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An unusual association: median arcuate ligament syndrome and nutcracker syndrome

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Key words: Abdominal pain. Arcuate ligament. Celiac trunk.

CASE REPORT

A 23-year-old female with no disease history was referred due to epigastralgia, vomiting and a 20 kg weight loss over the past year.

General laboratory tests were performed and the results were normal. An abdominal ultrasound identified a reduced aortomesenteric angle, which prompted a subsequent angio-CT scan. This scan confirmed a reduced distance between the superior mesenteric artery and the aorta, with left renal vein stenosis that was suggestive of nutcracker syndrome (NCS) (Fig. 1A). This was eventually ruled out by a urinalysis without proteinuria and a urology assessment.

Furthermore, an indentation on the superior aspect of the celiac trunk was observed with focal narrowing and post-stenotic dilation, which was suggestive of median arcuate ligament syndrome (MALS) (Fig. 1B and C). This prompted a Doppler ultrasound, which revealed an increased compression during expiration. This is a characteristic sign of MALS (Figs. 2 and 3). In view of the patient's symptoms, a laparoscopic section of the arcuate ligament was performed, which rendered the patient asymptomatic.

DISCUSSION

MALS results from the compression of the celiac trunk by the fibrous bands connecting the diaphragmatic crura on either side of the aortic arch, which give rise to the arcuate ligament. It is an uncommon condition, which is particularly associated with females (3:1) between 40 and 60 years of age.

The classic triad includes epigastralgia, vomiting and weight loss, and epigastric bruits are also characteristic (1). Doppler-US, angio-CT and angio-MRI are the diagnostic modalities of choice, which allow the visualization of the defect and of the collateral circulation when present (2,3).

Surgery is the treatment of choice for symptomatic patients, which involves ligament section together with arterial revascularization in the case of a permanent stenosis (1,3). Although NCS had no clinical relevance in our case, the unusual coexistence of both abnormalities should be highlighted.

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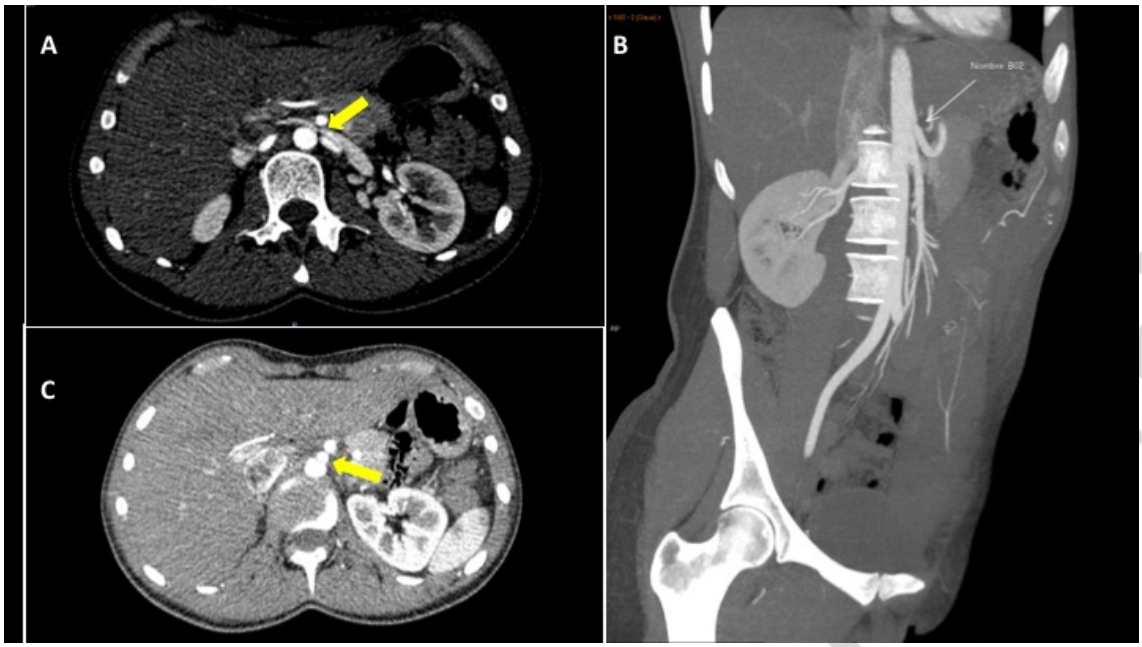

Fig. 1. A. Reduced distance between the superior mesenteric artery and aorta with a resulting left renal vein stenosis, which is suggestive of nutcracker syndrome. B. Indentation on the celiac trunk origin left by the arcuate ligament. Angio-CT reconstructions revealed a hook-shaped celiac trunk, corresponding to the so-called "hair bun and shawl" sign that may be seen in axial conventional CT images (C).

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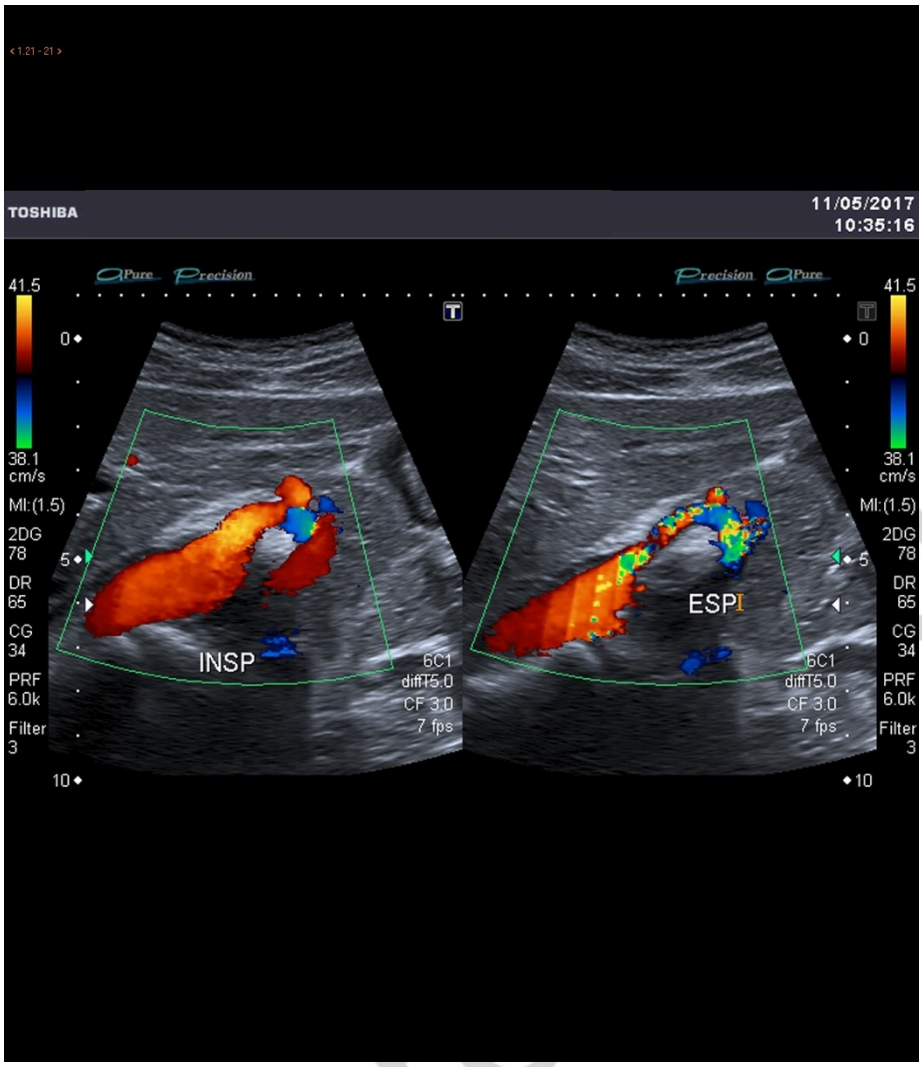

Fig. 2. B-mode, color Doppler ultrasonogram showing the proximal portion of the celiac trunk with a tortuous appearance and mildly reduced caliber, which is more obvious during expiration.

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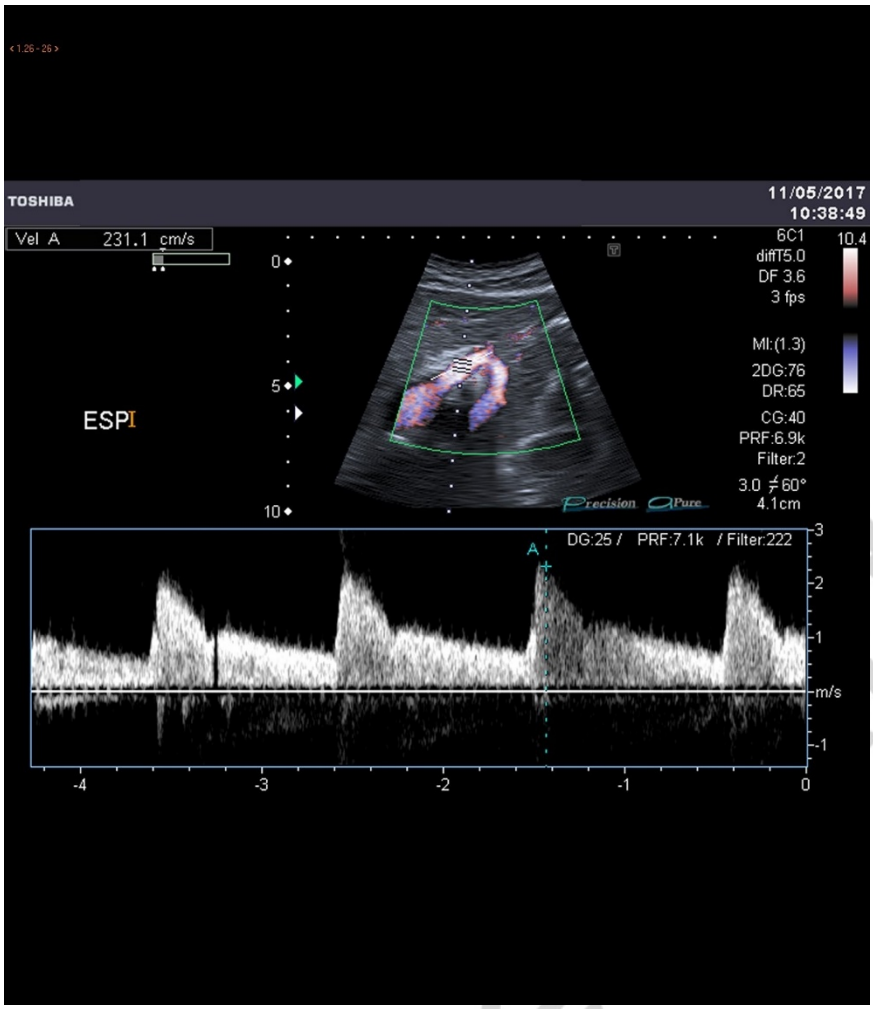

Fig. 3. Spectral Doppler ultrasonogram showing an elevated peak systolic velocity in the celiac trunk during expiration (above 200 cm/sec).