

Received: 2011.06.21
Accepted: 2011.08.24

A case of a spontaneous splenorenal shunt associated with the nutcracker syndrome

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Summary

Background:	Hypertension in the portal vein system stimulates the vascular system to develop a porto-caval collateral system, which occurs by adaptation of the already existing junctions between the porto-caval and the systemic circulation to changed conditions or by angiogenesis.
Case Report:	We present a case of a 42-year-old man with alcoholic cirrhosis of the liver and exacerbation of chronic pancreatitis, whose computed tomography of the abdominal cavity revealed a massive varicoid dilatation of a vein of the spleen with a splenorenal shunt and tortuous dilated venous vessels in the retroperitoneal space and mesentery.
Conclusions:	The unusual image of the mesenteric and retroperitoneal venous vessels in our patient resulted from the coexistence of collateral porto-systemic circulation and compression of the left renal vein.
Key words:	liver cirrhosis • portal hypertension • porto-caval collateral circulation • spontaneous splenorenal shunt • nutcracker syndrome • computed tomography (CT)
PDF file:	http://www.polradiol.com/fulltxt.php?ICID=882145

Background

Portal hypertension is defined as an increase in the blood pressure in the portal vein of over 12 mmHg and in the gradient between pressure in the portal vein and pressure in the inferior caval vein of over 2–5 mmHg. The increased pressure in the portal system may result from a higher resistance, an increased flow, or both [1]. In the European population, the main cause of portal hypertension is posthepatic and alcoholic cirrhosis [2].

Additional connections of the portal system with the systemic circulation constitute a significant feature of the portal system and are formed by gastro-esophageal veins, anal veins, periumbilical veins, recanalized falciform ligament, splenic venous bed, left renal vein and vessels of the retroperitoneal space [3]. The development of the porto-caval collateral system is a reaction of the vascular system to high portal pressure which consists in adaptation to the changed conditions by existing vessel junctions or in creation of new vessels in angiogenesis. The formation of the collateral circulation induces a decrease in portal pressure, load of cardiac muscle and degree of penetration

of intestinal toxins, without liver enlargement which can result in complications such as cardiomyopathy or hepatic encephalopathy [4].

Case Report

A 42-year-old male, with alcoholic cirrhosis of the liver of grade B/C according to the Child-Pugh classification, chronic pancreatitis, post-hepatitis B and history of surgery of pyloric ulcer perforation, was admitted to the gastroenterology ward with severe stomach-ache and vomiting.

Laboratory tests revealed an increase in the level of amylase, liver enzymes and C-reactive protein. The ultrasound examination raised a suspicion of inflammatory tumor of the pancreatic head with infiltration of the duodenum. The patient was subjected to computed tomography, which confirmed the presence of the inflammatory pancreatic tumor in the course of chronic pancreatitis with duodenum compression, which caused a temporary obstruction.

The computed tomography also revealed a massive varicoid dilatation of a vein of the spleen and area around it, with



Figure 1. CT examination, MPR reconstruction, coronal view showing dilatation of the proximal left renal vein; abnormal, dilated vessels in the retroperitoneal space.

a splenorenal shunt, dilatation of the superior mesenteric vein to the width of 1.6 cm, and varices in the lower part of the oesophagus. The portal vein and its main branches had a correct size; the diameters were 1.3 cm and 0.9 cm, respectively. In the retroperitoneal space and mesentery, there was a conglomerate of tortuous veins dilated to an average diameter of 3 cm, joined with the dilated left renal vein and superior mesenteric vein (Figure 1). The left renal vein was dilated to 3.2 cm in renal hilus and much narrower in its further course, which was probably caused by its compression between the aorta and the superior mesenteric artery (the nutcracker syndrome) (Figure 2).

A conservative therapy supplemented by electrolytes and proteins, and parenteral nutrition was introduced, which resulted in a significant recovery.

Two months later, a control CT examination was conducted. It showed a haemorrhage to the cyst in the head of the pancreas and swelling of the mesentery with increased density of the mesenteric adipose tissue. The anomalies of the abdominal organs and vessels remained unchanged.

Discussion

In hepatic cirrhosis, the collateral porto-caval circulation develops in several regions. Clinically, the most important ones are the oesophageal and gastric varices. Bleeding from ruptured oesophageal varices is one of the most frequent and life-threatening complications [3]. The collateral circulation in the abdominal integument or anus is less significant. Venous junctions in the retroperitoneal space seem to be hemodynamically important in portal system compensation but they are less accessible and less frequently examined [5]. Another way of decompression of the congested portal system is the formation of a spontaneous splenorenal shunt. Similar splenorenal connections are created in the surgical treatment of portal hypertension, i.e. Warren's operation. It is one of the procedures that decompresses the system selectively and serves to reduce hepatic

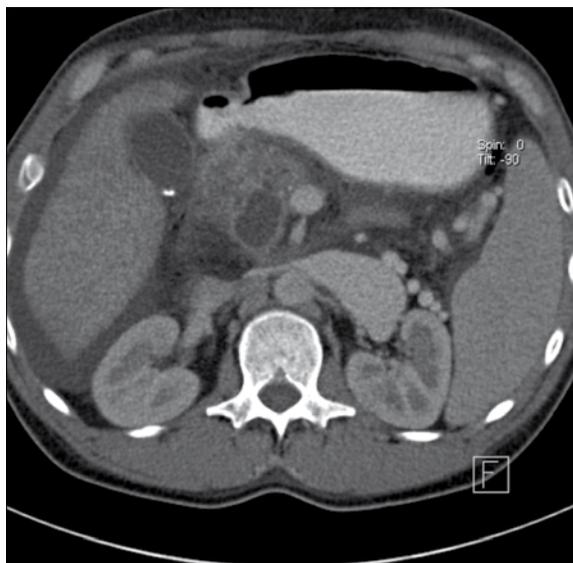


Figure 2. CT examination, axial view showing dilatation of the proximal left renal vein and compression of the left renal vein between aorta and SMA (nutcracker syndrome). Perihepatic fluid collection and splenomegaly.

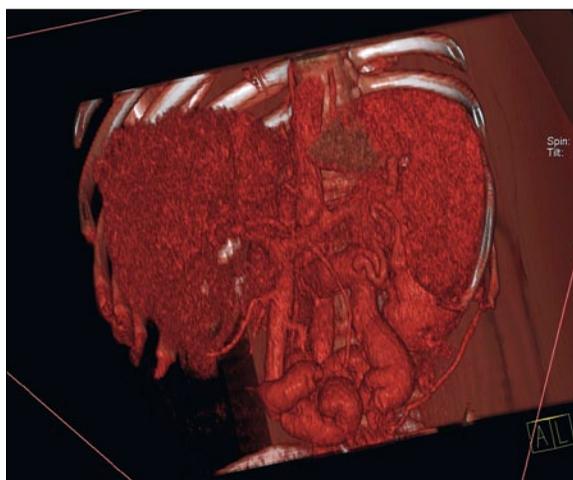


Figure 3. CT examination, VRT reconstruction showing splenorenal shunt, dilatation of the proximal left renal vein and tortuous, dilated collateral vessels in the retroperitoneal space, draining into the wide mesenteric vein.

encephalopathy and preserve liver functions [6]. A large, spontaneous splenorenal shunt may manifest as a palpable, hard, non-pulsating mass in the upper left quadrant of the abdomen [7]. It is often accompanied by splenomegaly, although a close correlation between them has not been demonstrated [8].

Spontaneous splenorenal shunts often appear in patients with hepatic cirrhosis. A. Von Herbay et al. using the color Doppler sonography found spontaneous portosystemic shunts in 38% of patients from a group of 109 cases with hepatic cirrhosis. These were mostly splenorenal shunts (21%) [9]. In the presented case, this mechanism was most probably responsible for the correct diameter of the portal vein. The atypical image of the venous cluster in the retroperitoneal space and mesentery was connected with

obstructed blood flow from the renal vein to the inferior vena cava, and to its compression between the abdominal aorta and the superior mesenteric artery (probably additionally raised by the surrounding tissue turgescence in the course of aggravation of chronic pancreatitis) (Figure 3).

The above-mentioned compression of the left renal vein, known as the nutcracker syndrome, leads to the dilatation of all renal vein tributaries, with secondary formation of a plexus of abnormal renal mucosal varicosities whose rupture is responsible for hematuria. Other symptoms which can accompany the described pathology include: left-sided varicocele, orthostatic proteinuria, hypertension, abdominal pain or chronic fatigue [11]. Treatment, stenting or open surgical interventions should be based on the occurrence or lack of the symptoms, their intensification and expected reversibility with regard to patient's age and medical condition [12].

Modern diagnostics of portal hypertension is primarily based on three methods: Doppler ultrasonography with vascular-flow imaging, dynamic helical computed tomography and angiography. Multi-Slice Computed Tomography

(MSCT) is one of the most important methods of imaging diagnostics of portal hypertension. It enables a precise evaluation of the abdominal parenchymal organs in the arterial, venous and parenchymal phase, evaluation of vascular structures for their width, patency or even the direction of the flow, and different types of collateral circulation. Thanks to the volume rendering technique (VRT) and multiplanar reformats (MPR), it enables spatial visualisation of the examination range. As a non-invasive and repeatable method, simple to interpret and clear for the surgeon, it allows to quickly establish the diagnosis and monitor treatment precisely [13].

Conclusions

1. The unusual image of mesenteric and retroperitoneal venous vessels results from the coexistence of collateral porto-systemic circulation and compression of the left renal vein.
2. The computed tomography examination in cirrhosis of the liver enables to estimate the degree of advancement of the porto-caval collateral system and possible complications of cirrhosis (secondary HCC).

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