Radiology Case Reports 16 (2021) 3534–3539

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Case report

May-Thurner syndrome with inferior mesenteric vein drainage and porta system - Case report ☆

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ARTICLE INFO

Article history:
Received 10 August 2021
Revised 17 August 2021
Accepted 19 August 2021

Keywords:
Case report
May-Thurner
extremity deep vein thrombosis
Iliac vein
Venous stent
Inferior mesenteric vein drainage

ABSTRACT

We present the case of a 12-year-old girl with a history of vascular anomalies in the lower pelvic limbs and back, who developed unilateral deep vein thrombosis of the left lower limb after her pubertal development, she was diagnosed with May-Thurner syndrome with an abnormal venous drainage of the pelvic structures through the superior hemorrhoidal veins to the inferior mesenteric vein towards the porta system, this being a chronic manifestation. This kind of behavior has not been documented in the reviewed medical literature. Secondarily, balloon angioplasty was performed without breaking the stenotic ring. As a second attempt, it was decided to place the venous stent, with satisfactory resolution of the symptoms. There are controversies about the indications for the use of anticoagulants and antiplatelet agents, or the indications to place a venous stent in children. We must consider an approach to for effective therapeutic treatment in these cases is to control bleeding, the main goal being trying to avoid ulcerations in the lower limb due to venous insufficiency with irreversible affectation of the valvular system.

May-Thurner syndrome (MTS) is a clinical situation that leads to compression of the left common iliac vein as it passes between the right common iliac artery and the axial skeleton, commonly at the level of the L5 vertebral body or the sacral promontory [1]. This partial obstruction is induced through two mechanisms: by its anatomical orientation, that causes physical entrapment, and by hypertrophy of the intimal layer secondary to chronic compression [2].

May and Thurner described constant changes in the venous intimal layer in 22% of 430 cadavers examined, and characterized the alteration of venous flow as an increase in local vascular resistance produced by changes in the wall, explained by the transmission of pulsatile force of the ad-

☆ Competing interests: The authors of this case report declare that they have no conflict of interest.

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https://doi.org/10.1016/j.radcr.2021.08.045

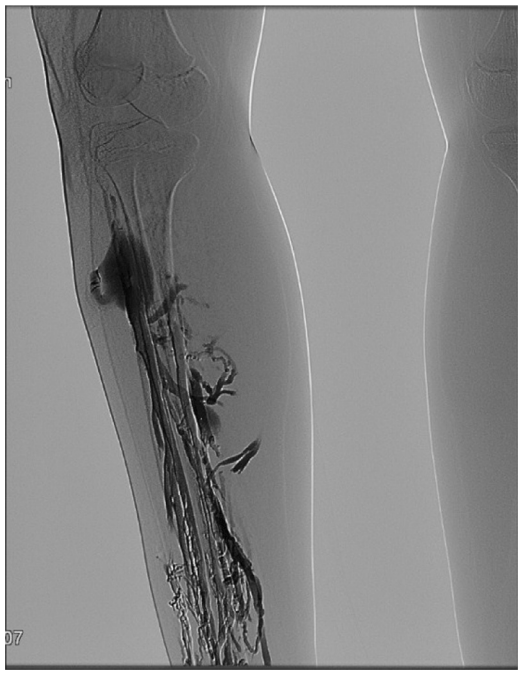

Fig. 1 – Phlebography of the right leg, with venous ectasia, malformed secondary to vascular anomaly. Some valves of the deep venous system are insufficient and there is bulging in these.

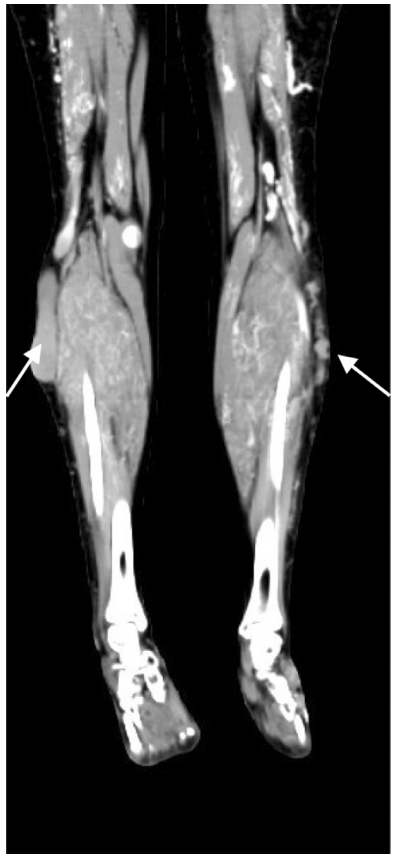

Fig. 2 – Contrast tomography of the lower members in coronal cut, where common vascular venous anomalies are observed in both legs, with muscular affectation. (Arrows).

jacent artery that causes intermittent friction between the vein walls, which successively stimulates the endothelium towards proliferation and partial obstruction of the lumen [2].

The typical but rare manifestation is deep vein thrombosis (2%-3% of patients with May-Thurner will develop it) [3]; most will have manifestations such as chronic venous insufficiency, characterized by pain, edema, varicose veins or venous ulcers. The degree of severity of the clinical manifestations depends on the efficiency of the collateral circulation of the pelvis to bring the venous return from the lower extremity to the inferior vena cava. However, atypical onsets have been described, from spontaneous retroperitoneal hematoma associated with ruptured iliac vein, lipodermatosclerosis, vascular accidents in patients with persistent foramen ovale, other congestive pelvic syndromes, and chronic pain [4].

12-year-old female patient, Hispanic-American, with an O+ hemotype, weighing 60 kg, height 162 cm, BMI 23 kg/m^2, elementary school student, with a history of exanthematous disease (Varicella Zoster) at 7 years old. Surgical and traumatic history and transfusion denied, she has a family history of diabetes mellitus and hypertension, with an apparently-healthy older brother.

From birth, common venous malformations were observed (according to the classification of the International Society Study Vascular Anomalies ISSVA 2018), with affection of both pelvic limbs and back, treated with sclerotherapy at the age of 2-year-old, on a monthly basis (lauromacrogol 3% with dilution at 0.75%). She started her menarche at age 10 (Fig. 1.)

She presented 2-month symptoms of evolution, characterized by asthenia, adynamia, pallor of integuments and lipothymia, accompanied by transvaginal bleeding and rectorrhagia, leading to a decrease in hemoglobin of up to 8.3 g/dL (anemia grade II, WHO Classification), later presenting an increase in the diameter of the left lower extremity associated with pain, edema, heat, and walking limitation, which yielded to the consumption of NSAIDs, sitting, elevation of the limb, and the use of compression socks.

On physical examination, small bluish-looking warty lesions (which peel and bleed regularly) were observed on both knees, fingers, and lateral sides of the legs. (Fig. 2.) There are also small telangiectatic lesions that appear after menarche on genitalia and anal region. This corresponds to the venular vascular anomaly and involvement of the superficial venous system, conditioning increasing dilations that are difficult to treat with sclerosis.

In the targeted exploration for consultation, the lower left limb, at the level of the infracondylar region, was found with an increase in its diameter with associated pain, edema and swelling of the limb. The patient also showed limitation in the arches of movement, being unable to maintain footing position.

Due to the clinical suspicion and the age, in addition to her antecedent of pre-existing venous malformation, CT angiography and phlebography (Figs. 3 and 4) were performed. In the usual protocol, in the search for this syndrome, the first step

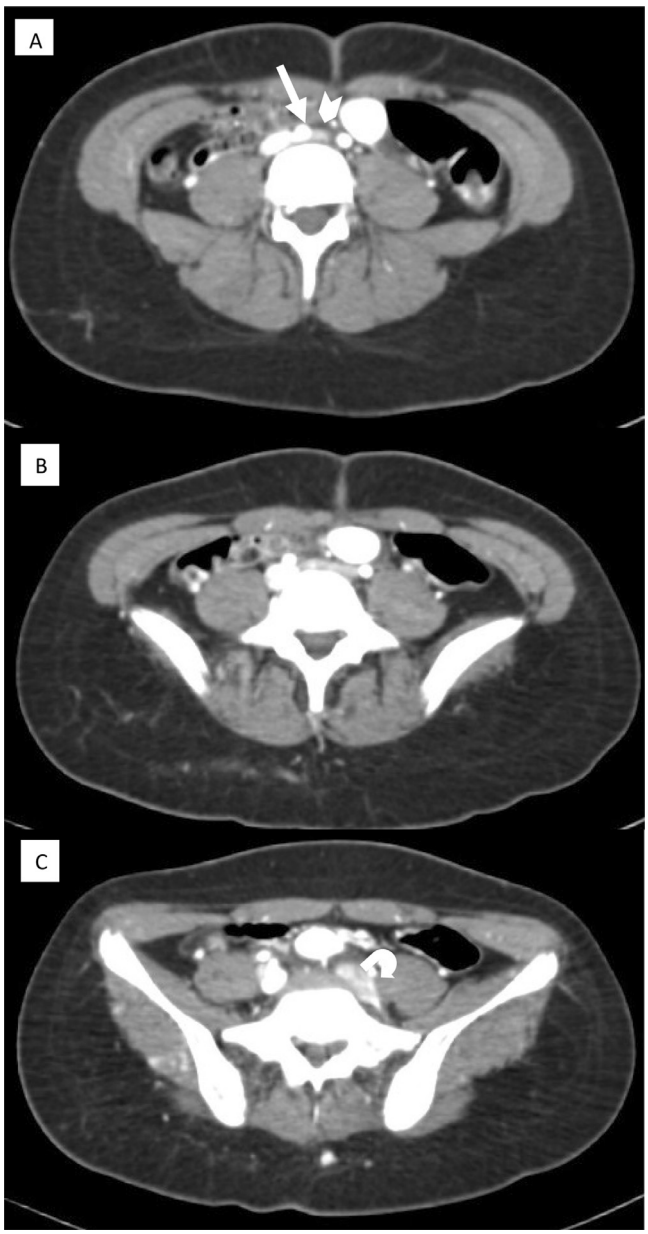

Fig. 3 – Sequences of axial tomographies of the abdomen in contrast phase where the compression by the common right iliac artery (Thick Arrow) on the left iliac vein is observed (Arrowhead). In figure C, the filling defect is observed regarding partial venous thrombosis. (Curved Arrow).

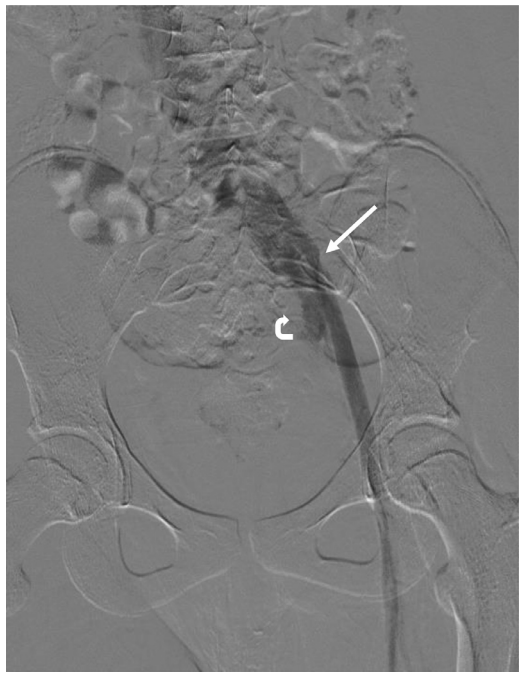

Fig. 4 – Phlebography with digital subtraction in AP. The left external iliac vein is observed, with reduced flow and turbulence at the expense of thrombus attached to the medial wall (*Thick Arrow*), as well as extrinsic compression of the common left iliac vein and venue reflux of the left internal iliac vein. (*Curved Arrow*).

to perform is an ultrasound, at the time of the initial consultation; however, our ultrasonography equipment was damaged),

CT showed: extrinsic compression of the left common iliac vein by the right common iliac artery in probable relation to TMS, which conditions venous congestion of the left limb, and reflux to the internal ilian vein with pelvic congestive venous vessels (Fig. 3.) The drainage predominantly of the superior rectal veins ascending through the inferior mesenteric vein confluence with the splenic vein which was overloaded, generating splenomegaly, at this point without evidence of liver involvement (Fig. 5).

After these findings, 18 mm x 40 mm balloon angioplasty was performed, observing a decrease in stenosis after the procedure without breaking the ring due to the large diameter (Fig. 6). There was redistribution of blood flow to the inferior vena cava, an increase in its diameter and a slight leak towards the internal iliac vein. The patient presented partial clinical improvement, with decreased abnormal uterine bleeding and cessation of rectorrhagia. It was decided to place a 24 mm x 70 mm x 75 mm "wallstent.uni" stent with its distal end in the proximal region of the inferior vena cava at the level of the lower platform of the vertebral body of L4, the proximal end exceeding the site of stenosis at the level of the external iliac vein (Fig. 7). Progress was satisfactory, stopping the metrorrhagia and without data of complications secondary to the procedure. A decrease in the caliber of the affected pelvic and hemorrhoidal veins was also observed.

Two months after the placement of the stent, the uterine bleeding presented a new increase in volume, requiring admission to the gynecology service, where it was classified as dysfunctional uterine bleeding, grade I anemia (Hb of 10 g/dL). Patient was evaluated by the pediatric hematology service, which found a family history of dysfunctional uterine bleeding, including a hypothesis that the bleeding may be due to a deficiency of Von Willebrand Factor-type genetics, which was discarded since the VW factor had a concentration of 74.6% (Deficiency <40%).

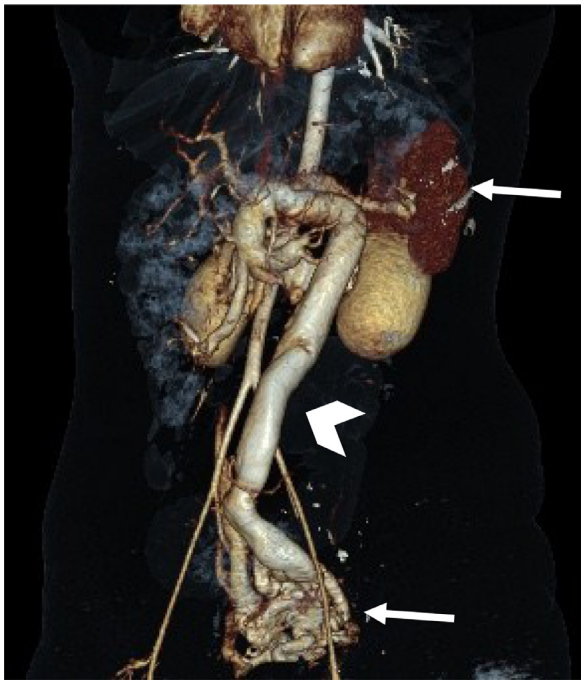

Fig. 5 – Reconstruction in 3 dimensions (3D) where dilation of the aneurysmatic aspect of the inferior mesenteric vein is observed (Arrowhead), associated with pelvic venous congestion and splenomegaly. (Arrows).

With suspension of Rivaroxaban, management for menstrual pain was given with mefenamic acid, plus a combination of ferrous fumarate 350 mg (equivalent to 115 mg of elemental iron), folic acid 1 mg, Vitamin B12 25 mcg, Vitamin C 600 mg, Vitamin E (D-succinate alpha-tocopherol) 25.83 mg, as well as the use of progestogens of the Desogestrel type, no criteria were presented for transfusion of blood components. The patient was discharged one week after admission due to bleeding improvement, and she did not have new episodes in outpatient follow-up. The patient underwent a basic genetic study with a 46XX Karyotype, 14 pstk +, 15 pstk + plus nonspecific numerical chromosomal alterations.

Discussion

The case that we presented was a diagnostic and therapeutic challenge, because May-Thurner syndrome has a low incidence, and does not usually have a presentation so early in childhood. The anthropomorphic characteristics of the patient and the expectation of growth conditioned the size of the stent selected to overcome iliac compression. The use of the endovascular management resolved the compression and the thrombotic syndrome, but the persistence of uterine hemorrhages limited anticoagulant management. Although gynecological symptoms have improved, the patient has not been anticoagulated again. Antiplatelet management is being used without unfavorable reactions.

The patient is still under follow-up by our department, with management for the venous-type vascular anomalies,

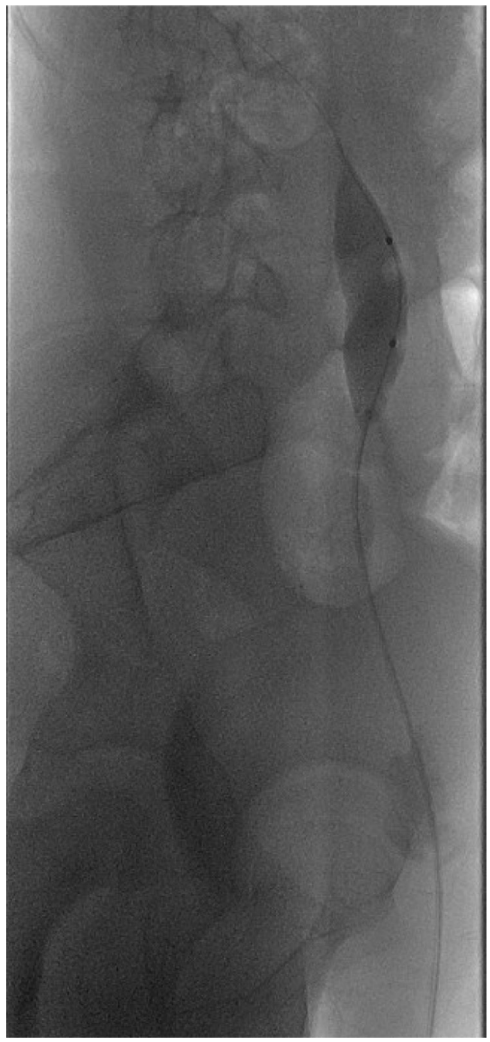

Fig. 6 – Phlebography corresponding to plasty performed on stenosis presented by the common left iliac vein, with partial reduction of stenosis. It is molded and could not break the fibrous ring.

without presentation of new thrombotic events. With a close follow-up, we have evaluated the compromise presented by a pelvic venous drainage due to the inferior mesenteric vein with significant ectasia, which at the time of the CT angiography study did not present data of portal hypertension, but which could result in liver involvement and later in a cardiopulmonary compromise. Just as the syndrome derived from symptoms of chronic venous insufficiency, or from post-thrombotic syndrome, which have not been manifested, were not ruled out, these remain latent because the patient does not have adequate adherence to antithrombotic measures such as the use of compression stockings, and her weight and body mass index have been increasing. An evaluation by the cardiology department is still pending, as well as a more specific genetic evaluation that has not been carried out due to financial limitations of the patient.

In the individual case series review carried out by V. Hansrani and R. Goldman, which included 32 patients (20 female patients and 12 male), only two associated manifestations of

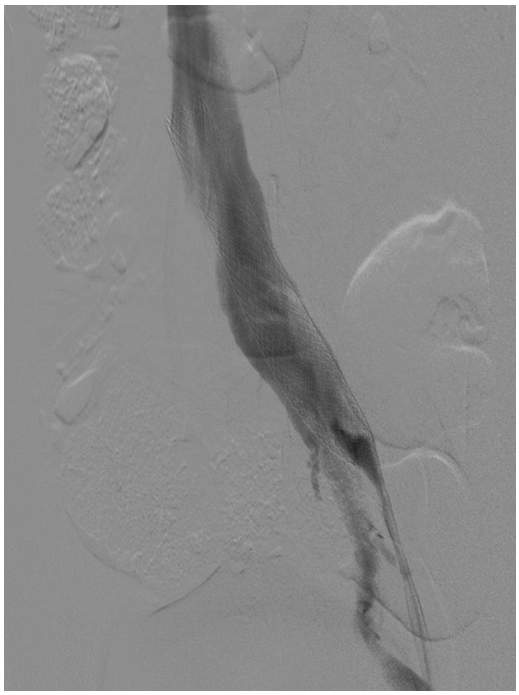

Fig. 7 – Phlebography corresponding to the release of the venous stent on the left iliac vein.

the superficial venous system of variceal type, both in women of the age of 17 years. In these same reviews, some patients had anticoagulation schedules for life, others from 10 days to 9 months, and some were not anticoagulated, without clear results [5,6].

Menometrorrhagia and rectal bleeding could be due to the compression of the external iliac vein, conditioning drainage of the venous flow towards the pelvic-lumbar system, the hemorrhoidal system and the inferior mesenteric vein, and as a consequence of the increase in pressure in these systems. These manifestations were observed.

A review of both surgical and radiological literature reveals that patients with symptomatic compression of the iliac vein tend to be young women with a mean age of 18-30 years, who present with persistent edema of the left pelvic limb and other findings related to post-thrombotic syndrome, either due to chronic venous flow obstruction, compression due to iliac vein stenosis or subsequent to iliofemoral thrombosis [7].

MTS is diagnosed in 2%-5% of all patients evaluated for chronic venous insufficiency of the lower extremity. In addition to the classic presentation, several variations have been described in the literature, such as right-sided disease and compression by other anatomical structures, including the bladder and kidney [8]. The main objective of MTS treatment is to resolve the extrinsic mechanical compression of the iliac vein. Initially, the non-invasive treatment was based on thrombolytic therapy and anticoagulation; however, this does not resolve or prevent hyperplasia of the vessel wall and does not eliminate the existing membranes and synechiae [1,9]. Berguer *et al.* were the first to describe treatment with catheter-guided thrombectomy and subsequent stenting in these patients. It is known that when compression of the left common iliac vein is >70%, the risk of recurrence and post-thrombotic syndrome is very high [9].

Grunwuald and Cols. describe that there are prospective and retrospective studies that suggest that endovascular treatment prevails, since, unlike isolated anticoagulation, it resolves mechanical compression when implanting a stent and achieves clinical dissolution of thrombi, if present, when administering local thrombolytic agents. In this way, a good long-term prognosis is ensured with low complication rates, thus leaving surgical interventions as alternative treatments for when endovascular therapy fails [10,11,12]. Endovascular treatment was chosen for the comprehensive management of our patient, requiring the placement of a stent, which significantly improved her symptoms and also the compression of the iliac vein.

Endovascular therapy for the treatment of MTS in adolescent patients was the most widely used, safe, and effective therapy for relieving venous obstruction. This is the same management that was performed on the patient, but with a 60-day anticoagulation scheme due to the aforementioned complications, in relation to the fact that patients who had thrombophilic disorders presented worse results. In our case, this could not be determined due to the economic limitation for a more specific genetic study.

TMS is a progressive disease with the risk of triggering severe long-term complications, being described in the literature: DVT/PE, ischemic cerebrovascular disease in the context of paradoxical embolism and post-thrombotic syndrome, for which its early diagnosis and treatment is imperative [2,13].

Conclusions

In the treatment of May-Thurner syndrome, there is little experience and there is a deficiency in the documentation regarding the pediatric population. It is important to determine its late manifestations and complications, in order to establish a therapeutic course, including the management of long-term anticoagulation and/or antiplatelet aggregation, deciding whether to use a venous stent and its close monitoring. It should be noted that these patients could also have deep vein involvement with damage to the valvular system that will lead to venous insufficiency that, in a period of time, will present ulcerations in the limb if it is not managed properly. This case shows that May-Thurner syndrome is an abnormal anatomical variant that goes unnoticed even into adulthood, but its severity will affect the clinical manifestations and hemodynamic compromise, with a decrease in life expectancy and quality of life. We know that in the case of our patient, improving the venous outflow will help us in the management of the vascular anomaly for the closure of the selectively failing system.

The diagnostic challenge of pediatric or adolescent patients with this entity persists because of how silent it remains, until severe clinical manifestations occur.

Patient consent

The informed consent of the patient was not included, since her identity and personal data were not exposed,

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