

Abdominal Vascular Compression Syndromes and Imaging Features.

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Abstract

Some intra abdominal structures like celiac artery, SMA and aberrant vessels may compress the adjacent hollow viscera, may be symptomatic or incidental imaging findings. These syndromes, though the etiopathogenesis is doubtful, will have some classical clinical findings. They are median arcuate ligament syndrome, May-Thurner syndrome, nutcracker syndrome, superior mesenteric artery syndrome, ureteropelvic junction obstruction, ovarian vein syndrome, and other forms of ureteral compression. Multidetector computed tomography (MDCT) is the imaging modality of choice, however interpretation of imaging findings should be carefully correlated with clinical findings.

Keywords: MALS- Median arcuate ligament syndrome, SMAS- Superior mesenteric artery syndrome, SMA – superior mesenteric artery, AMA - Aortomesenteric angle, AMD - Aortomesenteric distance

Introduction

Compression of the proximal celiac artery, transverse duodenum, left common iliac vein (CIV), left renal vein (LRV), ureteropelvic junction (UPJ) and ureter can occur due to their close anatomic relationship to adjacent ligaments as well as bony and vascular structures.

Anatomic or morphologic findings that predispose to such compression may occasionally be encountered in asymptomatic patients who undergo imaging for unrelated causes. Thus, caution should be exercised to avoid overdiagnosis of these syndromes. It is important that the diagnosis of these syndromes not be based on imaging findings alone.

Multidetector computed tomography (MDCT) is the imaging modality of choice for many of these syndromes due to high contrast, high spatial and temporal resolution, capacity for obtaining isotropic data sets that allow multiplanar, two-dimensional and three-dimensional (3D) postprocessing, remarkable accuracy, widespread accessibility, speed, and relative noninvasiveness.

Ultrasonography (US) is largely operator, patient, and region dependent, although duplex US can provide information on the hemodynamic significance of vascular compressions.

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We report six cases, i.e. two cases of median arcuate ligament syndrome (MALS), a case of superior mesenteric artery syndrome (SMAS), two cases of superior mesenteric vein (SMV) tributary compressing duodenum between SMA and aorta and a case of retrocaval ureter with classical imaging and clinical history.

Our aim is to familiarize radiologists with the MDCT appearance of these syndromes and the added benefit of MPR in diagnosis.

Median Arcuate Ligament Syndrome

It is also known as celiac artery compression syndrome or Dunbar syndrome, a rare entity characterized by narrowing of the proximal celiac trunk by the median arcuate ligament, which courses superiorly over the origin of celiac artery.

Harjola first described this syndrome in 1963 [1] and then Dunbar [2] in 1965.

It typically affects young patients of 10-40 years age and females are affected more. Post-prandial epigastric abdominal pain, vomiting and weight loss are the classic clinical symptoms. Compression of celiac artery by the ligament does not produce symptoms, probably due to collateral supply from the superior mesenteric artery in 10 to 14 % of the pts.

Doppler ultrasound [4] at compressed or narrowed segment of celiac artery reveals variation of peak systolic velocity (PSV) during respiration with a marked increase during expiration with PSV greater than 200 cm/s. A greater than 3:1 ratio of PSV in the celiac artery in

expiration compared with the PSV in the abdominal aorta just below the diaphragm is another useful criterion to diagnose MALS.

MDCT showing median arcuate ligament thickness of more than 4 mm³ is considered abnormal. Focal narrowing with hooked appearance is the hallmark which can help distinguish this condition from other etiologies of celiac artery stenosis such as atherosclerosis. Other findings include poststenotic dilatation and collateral vessels such as pancreaticoduodenal arcade from superior mesenteric artery.

We report two cases of of MALS in 26 year and 35 year old male patients showing moderate to severe narrowing of celiac artery by median arcuate ligament with post stenotic dilatation (fig. 1 and 2).

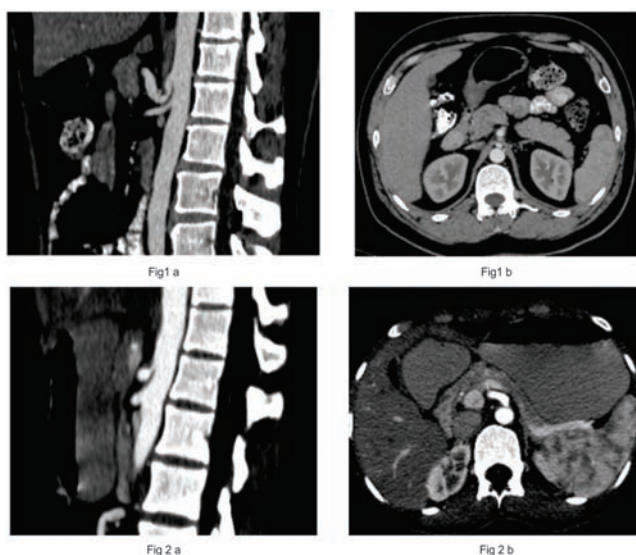


Fig 1 and 2- [a] : Oblique sagittal and [b] axial contrast MDCT images showing celiac artery compression by the median arcuate ligament with post stenotic dilatation.

Superior Mesenteric Artery Syndrome

It is an uncommon entity of duodenal obstruction (third part), extrinsically compressed between SMA and abdominal aorta due to reduced aortomesenteric angle (AMA). It is similar to MALS in affecting young patients and females.

Incidence of this condition is 0.013-0.3%. Von Rokitanski first described the condition in 1861. Later, Wilkie described in detail.

Risk factors include rapid weight loss which decreases fatty pouch between SMA and aorta, corrective scoliosis surgery which causes lengthening of the spine and hip or body cast that applies external abdominal pressure, abnormal high fixed position of ligament of treitz and low origin of SMA. There is controversy in etiopathogenesis and the relationship between anatomical findings and clinical symptoms is not well established.

Aortomesenteric angle (AMA) and the aortomesenteric distance (AMD) are reduced in the patients with risk factors [5,6].

The normal AMD is typically 10 to 28 mm and is measured at the level of the horizontal part of the duodenum as it travels between the abdominal aorta and SMA [6,7]. Aortomesenteric angle normal value is 25 to 60 degree. AMA of 6 to 15 degree and AMD of 2 to 8 mm is diagnostic of superior mesenteric artery syndrome (SMAS).

Patients will present with postprandial abdomen pain, vomiting, weight loss which relieves while lying on left lateral and prone position.

Barium study shows dilated stomach, dilated second part of duodenum and vertical cut off at third part of the duodenum with no mucosal irregularities and the obstruction relieves while lying in prone position. MDCT will confirm the above findings.

Mega duodenum is the differential diagnosis which will have similar radiological findings.

We report a case (fig.3) of 47 year female presenting with pain abdomen and vomiting. MDCT showed reduced AMA and AMD causing compression on third part of duodenum.

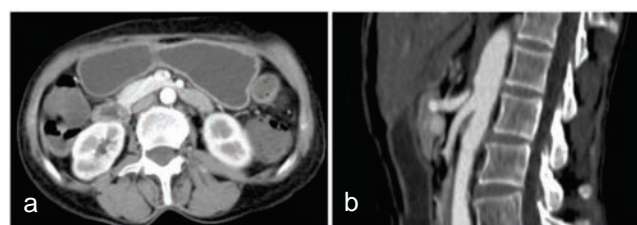


Fig. 3: MDCT images axial section (a) showing reduced distance between SMA and descending aorta causing compression over the third part of duodenum and sagittal section (b) showing acute angle of SMA with aorta and reduced aortomesenteric distance.

We report a case of 60 year old female presenting with pain abdomen, vomiting and weight loss. Barium study showed dilated stomach, first, second part of duodenum with vertical cut off of third part of the duodenum with normal mucosa and the obstruction was relieved while lying in prone position. MDCT (fig. 4) showed reduced AMA with third part of duodenum compressed by tributary of SMV between SMA and aorta.

We report a case of 46 year male where (fig. 5) MDCT showing reduced space between aorta and SMA with two SMV tributaries coursing posterior to SMA at L3 level between superior mesenteric vessels and aorta, causing compression on third part of duodenum with proximal dilatation of the duodenum and stomach.

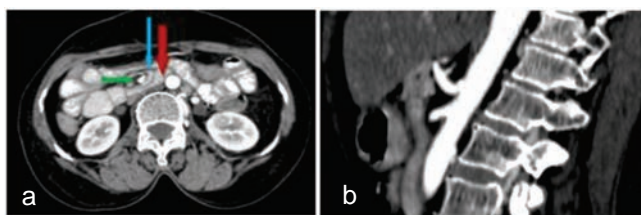


Fig. 4: MDCT images- axial section (a) showing compression of third part of duodenum (red arrow) between SMA (green arrow) and aorta by tributary of SMV (blue arrow) and sagittal section (b) showing reduced AMA.

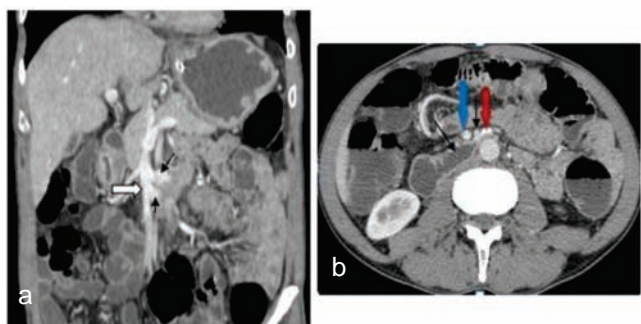


Fig. 5 : MDCT images oblique coronal section (a) showing two tributaries (black arrow), one below the other draining into SMV (white arrow) and axial section (b) showing abrupt tapering of third part of duodenum (oblique black arrow) by inferior tributary (red arrow) of SMV (blue arrow) shown in oblique coronal between SMA (vertical black arrow) and aorta.

Retrocaval Ureter

A preureteral vena cava and circumcaval ureter or preureteral vena cava, a rare congenital anomaly where ureter classically courses medially at L3 behind the inferior vena cava (IVC) winding around it and then passes laterally in front of it to then course distally to the bladder. Hydronephrosis is due to kinking of the ureter, a ureteric segment that is adynamic or compression against the psoas muscle.

Retrocaval ureter was first reported by Hochstetter [6]. Abeshouse and Tawkin (1952), Muller and Engel (1952), Goodwin et.al (1957)[7] and Rowland et.al (1960) have described the radiological features of circumcaval ureter. The IVC normally develops from the posterior cardinal, subcardinal and supracardinal veins, which undergo sequential development, anastomosis and regression to become the inferior vena cava and azygos venous system. Normally, the right subcardinal vein forms the pre-renal IVC, the subcardinal-supracardinal anastomosis forms the renal segment and the right supracardinal vein forms the post-renal IVC. In a circumcaval ureter, there is anomalous development of the infrarenal IVC from the right posterior cardinal vein that is embryologically more medial.

Retrocaval ureter may be asymptomatic or may present

with flank pain, UTI, calculus or hematuria usually at 3rd - 4th decade in male patients.

Prevalence rate of about 0.9 in 1000 with a male to female ratio of 2.8 [8].

Salonea [8,10] described two types of retrocaval ureter. Type I - the ureter crosses behind the inferior vena cava at the level of the third lumbar vertebra and it has an 'S' or fish hook type shape at the point of obstruction. Marked hydronephrosis is seen in 50% of patients. In the less common Type II, the crossover occurs higher at the level of the renal pelvis.

Dilated medial deviation of the ureter at L3 with hydronephrosis and 'S' or sickle shaped deformity at the level of displacement is the classical IVU findings but retroperitoneal fibrosis, a retroperitoneal mass, previous surgery must be kept in mind as differential diagnoses [9]. Retrograde pyelography was the old imaging modality.

Spiral CT delineation of the ureter and its course in relation to the IVC and location of IVC lateral to the right pedicle of the L3 vertebra were diagnostic of circumcaval ureter.

We report a case of retro caval ureter as shown in fig. 6.

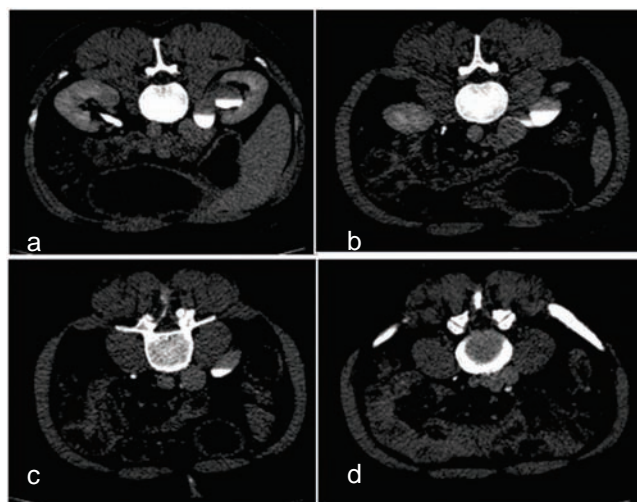


Fig.6 : Sequential MDCT axial sections in craniocaudal direction - (a) showing dilated renal pelvis and proximal ureter (b) showing compressed ureter coursing posterior to IVC (c) ureter seen on the left lateral aspect of IVC (d) ureter seen anterior to IVC

Other abdominal vascular compression syndromes

Anatomic compression of the left renal vein between SMA and aorta is described as anterior nutcracker syndrome or if the left renal vein has a retroaortic or circumaortic course, between the aorta and the underlying vertebral body resulting in its compression, it is described as posterior nutcracker syndrome.

May-Thurner Syndrome (also known as iliac vein

compression syndrome / Cockett syndrome), consists of obstruction of the left common iliac vein when it passes between the right common iliac artery and the spine.

Conclusion

The abdominal vascular compression syndromes discussed here are uncommon and are potentially easily missed on radiologic examinations, particularly in a nonspecific and vague clinical setting. Hence, knowledge of the typical imaging findings and associated clinical symptoms is essential so that they can be carefully sought and excluded. However, because these findings may also exist in healthy individuals as anatomic variants, it is important to correlate radiologic findings with clinical symptoms to identify the subset of patients who will benefit from treatment.

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