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The Posterior Nutcracker Syndrome as a Rare Cause of Abdominal-Pelvic Pain in Pregnancy of 32 Weeks of Amenorrhea: About a Case

M. Bendahhou Idrissi[1], H.Mochtari[1], K. Saoud[1], N. Mamouni[1], S. Errarhay[1], C. Bouchikhi[1], A. Banani[1], M. Ezzahi[2], M. Boubbou[2]

[1] Department of Gynecology and Obstetrics, Hassan II University Hospital, Fez, Morocco.
[2] Mother and Child Radiology Department

Abstract:- Nutcracker syndrome includes all manifestations related to venous stasis induced by stricture of the left renal vein (LRV) in the fork formed by the abdominal aorta and the superior mesenteric artery (SMA), leading to stenosis of the aorto-mesenteric part of the left renal vein and dilation of its distal part. The symptomatology is dominated by lumbar, abdominal and pelvic pain and microscopic hematuria. This syndrome can be aggravated during the pregnancy from 16 SA, this is due to the increase in renal plasma flow. Its diagnosis is based essentially on modern imaging means (CT scan, Doppler ultrasound, phlebography) and its treatment is controversial. We report the case of a woman aged 37 years, admitted with acute abdominal-pelvic pain with vomiting complicated by severe hypotension in a pregnancy of 32 weeks of amenorrhea in whom imaging revealed an inaugural nutcracker syndrome discovered for the first time during this pregnancy.

Keyword:- *Nutcracker Syndrome, Left Renal Vein, Pelvic Varices, Superior Mesenteric Artery, Ascites, 3rd Trimester Pregnancy.*

I. INTRODUCTION

The nutcracker syndrome encompasses all clinical manifestations related to venous stasis induced by stricture of the left renal vein (nutcracker phenomenon): either between the aorta and the superior mesenteric artery (anterior nutcracker syndrome), or between the aorta and the spine when the left renal vein passes abnormally behind the aorta (posterior nutcracker syndrome [1]). Pelvic venous stasis is associated in at least half of the cases, due to distension of the left ovarian (or spermatic) vein, which drains into the left renal vein. An aggravation of this ovarian distension has been described during pregnancy, particularly during the 2nd and 3rd trimesters [6].

Although this syndrome, described as early as 1950, should be familiar to urologists and nephrologists because of its repercussions on the left kidney (hematuria, proteinuria, orthostatic hypotension), it should be better known by gynecologists and obstetricians. It can indeed be revealed by atypical abdominal pain during pregnancy, often leading to a misdiagnosis of acute unexplained peritoneal pain, leading to the erroneous incrimination of a visceral etiology. We report the case of a 37 year old woman who presented with acute abdominal pain with vomiting complicated by severe hypotension in a pregnancy of 32 weeks' gestation. The diagnosis was retained in view of the scanographic signs.

II. CASE REPORT

This is a 37 year old patient, without any notable pathological history, admitted for abdominal pain associated with vomiting during a pregnancy of 32 SA, G5P4, the first 3 pregnancies well followed with normal course, carried to term with vaginal delivery, the 4th pregnancy followed, The fourth pregnancy was marked by the onset of vomiting that was resistant to treatment throughout the pregnancy, complicated by the appearance of ascites in the third trimester, for which a CT scan was indicated by the patient's attending gynecologist, but not performed by the patient. The pregnancy was carried to term with a vaginal delivery, and the evolution in the post-partum period was marked by a spontaneous improvement in clinical symptoms and the disappearance of the ascites The 5th pregnancy is the current pregnancy estimated at 32 SA poorly monitored marked by the installation of episodes of vomiting for 2 months until a week before his admission to the emergency room where the patient presented an aggravation of the symptomatology by the increase of episodes of vomiting and the occurrence of abdominal pain motivating his consultation in our training for Management. In whom the examination on admission to the emergency room found a conscious patient, hypotensive to 90/50mmHg, tachycardic to 110bpm, apyretic to 37ºC, The rest of the somatic examination was without particularities especially cardiac and pulmonary.

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An abdominal ultrasound was requested, but it did not reveal anything other than ascites, especially no hepatopathy. Given the hemodynamic instability and unexplained peritoneal suffering, a surgical cause was evoked, hence the decision to perform an abdominal CT scan C-/C+ given the unavailability of abdominal MRI in emergency.

Abdominal and pelvic CT showed a monofetal pregnancy responsible for a mass effect on the inferior vena cava, which was collapsed (Figure 1), with individualization of multiple pelvic serpiginous venous structures, extended perirenally, related to varicose veins, draining into the ovarian veins dilated because of the pregnancy (Figure 2).

The CT scan also showed the passage of the left renal vein between the anterior aspect of the vertebral body of L2 and the subrenal abdominal aorta, compatible with a nutcracker syndrome in its posterior variant (Figure 1), with intraperitoneal effusion of moderate abundance at the perihepatic level, peri-splenic level, the two parieto-colonical gutters and at the pelvic level.

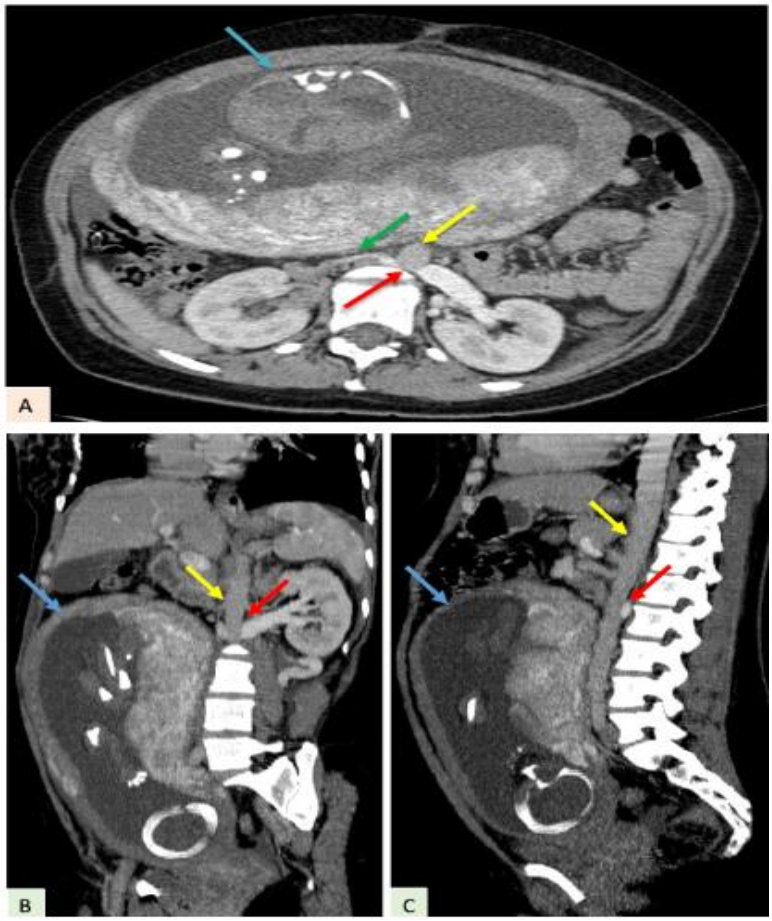

Fig 1: Axial (A) coronal oblique (B) and sagittal (C) sections of the abdominal-pelvic CT scan of our patient after injection of PDC (contrast medium), objectifying the passage of the left renal vein (red arrows) between the posterior aspect of the aorta (yellow arrows) and the anterior aspect of the vertebral body, which then drains into the inferior vena cava (green arrow) which is collapsed due to compression by the gravid uterus (blue arrows).

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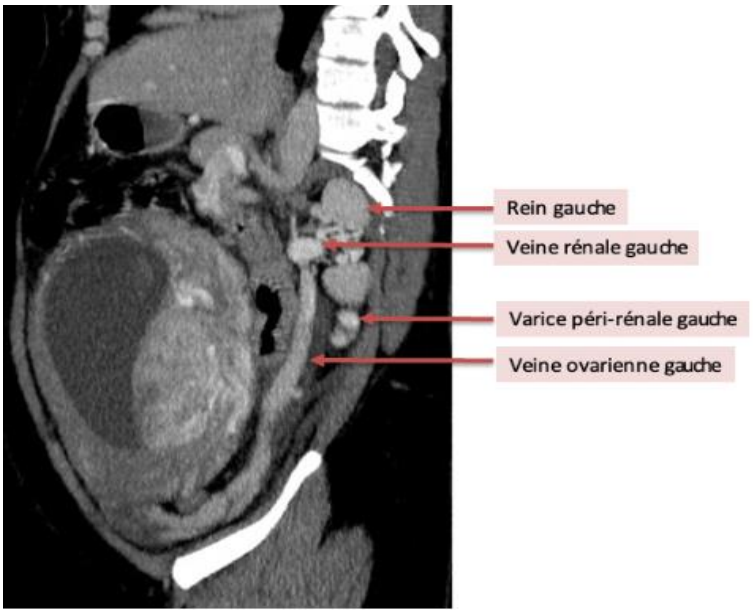

Fig 2: Sagittal section of our patient's abdominal-pelvic CT scan after injection of PDC, with MIP (maximal intensity projection) reconstructions, showing a dilated left ovarian vein, flowing into the left renal vein, with peri-renal varices.

III. DISCUSSION

The left renal vein must pass in front of the aorta to join the inferior vena cava, slipping under the superior mesenteric artery and behind the third portion of the duodenum. The left renal vein is therefore much longer (6 to 10 cm) than the right (Figure 3). The left renal vein also receives the left adrenal vein, the gonadal vein (ovarian or spermatic), the ureteral vein, the second lumbar vein, and sometimes the third (Figure 3). These veins normally have anti-reflux valves (ostial, inconstant), whose effectiveness may be altered in the event of overpressure in the left renal vein [4] or following repeated pregnancies, and this was the case in our patient who was multiparous 5th gesture.

Nutcracker syndrome (NSS) refers to compression of the GRV as it passes through the aorto-mesenteric clamp. It is a rare entity, but probably underestimated. Its prevalence is higher in young subjects between 30 and 40 years of age, with a more frequent involvement in women [1,2]. Entrapment syndrome can be divided into 3 types: anterior, posterior and mixed (Figure 4) [3]. Anterior SCN: found in the majority of cases, corresponds to compression of the RVG, normally located between the aorta and the MSA; whereas the posterior variant, refers to the passage of the RVG retro-aortically in the small space between the posterior aspect of the abdominal aorta and the anterior aspect of the spine [4] (Figure 4) and this is the case of our patient who presents with a nutcracker syndrome in its posterior variant (Figure 1).

The pathophysiology of SCN remains unknown, but several hypotheses have been put forward: anatomical variants [5,6]; duplicity of the left renal vein; ectopic or horseshoe kidneys; ectopic birth of the spermatic and ovarian arteries may also constrict the renal vein; All situations of overload/hyperpressure of the venous network (vena cava more than portal) may contribute to the increase or appearance of signs. Blood drainage occurs via retroperitoneal collaterals and countercurrently via the ovarian (or spermatic) vein to the pelvic venous network and then the vena cava. Although more a consequence than a cause of nutcracker syndrome, stasis in the pelvic network may therefore also facilitate its expression. The female predominance of nutcracker syndrome in adults could be explained by the fact that venous overpressures related to pregnancy can alter the valves of the gonadal veins [7]. A nutcracker syndrome may even occur on the right side during pregnancy due to compression of the right renal vein by the gravid uterus [6]. Extrinsic causes of renal vein compression can induce secondary nutcracker syndromes, including pancreatic cancer, retroperitoneal tumors, and para-aortic adenopathies [6].

The most common clinical manifestations are varied, induced by venous stasis upstream of the RVG:

- Asymptomatic microscopic hematuria or even macroscopic hematuria, our patient had microscopic hematuria.
- Orthostatic proteinuria: This can be massive (more than 400 mg/dL after standing for at least 15 minutes) [7,15], and is due to lysis of red blood cells in the ureter [6],
- Gynaecological signs have also been reported including dyspareunia, post-coital pain [16], unexplained heaviness or dysmenorrhoea [7].
- Cardiovascular and adrenal signs: Stasis in the renal vein may adversely affect the left adrenal gland. Insufficient cortisol secretion has been reported, which may facilitate a picture of chronic fatigue and orthostatic discomfort. The nutcracker syndrome is thought to account for 70% of severe orthostatic hypotensions [17].

- Digestive signs: The third portion of the duodenum may also be pinched (lower than the renal vein) between the aorta and the superior mesenteric artery, which may cause the combination of epigastric pain, nausea and weight loss with the other signs of nutcracker syndrome [18]. This is referred to as Wilkie's syndrome [2]. Abdominal pain can sometimes be severe enough to induce repeated syncope [19]. In this case, it is necessary to rule out stricture of the celiac trunk by the arcuate ligament (Dunbar's syndrome) [2].

All these symptoms are highly variable and sometimes difficult to correlate with anatomical findings: some subjects with marked compression of the RVG are completely asymptomatic [8]. The diagnosis of SCN relies essentially on modern imaging means. The multibarrier scanner, with its multiplanar acquisitions, offers a definite advantage for the establishment of the diagnosis by objectifying different criteria in particular: compression of the RVG in the space formed by the superior mesenteric artery and the aorta, distension of the gonadal veins and pelvic congestion. Some authors have therefore attempted to validate these criteria, including Kim et al. designating [9] Bec's sign very suggestive on axial sections (compression of the RVG in the aorto-mesenteric fork) with a specificity of 88.9%; angulation between the aorta and the MSA (<41°) with a specificity of 55.6%, RVG diameter ratio (hilar-aorto-mesenteric ratio) >4.9 with a specificity of 100%; gonadal vein distension and pelvic congestion.

Despite the great contribution of CT in the diagnosis of Nutcracker syndrome by its high accuracy of anatomical parameters and its non-invasive character, it presents non-negligible constraints nowadays such as exposure to radiation and risks of allergy to contrast media [6]. If the diagnosis remains uncertain, the pressure gradient between the RVG and the inferior vena cava (IVC) plays an important role in the diagnosis of nutcracker syndrome, it is measured during a Doppler examination but phlebography remains the most accurate method (both remain the gold standard) [10]. According to literature reviews, 1 mmHg corresponds to the normal pressure gradient, Beinart et al. designated that a pressure gradient of 1 mmHg or more indicates RV hypertension [11,12]. Doppler ultrasonography is a technique that can be very useful to confirm the diagnosis if it can be shown that the ratio of maximum RVG velocities at the level of the stenosis to maximum upstream distension is greater than or equal to five (sensitivity 69-90% and specificity 89-100%) [13].

Regarding the treatment, it remains very controversial. In addition to surgery (transposition of the RVG and/or AMS) indicated in case of severe pain and massive hematuria, some teams perform an endovascular treatment. Otherwise, therapeutic abstention is the rule, as in the case of our patient (where the symptomatology becomes severe during pregnancy) [14].

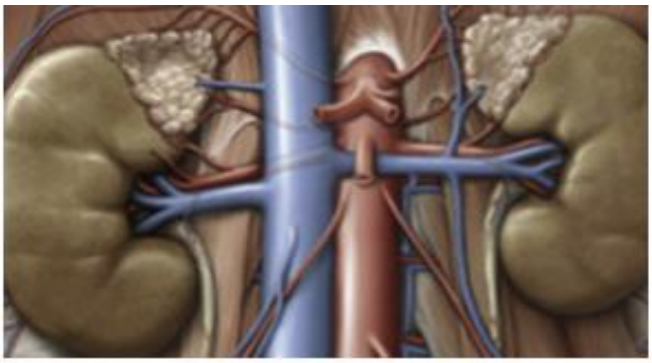

Fig 3: Differences between the left and right renal veins. The left renal vein, which is much longer than the right, must pass in front of the aorta and under the superior mesenteric artery to join the vena cava, which can "pinch" it. The other difference is that the left renal vein receives the left adrenal vein and especially the left genital vein (ovarian or spermatic), as well as the drainage veins of the lumbar veins.

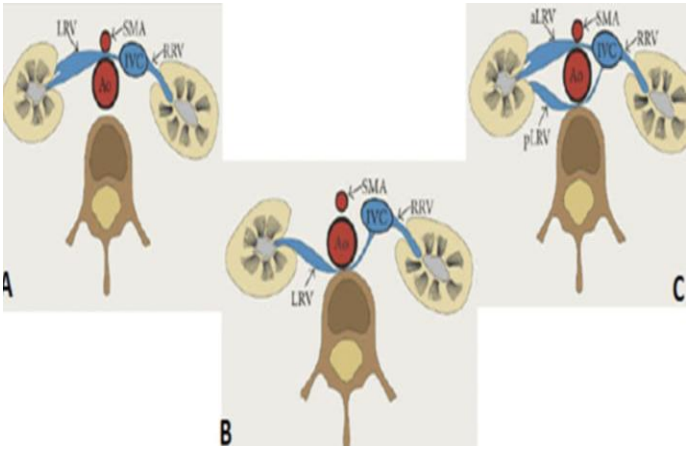

Fig 4: Anatomic diagram showing the different variants of Nutcracker syndrome; A) anterior variant (most frequent) corresponds to compression of the normally located GRV by the aorta and MSA; B) posterior variant designating retro-aortic passage of the GRV in a small space between the abdominal aorta and the vertebral column; C) mixed variant, including the combination of both forms in the context of a circum-aortic or double left renal vein.

IV. CONCLUSION

The Nutcracker syndrome is a rare entity, to be considered in the diagnostic range of rare etiologies of unexplained abdominal pain in young subjects, especially if it occurs during the 2nd or 3rd trimester of pregnancy and is accompanied by hematuria. However, this diagnosis should only be made on the basis of a set of clinical arguments and the demonstration of a marked excess of pressure in the left renal vein, as the management of this syndrome is not yet well codified (surgery, endovascular treatment or abstention).

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