# Portosystemic encephalopathy in an 86-year-old patient : a clinical challenge

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## Abstract

**Abernethy malformations are rare vascular abnormalities, classified into two types : type 1 if the portal vein is absent, type 2 when the portal blood is diverted into vena cava through a hypoplastic portal vein. These conditions present symptoms related to portosystemic shunting, and usually become clinically evident in children or young adults. Here we report the first case of Abernethy malformation diagnosed in an 86-year-old female patient affected by portosystemicencephalopaty.** (Acta gastroenterol. belg., **2016**, 79, **58-59**).

## To the Editor,

Congenital extra-hepatic portosystemic shunts (Abernethy malformations) are rare vascular malformations, classified into two types : type 1, if the portal vein is absent and the entire portal blood drains into the vena cava ; type 2, when the portal blood is diverted into the vena cava through a side-to-side extra-hepatic communication with a hypoplastic portal vein (1). These conditions may be asymptomatic, but clinical presentation is usually related to portosystemic shunting (hyperammoniemia, hepato-pulmonary syndrome), to the presence of other congenital abnormalities and to the increased neoplastic risk. Diagnosis is usually made in childhood, and only few cases affecting adults have been described (2-3).

An 86-year-old woman came to the emergency room for confusion and dyspnoea. Her personal medical history was positive for type II mellitus diabetes, hypothyroidism, and for a recent episode of deep venous thrombosis treated with success. An electrocardiography showed a chronic atrial fibrillation, while a neck Doppler ultrasound (US) revealed the presence of bilateral plugs in the internal carotid artery (stenosis : 50%). After an episode suggestive for transient ischemic attack, the patient was hospitalized in Stroke Unit ; cranial Computed Tomography (CT) scan showed no evidence of acute events. During observation, other episodes of confusion occurred ; thus, in order to evaluate a metabolic cause of the neurological presentation, ammonium was measured,

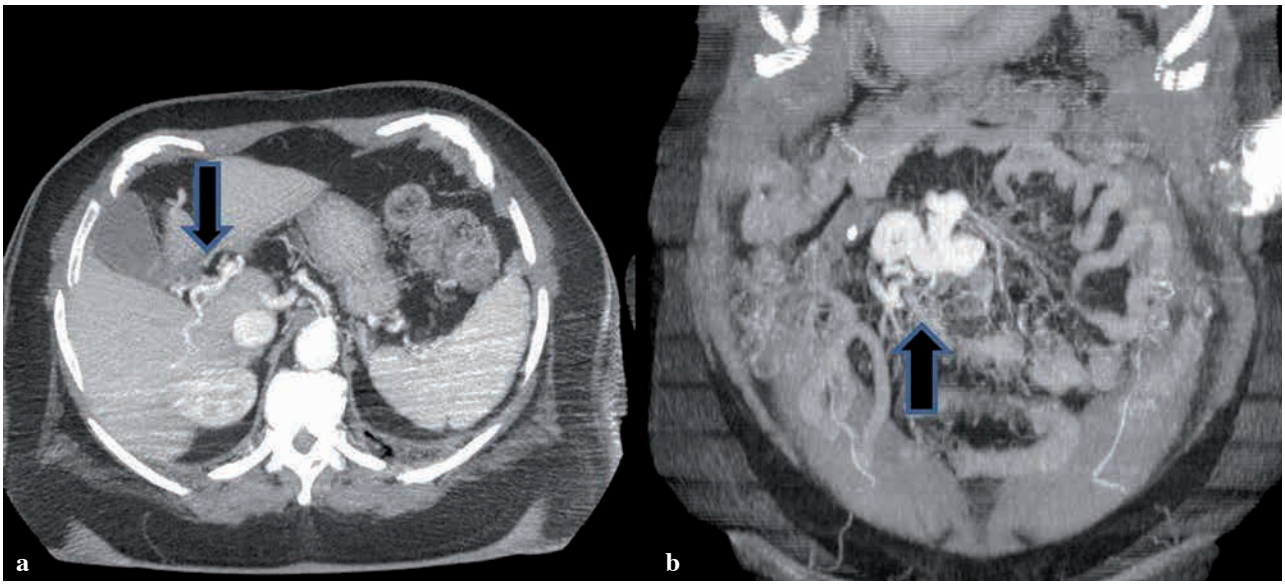


Fig. 1. —

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Submission date : 07/08/2015
Acceptance date : 17/08/2015

amounting for 174 umol/L (normal value : 11-51 umol/L). As the situation was compatible with hepatic encephalopathy, an abdominal US was performed to examine the liver features, but it was negative, as well as liver functional tests. A subsequent abdominal contrast-enhanced CT scan confirmed the normal appearance of the liver, but showed the presence of multiple collateral circles draining blood directly into the vena cava, in the absence of the portal vein. In details, at the hepatic hilum several vascular structures were visible, but neither portal vein nor portal vein branches inside liver parenchyma could be visualized (figure a, *arrow*) ; in the figure b, coronal maximum intensity projection CT angiogram image, with contrast material-enhancement, revealed complete portosystemic shunts that did not perfuse the liver via the portal vein. The aberrant anastomosis between the mesenteric superior vein and the splenic vein was identified, flowing through a common trunk by-passing the liver and draining into the inferior vena cava (*arrow*). Thus the diagnosis of Abernethy Syndrome type 1 was made.

The peculiarity of this case is the age of diagnosis (86 years) of Abernethy malformation ; in fact, a revision of the literature showed a total of 78 cases reported so far : 59 were aged < 18 years, 13 were represented by young adults (age : 19-30 years old), and only 6 were adults (age 31-68 years old). As our patient had remained asymptomatic for years, although having a congenital malformation, the message of this report is that when a clinical presentation with hepatic encephalopathy occurs in the absence of any radiological and biochemical sign of chronic liver disease, regardless the age of the patient, a congenital portosystemic shunt has to be suspected. This condition can be cured interventionally, even with liver transplant, in patients with good performance status ; however, our case was 86 years old, thus we only managed the complications of Abernethy malformation by enemas, lactulose and norfloxacin, in order to regularly move the patient bowel and treat the portosystemic encephalopathy ; she had a good response, episodes of neurological impairment decreased, no other related complications occurred.

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