RESIDENT'S CORNER

Gross hematuria caused by nutcracker syndrome and segmental pseudoaneursym

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Nutcracker syndrome is a rare disorder characterized by extrinsic compression of left renal vein by the superior mesenteric artery and aorta. Prevalence of the disease is unknown, but presents most commonly with gross hematuria and flank pain. Diagnosis requires a high

Introduction

Nutcracker syndrome is a rare cause of gross hematuria first pathologically described in 1937 by Grant and clinically in 1950 by El-Sadr and Mina.^{1,2} Impeded outflow from the left renal vein into the inferior vena cava is caused by extrinsic compression between the superior mesenteric artery and aorta resulting in left renal vein hypertension. Presentation consists of several symptoms not limited to gross hematuria, left flank pain, pelvic congestion syndrome, and/or varicocele. Imaging beginning with renal ultrasound and confirmed with renal angiography is the key to diagnosis. We present a case of 29-year-old female recently recovered from endometrial ablation with persistent gross hematuria secondary to nutcracker syndrome, segmental artery pseudoaneursym, and pelvic congestion syndrome.

index of suspicion and treatment consists of a wide range of vascular surgical options with a more recent focus using an endovascular approach. We present a case of a 29-year-old female with continuous gross hematuria and flank pain from a segmental artery pseudoaneursym secondary to nutcracker and pelvic congestion syndrome.

Key Words: pseudoaneursym, nutcracker syndrome, hematuria, pelvic congestion syndrome

Case report

A 29-year-old female with a history of Gaucher's disease presented to a community hospital with gross hematuria and left flank pain. The patient had undergone endometrial ablation approximately 1 week prior for menorrhagia. Her hemoglobin at presentation was 7 g/dL. The gross hematuria resolved spontaneously on hospital day one and the patient was sent home. The gross hematuria recurred approximately 2 days later with clots. A local urologist placed a Foley catheter and transferred the patient to the local hospital. The patient underwent cystoscopy with clot evacuation during which blood was seen effluxing from the left ureteral orifice. CT abdomen and pelvis revealed only an enlarged left kidney with a delayed nephrogram and absence of hydronephrosis. The patient was maintained on continuous bladder irrigation, while her hemoglobin trended down to a nadir of 5.63 g/dL. At this point the patient was transferred to University Hospital after being transfused with three units of packed red blood cells. The following morning the patient

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underwent a combination case with urology and gynecology. Cystoscopy, left retrograde pyelogram, and left ureteroscopy revealed a filling defect in the left upper pole with a tuft of tissue in the left upper pole calyx consistent with a necrosed papilla. The papilla was laser ablated and left ureteral stent placed. Gynecological evaluation revealed no abnormalities. The patient's gross hematuria resolved, her hemoglobin remained stable, and catheter was removed.

Four days later the patient presented again to the same community hospital with gross hematuria and left flank pain. A Foley catheter was placed, hemoglobin nadired to 6.3 g/dL. The patient was transfused with packed red blood cells and imaging ordered. CT abdomen and pelvis revealed delayed nephrogram of the left kidney and extensive vascular collateral vessels surrounding the uterus consistent with pelvic congestion syndrome. Interventional radiology service was consulted and a left renal arteriogram and pelvic angiogram were performed. A left superior segmental artery pseudoaneursym was found, that was subsequently embolized using tornado coils, Figure 1. In addition, external compression of the left renal vein by the superior mesenteric artery and aorta was visualized, along with an engorged left gonadal vein



Figure 1. Left renal arteriogram showing left superior segmental artery pseudoaneursym.



Figure 2. Left renal and pelvic angiogram showing engorged left gonadal vein.

that was responsible for the primary drainage of the left kidney, Figure 2. These findings were consistent with nutcracker and pelvic congestion syndrome. The patient underwent left renal vein endovascular stenting with resolution of gross hematuria. The patient has returned for outpatient follow up 1 month post discharge with no further occurrences of hematuria.

Discussion

Overview

The first pathologic report of nutcracker phenomenon was in 1937 by Grant,¹ and in 1950 the first clinical report was published in the literature by El-Sadr and Mina.² Nutcracker syndrome typically occurs when the left renal vein becomes compressed between the abdominal aorta and the superior mesenteric artery. Normally, the left renal vein passes anterior to the aorta at an angle of approximately 90°. In cases of nutcracker syndrome, the superior mesenteric artery is often noted to branch from the aorta at a more acute angle. As a result of this obstruction, there is impaired blood outflow to the inferior vena cava, and a variety of clinical symptoms may develop.

There are several theories of the causes of nutcracker syndrome including posterior left renal ptosis, an uncharacteristically high course of the left renal vein, and any unusual branching of the superior mesenteric artery from the aorta.³ Any other tumors, vascular abnormalities, or extensive lymphadenopathy can additionally cause compression of the left renal vein. Patients tend to be children and young adults, although cases have been reported from childhood to the seventh decade, and a slight predisposition for women has been found. This syndrome is also likely not a hereditary occurrence.

Clinical features

The classic symptoms of nutcracker syndrome include left flank pain with either gross or microscopic hematuria. The frequency and severity of symptoms can vary greatly which often contributes to missed diagnosis. Hematuria is the most common reported symptom, followed by pain. Pain is typically described as originating in the abdomen or the flank, and it may radiate to the posteromedial thigh. Flank pain results from ureteral colic as a consequence of passing blood clots.

Women often present with symptoms of pelvic congestion such as dysmenorrhea, dyspareunia, dysuria, lower abdominal pain and pelvic pain. In one study, out of 51 patients with pelvic congestion, 9 of these patients were diagnosed with nutcracker syndrome.⁴ While cases have been reported of the acute improvement of symptoms, the interruption of the gonadal veins may more commonly exacerbate the patient's symptoms, as these gonadal veins become channels for the outflow of blood from the compressed renal vein. Males can present with varicoceles that result from the reflux of blood from the compressed renal vein to the gonadal vein. Additional symptoms include pelvic varicosities, orthostatic proteinuria and anemia resulting from hematuria. Lower body mass index in patients has been found to correlate positively with Nutcracker phenomenon, as well as lower levels of retroperitoneal fat.5

Diagnosis

Diagnostic criteria have not been clearly established, but as in most pathological conditions, the primary initial diagnostic tests should be a thorough history and physical exam. Other more common kidney conditions often need to be ruled out when the presenting symptoms include hematuria. As a result, diagnostic tests including urinalysis, urine culture, urine cytology, cystoscopy to attempt to localize source of hematuria, CT urogram, and possible renal biopsy may be indicated. In addition, imaging modalities such as renal angiography, MRI and MR angiography can play an important role in diagnosis. Doppler ultrasound is the preferred initial diagnostic modality as it is a noninvasive test with no radiation exposure. With this imaging, the clinician can visualize an enlarged left renal vein diameter as well as the orientation of the SMA and the aorta.

Normal CT is inadequate for diagnosis with CT angiography typically the next imaging modality used. CT angiography can demonstrate delayed venous washout from the affected left kidney in a patient with nutcracker syndrome, as noted in our case.⁶ MR angiography can provide a radiation-free test that is also non-invasive that can be used in place of CT angiography. Multiphase CT urography is also helpful for diagnosis, and also has the capability to diagnose other possible causes of hematuria. Venography is not commonly performed despite the notion that it may be a useful diagnostic test.³

Treatment

There is a wide variety of treatment options based on individual patient symptoms and morbidity, ranging from observation to nephrectomy. In children under 18, observation is generally favored because nearly 75% of these patients will have complete resolution of hematuria.7 Interventions should be limited to patients who have very serious or persistent symptoms. Interventions include open surgical repair and endovascular stenting. Previously, open surgical repair had been the standard of care, but as technology has developed, endovascular intervention has become first line therapy. While more complex surgical interventions have been reported in the literature, endovascular interventions still have been proven to be a therapy with good long term clinical and imaging results.8

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