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Major Pulmonary Arterial Hypertension Secondary to Congenital Portosystemic Intra Hepatic Shunt: A Case Report

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Abstract **Case Report**

Congenital portosystemic shunt, also known as Abernethy malformation, is a rare vascular malformation in which the portal blood drains towards the systemic circulation eluding the liver. These shunts arise through disturbances that intervene in the embryonic development of the splanchnic circulation, or when elements that are specific to the fetal circulation (especially those regarding the ductus venous) persist beyond the intrauterine life. We present the case of an 23 men years old, followed for ventricular septal defect (VSD) and pulmonary hypertension (PHA), who was addressed to our department to realize a Computed tomography pulmonary angiography (CTPA) for suspicion an acute pulmonary embolism (APE) due to a high d-dimer level. This examination does not show direct or indirect signs of pulmonary embolism but on the other hand objective a major pulmonary arterial hypertension associated with cardiomegaly secondary to a portosystemic intrahepatic shunt. An abdominal CT scan was requested in order to detail this vascular malformation and to objectify the presence of the portal trunk by the confluence of the spleno-mesaraic trunk and the inferior mesenteric vein with the demonstration of the communication of the right portal branch with inferior vena cava. Angio-CT should be the performed whenever a vascular malformation is suspected in order to establish a correct diagnosis, because portosystemic shunts carry a high risk of severe complications.

Keywords: case report, portosystemic shunt, pulmonary hypertension, vascular malformation.

INTRODUCTION

Congenital portosystemic shunt, also known as Abernethy malformation, is a rare vascular malformation in which the portal blood drains towards the systemic circulation eluding the liver [1, 3, 4]. These shunts arise through disturbances that intervene in the embryonic development of the splanchnic circulation, or when elements that are specific to the fetal circulation (especially those regarding the ductus venous) persist beyond the intrauterine life [1, 3, 5]. It is important to acknowledge congenital portosystemic shunts in order to closely follow-up these patients and identify any complications that might occur.

In this observatin we are reporting a case of a major pulmonary arterial hypertension secondary to Congenital portosystemic intra hepatic shunt.

CLINICAL REPORT

We present the case of an 23 men years old, followed for ventricular septal defect (VSD) and pulmonary hypertension (PHA), who was addressed to our department to realize a Computed tomography pulmonary angiography (CTPA) for suspicion an acute pulmonary embolism (APE) due to a high d-dimer level. This examination does not show direct or indirect signs of pulmonary embolism but on the other hand objective a major pulmonary arterial hypertension associated with cardiomegaly secondary to a portosystemic intrahepatic shunt. An abdominal CT scan was requested in order to detail this vascular malformation and to objectify the presence of the portal trunk by the confluence of the spleno-mesaraic trunk and the inferior mesenteric vein with the demonstration of the communication of the right portal branch with inferior vena cava.

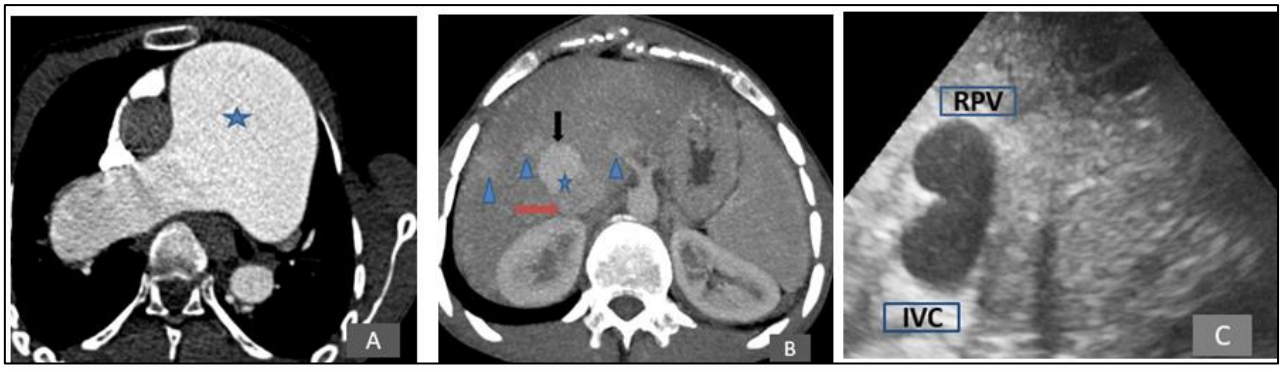

Figure 1: Axial CT image: (A) severe pulmonary hypertension (asterix). (B) porto-systemic shunt (asterix) between the right portal vein (RPV) (black arrow) and the inferior vena cava (IVC) (red arrow). There are associated nodular hepatic lesions (arrowhead) enhanced on portal time in relation to focal nodular hyperplasia. C: B-mode abdominal ultrasound showing communication between the IVC and the right branch of the portal trunk

DISCUSSION

From an anatomic point of view, portosystemic shunts are divided into intrahepatic shunts and extrahepatic shunts [1, 3-5]. For the intrahepatic ones the shunt originates from the portal branches, after the portal bifurcation. For the extrahepatic ones the shunt connects the portal trunk or its tributaries to a vessel belonging to the systemic circulation [4]. Type I portosystemic shunts are characterized by the complete absence of the portal vein, the portal blood draining entirely into the inferior vena cava [5]. Type I is further divided into IA—for which the splenic vein and the mesenteric vein separately drain into the inferior vena cava, and IB for which the portal vein itself drains into the inferior vena cava [4]. For type II portosystemic shunts the portal vein is unharmed, but there is an extrahepatic connection between the portal vein and the inferior vena cava [5]. Reviewing the literature, the malformation we identified in this case should be classified as intra hepatic portosystemic shunt, the patient has a portal trunk which drains into the inferior vena cava with its right portal branch.

The clinical presentation is reported to be variable, particularly in the pediatric population, ranging from asymptomatic to patients presenting symptoms related to different organs and systems—liver, respiratory system, central nervous system [3]. Characteristically, Abernethy syndrome patients have hypoxia and hepatopulmonary syndrome [5].

The abdominal angio-CT, considered the first choice examination [3], which was performed in our department, showed the presence of the portal trunk by the confluence of the spleno-mesaraic trunk and the inferior mesenteric vein with the demonstration of the communication between the right portal branch and inferior vena cava. In spite of the fact that the intestinal blood bypassed the liver and reached the systemic circulation through the shunt, which should have resulted in numerous complications (encephalopathy, pulmonary hypertension) [1, 7]. the patient was asymptomatic at all times, hence no treatment was performed, but he was followed-up closely in order to promptly identify any change in his status. In young patients with congenital portosystemic shunts, hyperammonemia without encephalopathy was reported. As possible explanations decreased sensitivity of the young brain to the effects of ammonia was proposed [8].

The presented patient was identified to have a nodular image in the left liver lobe, which was not biopsied, but imaging aspect was suggestive for focal nodular hyperplasia. Nodular hepatic lesions have been reported in patients with Abernethy malformation, most frequently benign. They appear to be caused by the increase of the arterial blood flow through the hepatic artery in context of the absence of the portal blood flow.

This circulatory malformation provokes the abnormal development and regeneration of the liver, hence the hepatic lesions [9, 10]. From a histologic point of view, focal nodular hyperplasia, regenereative nodular hyperplasia, hepatic adenoma, hepatoblastoma, and hepatocellular carcinoma have all been reported to be associated with the Abernethy syndrome [1].

Portosystemic shunts may result in complications such as portal hypertension, liver failure, encephalopathy (secondary to hyper-ammonaemia), development of hepatocellular tumors (either benign or malignant), hepatopulmonary syndrome, and pulmonary hypertension [11, 12]. Asymptomatic presentation, as in the presented case, is rare. Acknowledgment of these shunts is important before abdominal surgical interventions because there is a high risk of catastrophic bleeding when encountering unknown vascular malformations, possibly related to other anatomical malformations [12].

Treatment options depend on the shunt type and include conservative management or closure of the shunt—either trans catheter or surgical closure after balloon occlusion test, which helps to decide whether

there should be a 1-step or 2-steps closure of the shunt (small portal radicals, usually not visualized on ultrasonography, are allowed to enlarge by the 2-step procedure). In some cases, liver transplantation may be needed [12].

In our patient, given that he is persistently asymptomatic, we consider the conservative treatment to be the best choice.

CONCLUSION

Portosystemic shunts are a rare congenital abnormality without universal treatment guidelines. Abdominal ultrasound is usually the first examination performed that may detect a vascular malformation, but all lesions that have a vascular aspect should be further evaluated by angio-CT for a precise diagnosis. Early correct diagnosis is important for appropriate management, because portosystemic shunts carry a high risk of severe complications. Pulmonary hypertension.

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