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Case Report

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Retrocaval ureter and the diagnostic utility of dynamic scintigraphy: A case report

Sıla Ece Tiryaki, İrem İnanç, Dinçer Avlan

Abstract

Retrocaval ureter is a rare congenital anomaly characterized by the persistence of posterior cardinal veins in front of the ureter due to their failure to regress in early embryonic development. Its main symptoms are discomfort, right flank pain, hematuria, recurrent infections, fever, and stone formation. A seven-year-old male patient presented with hematuria. Urinary ultrasound revealed Grade 2 hydronephrosis in the right kidney and dilation in the right proximal ureter. Considering the possibility of ureteropelvic junction obstruction, dynamic renal scintigraphy was performed which showed radiopharmaceutical accumulated in the mid-segment of the right ureter, likely due to compression that did not pass distally. To confirm the diagnosis, retrograde pyelography was performed perioperatively, and it was consistent with the retrocaval ureter. Laparotomy showed that the ureter ran behind the inferior vena cava from its mid-segment. A ureteroureterostomy was performed, the ureter was positioned anterior to the inferior vena cava, and a ureteral stent was inserted. In conclusion, renal scintigraphy traditionally provides information about the degree of obstruction and renal function. However, as in the presented case, the examination of dynamic renal scintigraphy images showing the accumulation of radiopharmaceutical substance in the mid-segment of the ureter and the absence of the distal ureter may indicate a retrocaval ureter. It is, therefore, of utmost importance for clinicians to review the images obtained from dynamic scintigraphy in cases of obstructive uropathy and recognize the possibility of retrocaval ureter.

Keywords: Retrocaval ureter, scintigraphy, ureteral obstruction.

Retrocaval ureter, also known as circumcaval ureter or pre-ureteral vena cava, is a rare congenital anomaly characterized by the persistence of posterior cardinal veins in front of the ureter due to their failure to regress in early embryonic development.[1,2] It has an autopsy incidence of 0.9 per 1,000, with a predominance on the right side. On the left side, it is associated with partial or complete situs inversus anomalies or inferior vena cava duplications.[2]

With multiple variants defined in the literature, in most cases, the main symptoms include discomfort, right flank pain, hematuria, recurrent infections, fever, and stone formation that manifest in the third or fourth decades.[1,2] While some patients remain asymptomatic throughout their lives, patients who present with obstruction symptoms or kidney function deficits require surgical intervention.[2] There are several open surgical methods; in particular, ureteroureterostomy has been considered gold standard, since it was first used to treat a patient with a retrocaval ureter by Anderson and Hynes in 19492,.[3] Recent studies have also suggested laparoscopy as a less invasive method with significantly prolonged operation duration.[2]

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In this article, we report a case of retrocaval ureter diagnosis which was confirmed by renal scintigraphy and was successfully treated with ureteroureterostomy.

CASE REPORT

A seven-year-old male patient presenting with hematuria was referred to the Department of Pediatric Surgery. Urinary ultrasound revealed

Grade 2 hydronephrosis in the right kidney and dilation in the right proximal ureter. No significant pathology was reported in the left kidney and ureter. Considering the possibility of ureteropelvic junction obstruction, dynamic renal scintigraphy (Tc-99m mercaptoacetyltriglycine [MAG3]) was performed. It was reported as a partial response to diuretics in the right kidney, with ongoing retention in the renal pelvis and dilated 1/2 proximal ureter. In the dynamic scintigraphy images, the radiopharmaceutical was found to be accumulated in the mid-segment of the right ureter, likely due to compression, and did not pass distally (Figure 1). To confirm the diagnosis, retrograde pyelography was performed perioperatively and it was consistent with the retrocaval ureter (Figure 2). Subsequently, a laparotomy was performed. During laparotomy, it was discovered that the ureter ran behind the inferior vena cava from its mid-segment (Figure 3). An open ureteroureterostomy was, then, performed, and the ureter was positioned anterior to the

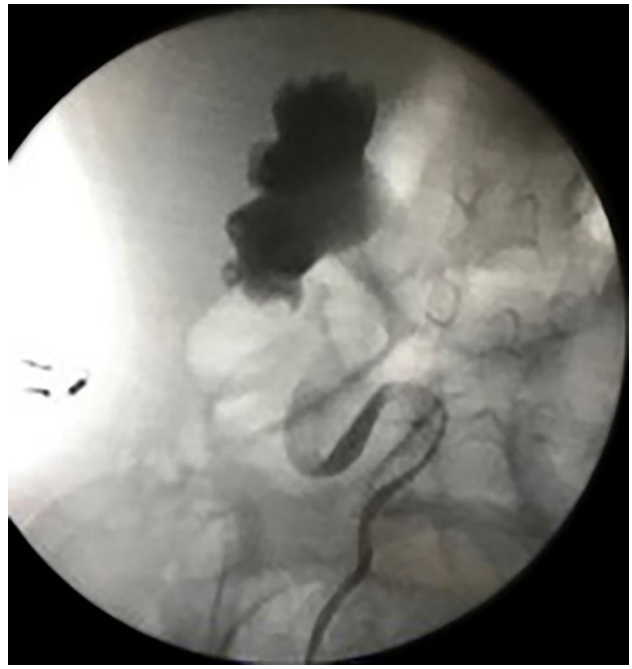

Figure 2. Retrograde pyelography image.

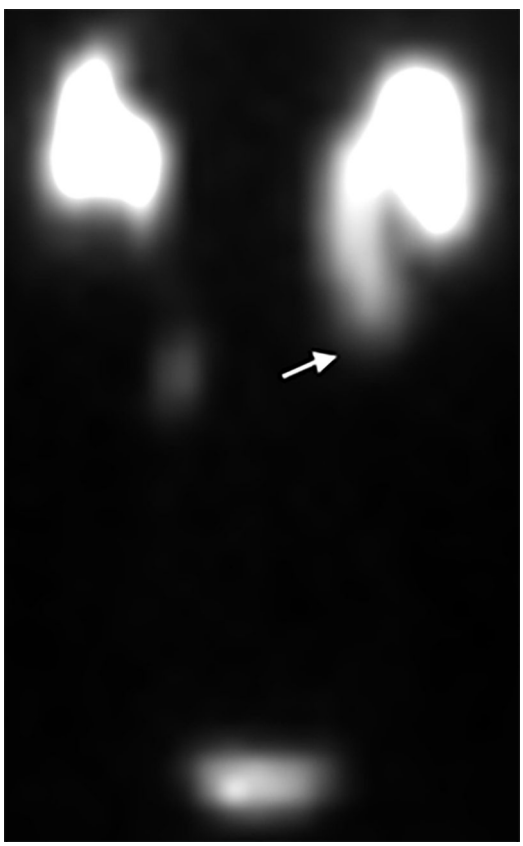

Figure 1. Reverse J sign in MAG3 (arrow).
MAG3: Mercaptoacetyltriglycine.

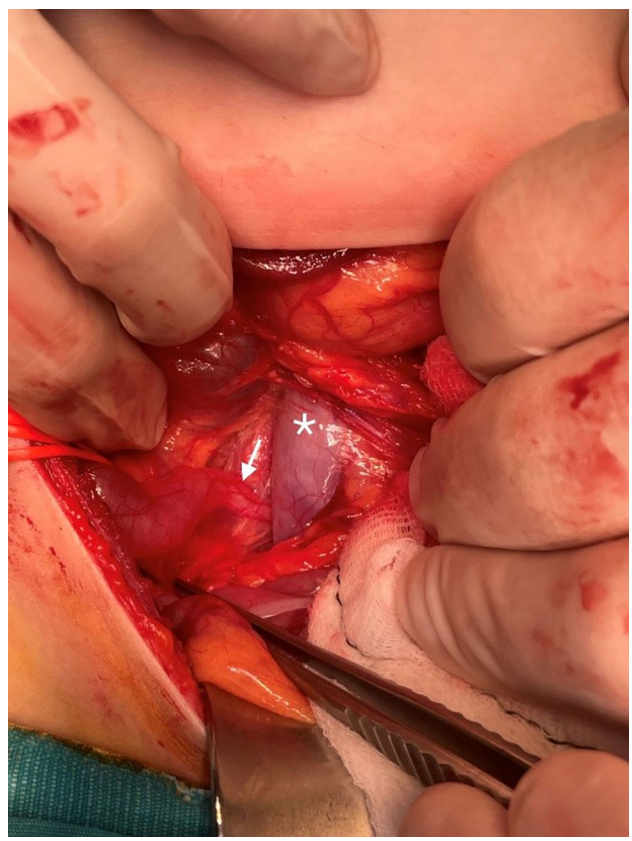

Figure 3. Intraoperative image of vena cava and ureter.
Arrow: right ureter and asterisk; * Vena cava inferior.

inferior vena cava, and a ureteral stent was inserted to preserve the anastomosis. The stent was removed three weeks after surgery and control ultrasound showed regression of hydronephrosis. A written informed consent was obtained from the parent of the patient.

DISCUSSION

Obstructive uropathy caused by the ureter being compressed by the inferior vena cava results in findings of moderate hydronephrosis and dilation in the proximal ureter on ultrasound. Currently, intravenous magnetic resonance imaging-urography (MRI-urography), three-dimensional (3D)-computed tomography (3D-CT), and retrograde pyelography are the methods used for diagnosis, with the abrupt narrowing and interruption of the ureter in the mid-segment being described as a "fishhook" or "reverse J" sign.[4-6] Renal scintigraphy traditionally provides information about the degree of obstruction (diethylenetriamine pentaacetate [DTPA] or MAG3) and renal function (dimercaptosuccinicacid [DMSA]).[7] However, as in the presented case, the examination of dynamic renal scintigraphy images showing the accumulation of radiopharmaceutical substance in the mid-segment of the ureter and the absence of the distal ureter may be indicative of a retrocaval ureter. As a result of scintigraphy images, renograms, and clinical features suggesting obstruction, many causes, such as foreign bodies, inflammation, tumors, and anatomical abnormalities that cause obstruction, may be suspected. However, as in our case, the diagnosis should be confirmed via a CT or an MRI scan, the gold-standard imaging technique.[2] In this case, scintigraphy is not mentioned as the gold-standard imaging method, but as a provider of findings that can direct clinicians to a retrocaval ureter diagnosis in mid-obstruction findings.

Scintigraphy remains a preferred method due to minimal complications and low costs.[8] It is also preferred in cases where CT and MRI scans should be avoided, such as claustrophobia and allergy to contrast matter. Also, it can be preferred, particularly in pediatric patients, due to lower-dose radiation exposure compared to a CT scan.[8]

In conclusion, it is of paramount importance for clinicians to review the images obtained from dynamic scintigraphy in cases of obstructive uropathy and recognize the possibility of retrocaval ureter, among other differential diagnoses, while encountering such images.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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