Clinical Imaging 38 (2014) 340–345

Contents lists available at ScienceDirect

Clinical Imaging

journal homepage: http://www.clinicalimaging.org

Right nutcracker syndrome associated with left-sided inferior vena cava, hemiazygos continuation and persistant left superior vena cava: a rare combination ☆

Adalet Elcin Yildiz [a,*], Fatma Semsa Cayci [b], Sinan Genc [a], Nilgun Cakar [b], Suat Fitoz [a]

[a] Department of Radiology, Ankara University School of Medicine, 06100 Ankara, Turkey
[b] Department of Pediatric Nephrology, Ankara Child Health, Hematology, Oncology, Education and Research Hospital, Ankara, Turkey

ARTICLE INFO

Article history:
Received 14 October 2013
Received in revised form 3 December 2013
Accepted 12 December 2013

Keywords:
Nutcracker syndrome
Left-sided inferior vena cava
Doppler US
MR angiography

ABSTRACT

The term nutcracker syndrome refers to compression of left renal vein between aorta and superior mesenteric artery causing renal venous hypertension. Right nutcracker syndrome associated with a left-sided inferior vena cava is an extremely rare anomaly. Reported two cases in English literature were diagnosed by ultrasonography and computed tomography angiography in adulthood. Herein, we present a case of right nutcracker syndrome with left-sided inferior vena cava and hemiazygos continuation in a 12-year-old girl.

1. Introduction

Nutcracker phenomenon is a well described anatomic anomaly which is usually associated with left renal vein (LRV) compression between the superior mesenteric artery (SMA) and aorta. Nutcracker phenomenon is called "nutcracker syndrome" when the compression of the renal vein becomes symptomatic as a result of renal venous hypertension, hilar and ureteral varices, and disruption of the small renal veins into the collecting system. The most commonly related symptoms are hematuria, proteinuria, flank pain, varicocele and chronic fatigue syndrome [1–3].

As well as its classical form there are also uncommon causes that result in compression of LRV, such as posterior nutcracker phenomenon (compression between aorta and vertebral column), compression between SMA and right renal artery or compression of a left-sided inferior vena cava (IVC) between SMA and aorta (Fig. 1A-D) [4–6]. On the other hand, not only the LRV but also the right renal vein (RRV) can be compressed and result in the right nutcracker syndrome. In English literature, there are a few case reports of right nutcracker syndrome; one was associated with pregnancy and two of them were associated with a left-sided IVC [1,2,7]. Related to the type of IVC anomaly-associated renal vein compression may be on either side; right or left (Fig. 1D-F) [1,2,6].

We reported here a case of a child with right nutcracker syndrome associated with left-sided IVC and hemiazygos continuation diagnosed by Doppler ultrasonography (US) and magnetic resonance angiography (MRA) to avoid risk of radiation in this age group.

2. Case report

A 12-year-old girl was admitted to the hospital for the evaluation of microscopic hematuria. The patient had been followed up for intermittent microscopic hematuria episodes for the previous 7 years. She had been examined with renal sonography 6 times with normal abdominal findings. Past medical history was unremarkable. There was no history of gross hematuria, trauma, urinary tract infection, arthralgias, or skin rashes. Family history was negative for hematuria, deafness, rheumatologic disease, or end-stage renal disease. The physical examination, ear and audiological examinations were entirely normal. Laboratory test results of complete blood count, serum electrolytes, and renal function tests were entirely in normal limits. Urine red blood cell count was 10–33 per high-power field. Remaining urine test results were normal. There was not any abnormality on kidney-bladder ultrasonography. On the basis of clinical and laboratory findings, she was referred to radiology department for renal Doppler US. At initial evaluation, renal Doppler US showed the absence of a right-sided IVC except its suprahepatic segment. However, at the left side of the aorta, there was a persisting left IVC being formed by right and left common iliac veins. Both the renal veins were draining into the left-sided IVC and right renal vein was compressed while passing between aorta and SMA. The diameters of RRV at the hilar and aortomesenteric stenotic portion were 7.7 and 1.1 mm, respectively (Fig. 2A, B). At spectral Doppler US

☆ No funding was received from any source. The authors declare no conflicts of interest.
* Corresponding author. Department of Radiology, Ankara University School of Medicine, 06100 Ankara, Turkey. Tel.: +90 505 8053 836.
E-mail address: aelcindr@gmail.com (A.E. Yildiz).

0899-7071/$ – see front matter © 2014 Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.clinimag.2013.12.007

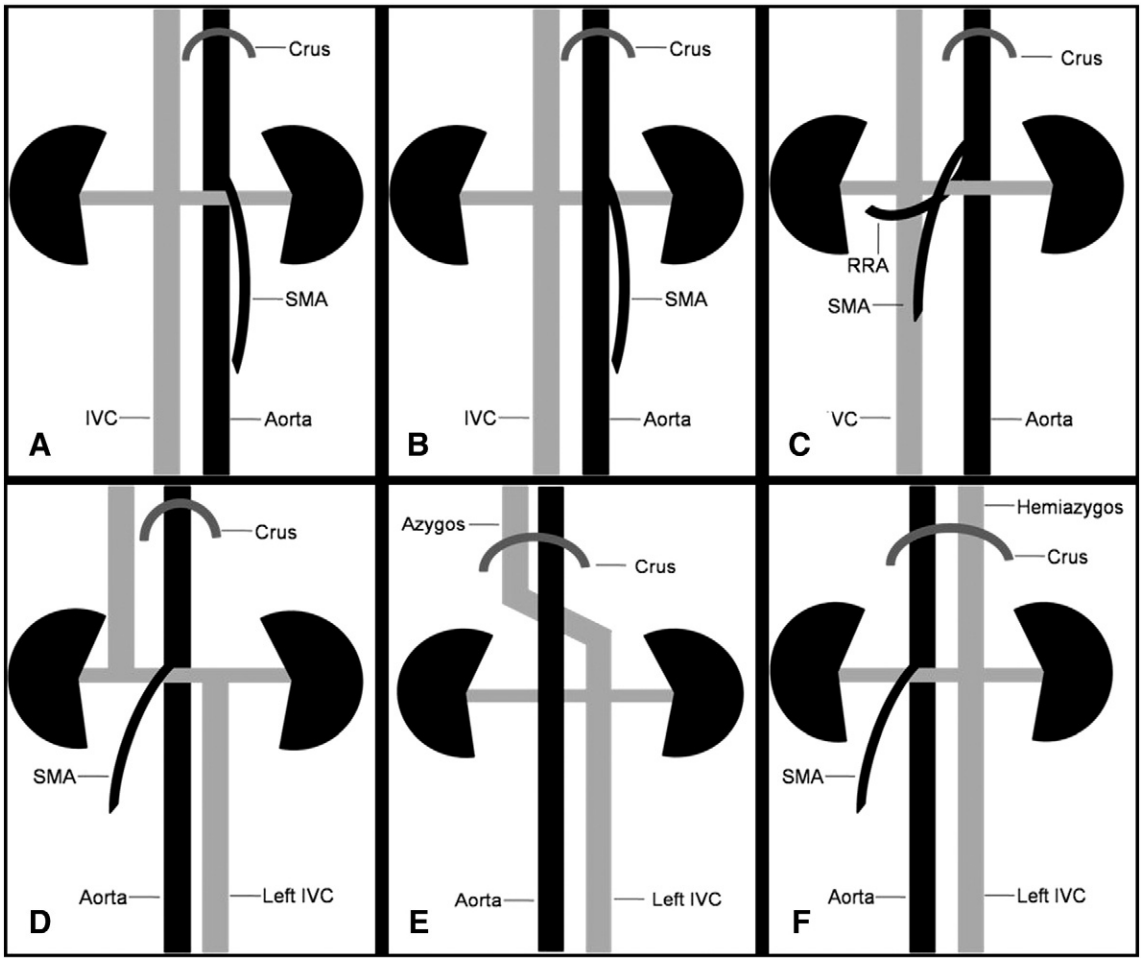

Fig. 1. Illustrations (A-F) show common and rare forms of nutcracker syndrome. In classical form (A) left renal vein is compressed between SMA and aorta. In posterior nutcracker phenomenon (B) left renal vein is compressed between aorta and vertebral column. Left renal vein compression between SMA and right renal artery (C) and compression of a left sided IVC between SMA and aorta (D) are rare types of left nutcracker syndrome. Compression of right renal vein between aorta and vertebral column and left-sided IVC between aorta and vertebral column (E) is a form of bilateral nutcracker syndrome. Compression of right renal vein between SMA and aorta associated with left-sided IVC (F) is another rare form of left nutcracker syndrome.

analysis of the RRV at the hilar portion and aortomesenteric stenotic portion, the peak velocities were 21.9 cm/s and 103.9 cm/s, respectively (Fig. 3A, B). SMA angle measurements in the supine and upright positions were 28° and 11°, respectively (Fig. 4A, B). The diameter and peak velocity ratios between dilated and stenotic portions of the RRV (7 and 4.7, respectively) and SMA angle were consistent with nutcracker phenomenon. The suprarenal segment of the left-sided IVC could be traced to the diaphragmatic level on the left side of midline. For further evaluation of this rare vascular anomaly at both sides of diaphragm we also performed contrast enhanced MRA. MRA was performed by using a 1.5 T MRI system. In addition to Doppler US findings, MRA revealed that left sided IVC continuation with a dilated hemiazygos (Fig. 5). The hemiazygos was draining into persistent left superior vena cava (SVC) with the left brachiocephalic venous system and finally to the left coronary sinus (Fig. 6). As the patient is a child and had not experienced any symptom with the episodes of microscopic hematuria, no intervention was planned and conservative management with regular follow-up was recommended.

3. Discussion

In normal anatomical formation, IVC lies on the right side of aorta and comprise four segments: hepatic, suprarenal, renal, and infrarenal. Embryogenesis of the IVC is a complex process which may result in different variations with renal vein anomalies. In the embryonic life during the sixth to eighth week of gestation, three pairs of fetal veins—posterior cardinal, subcardinal, and supracardinal veins—develop, regress, and form anastomoses between each other in an order and finally form the infrarenal IVC on the right side [8].

Although anomalies of IVC are rare, there are almost 14 theoretical variations in the anatomy of the infrarenal IVC described by the investigators. Left-sided IVC anomaly with a type of joining with the left renal vein and crossing anterior to the aorta to unit with the right renal vein and form a normal right-sided pre-renal IVC has a prevalence of 0.2–0.5% [7]. Persistence of the left supracardinal vein with regression of the right supracardinal vein leads to this anomaly. Besides, a left-sided IVC can also show an azygos or hemiazygos continuation. If a hemiazygos vein continuation occurs it can either drain into the azygos or join the coronary vein by a persistent left SVC or drain into left brachiocephalic vein by an accessory hemiazygos continuation [7]. If the left-sided IVC is compressed while passing between SMA and aorta it may result in nutcracker syndrome as reported with a few cases in the literature (Fig. 1D) [6,9,10]. Recently, two other types of left-sided IVC anomalies associated with nutcracker syndrome were reported in the literature: a left-sided IVC with azygos continuation and a left-sided IVC with hemiazygos continuation, the latter one is similar to our case [1,2]. In the first case reported by Luo et al. [1], nutcracker syndrome was obtained on both sides: right renal vein