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The roles of the liver and pancreas in renal nutcracker syndrome

Seong Jong Yun[a,b,1], Deok Ho Nam[a,*], Jung Kyu Ryu[a,2], Ji Su Kim[a,3]

[a] Department of Radiology, Kyung Hee University Hospital at Gangdong, Kyung Hee University School of Medicine, 149 Sangil-dong, Kangdong-gu, Seoul 134-727, South Korea

[b] Department of Radiology, Graduate School of Medicine, Kyung Hee University, Hoegi-dong, Dongdaemun-gu, Seoul 130-701, South Korea

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ABSTRACT

Introduction: To assess the frequency and significance of presence of the liver and pancreas at the left renal vein (LRV) level in patients with suspected renal nutcracker syndrome (NCS).

Materials and methods: We included 101 patients with hematuria who underwent urography three-dimensional CT between April 2009 and November 2013. These patients were divided into NCS ($n = 25$) and non-NCS ($n = 76$) patients according to the following CT criteria: (1) the presence of beak sign and (2) hilar-aortomesenteric left renal vein diameter ratio >4. Patients were grouped according to the presence of the liver and pancreas at the LRV: group LP (both liver and pancreas), group L (only liver), group P (only pancreas), and group O (neither liver nor pancreas). The difference in the frequencies of groups was analyzed between NCS and non-NCS patients. Multivariate analysis was used to determine the independent factors between NCS and non-NCS patients.

Results: The frequencies of group LP, group L, group P, and group O in NCS vs. non-NCS were 88% vs. 5.3% ($p < 0.001$), 4.0% vs. 2.6% ($p = 0.75$), 4.0% vs. 11.8% ($p = 0.45$), 4.0% vs. 80.3% ($p < 0.001$), respectively. Multivariate analysis demonstrated that group was a predictor for differential diagnosis between NCS and non-NCS ($p = 0.022$), and group LP was an independent factor for the presence of NCS (odds ratio, 43.8; 95% confidence interval, 3.8–500.3; $p < 0.002$; reference, group O).

Conclusion: The presence of the liver and pancreas at the level of the LRV was frequently found in NCS and was the independent factor for NCS.

1. Introduction

Nutcracker syndrome (NCS) indicates entrapment of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the aorta and presents with characteristic clinical symptoms [1]. The most commonly observed clinical symptom of NCS is hematuria, which is attributed to the rupture of thin-walled varices as a result of elevated venous pressure in the collecting system. The incidence of NCS is poorly characterized, although it usually occurs in adolescence or young adulthood and affects females more frequently than males [2].

Left renal venography is the most reliable method for diagnosis of NCS, but it is not commonly performed, particularly in children [3]. Recently, multiphase computed tomography (CT) urography has been shown to be a suitable method for patients with suspected NCS [3,4] because it clearly depicts the LRV and related structures around the aorta and SMA as well as the collateral circulation around the left renal hilum [4].

The etiology of the nutcracker phenomenon is not fully recognized. The most popular theory is that an acute aorto–SMA angle (<90°) causes compression of the LRV, leading to venous congestion of the left kidney [5]. However, abnormal origin [3] or abnormal branching [6] of the SMA from the aorta, ptosis of the left kidney [7], and preaortic fibrous tissue [8] have also been described as causes of NCS. Rarely, posterior NCS can be caused by a circumaortic left renal vein [9]. Buschi et al. [10] reported that LRV compression occurred even with a larger aorto–SMA angle in a patient with duodenal interposition. Based on our experience, we suggest that LRV compression may be induced by the liver and pancreas, which compress the SMA and LRV against the aorta. However, to

* Corresponding author at: Department of Radiology, Kyung Hee University Hospital at Gangdong, Kyung Hee University School of Medicine, 149 Sangil-dong, Kangdong-gu, Seoul 134-727, South Korea. Tel.: +82 2 440 6211; fax: +82 2 440 6932.

E-mail addresses: zoomknight@naver.com (S.J. Yun), namjindan@daum.net, dukenam23@naver.com (D.H. Nam), oddie2@naver.com (J.K. Ryu), js830808@hanmail.net (J.S. Kim).

[1] Tel.: +82 2 440 6933.
[2] Tel.: +82 2 440 6188.
[3] Tel.: +82 2 440 6929.

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our knowledge, there are no reports on whether the presence of the liver and pancreas in the anterior aspect of the SMA may contribute to compression of the LRV.

The purpose of this study was to assess the frequency and significance of the presence of the liver and pancreas at the LRV level in NCS patients.

2. Materials and methods

2.1. Patient selection

The institutional review board at our hospital approved this retrospective study, and the requirement for informed consent was waived. Collection of data and information was performed by one independent radiologist (J.S.K.) based on PACS (Piview Star, Infinitt Healthcare, Seoul, Korea) and electronic medical records. According to our institute's routine protocol as of April 2009, adult (over 18 years) patients with hematuria initially underwent urographic three-dimensional (3D) CT, but pediatric patients (<18 years) underwent urographic 3D CT either initially or two weeks later after conservative treatment for diagnosis including NCS. The inclusion criteria were age 40 years or younger and urographic 3D CT for evaluation of hematuria between April 2009 and December 2013 at our hospital. The exclusion criteria was another definite cause of hematuria such as trauma, stone, or neoplasm. Of the total 206 patients with hematuria, 105 patients were excluded for other causes of hematuria including trauma ($n = 11$), renal or ureter stones ($n = 45$), acute pyelonephritis ($n = 13$), cystitis ($n = 11$), renal cell carcinoma ($n = 8$), transitional cell carcinoma ($n = 9$), renal tuberculosis ($n = 4$), and idiopathic ureteropelvic junction obstruction ($n = 4$). The remaining 101 patients (mean age, 26.2 ± 8.8 [standard deviation] years; age range, 8–40 years) enrolled in this study included 62 male patients (mean age, 24.5 ± 8.6 years; range, 8–40 years) and 39 female patients (mean age, 28.8 ± 8.8 years; range, 10–40 years).

2.2. Imaging acquisition

Quadruple-phase (precontrast, corticomedullary, nephrographic, and excretory) scanning was performed using a 16-slice ($n = 70$) or 64-slice ($n = 31$) multidetector CT scanner (Brilliance 16 and 64; Philips Medical Systems, Cleveland, OH, USA). Scanning was performed after infusion of 2 mL/kg of iopamidol (Pamiray 370; Dongkuk Pharm, Seoul, Korea) through the antecubital vein at a rate of 4 mL/s. The scan range was from the lung base to the lesser trochanters of the femurs in the craniocaudal direction on portal phase and from the diaphragm to the lesser trochanters of the femurs in the craniocaudal direction on pre, arterial, and delayed phases.

CT examinations using a 16-slice MDCT were performed with the following parameters: tube voltage, 120 kVp; tube current, 200 mAs; collimation, 16×1.5 mm; pitch, 1.188; rotation time, 0.75 s; field of view, 350 mm; matrix, 512×512; slice thickness, 5 mm; reconstruction interval, 5 mm. The parameters for the 64-slice MDCT scanner were as follows: tube voltage, 120 kVp; tube current, 150 mAs; collimation, 64×0.625 mm; pitch, 0.891; rotation time, 0.75 s; field of view, 350 mm; matrix, 512×512; slice thickness, 5 mm; reconstruction interval, 5 mm. For corticomedullary phase scanning, a delay of 15 s was fixed after aorta attenuation reached 200 Hounsefield units according to bolus tracking. For nephrographic phase scanning, a delay of 55 s was fixed after corticomedullary scanning. For excretory phase scanning, a delay of 5 min was fixed after infusion of the contrast media.

Fig. 1. Measurement of left renal vein (LRV) diameter ratio. The diameter of the LRV in the aortomesenteric space (line A) and left renal hilum (line B) were measured, and ratios were calculated as B/A.

2.3. Reference standard of NCS on CT

For our study, urographic 3D CT was used as a reference standard. Two board-certificated radiologists (D.H.N. with 14 years of experience and J.K.R. with 12 years of experience) interpreted the urographic 3D CT data using a PACS and categorized the patients as NCS or non-NCS by consensus with reference to previously published CT findings for NCS [10,11]. The diagnostic criteria were as follows: (1) ratio of LRV diameter >4.0 (Fig. 1) and (2) the presence of the beak sign in all phase. Patients who satisfied both criteria were diagnosed with NCS (Fig. 2). The aortomesenteric angle was not included in the diagnostic criteria because sagittal reformatted images were not routinely obtained. In axial images of the corticomedullary phase, the LRV diameter was measured at both the hilum and aortomesenteric space (AMS). Beak sign was defined as an acute angle formed by the anterior and posterior walls of the narrowing segment of the LRV at the AMS [11].

2.4. Imaging analysis

All axial CT scans of the corticomedullary phase were evaluated retrospectively by one independent radiologist (S.J.Y.) who was not involved in image evaluation. The reviewer was unaware of any information including final clinical diagnoses or prevalence of NCS. The reviewer performed the evaluations in random order irrespective of CT study date using PACS and evaluated the following at the LRV level: (1) whether or not liver or pancreas was visualized; (2) whether or not liver or pancreas crossed the aorto–SMA line (Fig. 3); (3) the shortest distance of the AMS (D_{AMS}); (4) the shortest distance from the anterior abdominal wall to the posterior abdominal wall (D_{APW}), the shortest distance from the aorta to the anterior abdominal wall (D_{AW}), the shortest distance from the aorta to the posterior abdominal wall (D_{PW}) and the ratio of D_{AW} to D_{PW} (R_{APW}); and (5) the oblique origin of the SMA. Definitions for each value are presented in Figs. 4 and 5. According to the presence of liver and pancreas at the LRV level, patients were divided into four groups; (1) group LP: both liver and pancreas were visualized and crossed the aorto–SMA line; (2) group L: only liver was visualized and crossed the aorto–SMA line; (3) group P: only pancreas was visualized and crossed the aorto–SMA line; and (4) group O: neither the liver nor the pancreas were visualized, or either liver or pancreas was visualized but did not crossed the aorto–SMA line (Fig. 6). Evaluation of D_{APW}, D_{AW}, D_{PW}, and