and splenic vein junction left renal vein portocaval shunt (Figure 1). Given these radiological findings, a hypoplastic PV with a portocaval shunt (AM type 2) was suspected. Conservative treatment was continued for the patient, and after 1 month, abdominal pain and nausea became mild. At 3-month follow-up, no change was observed on USG. First, the alternative treatment option of non-surgical endovascular treatment should be considered for the patient. If this treatment fails, liver transplant may be considered. Informed consent was taken from the patient before writing this report.

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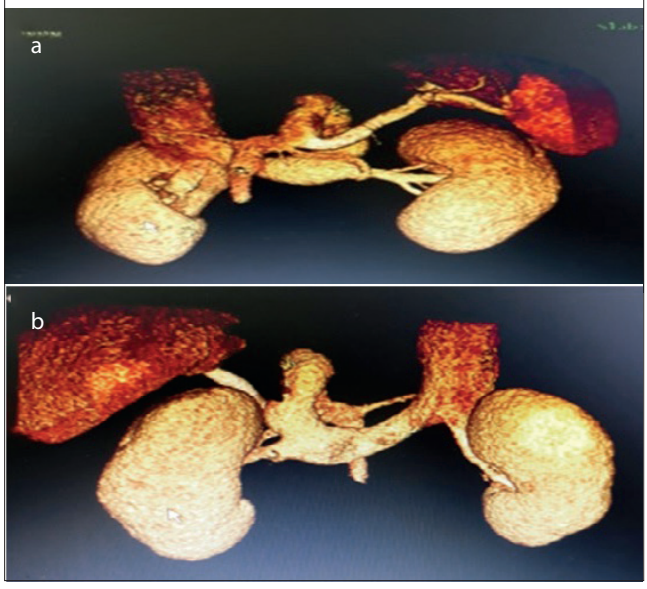
Figure 1. a, b. CT axial sections showed SMV and the splenic vein merging into the left renal vein. PV is thin-walled and hypoplastic

CT: computed tomography; SMV: superior mesenteric vein; PV: portal vein



DISCUSSION

Congenital anomalies and vascular shunt diseases have been reported to occur together (6). AM can be anatomically classified using radiological imaging modalities. AM type 2 portosystemic shunt is characterized by the presence of a patent intrahepatic portal venous supply and a partial shunt (7). Type 1 can be further subclassified into type 1a and type 1b. Type Ia is characterized by separate drainage of SMV and the splenic vein into systemic veins; in type 1b, SMV and the splenic vein join to form a short extrahepatic PV, which drains into a systemic vein. This patient had AM type 2 with a side-to-side portocaval shunt between the left renal vein and the splenic vein. Congenital vascular malformations are frequently associated with congenital anomalies. Other anomalies have also been reported in patients with AM; these include chromosomal anomalies such as Down syndrome and structural anomalies of the cardiac defects, biliary atresia, polysplenia, and situs inversus (8-10). Hepatic shunt can also frequently present with hypoglycemia. This is attributable to the effect of defective glucose uptake and defective insulin secretion due to reduced hepatic degradation of the normal quantity of the secreted insulin (11, 12). AM can now be diagnosed using noninvasive abdominal imaging modalities such as USG, CT, and magnetic resonance imaging (MRI) (13). The imaging findings in patients with AM with hepatocellular carcinoma do not appear to be typical, that is, hypervascularity on the arterial-phase images with washout on delayed phase (14). Patients who do not exhibit typical findings of a benign lesion, i.e., lack of arterial enhancement or arterial enhancement without washout, should be closely followed up. Two groups according to the type of shunt those should be offered shunt closure either interventional embolization or surgical whereas those with type 1 shunts should be liver transplanted (15, 16).

CONCLUSION

Abernethy malformation is a rare congenital vascular malformation that can be diagnosed using abdominal imaging modalities (USG, CT, and MRI). We presented the case of a male who was incidentally diagnosed with AM type 2. Endovascular treatment should be the first-line treatment for this type of AM; if it is not successful, liver transplant should be considered.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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