Splenic vein enlargement, a rare cause of nutcracker syndrome

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Abstract

Nutcracker syndrome refers to symptomatic compression of left renal vein. There are many reasons for that. We report a case that the enlargement of splenic vein has caused to nutcracker syndrome. Results implies that not firm venous structures can be cause of nutcracker syndrome.

Key clinical message

Abnormal enlargement of the splenic vein is one of the etiologies of Natcracker syndrome that should be considered when examining the causes of this syndrome. Because knowing rare etiologies can help correctly diagnose Natcracker syndrome and reduce its mortality.

Introduction

Nutcracker syndrome (NCS) is the symptomatic clinical condition in which of left renal vein (LRV) becomes compressed usually between abdominal aorta and superior mesenteric artery (SMA). NCS is important because secondary chronic LRV hypertension may lead to chronic renal disease or renal vein thrombosis (1). The most common cause of NCS is the short distance between SMA and abdominal aorta (1-4). Other reported causes are retroperitoneal pathologies as tumors or lymphadenopathies. Venous causes are very rare but splenic vein enlargement has not been reported. (1,3,5). NCS can happen at any age from childhood to seventh decade (6-11) with peaking spread in middle age adults (10,11). In this case, NCS is secondary to compression effect of enlarged splenic vein on LRV.

Case history

A 55 y/o woman, known case of hairy cell leukemia complained from intermittent abdominal pain (mainly in left flank), nausea, weight loss and loss of appetite from 3 months ago. The patient‘s family history was unremarkable and has undergone chemotherapy treatment from four months ago. Physical examination positive findings were huge splenomegaly & cachexia. Her height was 140 centimeter and her weight was 35 kilograms (BMI=17.85). Vital signs were normal. Laboratory positive finding was: HB= 8.1 g/dl. Urine analysis was normal.

In abdominal ultrasound, Heterogeneity and increase in splenic parenchymal echogenicity, huge splenomegaly as spleen span of 210 mm was detected.

Computerized tomography scan (CT scan) confirmed huge splenomegaly and heterogeneity of splenic parenchymal density that indicated splenic infarction. Splenic vein was markedly enlarged in diameter of 19 mm which had compression effect on LRV. Marked prominency of both gonadal veins and congestion of pelvic veins bilaterally was detected which represents that left gonadal vein and bilateral pelvic veins are

drainaged via right gonadal vein and this is secondary to LRV compression. These findings lead us that NCS had been occurred.

CT scan showed the normal angle and distance between the Abdominal Aorta and SMA. No retroperitoneal pathology was shown. Both kidneys were normal. Mild ascites, Mild hepatomegaly and evidence of secondary portal hypertension were also noticed. The patient was referred to surgery department for splenectomy to reveal her symptoms.

Discussion

NCS should be considered in differential diagnosis of patients with intermittent flank pain. According to this case, abnormally enlarged venous structures such as splenic vein which don't have a firm consistency can be mentioned as unusual causes of NCS.

The causes of NCS are divided to three main groups:

1. Arterial causes: the most common cause of NCS is proximity of SMA to abdominal aorta (1-4). Also other arterial causes are abdominal aortic aneurysm, overarching testicular artery and ectopic ventral right renal artery

2. Retroperitoneal tumors or pathologies such as pancreatic neoplasm, chronic pancreatitis, para-aortic lymphadenopathy and decreased retro-peritoneal mesenteric fat tissue

3. Venous causes (that are very rare): LRV duplication, left renal ptosis, Left-sided IVC, hemi-azygos continuation and persistent left superior vena cava combination (as high pressure veins) (1,3,5). Splenic vein enlargement has not been reported as a cause of NCS and this is the first time that an enlarged organic vein is reported as a cause of NCS.

All of the mechanisms involved in LRV compression lead to LRV outflow obstruction (1).

In our case SMA syndrome was one of the differential diagnoses due to rapid weight loss and nausea but imaging data ruled it out (12). CT scan findings showed huge splenomegaly, splenic infarction, and enlarged splenic vein with compression effect on LRV (NCS) (Figure 1). Secondary prominency of both gonadal veins and pelvic congestion were also noticed that represented drainage of these veins through right gonadal vein in the setting of compression of LRV in NCS (Figure 2). The interesting and unique point of our case is a compression effect on LRV that has been made by an enlarged venous structure (enlarged splenic vein) without having a firm or muscular consistency, instead of the arteries or other solid pathologies. In a similar case report, the massively dilated common bile duct which hasn't a firm structure was reported as an unusual etiology for NCS (13). The most common symptoms and signs of NCS are abdominal pain, left flank pain and hematuria (14).

As the another interesting point, abdominal and left flank pain existed in our case while hematuria as a main clinical manifestation of NCS wasn't obtained (15).

NCS is an important diagnosis due to the significant morbidity associated with it, including the risk of chronic renal disease from long-term LRV hypertension and thrombosis (1).

Knowing about the rare etiologies can help in accurate diagnosis of NCS, which will lead to a reduction of its morbidity. To our knowledge, abnormal enlargement of a venous structure has not been reported as an etiology for NCS.

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