A Case of Posterior Nutcracker Syndrome Revealed in the Aerospace Environment

Connie Y. Chung; Mark E. Lytle; Roselyn W. Clemente Fuentes

BACKGROUND: Nutcracker syndrome is caused by a rare anatomic variant where the left renal vein is trapped between the aorta and the superior mesenteric artery. Posterior nutcracker syndrome is an even rarer entity, characterized by the retro-aortic positioning of the renal vein, causing compression between the aorta and spinal vertebrae. Symptoms include microscopic or frank hematuria, flank pain, varicocele, pelvic congestion syndrome, and abdominal pain. A search of the literature did not reveal prior cases of nutcracker syndrome that became symptomatic and diagnosed secondary to the unique stressors of high gravitational force (G force) in the aviation environment.

CASE REPORT: A 25-yr-old man training as an F-16 flight test engineer presented with left scrotal/testicular pain, varicocele, and intermittent gross hematuria. After an extensive workup, he was diagnosed with posterior nutcracker syndrome and underwent a left varicocele ligation with spermatic cord denervation. He was eventually able to be returned to flying duties with limitation to non-high performance aircraft.

DISCUSSION: This case is particularly unique as its diagnosis was dependent on exposure to high G force conditions that may have otherwise remained asymptomatic without this environmental stressor. Education on the diagnosis of nutcracker syndrome as a differential in the setting of hematuria and pain is an important lesson learned. This case also illustrates the necessity of considering the effects of the stressful environment of high G force on even overall healthy individuals. Fortunately, due to the collaboration of medical-surgical expertise and familiarity with the requirements for operational readiness, this patient was able to resume his aviation career, albeit in a different capacity compatible with his condition.

KEYWORDS: posterior nutcracker syndrome, renal vein entrapment, renal vein compression, hematuria, proteinuria, varicocele, G force.

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Nutcracker syndrome, first described in 1950, refers to the entrapment of the renal vein, typically the left, between the aorta and the superior mesenteric artery.[7] Posterior nutcracker syndrome is an even rarer variant, characterized by the retro-aortic positioning of the renal vein causing compression between the aorta and the spinal vertebrae. Incidence is not well defined, with rare mentions in case reports and 0.5–3.7% postmortem discovery in cadavers.[5] Patients range from pediatric to geriatric. However, the symptomatic patients are most often diagnosed in their 20s or 30s.[9] Symptoms include microscopic or frank hematuria (the most common), as well as flank pain, varicocele in men, pelvic congestion syndrome in women, and abdominal pain.[1]

Imaging used to make the diagnosis include renal ultrasound with or without Doppler flow, computed tomography, magnetic resonance imaging, phlebography measuring pressure gradients, venography, and computed tomography venography.[8] Management ranges from conservative with mainly observation—especially if the finding is incidental and asymptomatic—to surgical or endovascular management.[4] While rare, it is also thought to be underreported overall due to its often asymptomatic nature and incidental findings on imaging from retrospective review studies.[5] This case study explores how an otherwise undiscovered condition became symptomatic and was subsequently diagnosed under the unique stressors of the military aviation environment.

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CASE REPORT

A 25-yr-old man was training as a flight test engineer in the F-16 when he presented to the flight surgeon noting worsening left scrotal/testicular pain and varicocele. Symptoms initially started after flying higher G sorties (7–9 G_z), resolved between flights, and did not occur with lower G sorties (< 7 G_z). After several months, his symptoms became more consistent and present in between flights. The pain was described as a constant dull ache with intermittent exacerbations with activity and heavy lifting. Additionally, he started to develop associated intermittent gross hematuria without clots after flying higher G sorties (7–9 G_z). Once he developed hematuria and persistent pain, the patient sought out medical evaluation. Physical exam was significant for dilated veins palpable mainly on the left side of the scrotum with associated tenderness on palpation and a well-healed left subinguinal scar. Testicular exam was otherwise normal with no inguinal hernia noted. He had a history of left subinguinal varicocelectomy as an adolescent with a goal of fertility preservation. The patient did not have any other antecedent history of trauma, surgery, genitourinary infection, nephrolithiasis, genitourinary malignancy, chemical exposures, or smoking history.

Testicular ultrasound revealed a left grade III varicocele. He was started on ibuprofen 800 mg every 8 h as needed for pain and instructed to wear scrotal support. However, his left testicular pain gradually worsened and he was placed on tramadol 50 mg every 8 h for improved pain control. He was evaluated by urology, who performed a cystoscopy for evaluation of the hematuria, which was normal. CT urogram revealed two separate left renal veins, one passing along the normal anatomic course anterior to the aorta (**Fig. 1**) and a second running posterior to the aorta (**Fig. 2**).

Management options included a repeat varicocelectomy, a major vascular reconstruction, renal auto transplant, or endovascular stent. Due to the high-risk nature of these procedures, the patient was referred to vascular surgery, who noted that it appeared that the retro-aortic left renal vein had been compressed over time and was the etiology of the recurrent varicocele and symptoms. The aberrant renal vein had been subjected to aortic compression, leading to increased pressure in the renal and gonadal veins, leading to varicocele creation. Although this would ordinarily be a more indolent development, the addition of high G force maneuvers requiring anti-G straining maneuvers (AGSM), causing increased intra-abdominal pressure, led to a faster progression of the varicocele and symptoms.

Due to the complexity of his case, the patient was sent to a high-volume surgical center for secondary vascular surgery evaluation. While waiting for the follow-up appointment, his pain started to progressively worsen. The patient was seen by a pain management specialist who started him on Percocet 5/325 mg every 8 h as needed and gabapentin 300 mg every 8 h, which controlled the pain. At his second vascular surgery consult, the vascular surgeon recommended interventional radiology evaluation to further assess the venous anatomy and to determine the extent of compression of the retro-aortic renal vein. The venogram demonstrated two retro-aortic renal veins and one larger anterior renal vein. The left gonadal vein emptied into the large anterior aortic left renal vein and did not demonstrate reflux, significant pressure gradient, or presence of collateral vessels. However, the retro-aortic renal veins did demonstrate collateral vessel formation, with a 4-mm pressure gradient. With regard to these findings, the surgical specialist concluded that the retro-aortic renal vein was likely contributing to his intermittent hematuria, but was less likely the cause of his recurrent varicocele and pelvic/scrotal pain.

Based on these results and the course of his symptoms, it was suspected that exposure to high G force stress provoked the renal congestion. The scheduled abdominal diagnostic laparoscopy and possible renal vein vascular bypass was cancelled. The results and the fact that major vascular reconstruction would ultimately disqualify him from flight test engineer flying duties were discussed with the patient. If his pain improved and hematuria was managed without being provoked by high G force exposure, he might still be fit to fly low G airframes.

After shared decision-making, the patient underwent a left varicocele ligation with spermatic cord denervation, which led to significant improvement in his pain overall. He initially had

Fig. 1. The arrow points to the anterior left renal vein on the patient's CT-urogram.

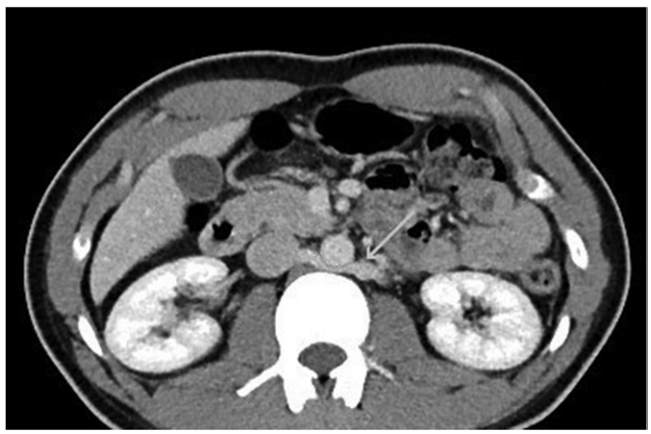

Fig. 2. The arrow points to one of two posterior left renal veins on the patient's CT-urogram.

some mild residual pain with activity that gradually improved over a few months, at which time he stopped pain medication use all together. Eventually, he was able to perform all activities without limitations or pain. Additionally, he denied any further episodes of gross hematuria and remained free of associated flank pain, nausea, vomiting, dysuria, urinary tract infections, or new voiding symptoms. He was cleared to participate in worldwide deployment from a urologic and vascular standpoint. The patient was submitted for a flying waiver to return to flight test engineer flying duties and he was approved to return to fly by the waiver authority with the caveat of a restriction to non-high performance aircraft and close annual follow-up with urology.

DISCUSSION

Posterior nutcracker syndrome is a rare condition with few documented case studies on literature review. This case is particularly unique as its diagnosis was suspected to be dependent on exposure to high G force conditions and may have otherwise remained asymptomatic without this environmental stressor. This patient underwent an extensive work-up, which is typical for this diagnosis.[1] Education on the diagnosis of nutcracker syndrome as a differential in the setting of hematuria and pain is an important lesson learned. Additionally, nutcracker syndrome is typically thought to be secondary to a congenital anomaly; however, acquired cases have been observed during pregnancy, after kidney transplant, and concurrent to aortic aneurysm.[1] Additionally, factors that increase intra-abdominal pressure such as prolonged straining and increased gravitational forces have been tied to an increased incidence of varicocele.[10]

A search of the literature did not reveal any similar cases of nutcracker syndrome that were possibly exacerbated and diagnosed secondary to stressors of the high G force environment. However, there were some studies that evaluated how the vascular system is affected by acceleration forces.[3] With acceleration, the pressure gradient in the hydrostatic column along the head to foot axis increases, in turn increasing blood pooling in the pelvis, lower extremities, and likely the testicular venous system. To counteract that, AGSM work to reduce the venous pooling below the heart and improve cerebral perfusion. A study done on subjects both with and without varicocele demonstrated marked increased venous pressure in the variceal veins with the Valsalva manuever.[6] The isometric contraction of the abdominal, buttock, and lower extremity muscles increases the intrathoracic pressure, also hypothetically increasing venous reflux and pooling to the testicular veins, similar to the mechanism of a Valsalva maneuver. A case report on scrotal hematoma precipitated by centrifuge training noted that varicocele rupture has been associated with increased intra-abdominal pressure, which is also a component during AGSM.[2] Increased physical strength training to maintain the physical endurance and conditioning to tolerate higher G_z may also be an aggravating factor.[3]

This case also illustrates the necessity of considering the effects of the stressful high G force environment on even overall healthy individuals. In this case, management decision-making was done with consideration for the military duty implications. Fortunately, due to the collaboration of medical-surgical expertise, familiarity with the requirements for operational readiness, and understanding of the physiological effects of the high G force environment, this patient was able to resume his aviation career, albeit in a different capacity compatible with his condition.

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