Pelvic Congestion Syndrome: Underdiagnosed, Misdiagnosed, and Undertreated…or a Misnomer?

A discussion of clinical findings related to pelvic venous disease, emphasizing the need for greater suspicion of disease, as well as minimally invasive percutaneous treatment techniques.

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Pelvic congestion syndrome (PCS) is a disease that is often undiagnosed in women with pelvic pain. Chronic pelvic pain affects approximately 15% of premenopausal women in the United States. The lack of diagnosis seems to be related to limited awareness of this disease process by both physicians and patients, coupled with incomplete data in the literature. This may explain why other common causes of pelvic pain and discomfort are widely known (ie, endometriosis, fibroids) and are therefore diagnosed and treated appropriately, whereas pelvic venous congestion—or rather, pelvic venous disease as it may be considered—is rarely included in the differential diagnosis. Chronic pain caused by venous disease is a significant burden on a patient's quality of life as well as health care utilization.[1,2] As a result, greater awareness of and suspicion for PCS/pelvic venous disease is essential.

CLINICAL PRESENTATION

PCS usually occurs in premenopausal, multiparous women. Chronic pelvic pain is diagnosed after pain has been present for at least 6 months. Patients with classic PCS from gonadal vein reflux often report lower pelvic pain, dyspareunia, postcoital pain, and/or bladder irritability or urgency. This pain is exacerbated by prolonged standing and can radiate to the posteromedial thigh or buttocks. Patients with pelvic venous disease caused by May-Thurner compression or other pelvic venous obstructive disease often present with left lower quadrant and/or groin pain and left lower extremity swelling. Not uncommonly, patients can have sacral pain and even sciatica. This pain is also worse at the end of the day or with standing. Labial varices and superficial venous insufficiency often coexist and can be related to either cause of pelvic venous disease. Given various well-known causes of chronic pelvic pain, PCS is a diagnosis of exclusion. Other entities such as endometriosis, uterine abnormalities (eg, leiomyomata, adenomyosis, arteriovenous malformations), pelvic inflammatory disease, and adhesions are usually considered first.[2,3]

It is time to start thinking about pelvic pain differently. With the advent of advanced ultrasound techniques, cross-sectional imaging, and careful questioning of patients, a diagnosis of pelvic venous disease can be moved higher or lower on the list of differential diagnoses and may in fact warrant first consideration in many patients.

Clinicians should also be aware of other associated manifestations of PCS that are not necessarily pelvic related, including generalized lethargy, depression, headaches, and nausea.[4] Although these symptoms are nonspecific, they should not be discounted, as they are a major cause of a decreased quality of life.

On physical examination, women may have varicosities involving the vulva, perineum, buttocks, or thighs. With May-Thurner syndrome, asymmetric left-sided swelling may be seen as well. The etiology of PCS is unclear; however, it has been reported to be related to a constellation of retrograde blood flow secondary to valve dysfunction and venous hypertension. However, Nutcracker syndrome or left renal vein compression between the superior mesenteric artery and the aorta is highly prevalent in these patients and may explain the elevated venous pressures and reflux in the majority of

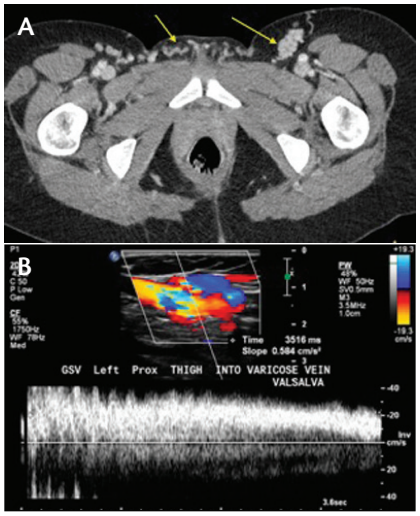

Figure 1. CT with contrast demonstrating multiple superficial varicosities in midline and left groin (arrows) (A). Color Doppler ultrasound of left great saphenous vein demonstrating insufficiency (B).

cases, as reflux occurs in the left gonadal vein 85% of the time.[5,6] The presence and significance of May-Thurner compression or pelvic venous obstructive disease is just beginning to enter the discussion of pelvic venous disease, and the relationship between pain and reflux in the gonadal vein versus elevated venous pressures into pelvic varices from iliac vein compression is not well understood.

Finally, it is not uncommon for patients to also have an element of lower extremity venous insufficiency (Figure 1). Treatment of lower extremity superficial venous insufficiency should be considered to treat leg symptoms and should be performed after treating pelvic venous disease. Treating lower extremity reflux will not improve pelvic pain and can, in fact, exacerbate symptoms by removing the decompression pathway and further elevating pelvic venous pressures.

IMAGING

In addition to clinical history, imaging plays an important role in the clinical pattern of pelvic venous disease. Ultrasonography is usually the first-line imaging modality when PCS is suspected and can demonstrate dilated gonadal veins (> 6–8 mm) and refluxing paraovarian varicosities. Ultrasonography can also identify other causes of pelvic pain such as uterine and ovarian pathologies.

CT venography and magnetic resonance venography are used to evaluate prominent ovarian veins and dilated pelvic varicosities and identify pelvic venous obstructive disease. Criteria for suspected PCS on cross-sectional imaging are four or more ipsilateral pelvic veins with at least one pelvic vein > 4 mm or an ovarian vein > 8 mm.[5]

Ultimately, catheter-directed retrograde venography of the gonadal veins and pelvic venography are the modalities of choice for diagnosing PCS and can be used in cases that are indeterminate. Patients are placed in a reverse Trendelenburg position to document ovarian and pelvic vein insufficiency and uterine vein engorgement, or simply, a left renal venogram is performed and reflux into the ovarian vein is noted. Typically, ovarian veins affected by PCS are > 10 mm on conventional venography. Intravascular ultrasound (IVUS) can be used to identify iliac vein obstruction (Figure 2).

MEDICAL MANAGEMENT

Medroxyprogesterone acetate or goserelin can be used to provide symptom relief of pelvic pain before any intervention is attempted. These medications suppress ovarian function and increase venous contraction. However, it has been postulated that resistance may develop, thus decreasing long-term efficacy.[4]

ENDOVASCULAR THERAPY

Endovascular therapy for PCS includes both mechanical and chemical therapies. Embolization can be performed with endovascular occlusive devices such as coils, vascular plugs, absorbable gelatin sponges, or detachable balloons. Other agents, such as glue, have also been used for embolization.[3]

Although venous access may be achieved from an internal jugular vein or common femoral vein, we would recommend a jugular vein approach because it

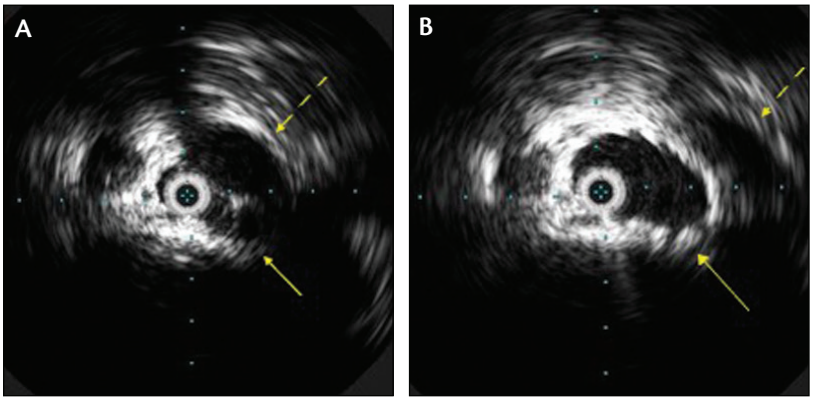

Figure 2. IVUS of the left common iliac vein demonstrating compression of the iliac vein (solid arrow) by the right common iliac artery (dashed arrow) (A). IVUS of the left common iliac vein after stent placement, showing improved iliac vein diameter (solid arrow). The right common iliac artery is shown for comparison (dashed arrow) (B).

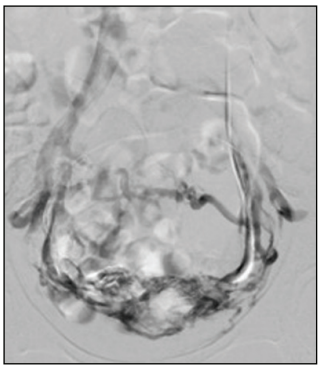

Figure 3. Catheter-directed retrograde venography of the left paraovarian vein demonstrating cross-filling collaterals.

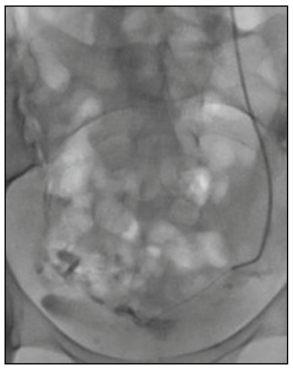

Figure 4. Catheter-directed sclerotherapy of paraovarian varicosities prior to coil embolization.

allows for evaluation of both gonadal reflux and pelvic venous obstruction. The left ovarian vein is usually the easiest to catheterize given its anatomy (draining into the left renal vein), whereas the right ovarian vein typically drains directly into the inferior vena cava just inferior to the right renal vein (Figure 3).

Sclerotherapy

Sclerosants are used as a chemical method for treating pelvic varicosities. Sclerotherapy is commonly used in conjunction with mechanical embolization. This method allows more distal treatment as well as direct sclerosis of the venous endothelium. Although no formal studies have been undertaken, experienced operators have noted that sclerotherapy may reduce symptom recurrence because sclerosants allow treatment of all branch vessels rather than only the main gonadal channel. This may also allow for the use of fewer coils and thus may reduce costs. Foam sclerotherapy, typically 1.5% to 3% sodium tetradecyl sulfate mixed with air (1:2 to 1:4) with or without a contrast agent, is most commonly used (Figure 4).[7]

Embolization

Coil embolization is the most common mechanical method of minimally invasive treatment for gonadal vein reflux (Figures 5 and 6). The number of coils varies by operator and institution. A recent study by Guirola et al demonstrated that there were no differences in treatment outcomes with the use of vascular plugs compared with coils; however, fewer plugs were used, leading to a lower associated radiation dose.[8] The use of foam can decrease the number of coils needed and lead to similar reductions in radiation dose.

It is not always clear whether unilateral and bilateral gonadal vein embolization is required to control symptoms. More commonly, unilateral left ovarian vein embolization is performed and is sometimes the only treatment required. Maleux et al found no statistical difference in symptom relief in patients who had unilateral or bilateral ovarian embolization—some level of improvement was seen in approximately 68% of treated patients, 58.5% of whom had total relief.[3] Given this fairly high rate of persistent or recurrent symptoms in patients treated with gonadal embolization alone, it is becoming clear that sclerotherapy and iliac venous stenting should be considered to resolve venous obstruction and decrease venous hypertension.

If patients develop recurrent symptoms, one may need to interrogate the internal iliac veins for cross-filling pelvic collaterals, which can be performed by using balloon occlusion. Collaterals from the internal iliac veins can be treated with similar methods as those used for other pelvic varicosities. Overall technical success of embolization is up to 99% in these cases, with fewer than 8% of patients experiencing recurrence. Rare complications are related to nontarget embolization of coils, vessel perforation, or thrombophlebitis.[4] There are no reports on the effect of embolotherapy on fertility.[5,6]

Figure 5. A single fluoroscopic image showing left and right ovarian vein coil embolization.

However, results in the literature may be misleading. The collaterals may be representative of decompression pathways related to May-Thurner syndrome, and perhaps stenting of the

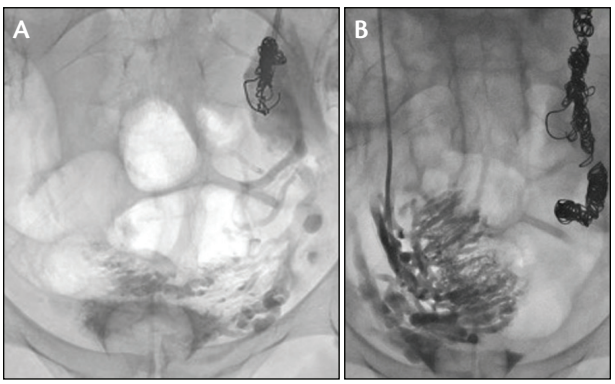

Figure 6. Catheter-directed sclerotherapy of bilateral paraovarian varicosities (A) with subsequent coil embolization (B).

iliac veins should be considered. This concept is new in recent years and not widely adopted. Studies evaluating the causal relationship of iliac vein obstruction versus gonadal vein reflux in pelvic pain are needed.

CONCLUSION

Pelvic venous disease (including PCS and May-Thurner syndrome) is prevalent in patients with chronic pelvic pain. Women who have persistent intermittent pain with no other underlying pathology should be considered for treatment, which consists of sclerotherapy, embolization, and iliac venous stenting. Interventionalists should be familiar with the symptoms, workup, and treatment of this disease, as thoughtful treatment can significantly improve the patient's quality of life. ■

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