

Lower gastrointestinal bleeding as a form of presentation in an adult case of Abernethy syndrome

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ABSTRACT

We report the case of a 29-year-old patient who presented with a short history of lower gastrointestinal bleeding. A diagnosis of Abernethy syndrome was made (congenital extrahepatic portosystemic shunt) after this uncommon clinical presentation. The prevalence of this congenital malformation is very low and usually manifests during the pediatric age, according to previously published reports.

CASE REPORT

A 29-year-old female presented with rectal bleeding. Colonoscopy revealed rectal varicose veins and additional studies were performed. Doppler ultrasound and abdominal computed tomography angiography showed a hypoplastic portal system, with a colateral portosystemic vein in confluence between the superior mesenteric vein and splenic vein, connecting to the left hipogastric vein via its distal end (Fig. 1). Two hepatic hemangiomas were diag-

nosed via abdominal magnetic resonance imaging (MRI) (Fig. 2). Type II Abernethy syndrome was confirmed based on these findings.

DISCUSSION

Abernethy syndrome is a congenital malformation characterized by agenesis or hypoplasia of the portal vein, causing an extrahepatic portosystemic shunt. It is classified in two types according to vascular relations (Table 1).



Fig. 1. Computed tomography angiography showing a portosystemic collateral vein.

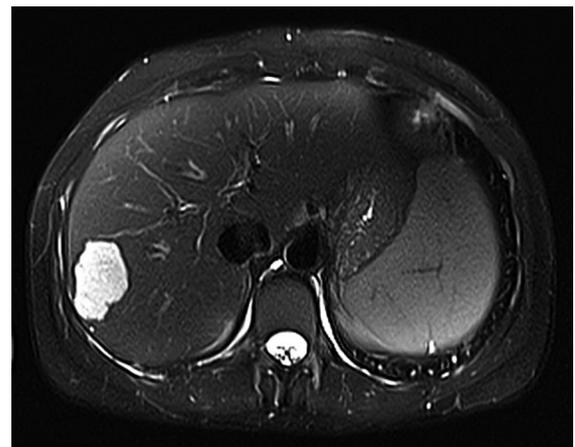


Fig. 2. Liver MRI revealed two hepatic hemangiomas in segments 6 and 7.

Table 1. Extrahepatic portosystemic shunt and vascular relations

Extrahepatic portosystemic shunt	Vascular relations
Type I	<ul style="list-style-type: none"> – The superior mesenteric and splenic vein drain separately into the inferior cava vein – The superior mesenteric vein and splenic vein form a common trunk before draining into the inferior cava vein
Type II	Hypoplasia of portal vein system

Type I is more frequent in females and is usually associated with vascular malformations and hepatic tumors. Type II has no sex predominance and no other malformations have been described. Hepatic encephalopathy and an altered liver profile are the most frequent clinical manifestations. Computed tomography angiography is the main radiological diagnostic test. Type II treatment in oligosymptomatic patients is conservative. Liver transplant is the only treatment described for type I Abernethy syndrome.

RECOMMENDED REFERENCES

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