Pelvic venous congestion syndrome, varicocele and duodenal compression, due decreased aortomesenteric angle: Pictoral review.

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Learning objectives

1. Illustrate the pathologies associated to the reduction of the aortomesenteric angle, including Superior Mesenteric Artery syndrome and Nutcracker syndrome, as manifested by:
   a. Duodenal compression
   b. Pelvic venous congestion
   c. Varicocele

2. Review the pathophysiologic characteristics and clinical findings of Superior Mesenteric Artery Syndrome and Nutcracker syndrome.

3. Highlight the imaging findings in the evaluation of these pathologies.
Superior mesenteric artery syndrome (SMAS) is an uncommon gastrovascular disorder resulting obstruction of the proximal intestinal. Affected patients are characterized by a decreased angle between the overlaying superior mesenteric artery and underlying abdominal aorta, resulting in compression of the third portion of the duodenum.

Nutcracker syndrome is other pathology caused by the same anatomic feature, the reduction in the aortic mesenteric angle (AMA), causing compression of the left renal vein, Resulting in an increase in venous pressure secondary to the obstruction of venous outflow into the inferior vena cava, Which may manifest as varicocele or Congestive Pelvic syndromen in men and women, respectively.
Findings and procedure details

Anatomy and pathogenesis

The superior mesenteric arises from the anterior surface of the abdominal aorta, approximately 1 cm below the coeliac trunk at the level of L1-L2., anteriorly, it is crossed at its origin by the splenic vein and body of the pancreas; it is separated posteriorly from the aorta by the left renal vein LRV, retroperitoneal fat and the second portion of duodenum. The angle formed with the aorta, known as aortomesenteric angle (AMA), has a normal range of approximately 28-65°, with and the normal aortomesenteric distance (AMD) is 10-30 mm (1).

The SMA is rare syndrome typically occurs secondary to rapid weight loss, due to any cause. Significant weight loss decreases the amount of retroperitoneal fat between the superior mesenteric artery and aorta, thus removing a major source of cushioning for the duodenum and allowing duodenal compression. Weight loss can be attributed to a number of causes including dietary disorders such as: anorexia nervosa or malabsorption; severe wasting states such as burns, cachexia, and endocrine disorders; spinal deformity due to scoliosis or severe trauma; mechanical compression due to retroperitoneal tumors; and even bariatric surgery. The clinical presentation of SMA syndrome is variable and nonspecific and includes nausea, vomiting abdominal pain, and weight loss. Fig. 5 on page 12 Fig. 6 on page 11

Nutcracker syndrome

The asymptomatic compression of the left renal vein between abdominal aorta and superior mesenteric artery is known as nutcracker phenomenon. This phenomenon is characterized by impeded outflow from the left renal vein into the inferior vena cava due to an extrinsic compression. When the nutcracker anatomy produces clinical symptoms, it's known Nutcracker syndrome (NCS), resulting from increased pressure in the LRV and it may with massive gonadal vein reflux causing left-sided varicoceles Fig. 8 on page 13 in males and left-sided vulvar and pelvic varices in females, manifested by pelvic venous congestion syndrome Fig. 4 on page 15.

Both arterial (SMA) and venous (left renal vein) anomalies have been associated with NCS. Factors in relation to the superior mesenteric artery include abnormally low or lateral origin from the aorta, abnormal configuration of its origin or abnormal branching. Venous anomalies include abnormal course of the left renal vein posterior the aorta or bifurcation of the left renal vein with tributaries coursing in anterior and posterior the aorta. Other contributory or possibly factors that may cause this phenomenon are abnormal posterior situation of the left kidney with stretching of the left renal vein over the aorta and abnormally high course of the left renal vein.(5)
**Varicocele** consists of dilated veins in the pampiniform plexus, often visible or palpable as a conglomerate of dilated vessels Fig. 8 on page 13 and is associated with subfertility, however the precise mechanisms impairing spermatogenesis and fertility are still controversial (3). The nutcracker phenomenon has been implicated as a cause of varicocele by producing a persistent elevated renocaval pressure gradient and reflux down the internal spermatic vein thereby promoting the development of collateral venous pathways (4).

Clinical manifestations range from mild microhematuria to severe gross hematuria (which can result in anemia), resulting from the rupture of fragile varices into the collecting system, mild to severe orthostatic proteinuria, and left flank pain secondary to the passage of ureteral blood clots. (1)

**Pelvic congestion syndrome** is a common cause of chronic pelvic pain. Although the etiology is multifactorial (2), an important cause is left ovarian vein congestion secondary to compression of the left renal vein by the superior mesenteric artery. Other causes include retroaortic left renal vein and right common iliac vein compression Fig. 4 on page 15

The diagnosis of ovarian and pelvic varices is established by the identification of multiple dilated tubular structures with venous blood Doppler signal around the uterus and ovary.

There are three diagnostic criteria for establishing the diagnosis of pelvic congestion: (a) a tortuous pelvic vein with a diameter greater than 4 mm, (b) slow blood flow (approximately 3 cm/sec), and (c) a dilated arcuate vein in the myometrium that communicates between Pelvic varicose veins bilaterally (2). MR and CT are also used to diagnose pelvic varices.

**Findings and procedure details**

**Duodenal compression**

Initially, ultrasound may be used to demonstrate narrowing of the aortoesenteric angle Fig. 3 on page 9 Fig. 5 on page 12 The radiographic findings in upper gastrointestinal barium study are compression defect of the third portion of the duodenum Fig. 5 on page 12A, segmental dilatation of intestine proximal to the compression, vertical linear extrinsic compression of the third portion of the duodenum and delayed gastroduodenal emptying.
The CT findings include mid-transverse duodenal compression, dilated stomach and proximal duodenum, reduced aortomesenteric angle Fig. 5 on page 12, Fig. 3 on page 9 and aortomesenteric distance, evaluated in the angiographic phase.

**Nutcracker syndrome**

Non invasive imaging methods for diagnosis include Doppler ultrasonography, Fig. 7 on page 14, computed tomography angiography, and magnetic resonance angiography. Invasive techniques include retrograde venography.

In our experience, ultrasonography is often the first diagnostic approach in patients with suspected NCS.

Studies have reported significantly reduced AMD of up to 3 mm, which contrasts greatly with control groups where the average distance is 10-14 mm. Similarly, an AMA of less than 16° has been reported in patients with nutcracker syndrome. (1) A measure the left gonadal vein diameter above 5 mm is suggestive of compression. Pulsed Doppler evaluation demonstrates the most important findings: reversed caudal flow in the gonadal vein and acceleration of the flow velocity at the point of compression (greater than 100 cm/s).

CT angiography (CTA) and MR angiography (MRA), provide visualization of the anatomy. Both tests can demonstrate the precise LRV compression point and prestenotic dilatation of the LRV. The abrupt narrowing of the LRV with a triangular shape at the aortomesenteric portion is known as the **beak sign** (sensitivity 91.7% and specificity 88.9%).

Retrograde venography is the gold standard for the diagnosis of NCS. It not only confirms anatomic change, but also shows a pressure gradient across the area of entrapment. Reflux of contrast into gonadal vein and pooling of contrast into the renal vein can also be demonstrated.

**Treatment**

**Superior mesenteric artery syndrome**

Conservative initial treatment is recommended in all patients with superior mesenteric artery syndrome; this includes adequate nutrition, nasogastric decompression, and
proper positioning of the patient after eating. Enteral feeding using a double lumen nasojejunal tube passed distal to the obstruction under fluoroscopic assistance is an effective adjunct in treatment of patients with rapid severe weight loss and also eliminates the need for intravenous fluids and the risks associated with total parenteral nutrition. (8)

In some instances, both enteral and parenteral nutritional support may be needed to provide optimal calories. The patient can be started on oral liquids followed by slow and gradual introduction of small and frequent soft meals as tolerated. Metoclopramide treatment may be useful. (7)

Surgical intervention is indicated when conservative measures are ineffective. Conservative treatment should be instituted for at least 4-6 weeks prior to surgical intervention.

Options for surgery include a duodenojejunostomy or gastrojejunostomy to bypass the obstruction, or a duodenal derotation procedure to alter the aortomesenteric angle and place the third and fourth portions of the duodenum to the right of the superior mesenteric artery.

**Nutcracker syndrome**

For NCS, management options range from observation to nephrectomy, depending on the severity of symptoms. Conservative treatment is recommended for mild hematuria. For patients younger than 18 years, the best option is a conservative approach with observation for at least 2 years.

Interventions should be considered only when symptoms are severe or persistent, including severe unrelenting pain, severe hematuria, renal insufficiency, and failure to respond to conservative treatment after 24 months. Most interventions aim to decrease LRV hypertension and others are directed against pelvic venous reflux. A variety of surgical approaches have been used, including medial nephropexy with excision of renal varicosities, LRV bypass, LRV transposition with or without Dacron wedge insertion between SMA and the aorta, SMA transposition, renal-to-IVC shunt, renal autotransplant, gonadocaval bypass, and even nephrectomy for persistent hematuria.

External stenting with ringed polytetra uoroethylene graft interposition around the LRV and intravascular stenting have been applied relatively recently. Both balloon-expandable and self-expanding stents have been used in adult and pediatric patients. Intravascular stenting approaches in NCS were extrapolated from the stenting experience in May-Thurner and superior vena cava syndromes.
Treatment should be based on severity of symptoms and their expected reversibility with regard to the patient’s age and stage of the syndrome. Ligation of the collateral veins may increase renocaval pressure gradients, and ablation of pelvic venous collaterals should be combined with a procedure to relieve renocaval pressure gradients. Coil embolization of ovarian veins in patients with pelvic congestion syndrome and demonstrable pelvic varicoceles may provide symptomatic improvement in 56% to 98% of patients. (5)
Fig. 2: Normal aortomesenteric angle. Gray scale ultrasound in a 37 years-old woman.

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**Fig. 3:** Normal aortomesenteric angle. Color Doppler US image in a 34-years-old woman.

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**Fig. 1:** Normal content of Aortomesenteric Angle: Left renal vein (LRV), retroperitoneal fat (RF) and duodenum (D).

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Fig. 6: Case 3: Duodenal compression in SMA: 40 years old woman, with colon cancer, diagnosed 2 years ago. A) Midline sagittal reformatted MIP images from two different abdominal CT studies demonstrate a normal AMA (25°). B) Same patient 11 months later after development of consumptive syndrome, demonstrating a reduced aortomesenteric angle (17°), with secondary duodenal compression. Patient underwent nasojejunal tube passed distal to the obstruction under fluoroscopic assistance. placement.

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Fig. 5: Case 2: Duodenal compression in a 14-years-old man presented with abdominal pain, vomiting and oral intolerance following a corrective spine scoliosis procedure and weight loss. A) upper gastrointestinal study, after the ingestion of barium meal. The patient is prone. There is a compression defect in the second portion of the duodenum, produced by the superior mesenteric artery. The loop proximal to the duodenal compression is dilated. B) US image demonstrates a reduced aortomesenteric distance of 4.9 mm. C) Midline sagittal reformatted MIP image from an abdominal CT angiographic shows a reduced AMA. D) Three-dimensional image shows a reduced aortomesenteric angle.

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**Fig. 8:** Case 4. Left varicocele in Nutcracker syndrome: A 6 years-old children. A) Doppler color image, shows dilated pampiniform venous plexus, B & C) Image demonstrates reverse flux on the examined venous, D) The ascending left gonadal vein dilated. E, F, G & H) Reverse flux in the left gonadal vein.

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**Fig. 7:** Case 4. Nutcracker syndrome: Left varicocele in a 6 years-old children. A) Sagittal view demonstrating a narrow (19°) aortomesenteric angle. B) Doppler US shows left renal vein partially collapsed. C & D) Transversal view, showing left renal vein is compressed between the aorta and the superior mesenteric artery, Doppler US demonstrate turbulent flow. E & F) Velocity gradient from 20 to 84 cm/s, before and after compressed renal vein.
Fig. 4: Case 1: Pelvic congestion syndrome in a 38 years old female patient with chronic pelvic pain. A & B) Reverse flux in the left gonadal vein associated with multiple pelvic dilated veins. C) Sagittal view demonstrating a narrow (25°) aortomesenteric angle. D) Transversal view, showing left renal vein is compressed between the aorta and the superior mesenteric artery. E & F) Velocity gradient from 12 to 157 cm/s, before and after compressed renal vein.

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Conclusion

A reduced aortomesenteric angle is associated with uncommon pathologies due to duodenal and left renal compression, and the clinical findings are nonspecific. The first diagnosis approach may be performed with ultrasound evaluation allowing measurement of the aortomesenteric distance and angle.

Doppler ultrasound is particularly important in nutcracker syndrome to evaluate reversed caudal flow in the gonadal vein, flow acceleration at the point of stenosis, and dilated veins in the pampiniform plexus. The gold standard for diagnosis of nutcracker syndrome is retrograde venography for anatomic evaluation and demonstration of a pressure gradient across the area of entrapment.

MR and CT provide a global vision of anatomy with a different imaging perspective of these syndromes.
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