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Learning objectives

Review of vascular compression syndromes frequently in the abdomen.

Know the angio-CT imaging findings for diagnosis of these syndromes. Fig. 1 on page

Background

Certain abdominopelvic vascular structures may be compressed by adjacent anatomic structures or may cause compression of adjacent hollow viscera. Such compressions may be asymptomatic; when they are symptomatic, however, they can lead to a variety of uncommon syndromes in the abdomen and pelvis. Fig. 2 on page

This heterogeneous group of disorders nominees "vascular compression syndromes," since they all involve either the compression of vascular structures or the compression of hollow viscera by vascular structures.

The most common are: Fig. 3

- Median arcuate ligament syndrome,
- May-Thurner syndrome,
- Nutcracker syndrome,
- Superior mesenteric artery syndrome
- Retrocaval Syndrome,
- Ureteropelvic junction obstruction
- Ovarian vein syndrome.

Although classically arteriography has been the main tool in diagnosis of these entities, nowadays it is only used in doubtful cases and for those patients in whom endovascular treatment is indicated. Currently most used techniques are Doppler ultrasound, Angio-MRI and the Angio-multidetector CT (MSCT), being this last one particularly useful since it allows a full assessment of the anatomy and the resulting morphologic changes, thanks to its ability to produce multiplanar reconstructions.

Findings and procedure details

We review the radiologic findings of these rare and heterogeneous
entities and present our most representative cases.

**Medial arcuate ligament syndrome (MALS):** Fig. 3 on page 10

Narrowing of the proximal celiac trunk (CT) produced by the medial arcuate ligament (MAL) resulting in clinical symptoms such as postprandial epigastric pain and weight loss.

The compression may occur if TC has a more cephalic origin or if MAL is abnormally low, increasing during exhalation since the aorta and the TC move upward. The characteristic image will be a focal narrowing of the proximal celiac trunk "on hook", which may be more pronounced at the end of exhalation. This narrowing is displayed best on sagittal MIP and reconstructed images. Post-stenotic dilatation may be present in severe cases, and in some cases with compensatory circulation, usually through pancreatic-duodenal arcade.

Due to the large increase in abdominal CT exams, celiac trunk compression is occasionally and incidentally seen in patients who undergo this scan for other reasons. Diagnosis is difficult and relies on the presence of symptoms, imaging findings and exclusion of other causes.

MALS treatment remains mainly surgical, and often involves the medial arcuate ligament transection, in combination with some kind of surgical reconstruction of the celiac trunk. Endovascular stents and by-pass technique are also employed.

**Superior mesenteric artery syndrome (SMAS), or aorto-mesenteric clamp syndrome or Wilkie syndrome:** Fig. 4 on page 11

Rare cause of duodenal obstruction. It is caused by the compression of the third portion of the duodenum due to clamp formed by the aorta and the superior mesenteric artery (SMA).

The SMA origins at L1-L2 level and its is directed anterior and caudally, forming an angle with the aorta known as aorto-mesenteric angle (AMA). On the other hand, the third portion of the duodenum is surrounded by retroperitoneal fat, which helps to keep a wide distance between the SMA and aorta, usually called aorto-mesenteric distance (AMD). In normal conditions it must exceed the 28° and 10 mm, respectively.

SMA syndrome, occurs relatively infrequently, but it has been estimated at 0.1% - 0.3% of total population. It is more common in young women.

Factors that predispose to this syndrome are:

1. Conditions associated with the rapid and severe weight loss.
2. Patients who have undergone scoliosis surgical correction, in whom spine elongation reduces both AMD and AMA.

3. Conditions that involve external pressure applied to the abdomen.

4. Anatomic variants such as a variation on Treitz ligament insertion or a low origin of the AMS.

Symptoms include epigastric pain, postprandial (may be relieved by lying down or in left lateral decubitus position), nausea, vomiting, weight loss, resulting in a vicious circle with greater loss of retroperitoneal fat. All other causes of duodenal obstruction should be excluded before diagnosing SMA syndrome.

Barium studies show a dilated stomach and proximal duodenum with a vertical extrinsic impression on the third portion of the duodenum by compression, delay in gastroduodenal emptying, and relieved with postural change.

Multislice CT allows simultaneous evaluation of vascular anatomy, cross-sectional duodenal compression, and proximal dilatation, and it is therefore the best diagnostic test choice.

Treatment is usually conservative for acute and initial presentations, replenishment of fluid and electrolytes, mobilization of the patient in the prone position or left lateral decubitus position and weight gain. Surgical treatment is indicated if conservative measures failure and usually involves obstructed segment by-pass.

**May-Thurner syndrome or iliac vein compression syndrome:** Fig. 5 on page 12

It consists on obstruction of the left common iliac vein (LCIV) caused by the junction of the right common iliac artery (RCIA) being trapped between it and the underlying vertebral body. In addition, the pulsating force transmitted causes vein intimal hypertrophy, which eventually changes its light.

The typical symptom is swelling of the extremities, acutely due to left side iliofemoral venous thrombosis, or chronically due to venous congestion, without underlying thrombosis. Other chronic symptoms include chronic varicose veins and chronic venous stasis ulcers. Less frequent are more serious complications such as pulmonary embolism or Phlegmasia Cerulea dolens.
True May-Thurner syndrome prevalence is unknown, it should be relatively common, being suspected in young women with acute left leg iliofemoral thrombosis, often being overlooked in the differential diagnosis, especially if the patients have other risk factors.

Venography has been traditionally the diagnostic method, but it is an invasive procedure which takes time and may cause phlebitis. Angio-CT is recommended for diagnosis since it shows LCIV compression and thrombosis (if it exists), as well as helps to exclude other causes of venous compression.

Thrombolysis followed up by endovascular stent placement has a high success rate (95%). Oral anticoagulants are used to prevent recurrence of thrombosis.

**The Nutcracker syndrome (NCS):** Fig. 6 on page 13 and Fig. 7 on page 14

Refers to a set of symptoms due to increased of venous pressure in the left renal vein (LRV), secondary to flow obstruction in its arrival to the inferior vena cava (IVC). This produces an intra and extra-renal hypertension with venous colateral replacemente and left gonadal vein reflux.

Depending on their mechanism of production may be:

- Anterior, when LRV is compressed between superior mesenteric artery and the aorta, secondary to a decrease in DAM and AAM. In this way you can produce the above Nutcracker syndrome with a SMA syndrome simultaneously.

- Posterior, when left renal vein retro-aortic clutches between the underlying vertebral body and the aorta.

- Combined, when renal vein is duplicated and anterior branch is compressed between the aorta and superior mesenteric artery, while the posterior branch is trapped between the aorta and spine.

Nutcracker syndrome diagnosis remains clinical and should be done only when characteristic symptoms are present.

Most of the affected patients are young people with symptoms ranging from mild to severe, and may be exacerbated with physical activity.

The most common symptom is hematuria. Other manifestations are orthostatic proteinuria and left flank pain. Reflux can cause left gonadal vein varicocele in men, and pelvic varices and symptoms arised from vulvar and pelvic venous congestion.
MS-CT allows a simultaneous representation of meso-aortic anatomy and LRV compression, with its hemodynamics consequences (prestenotic dilated hilar and pelvic varicose veins, gonadal veins enlarged)

Expectant management in young or thin patients, or those with mild symptoms is preferred. Intervention is indicated in cases of symptomatic severity. Recently endovascular stent placement on LRV is performed with good results, although there are no long term studies. Stent migration, fibromuscular intimal hyperplasia, intra-stent stenosis and venous thrombosis are potential complications.

**Pyelo-ureteral or Ureteropelvic junction syndrome (UPJS): Fig. 8 on page 15**

It is the most common cause of neonatal hydronephrosis (1 for each 20,000), predominantly in males, affecting twice the left kidney, and being bilateral in 10-30% of cases.

The origin of this alteration may be primary, including intrinsic (functional) causes, or extrinsic ones such as aberrant vessels or renal cysts. It may also be secondary to inflammatory processes. Moreover, multiple causes can coexist and be multifactorial.

Vascular crossroads has been implicated in 11-79% of the cases, either by a renal artery or vein anormal branch, or by an accessory branch, which may arise from the aorta, iliac artery, or from the IVC. It is necessary that the vessel involved is adjacent to the ureteropelvic junction (UPJ).

Regardless of the controversy concerning the possible role of crossing vessels in the UPJ obstruction, its preoperative detection can be important for the urologist, since these vessels can be injured during therapeutic endopyelotomy, and its presence can dictate the surgical approach used.

Symptoms ranges from mild to severe, being generally slow and progressive. Patients may present with flank pain, hematuria, renal lithiasis, infections of the urinary tract and/or pyelonephritis.

Different imaging modalities have been used for the detection of vascular crossroads, demonstrating that MS-CT is the most accurated technique, showing us the transition zone between the vessels and the uréter, as well as the morphology and severity of the hydropephorsis.

If vascular crossroads are present, UPJS surgical treatment is aimed to provide symptomatic relief and preservation of any remaining kidney function, being the treatment of choice.
Retrocaval ureter syndrome: **Fig. 9** on page 16

Due to an abnormal development of the inferior vena cava (IVC). It is rare, with an incidence of 1/1000.

Clinical appears in adults, whose main symptom are cramping pain on right flank (coming to resemble a picture of renal lithiasis) or can also be manifested by urinary tract infection.

Excretory urography and ascending pyelogram show the typical "J" deformation and a proximal ureteral and calyceal system dilation of the right ureter.

MS-CT images helps identifying the cause by locating the transitional zone where the gauge of the middle third of the right ureter is decreased, site in which the ureter takes a medial and posterior direction, this segment is partially sorrounded by the inferior vena cava. Distal ureter presents normal size and continues its course downwards towards the bladder.

Surgical treatment is only reserved for those patients with significant symptoms and impaired renal function. In general, management is conservative and is aimed to prevent and avoid recurrent infections.

Ovarian vein syndrome (OVS): **Fig. 10** on page 17

Rare entity, whose existence has been controversial. It was first described by Clarke in 1964. It refers to an obstructive uropathy due to ureteral compression by an aberrant ovarian vein. This author describes acute cases in pregnant women and chronic cases in nonpregnant women, but mainly right side ureters. Subsequently, other studies reported left side cases and even bilateral appearence.

It is controversial if this syndrome represents a truely physiopathological entity, although in practice it seems to be an association between ureteral dilatation and ovarian vein, being more common in the right vein, usually displayed a ureteral notch with a mild degree of proximal expansion and a normal pelvic ureter.

The clinical picture is characterized by low back pain and other signs and symptoms of pyelonephritis in pregnant women. In nonpregnant patients back pain may be acute and intermittent.

Images for this section:
Vascular compression syndromes in the abdomen and pelvis

The most common are:

- Median arcuate ligament syndrome,
- May-Thurner syndrome,
- Nutcracker syndrome,
- Superior mesenteric artery syndrome,
- Retrocaval ureter syndrome,
- Ureteropelvic junction obstruction,
- Ovarian vein syndrome

Fig. 2
Fig. 3: Median Arcuate Ligament Syndrome in a 52-year-old man with postprandial pain, loss of appetite, and weight loss of more than 10 kgs in the last 6 months. CT angiographic: a) sagittal CT and b) Three-dimensional VR images shows significant stenosis of the proximal celiac artery (arrow) caused by the median arcuate ligament. Given the absence of any atherosclerotic changes in the proximal celiac artery. c) coronal MIP image shows collateral vessel. d) Photograph obtained during laparoscopic surgery shows the released fibers of the median arcuate ligament in the vicinity of the celiac artery and its branches.
Fig. 4: Superior mesenteric artery syndrome in a 33 year old woman with postprandial epigastric pain, intermittent vomiting and weight loss. CT angiography: a): Midline sagittal reformatted image shows a short aortomesenteric distance (AMD) and decrease of the aorto-mesenteric angle. Axial slices: b): where we can observe the gastric dilatation. c): narrowing of the duodenum as it passes between the aorta and mesenteric artery. The patient received conservative treatment and she improved their symptoms.
Fig. 5: May-Thurner syndrome. 42 year old woman with pain and swelling of the left leg. a): venography absence of left common iliac vein and varicose multiple collateral branches. The CT angiography study: b): axial, c): coronal and d): Volumetric reconstruction: shows the compression of the left iliac vein between multiple side right common iliac artery and the vertebral body (yellow arrow), and the presence of thrombosis left internal iliac vein (pink arrow).
Fig. 6: Anterior nutcracker syndrome in patients with left flank pain and hematuria: a): Study Angio CT Axial image showing the left renal vein between the aorta and the mesenteric (arrow). b): Image showing collateral vessel (circle). c): Reconstruction coronal MIP technique showing varicose veins in the pelvis as a result of venous stasis caused by obstruction.
Fig. 7: Posterior nutcracker syndrome in a male of 39 years old with hematuria and flank pain on the left side. Angio CT axial images: a) compression of the left renal vein between the aorta and vertebral body (arrow) and b) showing multiple collateral vessels secondary to compression (circle).
Pieloureteral Junction Syndrome (UPJ)

Obstruction at the renal pelvis and proximal ureter due to a crossing of a main renal artery or vein or accessory branch of the aorta, iliac artery, inferior vena cava.

Fig. 8: Pieloureteral Junction Syndrome (UPJ): in patients with recurrent urinary tract infections. CT angiography: a-b): axial and coronal images, c): volumetric-reconstruction showing ureteral compression by a retropelvic artery (arrow pink) Note the presence of a ureteral catheter bypass for easy viewing of the crossing. d-e): axial and coronal images of a second patient whose compression is produced by a polar artery (yellow arrow)
Fig. 9: Retrocaval syndrome: 55-year-old man with right-sided abdominal pain and hematuria. a): Urography across the catheter of external derivation image presents typically as dilatation of the upper third of the ureter exhibiting a classic "reversed J-shaped". b): CT urographic study show the abnormal course right ureter wrapping around the IVC. c): Three-dimensional VR
Fig. 10: The ovarian vein syndrome. a): Angio CT, Coronal and b): VR images showing an aberrant ovarian vein that leads to a lower renal polar vein producing an ureteral compression and hydronephrosis.
Conclusion

The radiologist should be aware of these entities are rare and difficult to diagnose because the clinical data are not always approximate.

Multidetector computed tomography (CT) is the imaging modality of choice for many of these the imaging modality of choice for many of these syndromes owing to its high contrast and high spatial and temporal resolution, capacity for obtaining multiplanar two-dimensional and three-dimensional image, remarkable accuracy, widespread accessibility, speed, and relative noninvasiveness.

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