Case Report

Combination between Dunbar Syndrome and May–Thurner Syndrome: A Rare Case Report

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Abstract

Dunbar syndrome (DS) and May–Thurner syndrome (MTS) are part of a group of rare vascular disorders known as "vascular compression syndromes." Dunbar's syndrome is caused by the median arcuate ligament of diaphragm, which, due to an abnormal course, causes celiac artery compression. MTS is caused by the left common iliac vein compression pushed against the spine by the right common iliac artery causing progressive flow congestion and leading to thrombosis. Ultrasound is the first-level examination for the diagnosis of these rare pathologies and allows to recognize vascular compressions and to obtain an estimate of stenosis degree. We describe a very rare case of DS and MTS combination in a young man with postprandial pain and left lower limb thrombosis.

Keywords: Cardiovascular abnormalities, celiac artery, Dunbar syndrome, May–Thurner syndrome, ultrasound

Introduction

Vascular compression syndromes include some uncommon vascular disorders caused by arterial or venous vessels compression by contiguous vascular or musculotendinous structures.[1] When the strictures are significant, they become symptomatic. In most cases, compressions are asymptomatic and, for this reason, are often not diagnosed. Imaging plays an essential role as it allows to identify vascular stenosis and contextually measure the stenosis degree whose knowledge is important to set up a correct treatment. Median arcuate ligament syndrome[2] or Dunbar syndrome (DS)[3] [Figure 1a] is caused by the median arcuate ligament (MAL) of the diaphragm, which, due to an abnormal course with an insertion lower, causes celiac artery (CA) compression. When stricture is severe, it manifests as a typical syndromic triad with weight loss, postprandial abdominal pain, and epigastric murmur. MAL can also compress the neural ganglion, which can accentuate epigastric pain, especially in the exhalation phases that cause the diaphragm to rise, the MAL to lower, and the CA to be compressed. The CA compression, although severe, is asymptomatic in most patients due to the splanchnic compensatory circles that connect the CA with the superior mesenteric artery (Riolan's arch, pancreatic-duodenal arches, Drummond's marginal artery).[4] In symptomatic patients, epigastric pain becomes chronic and, in the long term, causes weight loss as patients cut back on meals to avoid pain. May–Thurner syndrome (MTS), first described in 1957,[5] is caused by the left common iliac vein compression by the right common iliac artery (84% of cases),[6] left common iliac artery, or much more rarely by both arteries[7] [Figure 1b]. According to the authors, two causes predispose to the disease: the reduction of distance between right common iliac artery and spine and chronic pulsations of the right common iliac artery, which cause the formation of a "spur" in the wall of common iliac vein which progressively causes the lumen narrowing. The flow congestion causes edema in the lower limbs, pain, claudication, thrombophlebitis, thrombosis, and, in severe cases, pulmonary or cerebral embolism. Other causes of compression have been reported in the literature, induced by bladder, penile prosthesis, aneurysms, pseudoaneurism, endometriosis, and osteophytes. Compression can be symptomatic or asymptomatic; values

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>25% are reported in 66% of patients with MTS.[8] Ultrasound is the first-level examination for the diagnosis of these two rare diseases; it allows to easily locate the vascular compressions and to obtain a flowmetric estimate of stenosis degree. Computed tomography (CT) or magnetic resonance imaging is used to rule out other causes of compression (abdominal masses, aneurysms, and surgical outcomes) or to rule out serious complications such as pulmonary or cerebral embolism. Intravascular ultrasound (US)[9] allows direct evaluation of the endoluminal content and is currently increasingly used in the management of many vascular diseases such as dissections, aneurysms, pseudoaneurysms, thromboses, and stenoses for which it represents the current reference imaging. The treatment of these pathologies depends on the symptoms and the stenosis degree. Treatment of DS consists of laparoscopic or robotic surgery with open MAL release and celiac ganglionectomy;[10] surgery is the treatment of choice and causes symptom regression in 85% of patients. Endovascular stenting is indicated only in cases of relapse after surgical treatment (7%).[11] Vascular compression syndromes are generally present singly; their association is very rare, although cases of double[12] or triple[13] association have been described in the literature. We describe a rare case of DS and MTS combination in a young patient with deep-vein thrombosis of the left lower limb.

Case Report

A 38-year-old man was referred for our observation for excessive weight loss (12 kg in 6 months), swelling, and claudication of the left lower limb. The patient underwent CT and US of the left lower limb and abdominal vessels. A CT device (Optima 64 slice, GE Healthcare) and a May Lab "Nine" US device (Esaote Genova), with convex 3–7.5 Mhz and linear 5–14 MHz probe, were used. The study has been performed in forced inspiration and forced expiration phases. CT showed CA stenosis at the origin [Figure 2a] with the "hook" appearance in sagittal view in forced expiration [Figure 2b]; furthermore, he highlighted a severe compression of the left common iliac vein at the point of passage between the right common iliac artery and spine with thrombosis of the left common iliac vein and left femoral vein [Figure 2c and d]. In the US study, the following were measured: diameter and peak systolic velocity (PSV) measured in the stenotic tract of CA; flow ratio (FR): PSV measured in the stenotic tract of CA / PSV of abdominal aorta measured at the origin of CA. CA US showed stenosis at origin in inspiratory apnea (3.7 mm in caliber), more severe in expiratory apnea (2.6 mm) [Figure 3a and b and Video 1], PSV of 227.6 cm/s in inspiratory apnea, and 381.2 cm/s in expiratory apnea [Figure 3c and d and Video 2]. The RF between CA and abdominal aorta in expiratory apnea was 3.6: 1 (381.2/104.6) [Figure 4a and b]. In the US study of the left lower limb, the following were measured: diameter of the left common iliac vein and distance between the right common iliac artery and spine. The FR of the left common iliac vein was obtained from the ratio between PSV poststenosis and PSV prestenosis US of the left lower limb revealed common iliac vein thrombosis extending to the common femoral vein [Video 3] and regular flow in the right common iliac vein. The distance between the right common iliac artery and spine was

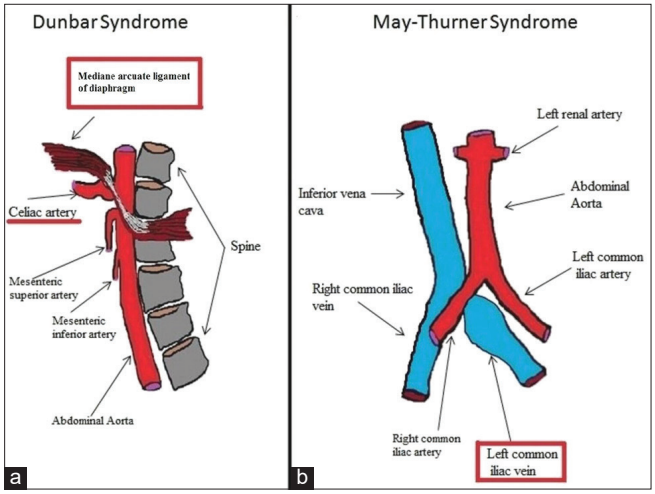

Figure 1: Summary diagram of the anatomical structures involved in patients with Dunbar syndrome and May–Thurner syndrome. (a) Patient with celiac artery compression by the median arcuate ligament. (b) Patient with left common iliac vein compression

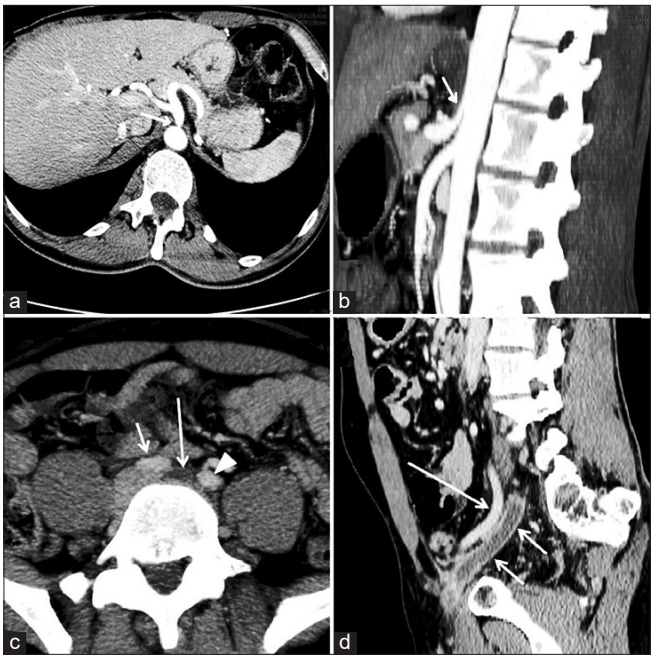

Figure 2: CT findings of DS. (a) CT scans in axial view performed in expiratory apnea show celiac artery stenosis at origin (arrow). (b) The CT view in sagittal plane shows the typical DS pattern with "hook" appearance due to celiac artery stenosis (arrow) and poststenotic dilatation. (c) CT scans in axial view show left common iliac vein stenosis caused by the right common iliac artery (short arrow) pushing it against the spine, with thrombosis of the prestenotic tract (long arrow). Left common iliac artery (arrowhead). (d) CT scan in sagittal view shows complete thrombosis of the left common iliac vein (short arrows). Left common iliac artery (long arrow). CT: Computed tomography, DS: Dunbar syndrome

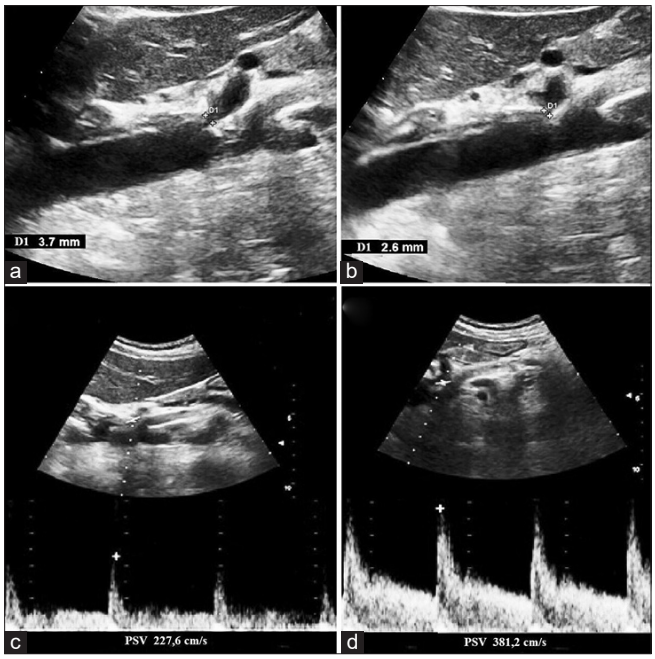

Figure 3: Ultrasound findings of DS. (a) Gray scale ultrasound of the CA, performed in inspiratory apnea, shows a stenosis at origin of CA (3, 7 mm in caliber), (b) which is accentuated in expiratory apnea (2, 6 mm). (c) The flow measurement of CA shows mild increase of peak speed velocity in inspiratory apnea (227,6 cm/s) and significantly increases in expiratory apnea (381,2 cm/s) (d). CA: Celiac artery

Table 1: Summary of ultrasound result

	Celiac artery	Right common iliac vein	Left common iliac vein	Abdominal aorta
PSV (cm/s) in inspiratory apnea	227.6			
PSV (cm/s) with expiratory apnea	381.2			
PSV (cm/s)		18	Not applicable	104.6
Caliber (mm) in inspiratory apnea	3.7			
Caliber (mm) with expiratory apnea	2.6			
Flow ratio	3.6:1		Not applicable	

PSV: Peek speed velocity

2.7 mm. The results are summarized Table 1. The US study was performed by an operator with 20 years of experience. The patient signed an informed consent form.

Discussion

The diagnosis of vascular compression syndromes is very difficult due to their rarity, little knowledge, and nonspecificity of symptoms. Imaging is essential to detect vascular compressions, to rule out other causes of compression, and to highlight the serious complications. The US represents the first-level examination and is important in these pathologies because it allows to obtain a flowmeter measurement of

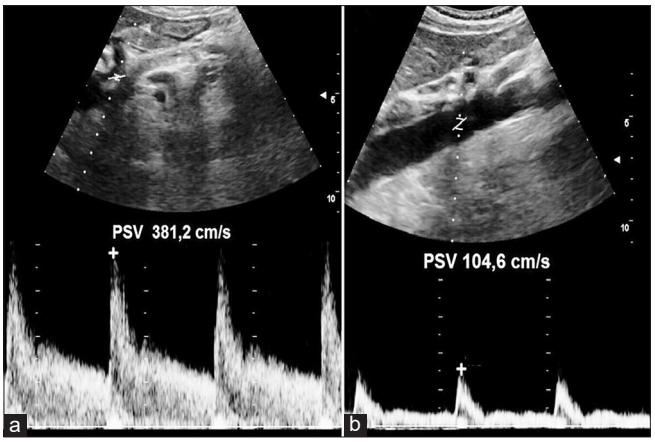

Figure 4: Ultrasound findings of DS. The flow ratio measuration is obtained from ratio between PSV of CA, measured in stenotic tract in expiratory apnea (a) and PSV of abdominal aorta measured at CA origin level (b). DS: Dunbar syndrome, CA: Celiac artery, PSV: Peak speed velocity

stenosis degree, which facilitates a more accurate choice of treatment. Diagnostic difficulties increase when patients are asymptomatic; in fact, in almost all these cases, the vascular compressions are discovered accidentally during routine CT or US examinations.

The DS patients show a typical syndromic triad: epigastric pain, weight loss, and epigastric murmur which, although nonspecific, in the absence of other causes of stenosis, should lead to suspicion of CA compression by the MAL. Epigastric pain, as well as postprandial pain, can also occur in the phases of expiratory apnea and this is due to the compressive aggravation of CA induced by the ascent of abdominal organs and lowering of MAL; however, according to some authors, pain can be caused by the celiac plexus ganglia compression.[14] The finding of the "hook sign" in CT is not uncommon, even in scans performed on inspiratory apnea, but, in most cases, patients are asymptomatic. US is of fundamental importance in the DS diagnosis as it demonstrates the significant PSV increase in expiratory apnea and, at the same time, provides an estimate of stenosis degree thanks to the FR measurement. Asymptomatic patients with CA compression should undergo periodic US follow-ups to monitor the stenosis evolution. CT can estimate stenosis degree with morphological criteria (NASCET, ECST)[15] but cannot measure typical flow variations with breaths; instead, the US, thanks to the FR, provides us with a functional estimate of stenosis degree based on the significant increase of the PSV in forced expiration which can reach values higher than 300 cm/s. In our case, the discovery of CA compression was accidental; only after a more in-depth analysis of the clinical history did it emerge that the patient had been suffering from recurrent episodes for at least 2 years of postprandial epigastric pain, especially after large meals for which he had also performed a gastroscopic examination that was negative. The aforementioned symptomatology, although not recurring, nevertheless influenced his diet, causing him to

lose weight. In these patients, if DS is not diagnosed in time, it causes significant weight loss that could induce the reduction of perivascular adipose tissue and cause superior mesenteric artery syndrome, which would worsen their clinical condition. The therapy of choice of the DS is surgical and consists of decompression with a cut of the MAL and sympathectomy of the celiac ganglion with laparoscopic, robotic, or traditional surgery.[16] In our case, the patient underwent surgical treatment which resulted in a rapid epigastric pain regression. US confirmed complete thrombosis of the left common iliac vein and femoral vein, but, in this case, due to the complete thrombosis of the left common iliac vein, it was not possible to FR mensuration. Medical treatment of MTS with deep-vein thrombosis generally consists of long-term anticoagulants administration and thrombolytic drugs. If the thrombosis is not extensive, the treatment of choice should be endoluminal thrombolysis with US endoscopy (IVUS).[17] When stenosis is significant, the most effective treatment, in the absence of thrombosis, is vascular stenting of the left common iliac vein which reduces venous compression and chronic thrombotic episodes.[18] In our case, due to the thrombosis extension, we subjected the patient to thrombolytic and anticoagulant therapy with a drug regimen: warfarin (Coumadin) 4 mg for 6 months, 300 mg of aspirin per day, and 75 mg of clopidogrel (Plavix, Bristol-Myers Squibb and Sanofi) every day for 4 weeks. At the first control after 1 month, the CT showed a resolution of the left common iliac vein thrombosis; therefore, it was recommended to continue the therapy and periodic US follow-up until the complete resolution of femoral thrombosis. Asymptomatic patients with mild compression of the left common iliac vein should, in our opinion, undergo periodic US follow-up to prevent worsening of the stenosis. Asymptomatic patients with stenosis >50% should undergo long-term prophylaxis with anticoagulant drugs.

Conclusions

Early diagnosis in these patients is very important because it could allow us to prevent serious complications. It is advisable to choose the therapy with great caution based on the symptoms and stenosis extent, always choosing the treatment that offers the best long-term guarantees. Greater disclosure of these diseases can significantly contribute to limiting the false negatives. Failure to diagnose could expose you to serious health consequences for patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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