Review

Pelvic congestion syndrome: a current review

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Abstract: Pelvic congestion syndrome (PCS) is an obscure diagnosis thought to cause chronic pelvic pain (CPP) in premenopausal women. It is believed to be caused by pelvic vein insufficiency but its etiology is still being investigated. Diagnostic criteria have yet to be determined making its prevalence unclear. Moreover, its clinical presentation mimics that of other common conditions causing chronic abdominal and pelvic pain[1,2]. In this review we will attempt to summarize the key elements of the clinical features, diagnostic work-up and treatment options of this enigmatic and complex entity.

Keywords: Pelvic pain, Congestion, Incompetence, Reflux, Embolization, Chronic

EPIDEMIOLOGY OF PCS

Having no definite diagnostic criteria, the incidence of PCS is difficult to determine. According to current literature, up to 10% of women have ovarian varices, and 60% of them may develop PCS[3-5]. PCS is to be considered in the differential diagnosis of CPP which affects approximately 4-16% of women, and up to 30% of them are thought to have PCS if no other obvious pathology can be found[6-9].

ETIOLOGY OF PCS

The exact etiology of PCS is unclear. Multiple investigations have observed that insufficiency of the pelvic veins originates mainly from the ovarian veins, but may also originate from the internal iliac veins or other communicating branches found in the pelvis[5,10,11].

The pathophysiology of congestion is believed to be multifactorial as a result of valvular insufficiency, vein obstruction or hormonal changes[12]. Valvular insufficiency can be caused by congenital absence of the ovarian veins' valves, which is relatively common, reported in 13-15% of patients[13]. Alternatively, it may be caused by valvular incompetence seen more frequently in multiparous women as a result of the 50% increase in pelvic vein capacity during pregnancy, which may in turn lead to venous incompetence and reflux in the non-pregnant state[10].

The absence of PCS in post-menopausal women suggests the role of estrogen in premenopausal women as a venous dilator, an observation supported by the symptom relief or resolution that is observed following the initiation of a hypo-estrogenic state in these women[14,15].

Extrinsic compression of the left renal vein ("Nutcracker syndrome") which receives inflow from the left ovarian vein or compression of the left common iliac vein can also account for pelvic vein congestion (May-Thurner Syndrome)[16,17]. Whatever may be the cause for reflux and congestion, the clinical symptoms are probably attributed to the abnormal dilatation of the pelvic venous system, as stretch and stasis of the engorged ovarian and pelvic veins may activate selective pain receptors and cause release of neurotransmitters from the walls of the dilated vessel[2,18-20].

A genetic basis has not been established but some studies suggest certain genetic traits to be involved in venous pathology[21-23].

CLINICAL PRESENTATION

PCS is characterized by symptoms of CPP, such as intermittent or constant abdominal or pelvic pain, not limited to any period of the menstrual cycle or intercourse and not associated with pregnancy. It typically affects multiparous women of reproductive age. The pain is usually described as a dull ache or fullness that persists for more than six months, and is often exacerbated with prolonged standing, coitus, menstruation and pregnancy. Symptoms are most severe at the end of the day and diminish with supine positioning. The presence of vulvovaginal, perineal or limb varices is suggestive[24,25].

The physical examination may reveal ovarian point tenderness, cervical motion tenderness and uterine tenderness with direct palpation. One study by Beard et al.[26] reported that the combination of these symptoms with post-coital pain are 94% sensitive and 77% specific for PCS.

After excluding other possible causes for CPP including gynecological, urinary, gastrointestinal, musculoskeletal, neurological and even mental health disorders, the key finding in PCS is documentation of pelvic vein dilation or incompetence on imaging studies. Congested pelvic veins can be very painful and account for approximately one third of cases of CPP[27]. Even so, the hallmark feature of enlarged pelvic veins can also be found in asymptomatic women[28,29], emphasizing the challenging diagnosis.

DIAGNOSTIC WORKUP

Although documentation of pelvic vein dilation is necessary for accurate diagnosis, pelvic imaging supports the diagnosis but does not define it, as pelvic congested veins are a common and non-specific finding that can bae seen in many asymptomatic women as well[29]. In addition, to date, there are no validated measures to determine venous congestion or tortuosity[30].

Ultrasound

Pelvic ultrasound is usually the first line of imaging study in patients with suspected PCS. It is easy of perform, non-invasive and relatively low cost. Sonography is also used to exclude other potential causes for pelvic pain such as pelvic masses or uterine pathologies. With regard to PCS, ultrasound can evaluate dilation of pelvic veins and morphology of other pelvic structures such as the uterus or the ovaries that might suggest the diagnosis of PCS. The transvaginal approach (TVS) with Doppler is generally preferred due to better visualization of pelvic venous system[31]. Common findings in sonography of PCS are dilated left ovarian vein or pelvic venous plexuses, stasis (slow blood flow) or reversed blood flow and variable duplex waveforms in the varicoceles during the Valsalva's maneuver (implying valve incompetence)[6,7,32-34]. It is recommended to perform the transabdominal or transvaginal sonography in semi-supine or upright position since veins are flaccid when the patient is supine, and venous changes may not be detected. Another maneuver that can improve sensitivity is the Valsalva maneuver, but even with these techniques employed, imaging is still operator dependent and may render false results[10].

Venography

Catheter-directed venography is the diagnostic gold standard test for pelvic congestion[35,36]. A percutaneous jugular, brachial or femoral catheter is used to visualize the ovarian or internal iliac veins and assess for incompetence, congestion and retrograde filling[37]. It is more sensitive than ultrasound scan and thus, the Society for Vascular Surgery (SVS) and the American Venous Forum (AVF) guidelines recommend it as the test of choice for pelvic venous disorders[38]. Its main disadvantage is being an invasive procedure that carries potential risks and complications, however, using catheter venography provides a therapeutic opportunity for intervention (embolization or sclerotherapy) if indicated.

CT (Computed tomography) and MR (Magnetic resonance) Imaging

CT and MRI scans provide better imaging of pelvic congestion but are more expensive and do not allow therapeutic intervention[4,6,28,39]. Moreover, CT scan requires radiation and should be avoided when possible, especially in premenopausal women[36]. Specificity is also considered low for both modalities; however, both scans provide a good anatomical overview of the pelvic vasculature and surrounding tissues and can identify coexisting pathologies.

Contrast-enhanced MRI may become a leading imaging study for pelvic venous incompetence due to its superior imaging and, in contrast to CT, does not involve radiation exposure. Velocity-encoded phase contrast imaging and time-resolved MR angiography have significantly improved detection of venous reflux[40-42].

Laparoscopy

Diagnostic laparoscopy is often preformed as part of a CPP investigation[43]. Although it is useful for detecting conditions such as endometriosis or adhesions which are otherwise undetected, its role in PCS detection is less established. Characteristic pelvic venous changes can sometimes be visualized at laparoscopy[44], but since the procedure is performed with the patient in the supine position and requires insufflation of carbon dioxide gas, the pelvic varicosities are often drained or compressed and ultimately remain undetected[27,45].

MANAGEMENT OF PCS

Several treatment modalities have been proposed and evaluated, including medical, invasive or surgical approaches. All modalities have shown effective relief of symptoms, but no standard approach to treat PCS exists, and it is not clear which could be considered the best option[46]. Therefore, therapy is individualized based on the clinical presentation and symptoms.

Medical therapy can be considered for first line treatment as risks are low compared with invasive procedures. However, limited data exists from only few small randomized trials. These trials reported that treatment with medroxyprogesterone acetate, GnRH agonist (goserelin) and subcutaneous etonogestrel implant improved pain scores and venography scores[8,15,47]. Presumably, the hypo-estrogenic state causes venous constriction that alleviates congestion and offers symptomatic relief. However, this symptomatic relief typically lasts for several weeks and benefits are rarely sustained. For example, women treated with medroxyprogesterone acetate reported rapid return of pain after treatment cessation[15]. Some systemic side-effects and need for regular follow up also reduces compliance of the medical therapy. Patients refractory or non-compliant to medical treatment should be considered for invasive treatment.

Surgical treatment such as ovarian vein or pelvic vasculature ligation (whether using laparoscopic or laparotomy approaches) have shown symptomatic relief of PCS in up to 75% of symptomatic women[30]. Nevertheless, this evidence is supported only by observational data and case series, therefore the value of these treatments has not been established in clinical practice. Hysterectomy and bilateral salpingo-oophorectomy have also been performed to treat this condition, but are limited only to women who have completed childbearing. This radical approach does not always provide relief of symptoms and is only indicated in cases where less invasive techniques are unavailable or have failed[38]. The surgical approaches are accompanied by the potential for several complications such as high rates of residual or recurrent pelvic pain, esthetic damage and longer hospitalizations[48-50]. A randomized controlled trial showed that embolization was superior to hysterectomy and oophorectomy in providing symptomatic relief for PCS[51]. Finally, the most important risk related to gonadal vein ligation and oophorectomy remains the post-procedural loss of gonadal function and the need for hormonal replacement[52].

Radiological treatments are now being used more often for PCS as they provide good technical success rates and a minimally invasive approach. **Percutaneous embolization procedures** can be performed in an outpatient or day hospital setting, thus reducing patient's discomfort and costs[53,54]. A trans-catheter embolization of the ovarian veins can be achieved through femoral, jugular, subclavian and brachial routes, all with good technical success and low complication rates[33]. Regardless of the embolization agent used (coils, foams, glue or liquid sclerosants), clinical and technical success rates remain high for all. Complications are rare but may be variable and most commonly include coil migration, vessel perforation and local thrombophlebitis[55-57]. These complications can be reduced by using a combination of embolization agents and techniques. Additional complications related to drug administration for sedation, venous puncture such as hematoma or pneumothorax and those related to radiation exposure are noteworthy[35,58,59]. Long term symptom relief with embolization therapy has been suggested in several studies since the 1990s[48,51,55,60-62] with 60-100% of patients reporting significant relief lasting up to 72 months[55]. No gynecological complications, such as menstrual cycle changes or changes in hormonal levels, were noticed[60,61].

Specific treatments for pelvic vein compression syndromes ("Nutcracker" or "May-Thurner" syndromes) are available with the use of percutaneous stent placement or in combination with ligation or embolization[63-65].

SUMMARY

PCS is a common condition that negatively impacts women's daily life, with significant physical, psychological and sexual consequences. It must be considered in the differential diagnosis of CPP and if overlooked it may cause delay in correct diagnosis and treatment. Although PCS is multifactorial and still poorly understood, treatment modalities exist with encouraging success rates. The clinical practice guidelines of the society for vascular surgery (SVS) and the American venous forum (AVF) conclude that ovarian and pelvic vein embolization has become the standard of treatment for this condition with a grade 2B recommendation[38]. Nevertheless, to date, limited evidence for long-term efficacy of these treatments is available and further studies are required and called for to optimize treatment for this complex condition.

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