May-Thurner syndrome presenting as pelvic congestion syndrome and vulvar varicosities in a nonpregnant adolescent

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ABSTRACT

May-Thurner syndrome (MTS) refers to compression of the left common iliac vein (CIV) by the common iliac artery. Although this is typically manifested as acute left lower extremity deep venous thrombosis, MTS is a rare cause of pelvic congestion syndrome. A 17-year-old girl presented with a 5-year history of worsening painful vulvar and labial varicosities. Venography demonstrated pelvic collateralization and left CIV stenosis consistent with MTS. The left CIV was stented with complete radiographic resolution. At 1-year follow-up, the patient is asymptomatic with a widely patent stent. This case highlights the role of endovascular therapy for patients presenting with MTS causing pelvic congestion syndrome. (J Vasc Surg Cases and Innovative Techniques 2019;5:252-4.)

Keywords: May-Thurner syndrome; Pelvic congestion; Intravascular ultrasound

May-Thurner syndrome (MTS) refers to a condition in which the left common iliac vein (CIV) is compressed by the overlying right common iliac artery. It is often manifested as left lower extremity swelling, pain, and venous insufficiency in a chronic setting, but patients may also present acutely with a deep venous thrombosis (DVT) with phlegmasia. There is a paucity of literature about MTS as a cause of pelvic congestion syndrome (PCS) and vulvar varicosities. We describe a case of PCS secondary to MTS.

PCS is characterized by pain as well as by the presence of pelvic and vulvar varicosities, and it is a frequently underdiagnosed cause of chronic pelvic pain.[1] PCS is typically seen in pregnant and multiparous women due to the "the strain of increased circulatory volume and flow and to compression by the gravid uterus," which causes pelvic venous insufficiency.[2] Other less common causes of PCS include primary valvular insufficiency and central outflow obstruction. PCS can be caused by MTS by retrograde flow through the internal iliac veins and cross-pelvic collaterals.[1,2] We present the case of a 17-year-old wheelchair-bound girl with a history of cerebral palsy who presented to our clinic with a 5-year history of worsening vulvar varicosities. A publication consent form was signed by the patient.

CASE REPORT

Our patient, a nulliparous 17-year-old girl with a medical history of prematurity (28 weeks) resulting in neonatal seizures, cerebral palsy, and developmental delay, was referred to the vascular surgery clinic for evaluation of PCS. She presented with painful, worsening vulvar varicosities with frequent bleeding episodes. She denied history of current or prior DVT, pulmonary embolus, or anticoagulation therapy. She was being treated by her gynecologist with medroxyprogesterone acetate (Depo-Provera) injections along with a prolonged course of supportive measures without success. On examination, she was wheelchair bound with lower extremities shorter in proportion to age and body size. Her gynecologic examination revealed a purple hue to the labia majora bilaterally, with significant venous engorgement and exquisite tenderness to light touch. Her left leg was also noted to have swelling with a larger caliber in comparison to the right leg.

Preoperative workup included bilateral lower extremity venous duplex ultrasound examination that showed no evidence of femoral vein and external iliac vein DVT; but given her body habitus, the central veins could not be visualized. Computed tomography venography demonstrated no abnormalities in the gonadal venous vasculature, a compressed left iliac vein with large pelvic venous collaterals, and engorged veins in the labia (Fig 1). Given this, the decision was made to proceed to the operating room for venography and intervention. Despite a history of prematurity, no abnormalities in the patient's inferior vena cava (IVC) were noted on imaging.

Percutaneous left femoral vein access was obtained under ultrasound guidance, and iliocaval venography was performed. Initial images revealed substantial pelvic collateralization with crossover to the right iliac system and delayed emptying of contrast material into the IVC; late images of the same digital subtraction angiography run identified the collateralized varicosity that was feeding the vulvar varicosities (Fig 2). These findings were consistent with high-grade outflow stenosis or occlusion of the left CIV. Intravascular ultrasound confirmed synechiae and webbing in the stenotic CIV, diagnosing MTS.

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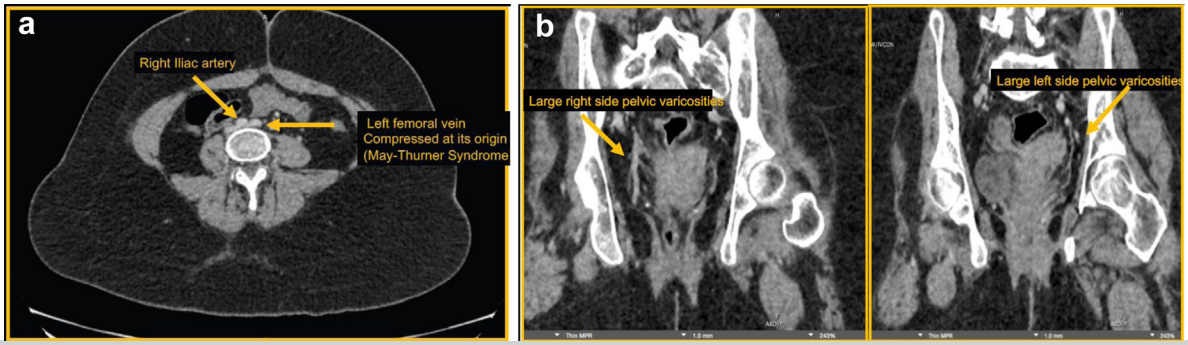

Fig 1. Computed tomography venography demonstrating flattening/compression of left common iliac vein (CIV) origin by an over-riding right common iliac artery **(a)** and bilateral large pelvic varicosities **(b)**.

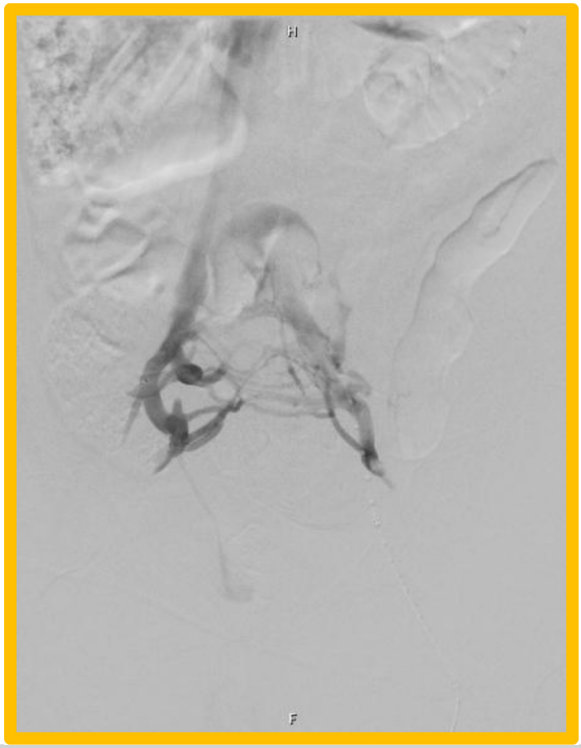

Fig 2. Intraoperative venography with digital subtraction angiography run demonstrating pelvic varicosities with reflux into the engorged veins of the labia.

An 18-mm Wallstent (Boston Scientific, Marlborough, Mass) was deployed in the left CIV, extending into the IVC distally. Completion venography demonstrated a widely patent iliac venous system with brisk flow of contrast material into the IVC and no filling of pelvic venous collaterals. There was no opacification of the previously noted vulvar varicosities (Fig 3). At the time of our intervention, the right venous system was also interrogated. Venography and IVUS demonstrated a normal right iliac vein without any evidence of pelvic collateral filling. Postoperative physical examination showed immediate decompression of her labial varicosities. She was discharged on warfarin for 6 months, followed by lifetime aspirin. The patient has remained asymptomatic at 1-, 3-, 6-, and 12-month follow-up.

Fig 3. Completion venography after left common iliac vein (CIV) stent placement with complete resolution of pelvic collaterals.

DISCUSSION

PCS is characterized by pelvic pain, pelvic varicosities, and atypical lower extremity varicosities. The primary cause of PCS is pelvic venous insufficiency, which refers to incompetence of the ovarian vein, internal iliac vein, or both,[2] resulting in venous hypertension in the deep pelvis, leading to the development of varicosities. Various theories have been implicated in the etiology of pelvic venous insufficiency. These include hormonal factors; venous obstruction, such as MTS or nutcracker syndrome (left renal vein occlusion); and extravenous factors, such as pelvic tumors, post-traumatic lesions, and endometriosis.[3] Whereas PCS can occur in nulliparous women, it is most commonly diagnosed in multiparous or pregnant women, with 18% to 22% of pregnant women developing vulvar varicosities.[4]

Treatment of PCS ranges from over-the-counter analgesics to surgical intervention. In terms of medical management, medroxyprogesterone acetate hormone therapy has been shown to be effective in reducing pain (50% reduction of pain from baseline in 73% vs 33% [n = 104]; $P < .0001$).[5] Pelvic vein ligation has been performed for PCS, with some evidence of pain reduction, but no large-scale randomized controlled trials have been performed to determine efficacy.[6] Hysterectomy and oophorectomy are also commonly performed procedures. Patients with secondary PCS due to MTS and nutcracker syndrome

who present with symptoms of PCS can be treated with endovascular techniques including stenting.

Patients with MTS can present with acute DVT, and the role of endovascular stent therapy for these patients is well described in the literature.[7-10] For patients presenting without DVT, including those who have varicose veins recalcitrant to therapy and those with symptoms of PCS, however, there are limited data regarding the role of stenting. A retrospective study by Ahmed et al[11] in 2016 investigated the link between PCS and MTS; within their cohort of eight patients with PCS and MTS, 87.5% of patients who underwent stenting experienced improvement in their symptoms.

Our patient was a 17-year-old nulliparous girl and thus was not a typical PCS patient. Whereas most pelvic venous insufficiency is caused by primary venous insufficiency stemming from the strain of pregnancy, our patient's underlying cause was structural in nature and arose from left iliac vein compression consistent with MTS. Given her radiologic imaging findings and symptomatic relief on iliac vein stenting, it appears that her vulvar varicosities were secondary to iliac venous outflow obstruction due to MTS.

CONCLUSIONS

PCS is often caused by primary venous insufficiency, but in rare cases, structural abnormalities such as MTS caused by left iliac vein compression can result in the development of pelvic pain and varicosities. In these patients, it is imperative to treat the underlying anatomic abnormality to alleviate symptoms. This case highlights the importance of endovascular therapy as an essential component to treatment of PCS in this group of patients.

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