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Pelvic Congestion syndrome - common, but underdiagnosed disease of women

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Abstract

Introduction: Pelvic congestion syndrome is defined as the presence of varicose changes in the ovarian veins and venous plexuses associated with chronic pelvic pain syndrome. The main problem with pelvic congestion syndrome is the low awareness of medical staff about this syndrome and the resulting very low recognition.

State of knowledge: Pelvic congestion syndrome is an important cause of chronic pelvic pain. The diagnosis is based on imaging examination with ultrasound as a first-line choice and correlation with clinical presentation. The aetiology is multifactorial and may result from a combination of genetic predisposition, anatomical abnormalities, hormonal, mechanical factors with venous wall weakness, reverse blood flow and hypertension. The incidence is strongly related to the number of pregnancies.

Conclusions: Pelvic congestion syndrome is one of the possible causes of chronic pelvic pain, thus differential diagnosis should necessarily include venous insufficiency during interviewing, clinical and ultrasound examination of patients. Among several treatment methods, intravascular embolization is a minimally invasive, effective and safe, currently recognised as a method of choice in treatment.

Key words: pelvic congestion syndrome; chronic pelvic pain; embolization; varicose veins; Interventional Radiology

Chronic pelvic pain syndrome - introduction

Chronic pelvic pain syndrome is one of the most common diseases in urology and gynaecology defined as a pain located in the pelvic area below the umbilicus, that lasts over six months and is severe enough to limit functioning, unrelated to menstrual cycle, pregnancy, local trauma or operations. It is a multifactorial disorder where pain may originate in any of urogynecological, gastrointestinal, musculoskeletal or nervous systems.

Besides the duration criterion, recognition of chronic pelvic pain syndrome requires an identification of incomplete relief by previous treatments, significantly impaired physical function, signs of depression (sleep disturbance, weight loss, loss of appetite), hypersensitive response to nociceptive stimuli and altered family roles. Management requires a holistic approach in order to provide patients with proper care [1, 2].

It has been reported that as many as 39% of women experience chronic pelvic pain at some time in their life. In this group, an estimated 30% have pelvic venous insufficiency. Ovarian vein dilatation is seen in 10% of women, up to 60% of whom may develop pelvic congestion syndrome (PCS). Pelvic varices occur as a result of ovarian or iliac venous incompetence and, due to anatomical reasons, are much more common on the left side. However, ovarian vein diameter may not correlate with ovarian venous reflux, with many normally sized ovarian veins exhibiting pathological reflux, and some large ovarian veins being competent. The true real prevalence is still unclear and large population based epidemiological studies are needed [3].

Pelvic congestion syndrome or pelvic venous reflux is one of the causes of chronic pain of the lower abdomen. PCS is said to occur as a result of retrograde flow in an incompetent ovarian vein. Overfilling (congestion) of the pelvic venous system has been implicated as a cause of dull chronic aching pain that usually is bilateral, worse at the end of the day after prolonged standing, premenstrually, and postcoitally. PCS has been reported to affect quality of life and personal relationships resulting in physical and psychological disorders, even leading to depression and anxiety [2, 4].

The better part of women with PCS are in premenopausal age (20-45 years) and typically multiparous - an incidence is strongly related with number of pregnancies. PCS is very rarely reported in postmenopausal women [5]. The aetiology of PCS is likely to be multifactorial, involving hormonal, genetic and mechanical factors. High risk for PCS concern patients with multiple pregnancies, anomalies in pelvic venous anatomy, history of pelvic pain in family, hormonal disorders with increased levels of oestrogens (polycystic ovary syndrome, oestrogen therapy), as well as varices of the lower limbs, phlebitis, prolapsed uterus, previous pelvic surgery, heavy lifting or prolonged standing. It may occur either alone or in combination with vulvar varicosities or lower extremity venous insufficiency. The fact that syndrome often affects young women with no predisposing risk factors suggests that inborn genetic features are involved in the underlying venous pathology, although genetic predisposition is uncertain [6]. Moreover, women with PCS are more likely to have a normal BMI with ovarian vein dilation compared with obese women [7].

Particularly, for the confirmation the diagnosis of pelvic congestion syndrome must be excluded other pathologies of pelvis such as: fibroids, adenomyosis, endometriosis, pelvic inflammatory disease, pelvic tumours, inflammatory bowel diseases and adhesions, pelvic arterial-venous malformations, portal hypertension, etc. [3]

Because of the variable manifestation and the limited appreciation of this condition by both clinicians and radiologists, PCS remains an under-diagnosed cause of chronic pelvic pain [8]. The purpose of review is to raise awareness among medical professionals about this condition.

Anatomy

In the case of PCS special attention is paid to the anatomy of the pelvic venous system. Blood from the uterus is drained through the interconnecting pelvic plexus. The plexus passes into two ovarian veins - the left one connects with the left renal vein and the right one connects directly to the inferior vena cava. The lower section of the plexus faces to the left and right internal iliac veins, which lead to the common iliac veins. Valves in the distal segment of ovarian veins are

present in most women, and presence of valves in iliac veins were found in approximately 10% of the population. The cause of PCS is associated with venous wall impairment and valvular insufficiency [9].

Diagnostic tools for pelvic congestion syndrome

Currently, the diagnosis of PCS remains challenging, given that there are not universally accepted criteria for enlarged pelvic venous vessels. In addition, many medical conditions have similar manifestations. In recent years, the Symptoms–Varices–Pathology classification has been published for pelvic venous disorders, encompassing three domains: (S) symptoms, (V) varices, and (P) pathophysiology. The (P) domain is composed of three subdomains, including the anatomy of the involved abdominal and pelvic veins, the associated hemodynamic abnormalities and the underlying aetiology. This instrument, once validated, could help to obtain homogeneous study groups with unified diagnostic criteria [10, 11].

Pelvic ultrasound is usually the first-line imaging modality in patients with suspected PCS. Ultrasound permits to exclude major pelvic masses, cystic changes in the ovaries, uterine pathologies and other potential causes of pain. Besides its repeatability, low cost and no need for special preparation, it has the advantage of allowing dynamic examination with provocative Valsalva manoeuvres or in standing position. Transvaginal ultrasound examination is preferred over transabdominal due to better visualisation of the pelvic venous plexus. The normal venous plexus appears as straight tubular structures with a diameter of 4 mm. Tortuous, dilated veins with >5mm (typically >6 mm) in diameter, slowed and reversed blood flow in the ovarian veins, dilated arcuate veins communicating with bilateral pelvic varicose veins or associated polycystic ovaries are the features that raise suspicion of varicosities and PCS [3, 6, 10].

Pelvic varices can be visualised efficiently with computed tomography (CT) and magnetic imaging resonance (MRI) as well. CT is characterised by the smaller diagnostic importance than other methods. Unlike ultrasound, it does not give information about hemodynamic changes in pelvic veins, but it has the capability to exclude other pelvic pathologies. In comparison to CT, magnetic resonance venography has the superiority because of the absence of radiation and therefore is a priority cross-sectional diagnostic method [3].

Position does influence the ability to detect pelvic venous pathology. Ultrasound evaluation in reports was performed in variable positions - the supine, reverse Trendelenburg position, semi-erect and upright positions. CT and MRI are obligatorily performed in the supine position. There is no consensus regarding positioning for noninvasive examinations, however clinicians should be aware of the role that position may have in the interpretation of all imaging studies [11].

Catheter-directed retrograde selective venography of ovarian and internal iliac veins is method of choice for the diagnosis of pelvic venous pathology. This is done to confirm the diagnosis, assess the venous anatomy, especially the collateral venous supply, allows observation of retrograde reflux and demonstrates the presence or absence of contralateral venous filling. Venography allows planning of embolization and coil selection. This minimally invasive diagnostic method can be performed under local anaesthesia. Venography can be done via cubital, jugular or femoral access, as well as through the direct injection of contrast to the uterine fundus with a needle inserted into the myometrium. It provides evaluating venous flow under fluoroscopy [3, 12]

Pathological findings of PCS can be identified on laparoscopies. Laparoscopy has the advantage of higher sensitivity than ultrasound and CT scans in the detection of pathologies that associate with PCS. In general, the common findings observed in women with pelvic pain via laparoscopy include pelvic inflammatory disease, ovarian cysts, pelvic adhesions, and endometriosis for which laparoscopy is the gold standard. However, it has been noted that due to the CO₂ insufflation used in the method, varices may collapse and remain overlooked. At all events authors suggest that it should be considered as a diagnostic method in some cases of chronic pelvic pain and suspicion of PCS [13].

Treatment

As the cause of PCS is believed to involve both hormonal and mechanical mechanisms, inducing

a hypoestrogenic state are primary goals of medication intervention. Clinical evidence regarding the efficacy of medication therapy of PCS remains limited, and available therapies have not been shown to produce long-term improvement.

Nonsteroidal anti-inflammatory drugs are an acceptable firstline treatment. They may offer some short-term relief while patients expect further investigations or a more permanent treatment. Nevertheless they are not a solution for the main cause of the complaint. Past investigators have demonstrated transient pain improvement with medroxyprogesterone with psychotherapy, gonadotropin-releasing hormone agonists, and the etonogestrel implant. Therapy with intravenous dihydroergotamine may be effective in decreasing the size of veins and alleviating pain symptoms, but treatment was accompanied by numerous side effects. Venoactive drugs, such as micronized purified flavonoid fraction has also been reported by multiple investigators to achieve significant reduction in pain within 2–4 weeks and improvement in venous outflow with relatively low rate of adverse effects [14, 3].

Ovarian vein ligation or resection, either retroperitoneal or laparoscopic has been performed for primary ovarian vein incompetence. Excellent results following extraperitoneal ovarian vein resection were reported in 57 patients with moderately severe symptoms, moreover some evidence suggests that bilateral ovarian vein resection may be more effective than unilateral approaches. Nevertheless surgery as an invasive method requires general anaesthesia and is associated with longer hospital stay, thus it is rarely used at present [15, 3].

Nowadays embolization of the incompetent veins and the periuterine venous plexus is used to obliterate refluxing varicose vein in minimal invasive way. Embolization for PCS was firstly performed in 1993 and has revolutionised the treatment of this condition [16]. Since then, many authors have used this procedure, with positive results therefore embolization became the method of choice for PCS treatment. In most patients it is performed via jugular or femoral access under fluoroscopy. An occlusion of varices is obtained with application of embolization coils, particles or glue. Both technique and the materials used for embolisation vary in publications and the choice of method belongs to interventionist [17, 18].

Due to its low rate of morbidity and complications it has largely replaced open surgical intervention for symptomatic ovarian vein incompetence. Another advantage of embolisation is a high effective rate in pain relief and possibility to repeat intervention in case of recurrence of symptoms. Ovarian vein embolization is generally performed under local anaesthesia and patients are usually discharged from the hospital at the same day [18, 19]. In case of a patient with severe kidney failure or allergic to iodine-containing contrast, a procedure can also be performed using CO₂ as an alternative contrast agent [20].

From the complications associated with embolization the most severe early complication is migration of embolisation material, usually to the pulmonary circulation. High diameter of varices, the use of glue and embolisation of left ovarian vein is associated with increased risk of migration. Retrospective evaluation of studies shows that during the last decade the incidence of this complication has been reduced from 4.2% to 1.6% as a result of increased experience and usage of detachable coils. Main late postprocedural side effect is the recurrence rate, which is approximately 5%. There is no clear evidence that reproductive function is affected. Moreover, no significant changes in sex hormone levels has been noticed [3, 18].

Summary

PCS is a common, but probably under-recognized, debilitating condition with a vascular aetiology that has protean manifestations in young, premenopausal, usually multiparous women. Low awareness of PCS among clinicians results in the frequent misdiagnosis of the condition, despite extensive imaging use. Once the syndrome has been accurately diagnosed, accurate treatment can improve symptoms in a high percentage of cases. The efficacy of noninvasive medical treatment is limited and does not show long-term improvement. Endovascular embolization remains a safe and effective procedure in relieving pain and improving quality of life and functioning, however thorough assessment requires large, randomised clinical trials.

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