

Treatment-refractory vulvodynia from nutcracker syndrome: A case report

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ABSTRACT

Background: Pelvic venous disorders are often undiagnosed due to the symptom variability and similarity to other disease presentations. 'Pelvic congestion syndrome' is a term often used as a diagnosis of exclusion, since there is currently no standardized diagnostic approach for pelvic venous disorders, which further delays treatment.

Case: A 25-year-old woman with treatment-refractory vulvodynia presented with symptoms that included left-sided vaginal wall pain, pruritis, dysmenorrhea, dyspareunia, muscle tension, and a chronic vaginal ulceration. Abnormal pelvic varices were discovered, and she was referred to vascular surgery for treatment of nutcracker syndrome causing ovarian vein reflux and abnormal engorgement of pelvic varices.

Conclusion: Patients presenting with signs of pelvic venous insufficiency such as vaginal pruritis, irritation, pain, recurrent vaginitis, or chronic ulcerations should be examined for pelvic venous disorders.

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1. Introduction

Chronic pelvic pain (CPP) is described as noncyclic pain lasting over six months; it occurs more frequently in females than males [1], which may be due to under-recognition or misdiagnosis of the condition in men. The incidence of chronic pelvic pain in the world is estimated to be around 39% and accounts for 15% of gynecologic visits [2].

One etiology of CPP that is often a diagnosis of exclusion is pelvic congestion syndrome (PCS). Classically, PCS is described as a noncyclic, dull aching or heaviness worsened with standing, and it is associated with multiparity, back or flank pain, dyspareunia, and dysmenorrhea [1]. It can be caused by increased pelvic vein capacitance during pregnancy, venous anomalies, lack of valves or presence of incompetent valves in the left ovarian vein, or obstruction of venous flow from compression or thrombosis [1]. The incidence of PCS is difficult to state accurately because it is frequently underdiagnosed, and it is often misdiagnosed due to variability in presentation, lack of a standardized diagnostic approach, and variability of underlying venous etiology. However, in CPP patients the prevalence of diagnosed PCS is reported to be 10–30% when no additional pathologies are identified [3].

Historically, pelvic venous disorders have been grouped into the nonspecific diagnosis of PCS. We now understand that PCS is an

imprecise term that encompasses many pelvic venous disorders, which can generally be divided into two categories: venous reflux and venous obstruction. Nutcracker syndrome is a syndrome of venous obstruction that occurs when the left renal vein (LRV) is compressed between the superior mesenteric artery (SMA) and abdominal aorta as the left renal vein crosses the abdominal aorta to enter the inferior vena cava (IVC), which sits to the right of the aorta [4]. This compression causes high venous pressures and leads to compensation by decompression through the left ovarian vein, thus causing abnormal pelvic flow and varices.

This case report describes a unique presentation of nutcracker syndrome that was difficult to diagnose because of symptoms mimicking vaginitis, vulvodynia, and pudendal neuralgia. This presentation demonstrates the need to consider venous insufficiency in patients with unknown sources of recurrent vaginitis and a nontraditional constellation of pelvic pain symptoms.

2. Case report

A 25-year-old healthy, nulliparous Caucasian woman with a history of von Willebrand disease, recurrent vaginitis, and a left Bartholin gland cyst was referred to a pelvic pain specialist due to 18 months of treatment-refractory vulvodynia. The patient thought the pain was due to a vaginal infection, yet all cultures were negative. She also had increased pain severity at the five o'clock position along her vaginal wall, stating that it was worsened by stress, menses, intercourse, and laying

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on her left side. She had left hip and lower back pain along with awareness of pelvic floor muscle tension exacerbated by strenuous activities and sitting or standing for extended periods. Her pelvic pain workup was consistently negative, despite multiple infectious vaginitis tests for *Candida albicans* and *Gardnerella vaginalis*. Upon physical examination, the patient had bilateral tenderness of her pelvic floor muscles and pudendal canals, a tender and erythematous vaginal introitus at the four to seven o'clock position, and an ulceration of unknown etiology along the left vaginal vestibule.

Due to vague symptoms possibly suggesting pudendal neuralgia, vulvar vestibulitis, and vulvodynia, she was prescribed a variety of vaginal suppositories, topical creams, and oral pain medications, which all provided only minimal relief. She also participated in physical therapy, received perineal muscle Botox injections, and had peripheral pudendal nerve blocks, which temporarily relieved the pain. None of the treatments permanently eased her discomfort, and her symptoms progressed towards increased vaginal pain, pressure, and tight pelvic floor muscles. Additionally, the vaginal ulcer persisted, and she continued to complain of left vaginal wall discomfort.

After 14 months without significant improvement, she was sent for a magnetic resonance neurogram (MRN) to evaluate the pudendal nerves. The MRN showed a thickened right pudendal nerve distal to the pudendal canal, but the most significant finding was venous congestion and engorgement of abnormal varices in the deep pelvis around the uterus, cervix, and vaginal cuff (Fig. 1). Due to these findings, she was referred to the vascular surgery department for further workup with venography of the bilateral renal veins, ovarian veins, and internal iliac veins. Venography of the left ovarian vein showed reflux into the pelvis and dilation of 9.6 mm indicating venous insufficiency. It also identified an abnormal mass of dilated parauterine varices (Fig. 2). Intravascular ultrasound revealed 75% compression of the left renal vein by the superior mesenteric artery, also known as nutcracker syndrome (Fig. 3). These studies confirmed the initial MRN findings of venous engorgement and a final diagnosis of nutcracker syndrome.

The patient was first scheduled for a left renal vein (LRV) transposition, patch venoplasty, and endovenectomy via midline incision to treat her nutcracker syndrome. Through a midline laparotomy, the peritoneum was reflected to the right, exposing the abdominal aorta, IVC, and LRV. Large collateral venous branches surrounding the LRV were

ligated and the left renal vein was clamped after systemic heparinization. The LRV had significant scarring and intraluminal synechiae, so an endovenectomy was performed with patch venoplasty prior to re-anastomosis of the LRV two centimeters caudally on the IVC to relieve compression from the SMA. Kidney function remained stable with a blood urea nitrogen (BUN) of 5 mg/dL on post-operative day two and 6 mg/dL on post-operative day three, with a serum creatinine stable at .7 mg/dL. Prior to the surgery, all laboratory values had been within normal limits.

Three weeks following the LRV transposition, patch venoplasty, and endovenectomy, she returned for left ovarian vein embolization. Anatomic stenosis of the LRV at the IVC was identified, along with significant collaterals around the left kidney. After venoplasty of the stenosis, decreased collaterals were noted. The origin of her left ovarian vein could not be identified, and it was presumed to have been occluded after the first procedure, so coil embolization was not possible. At her two-week and four-week follow-up visits, the patient reported decreased pain and overall improvement. She was advised that if symptoms returned or worsened, embolization could be attempted again.

Four months later she returned with burning vaginal and vulvar pain that was similar to that before the initial surgery and she requested a further attempt at embolization. She underwent a translumbar puncture and direct embolization of the LOV along with sclerosis of pelvic varices and cross pelvic collaterals. This intervention was successful and resulted in no further opacification of pelvic veins. A pelvic ultrasound scan one month after the procedure showed a normal uterus and ovaries with normal doppler flow and without masses. Three months following this procedure the patient continued to have predominantly left-sided pelvic pain, but her left-sided vaginal wall pain had resolved. To help with the pelvic pain, she had restarted pelvic physical therapy and medical therapy. If these more conservative measures proved to be unsuccessful and the pain was to persist, the patient intended to undergo a left iliac vein embolization.

3. Discussion

This case demonstrates the variability in presentation of pelvic venous disorders and treatment options. The concern is that a general term such as 'pelvic congestion syndrome' is an imprecise diagnosis

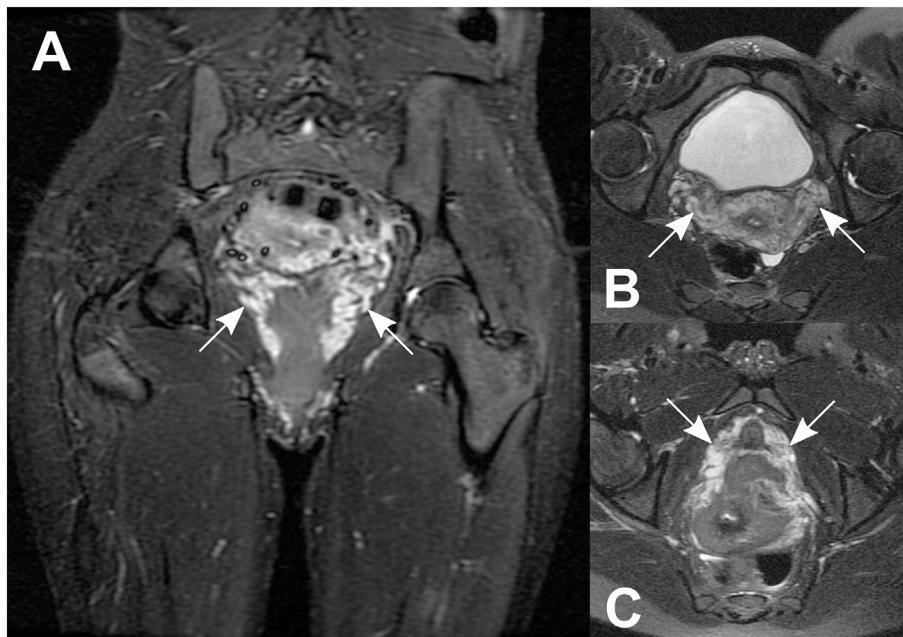


Fig. 1. T2-weighted MRN of the pelvis. A. Coronal section of the pelvis demonstrating bilateral venous engorgement surrounding the uterus, cervix, and vagina. B. Axial section of the pelvis with bilateral venous dilation around the uterine fundus. C. Axial section of the pelvis with bilateral venous congestion around the uterus and cervix.

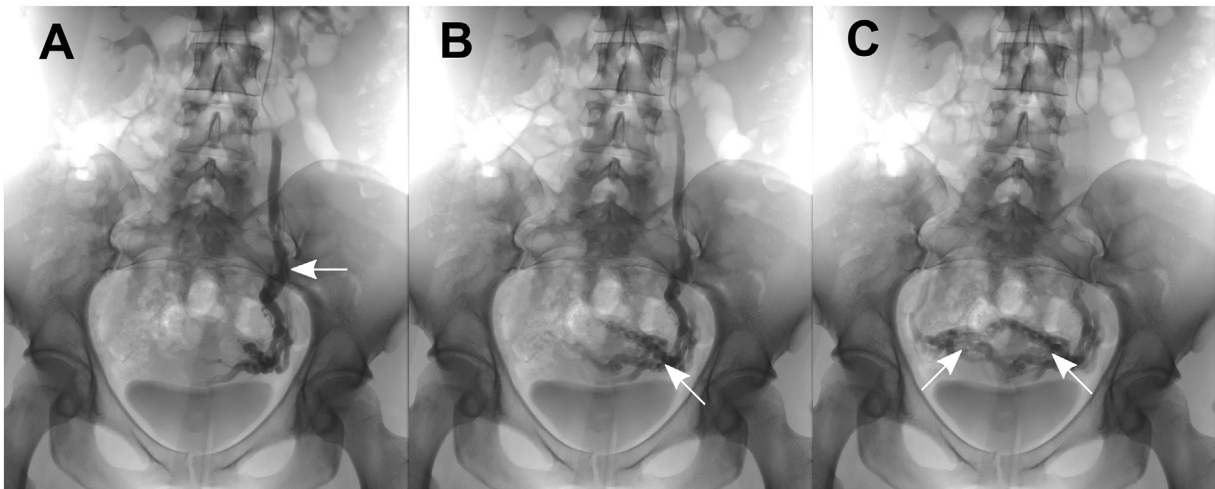


Fig. 2. Venography obtained from a catheter advanced through the right internal jugular vein to the inferior vena cava, left renal vein, and left ovarian vein where contrast was injected. A. Note the size of the left ovarian vein. B. Filling of the deep pelvic veins following contrast injected into the left ovarian vein. C. Significant bilateral congestion around the uterus.

for a true pelvic venous disorder, which can have multiple etiologies. Pelvic venous disorders can be separated into two categories: venous reflux or venous obstruction. Reflux-related pelvic venous disease is from reflux in the ovarian veins or internal iliac veins. Obstructive venous disorders encompass left renal vein compression, nutcracker syndrome, or iliac vein compression from the internal iliac artery, also known as May Thurner syndrome. Obstruction leads to an increased pressure gradient and ultimately causes reflux. Both reflux and obstructive etiologies can cause engorgement of the pelvic veins and abnormal pelvic varices.

This case is unique because our patient was young, previously healthy, and nulliparous. Many women present with a pelvic venous disorder following pregnancy, which is attributed to a 50% increase in pelvic vein capacity associated with valve incompetence and retrograde flow [5]. In a study of 151 asymptomatic patients, nulliparous women were found to have a 5% incidence of reflux compared to 44% in multiparous women [6]. Our patient's nulliparous status and young age makes her symptomatic presentation less common.

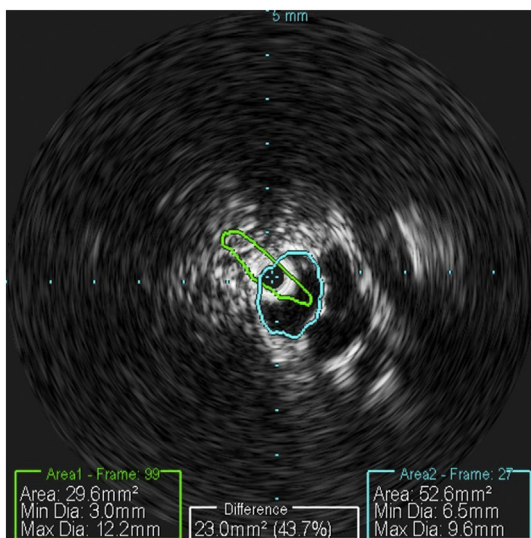


Fig. 3. Intravascular ultrasound of the left renal vein and opening of the left ovarian vein demonstrating diameter variability between the two. The left renal vein diameter (green, left) ranged from 12.2 mm to 3.0 mm due to compression from the superior mesenteric artery. The left ovarian vein diameter (blue, right) ranged from 6.5 mm to 9.6 mm. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

There is overlap in symptoms between vaginitis and chronic venous stasis, which complicated this case. Patients with venous stasis often present with aching, cramping, swelling, pruritis, and heaviness that can progress to ulceration, edema, or stasis dermatitis [7], and our patient had many of these symptoms. Our patient's most unusual symptoms include her remote history of recurrent infectious vaginitis, persistent sensation of vaginitis with irritation and pruritis, chronic vaginal ulcer of unknown etiology, and specific left-sided vaginal wall pain. She had persistent pelvic floor muscle tension despite extensive treatment, and the etiology of her pain remained uncertain. Our patient's presentation highlights the complex nature and presentation of pelvic pain, which is why diagnosing pelvic venous disorders continues to be difficult.

The diagnosis of nutcracker syndrome in our patient was made from an incidental finding on her MRN, which had been initially ordered to examine her presumed pudendal neuralgia. There is currently no standardized diagnostic workup for pelvic venous disease, likely due to the variability in presentation, but the gold-standard diagnostic test is catheter-based venogram and intravenous pressure measurement [8]. Transabdominal ultrasound is the initial study of choice, followed by venography, but CT and MRI studies may offer a more thorough analysis of pelvic anatomy [1]. To diagnose PCS initially, one study suggests implementing a systematic ultrasound method of examining the venous system transabdominally from the diaphragm to the perineum, after screening for other pathologies [9]. Ovarian vein diameter can also be used to diagnose pelvic vein incompetence, but there are conflicting recommendations in the literature, with ovarian vein diameter ranging from >8 mm by CT/MRI or 5 – >10 mm by venogram [10, 11]. While there are multiple diagnostic criteria that can be met, there is not an established or thoroughly researched guideline for how to methodically work up a suspicion of pelvic venous disorders, which enhances the complexity of diagnosis.

This case emphasizes the complexity of pelvic venous disease as well as variability in presentation, diagnosis, and treatment. Our patient's symptoms mimicked those of vaginitis, vulvodynia, and neuralgia, which confounded the underlying cause and delayed surgical treatment. If our patient's pain were to persist after the procedures and she were to continue to have pelvic floor tension and myalgia, the presumption would be that her chronic muscle tension was exacerbated by the stress caused by her surgical procedures. It is our hope that, now the underlying etiology of pelvic venous disease has been eliminated, treatments for her pelvic floor muscle tension will be effective in relieving her pain.

Contributors

Paulina C. Altshuler collected and analyzed patient data, completed a literature search, and composed the paper.

Brandon T. Garland allowed access to patient data and provided edits to the paper.

Michael E. Jorgensen provided edits to the paper and compiled the images.

Nel E. Gerig allowed access to patient data and provided edits to the paper.

Conflict of interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient consent

Written informed consent was obtained from the patient for usage of her medical history for this case report.

Institutional review board approval

The Rocky Vista University College of Osteopathic Medicine (RVUCOM) Institutional Review Board approved this project on June 28, 2017: HIRB 2017-0011.

Provenance and peer review

This case report was peer reviewed.

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