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Case Report

A man with the rare simultaneous combination of three abdominal vascular compression syndromes: median arcuate ligament syndrome, superior mesenteric artery syndrome, and nutcracker syndrome ☆,☆☆

Renato Farina[a], *Cecilia Gozzo*[a,*], *Pietro Valerio Foti*[a], *Andrea Conti*[a], *Tiziana Vasile*[a], *Isabella Pennisi*[a], *Massimo Venturini*[b], *Antonio Basile*[a]

[a] *Department of Medical and Surgical Sciences and Advanced Technologies "GF Ingrassia", Radiodiagnostic and Radiotherapy Unit, Catania, Italy*

[b] *Diagnostic and Interventional Radiology Department, Circolo Hospital, Insumbria University, Varese, Italy*

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ABSTRACT

Median arcuate ligament syndrome and superior mesenteric artery syndrome are well-known abdominal compression syndromes, the coexistence of which is rarely described in literature. In addition, due to the common pathogenesis, anterior nutcracker syndrome may occur simultaneously to superior mesenteric artery syndrome.

To our knowledge, this is the first case reporting combination of these 3 syndromes detected with ultrasound, Computed Tomography and upper gastrointestinal fluoroscopic exam.

A 69-year-old man came to our attention for rapid weight loss, postprandial epigastric pain and recurrent vomiting for at least 6 months. Doppler ultrasound showed both celiac artery and left renal vein stenosis with simultaneous left varicocele. Computed tomography showed a reduction of aortomesenteric space causing both left renal vein and duodenal stenosis, this latter confirmed by upper gastrointestinal fluoroscopic exam. The diagnosis of these three vascular compression syndromes (MALS, SMAS, and anterior NCS) has been formulated, based on clinical and imaging findings.

We assumed that the postprandial crises caused by median arcuate ligament syndrome may induce a reduction of meals consumption and progressive weight loss which can be a cause of anterior nutcracker syndrome and superior mesenteric artery syndrome onset.

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* Corresponding author.
E-mail addresses: radfaro@hotmail.com (R. Farina), ceciliagozzo91@gmail.com (C. Gozzo), pietrofoti@hotmail.com (P.V. Foti), conti.andrea90@gmail.com (A. Conti), tiziana_vasile@hotmail.it (T. Vasile), pennisi.isabella@gmail.com (I. Pennisi), massimo.venturini@uninsubria.it (M. Venturini), antodoc@yahoo.com (A. Basile).

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Doppler ultrasound, in expert hands, allows to accurately diagnosing these syndromes which are often underestimated. Failure to recognize it and inadequate treatment could have serious consequences for patients' health.

Introduction

The coexistence of median arcuate ligament syndrome (MALS) and superior mesenteric artery syndrome (SMAS) is rarely seen and described in literature [1,2]. Due to the common pathogenesis, SMAS and anterior nutcracker syndrome (NCS) may occur simultaneously [3]. Our literature research has shown that, until now, the combination of these 3 syndromes has never been described.

MALS, also known as Dunbar syndrome, is a vascular compression syndrome consisting in stenosis of the celiac artery (CA) which results compressed by the diaphragm median arcuate ligament (MAL) with a lower insertion [1–3]. This latter anatomical variant can be the underlying cause of postprandial epigastric pain and chronic ischemia exacerbated by exhalation due to the moving caudally of the MAL which increase the CA compression (Fig. 1a and b) [4].

Ischemia can also be asymptomatic if the arterial blood supply is well-compensated by vascular shunts between the

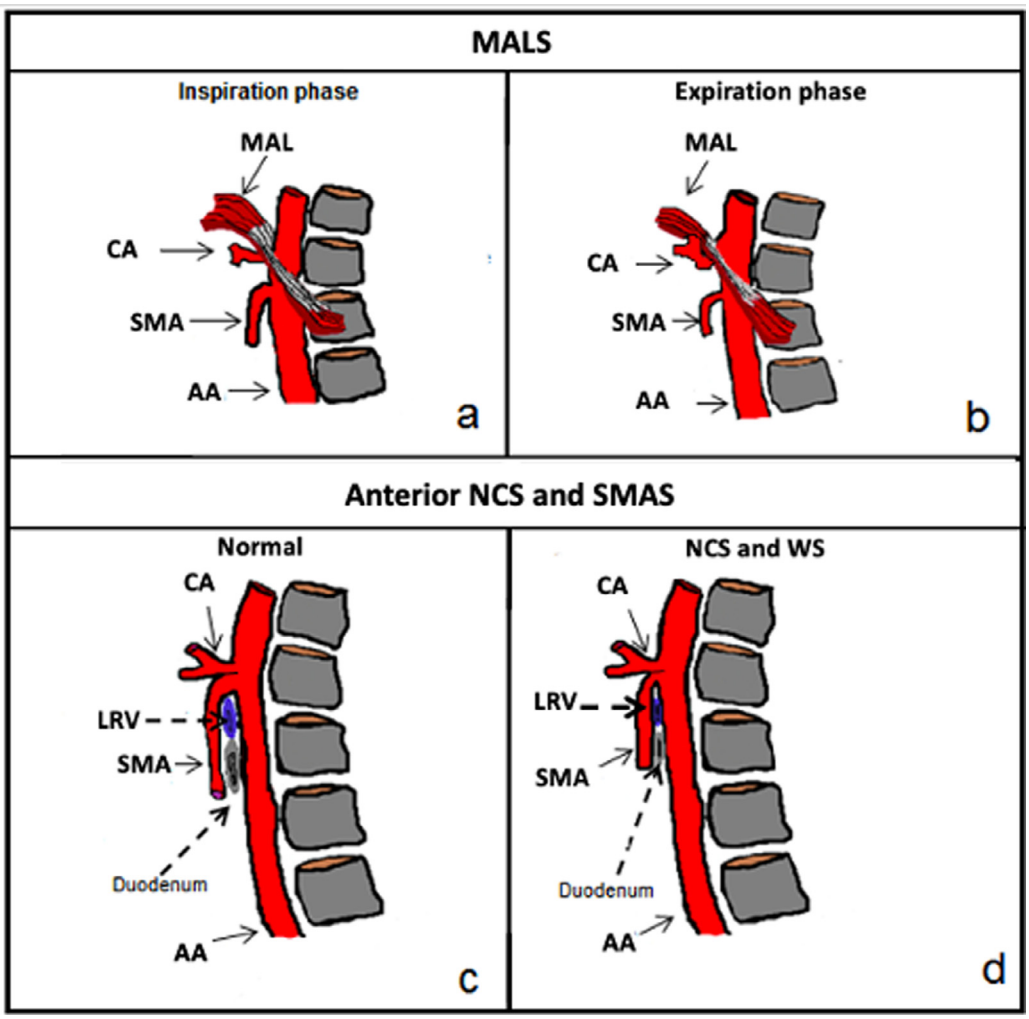

Fig. 1 – Drawings describing the main anatomical structures involved in median arcuate ligament syndrome (MALS), Nutcracker syndrome (NCS) and superior mesenteric artery syndrome (SMAS). Drawings on a sagittal plan showing the relationship between the celiac artery (CA) and the abdominal aorta (AA) in MALS respectively in (a) inspiratory apnea (b) and expiratory apnea; (c) the relationship between the SMA and the AA in healthy patient and (d) in patient with NCS and SMAS.

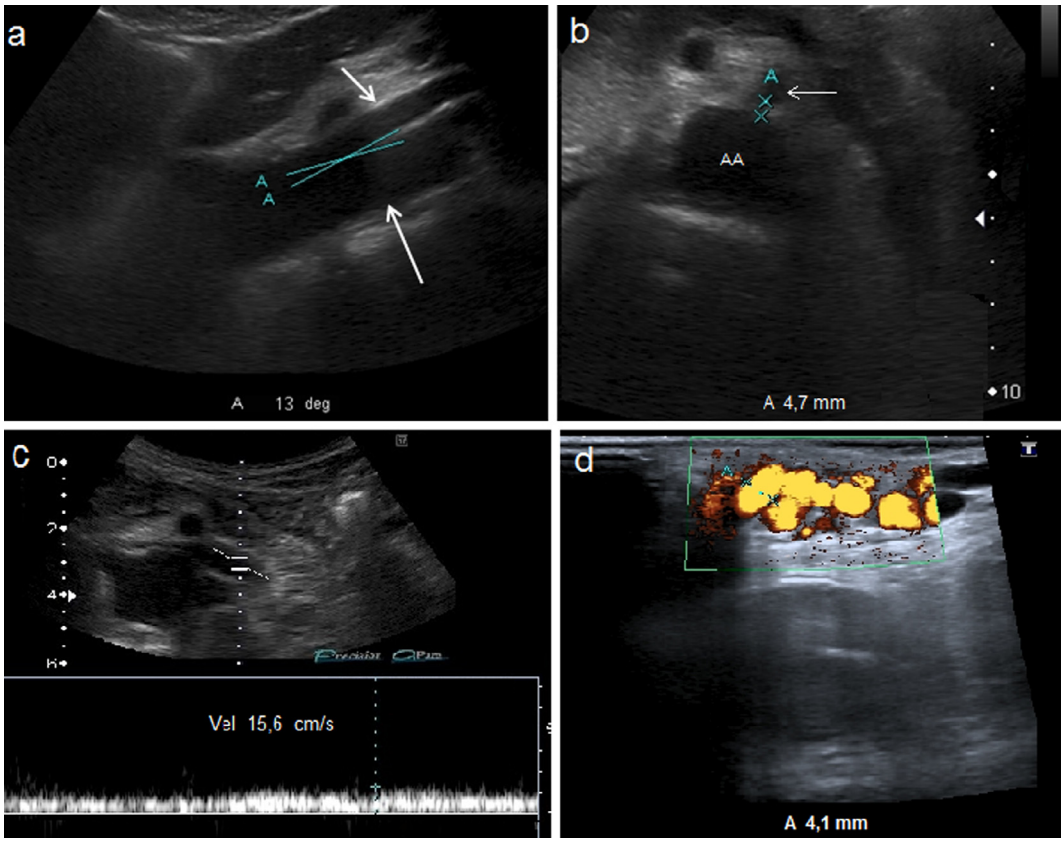

Fig. 2 – US findings of NCS. (a) Longitudinal B-Mode US scan of the abdominal aorta (AA, long arrow) and superior mesenteric artery (SMA, short arrow) with aortomesenteric angle measurement (13°); (b) transversal B-Mode US scan of the AA and SMA (arrow) with aortomesenteric distance (4,7 mm) measurement; (c) Peak Speed Velocity (PSV) measurement in the prestenotic tract of the left renal vein (LRV); (d) Power Doppler US of left pampiniform plexus showing varicocele.

CA and the superior mesenteric artery (SMA) [5]. Although some authors associate the onset of pain with the ischemia, others attribute it to compression of the celiac plexus fibers and ganglia [6,7].

The anterior NCS and SMAS (also known as Wilkie syndrome) may occur in case of reduction of the space between the SMA and the abdominal aorta (AA) (aortomesenteric distance <8 mm and aortomesenteric angle <22°) (Fig. 1c and d) with respectively compression of left renal vein (LRV) and third portion of duodenum [3,8,9].

Rapid and severe weight loss can be the underlying cause of these syndromes through the reduction of perivascular adipose tissue which normally acts as a fatty cushion in the aortomesenteric space [3]. NCS can be clinically manifest with hematuria, gonadal reflux, varicocele and, in more severe forms, irreversible damage to the left kidney caused by thrombosis of the LRV [3]; it may also have no clinical manifestations in so-called "nutcracker phenomenon" [10].

Whereas, SMAS may cause postprandial pain and vomiting related to subocclusive crisis [3].

Doppler ultrasound (US), in expert hands, can be the first imaging exam which allows recognizing CA stenosis in MALS and LRV stenosis in NCS. Computed tomography (CT) angiography may provide further details for diagnosis, such as the "hook sign" of CA typical for MALS or "beak sign" of the LVR typical for NCS [11,12].

Treatment depends on the severity of the vascular and duodenal stenosis; it can be conservative, surgical [13], or minimal invasive (eg, endovascular stenting) [14].

We report a case of combination of 3 abdominal vascular compression syndromes.

Case report

A 69-year-old underweight (body mass index of 17.01) man came to our observation complaining rapid weight loss (15 kg in 6 months), postprandial epigastric pain and recurrent episodes of vomiting for at least 6 months. US, CT, and upper gastrointestinal fluoroscopic exam were performed. The results of US study are summarized in Table 1.

US examination was performed with Aplio XG device (Toshiba) using a 3.5 MhZ convex and 7.5 MhZ linear probes. The US scans of upper abdomen showed: aortomesenteric angle of 13° (Fig. 2a) and aortomesenteric distance of 4.7 mm (Fig. 2b) representing an aortomesenteric space reduction; LRV stenosis with prestenotic dilatation, flux congestion

Table 1 – Summary of the results obtained by ultrasound examination.

Vascular structure	PSV (cm/s)		Flow-Ratio	Expiratory apnea		Inspiratory apnea	
	Prestenotic	Poststenotic		Diameter (mm)	PSV (cm/s)	Diameter (mm)	PSV (cm/s)
CA	-	-	3.6	14	246.1	22	183.6
LRV	15.6	42.4	2.71	-	-	-	-

PSV, peak speed velocity; LRV, left renal vein; CA, celiac artery.

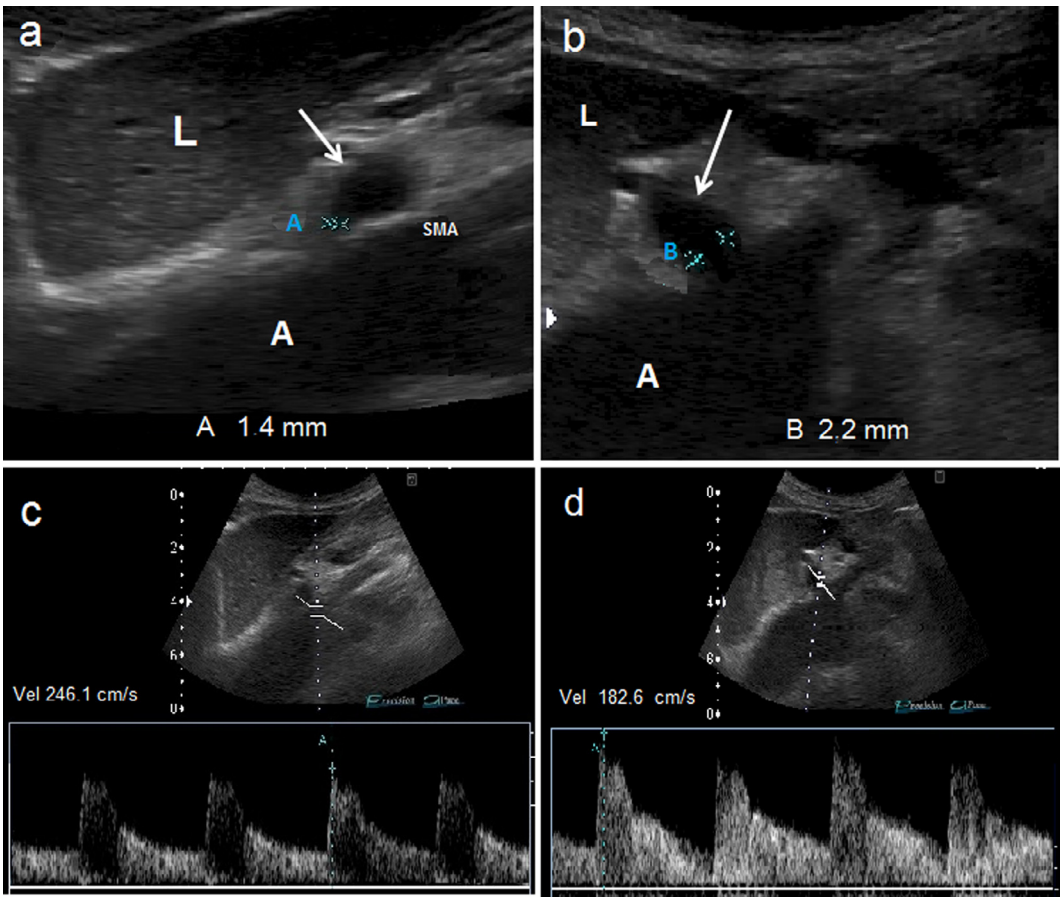

Fig. 3 – US findings of MALS. (a) Measurement of CA diameter in expiratory apnea (A: 1.4 mm). Poststenotic dilatation of CA (arrow). L: Liver. A: Abdominal Aorta. MSA: Superior Mesenteric Artery. (b) Measurement of CA diameter in inspiratory apnea (A: 2.2 mm). Poststenotic dilatation of CA (arrow). L: Liver. A: Abdominal Aorta. (c) Duplex Doppler US of CA. The PSV measurement in expiratory apnea in stenotic tract show increase of PSV (246.1 cm/s). (d) The PSV measurement in inspiratory apnea in stenotic tract show PSV reduction (183.6 cm/s).

with increased Peak Speed Velocity (PSV) in poststenotic tract (42.4 cm/sec) (Fig. 2c) and reduced PSV in prestenotic tract (15.6 cm/sec) (Fig. 2d); Flux Ratio (FR) (poststenotic tract PSV/prestenotic tract PSV) of 2.71; normal value (0.6) of left renal arterial resistive index (RI).

Doppler US of the pampiniform plexus revealed simultaneous left varicocele (III degree of Sarteschi). Doppler US performed in forced expiration showed a significant stenosis at the origin of the CA (diameter: 1.4 mm) (Fig. 3a) which decreased in forced inspiration (CA diameter: 2.2 mm) (Fig. 3b). Duplex Doppler US showed elevated flow in stenotic tract with a PSV in forced expiration of 246.1 cm/s (Fig. 3c) which decreased in forced inspiration (PSV of 183.6 cm/s) (Fig. 3d).

The FR between CA and AA measured during expiratory apnea was calculated (FR: 3.6).

In order to confirm the suspicion of abdominal compression syndromes, CT study with an Optima 64 slice CT scanner (GE, Healthcare) was performed. CT showed a stenosis of CA with "hook" appearance on sagittal reconstructions (Fig. 4a and b) typical for MALS. Axial CT multiplanar reconstructions

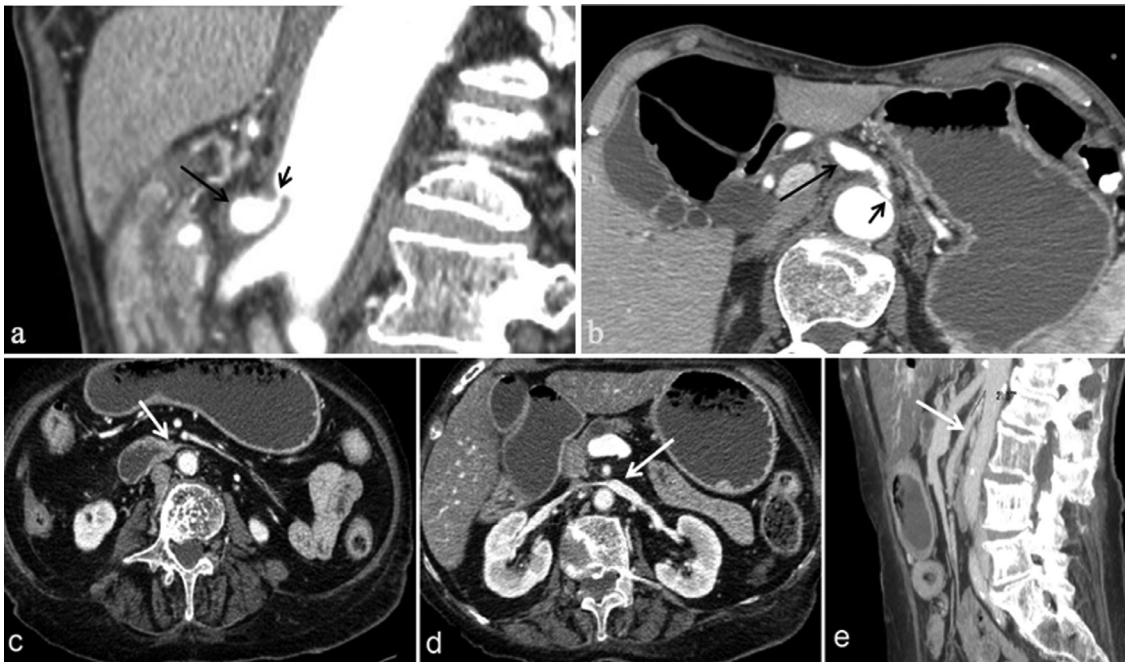

Fig. 4 – CT findings of MALS, SMAS and NCS. (a) Sagittal and axial (b) multiplanar reconstruction show a stenosis at the origin of celiac artery (CA, short arrow) with poststenotic dilation (long arrow) and the characteristic "hook sign"; (c) axial multiplanar reconstruction shows a stenosis (arrow) with prestenotic dilatation of the third portion of duodenum (SMAS); (d) axial multiplanar reconstruction shows LRV stenosis with dilation of prestenotic tract (arrow); (e) sagittal multiplanar and MIP reconstruction shows an acute angle (21°) between the superior mesenteric artery (SMA) (arrow) and the abdominal aorta (AA) reducing the aortomesenteric space with consequent compression of the LRV and duodenum.

showed compression of both third portion of duodenum (Fig. 4c) and LRV which appears dilated in the prestenotic tract with a "beak sign" typical for NCS (Fig. 4d); on sagittal reconstruction, reduction of the aortomesenteric angle (lower than 22°) was detected (Fig. 4e).

The upper gastrointestinal fluoroscopic study of the stomach and duodenum confirmed a narrowing at the third portion of the duodenum with no progression of contrast agent beyond the stenosis (Fig. 5).

Once the diagnosis of these 3 syndrome (MALS, SMAS, and anterior NCS) has been formulated, based on the imaging findings, a surgical treatment was proposed.

The patient underwent a laparoscopic MAL release and celiac ganglionectomy which resulted in a progressive resolution of postprandial symptoms; after treatment a high-calorie diet was prescribed. At 3-month US follow up, the aortomesenteric angle increased to 18° and no postprandial vomiting episodes were reported.

Therefore, it was decided to continue the high-calorie diet and US follow-up every 3 months.

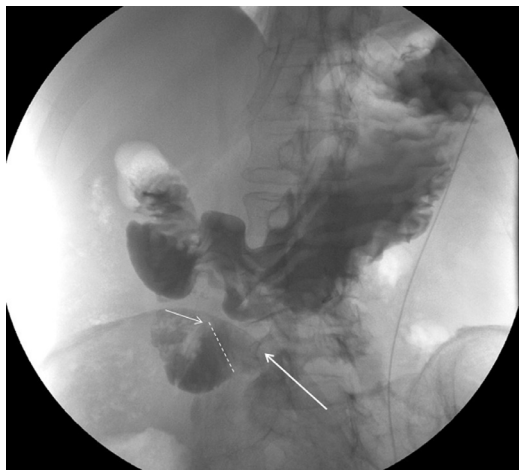

Fig. 5 – SMAS findings at upper gastrointestinal fluoroscopic exam. A conventional radiography after oral administration of contrast agent (Gastrografin) shows no progression of the opaque bolus beyond the third stenotic portion of the duodenum. The duodenum appears dilated (short arrow) due to a narrowing at the aortomesenteric space (long arrow) with no progression of the contrast agent.

Discussion

First, a search in MEDLINE literature through Pubmed was conducted using the keywords association "'median arcuate ligament syndrome/Dunbar's syndrome' AND 'superior mesenteric artery syndrome/Wilkie syndrome'" and very few article about the coexistence of MALS and SMAS were reported. Then the search of keywords "'nutcracker syndrome' AND 'median arcuate ligament syndrome/Dunbar's syndrome'" showed the same few results. Our literature research has shown that,

until now, the coexistence of MALS, SMAS and anterior NCS has never been described.

We assumed that in our patient, the postprandial crises caused by MALS have induced a limitation of meals consumption resulting in progressive weight loss; this latter caused the aortomesenteric space reduction with NCS and SMAS onset, responsible for recurrent vomiting and consequently further weight loss.

Although CT angiography is crucial to rule out other causes of compression, US examination may be the first imaging approach which allows formulating the suspicion of compressive syndromes.

Doppler US allowed accurately identifying CA and LRV stenosis and also giving estimation of stenosis degree through measurement of FR. This latter value is very useful for therapeutic planning and prophylaxis: in fact a venous FR of 2.5 cm/s corresponds to a stenosis of 50% which allows to choice a therapeutic conservative approach for NCS [15]. Meanwhile, the US evaluation of CA in expiratory apnea, allows the diagnosis of MALS in case of FR greater than 3:1 or a PSV greater than 200 cm/s [15].

Compared to CT, Doppler US shows numerous advantages due to its noninvasiveness, sensitivity, repeatability, lower cost, and lack of ionizing radiation.

Doppler US could guide in treatment choice and play an important role in follow-up.

The surgical treatment of MALS guarantees regression of symptoms in 85% of patients, while endovascular stenting is currently indicated only in cases of relapse of MALS (7%) [16].

The management of our case has first been focused on surgical resolution of CA stenosis in MALS; in addition a high calorie diet was prescribed to settle the NCS and SMAS [17].

After surgical treatment of MALS a 3-month US follow-up has been established.

Conclusions

The pathogenesis of MALS, SMAS and anterior NCS can be closely related since the progressive weight loss in MALS may cause aortomesenteric space reduction and consequently NCS and SMAS onset.

Doppler US, in expert hands, allows to accurately diagnose these syndromes and compared to CT shows numerous advantages due to its non-invasiveness, sensitivity, repeatability, low cost, and lack of ionizing radiation.

Knowing Doppler US findings could help in the diagnosis of vascular compression syndrome whose lack of recognition could result in serious health consequences.

Authors contributions

RF and CG: drafting the article, collecting and interpretation of data; RF, AC, TV, IP: data acquisition; PVF, MV, AB: All authors read and approved the final manuscript.

Ethical approval

"All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards".

Patient consent statement

The consent was obtained from the patient for the publication of this case report and accompanying images.

Clip1: US findings of NCS. Longitudinal B-Mode scan of the upper abdomen showing the reduction of the aortomesenteric angle and the distance between the SMA and the AA.

Clip2: US findings of MALS. Longitudinal B-Mode scan of the upper abdomen showing the stenosis of the CA at the origin with dilation of the post-stenotic tract.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2021.02.065.

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