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Appreciation of the treatment in adult patients with congenital portosystemic connections in relation with their symptoms

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INTRODUCTION
Portosystemic venous connections (shunts), also known as Abernethy syndrome, are rare malformations that anomalously connect the portal venous circulation and the central venous system (1). Depending on their location, they can be intrahepatic (intrahepatic portosystemic shunt [IPSS]) or extrahepatic; the latter is the most common presentation. Symptoms tend to manifest according to the amount of blood flow that the shunt leaves “unfiltered” by the liver.

The mechanisms behind these malformations remain elusive and therefore, the recommended treatment for these pathologies is still poorly defined (2-5). In children, when the pathology is symptomatic, it is associated with prominent alterations in cognitive development and requires treatment in all cases. However, there is less agreement about the recommended treatment for adult patients and therefore, treatments tend to be individualized depending on the symptomatology presented.
Here we present two cases of IPSS (which is the least common presentation of this pathology) and the treatments administered, which were based on the disease-associated symptoms presented by each patient.

**CASE REPORTS**

**Case report 1**
The case was a male patient aged 66 years with a history of diabetes mellitus, arterial hypertension (AHT) and dyslipidemia. He was under study for three weeks after presenting clinical manifestations of bradypsychia, poor concentration and confusion. The physical examination showed drowsiness, asterixis (flapping tremor) and splenomegaly. The presence of an organic brain pathology was ruled out as no analytical alterations were found. However, a characteristic pattern of encephalopathy appeared on the electroencephalogram (EEG). An abdominal Doppler ultrasound showed normal liver morphology and a dilation of the portal vein (PV) of 14.8 mm, which continued into a 23 mm fusiform dilation with a flow in the direction of the inferior vena cava (IVC). Computed axial tomography (CAT) and magnetic resonance imaging (MRI) scans confirmed the presence of a connection between the inferior vena cava and the right branch of the PV.

**Case report 2**
A 63-year-old female with a history of hypertension and dyslipidemia was studied due to chest pain, epigastralgia (upper abdominal pain) and anxiety. A cardiac MRI was performed which allowed us to discard the presence of heart disease. However, two liver masses were incidentally identified in the right hepatic lobe (the largest of which measured 33 x 24 mm). No analytical alterations were present. However, an abdominal ultrasound showed the presence of an aneurysmal lesion with a diameter of 2 cm in the area of the right PV branch that connected to the IVC and had portosystemic flow. A CAT scan confirmed these findings.

Both patients were diagnosed with IPSS Abernethy syndrome and we decided to treat the symptomatic patient (case 1). A percutaneous interventional radiology procedure was performed to retrogressively reach the IVC and the connection between these two
areas was identified and the shunt was selectively canalized. Both the portal and hepatic ends were sealed with self-expanding cylindrical vascular plug devices (Amplatzer™ vascular plugs [AVPs]) to completely isolate the shunt. The closure was confirmed by venography.

No complications occurred during the procedure and the patient showed a clinical improvement which allowed his discharge on the fourth post-surgical day. The follow-up at eighth months with ultrasound showed a complete closure of the shunt and a decrease both in PV diameter and in splenomegaly. To date, no clinical or complementary-test alterations associated with the shunt have been identified during follow-up.

The asymptomatic patient (case 2) continues to be controlled via outpatient consultations and has been in follow-up for four years. Thus far, he has not presented any symptoms or analytical alterations and ultrasound and CAT imaging tests indicate that the shunt remains stable.

**DISCUSSION**

IPSS is an abnormal and direct connection between the PV and IVC or hepatic veins whose diameter usually exceeds 1 cm and is extremely rare (5). It may be congenital in origin, or the result of a trauma, cirrhosis or PV hypertension. Park et al. suggested classifying IPSS into four categories based on their locations (Table 1) (6). According to this classification system, the cases reported here were type 3 (aneurysmal) and these IPSSs normally have a characteristically saccular or fusiform morphology.

The clinical manifestations of IPSS depend on the volume, duration and magnitude of the shunt. In fact, most IPSS spontaneously close during childhood without presenting clinical manifestations. If they remain open, patients do not usually present symptoms for decades or may even remain asymptomatic for their entire lives. In adults, IPSS should be suspected if neurological symptoms appear that are suggestive of hepatic encephalopathy, in the absence of signs of cirrhosis or liver disease (5,6). However, some cases have been described in which unspecific symptoms such as rectal bleeding were the first symptoms to arise (7).
The most valuable diagnostic tool is Doppler ultrasound (8). This type of analysis usually identifies blood-flow alterations such as anterograde flow in the shunt region or the loss of the normal venous waveform morphology at the level of the PV and adjacent central venous system. CAT (venous phase) and MRI imaging are used to confirm the diagnosis and establish the anatomy of the shunt. Angiography is used to plan the embolization.

Treatments for IPSS are not standardized and are determined by the presence of symptoms and risk factors (e.g., high flow during childhood [4]). Percutaneous embolization using stainless steel or platinum coils has been described as a treatment method and the experience with cases of shunts caused by portal hypertension due to liver disease has also been published (9). Evans et al. (10) advanced this procedure, describing the closure of both the portal and hepatic shunt origins with the placement of AVPs. AVPs allow a greater, more precise control over the closure and they less frequently become displaced. Despite this, they are not used routinely in every center (5). With regard to the possible complications derived from this procedure, Tanoue (1) reported PV occlusion caused by embolization and thrombosis of the right portal branch as a result of the manipulation process.

In conclusion, these two cases present our experience with a type of portosystemic shunt that is rare in adults and may be the cause of a neurological clinical picture of hepatic encephalopathy (9). Angiographic embolization with AVPs is a minimally-invasive procedure that allows the treatment of the shunt and results in almost immediate biomechanical and clinical improvements upon installation (1,9,10). The use of vascular plugs allows the rapid and safe occlusion of these shunts and is clearly beneficial for symptomatic patients (10), making it the treatment of choice in these individuals. In contrast, the treatment of shunts in asymptomatic patients is more controversial and in our case, we preferred to continue to follow-up the evolution of the patient (5). However, as very little data regarding the mechanisms and best treatment practices for this disease are available, larger studies which collect long-term results will be required to allow IPSS treatment to be standardized.

REFERENCES


Table 1. Classification established by Park et al. for portosystemic shunts

<table>
<thead>
<tr>
<th>Park classification for intrahepatic portocaval shunts</th>
<th>Description</th>
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<tbody>
<tr>
<td>Type 1 (most frequent)</td>
<td>Long connection between the right portal vein and the inferior vena cava</td>
</tr>
<tr>
<td>Type 2</td>
<td>Single connection between a terminal branch of the portal vein and the inferior vena cava</td>
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<tr>
<td>Type 3</td>
<td>Communication between a peripheral branch of the portal vein and a peripheral branch of the inferior vena cava</td>
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<tr>
<td>Type 4</td>
<td>Multiple diffuse branches between a peripheral branch of the portal vein and the hepatic veins in several segments of the liver</td>
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Fig. 1. Diagnostic images of the shunt from computed axial tomography (CAT) and ultrasonography imaging. Images 1 and 2 show evidence of the anomalous fusiform connection presented by case 1 in the venous phase of the CAT imaging. Image 3 shows Doppler ultrasonography images of the shunt in which the characteristic three-phase flow can be seen. Finally, image 4 shows the connection (shunt) in case 2 in the arterial phase of the CAT imaging, which is very similar to that presented in case 1.
Fig. 2. Images after the treatment of case 1, in which the complete isolation of the shunt with two Amplatzer™ plugs can be seen.