Endovenous thermal ablation of superficial truncal veins and compression therapy result in symptom relief in venous valve aplasia

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ABSTRACT

Venous valve aplasia (or valvular rarefication) is a rare cause of chronic venous insufficiency. In the present report, we have described the case of a 33-year-old man with severe symmetric lower leg edema and heaviness and pain in both lower legs. Duplex ultrasound demonstrated severe venous insufficiency in the superficial and deep venous system of both legs. Further imaging examinations supported the diagnosis of venous valvular aplasia. Treatment consisted of endovenous thermal ablation of the great saphenous vein and small saphenous vein as well as consistent compression therapy, resulting in a marked reduction of his leg edema, heaviness, and pain. (J Vasc Surg Cases Innov Tech 2023;9:101113.)

Keywords: Endovascular thermal ablation; Venous insufficiency; Venous valve aplasia

Venous valvular aplasia is defined as the congenital absence or decrease in the size and number of venous valves. Because proper valve function is crucial for maintaining unidirectional blood flow against gravity, affected patients will experience severe venous insufficiency with pronounced reflux in multiple veins of the lower limbs (ie, legs).[1] Epidemiologic data regarding this rare condition have remained scarce. To date, the literature has included merely a few case reports and case series.[2-4]

In the present report, we have described a case of venous valvular aplasia. The patient provided written informed consent for the report of his case details and imaging studies.

CASE REPORT

A 33-year-old man had presented to the phlebologic outpatient clinic with severe symmetric leg edema and heaviness and pain in both lower legs. Additionally, hyperpigmentation in the malleolar area was observed. However, no major varicose vein structures were visible (Fig 1, *A*). The onset of these symptoms and signs had occurred during the patient's childhood, and he reported that the complaints had been increasing during recent years. He had never experienced deep vein thrombosis and had not yet undergone venous surgery or intervention. He had no preexisting medical conditions and was not taking any long-term medications. The venous clinical severity score was 9 points for each leg. Duplex ultrasound examination revealed pronounced reflux of all superficial truncal veins (great saphenous vein [GSV] and small saphenous vein [SSV]) and the popliteal veins (VPOPs) and femoral veins (VFs) of both legs (Fig 1, *B*). For the left leg, the reflux duration was 1400 ms in the GSV, 700 ms in the SSV, and 1000 ms in the VF and VPOP. For the right leg, the reflux duration was 1100 ms in the GSV, 800 ms in the SSV, 1400 ms in the VF, and 1000 ms in the VPOP. No evidence was found of previous deep vein thrombosis. Additionally, no evidence of venous valves was observed. The deep veins appeared remarkably uniform; however, they would normally exhibit an irregular shape, indicating the valvular sinus dilatations. Therefore, venous valve aplasia was included in the differential diagnosis. In summary, the CEAP (clinical, etiologic, anatomic, pathophysiologic) classification was C2, C3, C4a, Ep, As, Ad, and Pr for the GSV above and below the knee, SSV, VF, and VPOP for both legs. Because the duplex ultrasound findings had revealed reflux in both legs in both the superficial and the deep veins without any signs or medical history of current or previous deep vein thrombosis, further imaging studies were performed to investigate the origin of this pronounced venous reflux. Magnetic resonance phlebography, descending venography, and intravascular ultrasound did not show evidence of a vascular malformation or proximal flow obstruction and excluded post-thrombotic vein changes (Fig 2). Descending venography confirmed the presence of reflux of the superficial and deep venous system in both legs. Furthermore, no venous valves could be detected using these imaging modalities. Photoplethysmography and invasive phlebodynamometry showed an increase in the venous volume and intravenous pressure during 10 tiptoe exercises, demonstrating considerable ambulatory

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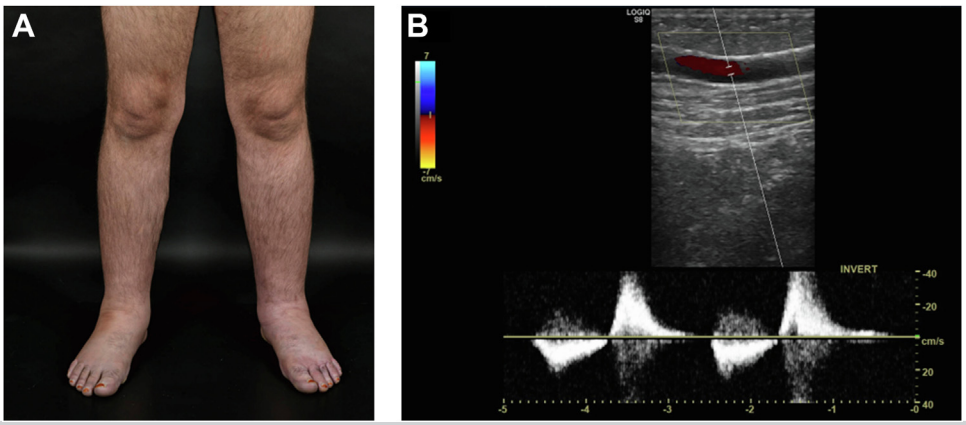

Fig 1. Clinical and duplex ultrasound findings for a patient with venous valve aplasia, including bilateral edema of the lower limbs (**A**) and pronounced reflux in the left great saphenous vein (GSV) after a calf squeeze (**B**).

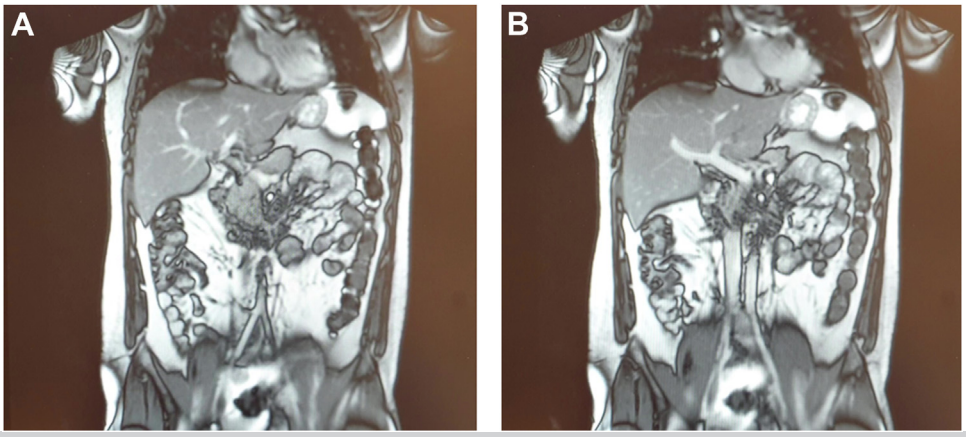

Fig 2. A,B, Magnetic resonance phlebography did not show any vascular malformations or proximal flow obstruction.

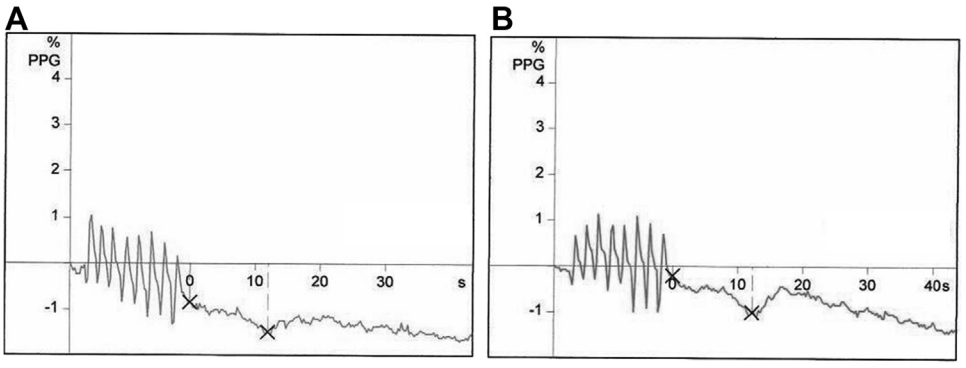

Fig 3. Photoplethysmography (*PPG*) of the left (**A**) and right (**B**) legs showing an increase in venous volume and intravenous pressure during 10 tiptoe exercises, demonstrating considerable ambulatory venous hypertension.

venous hypertension and resulting in failure of calf muscle pump function in both legs (Fig 3). In summary, these findings supported the diagnosis of venous valvular aplasia.

After a literature review and interdisciplinary consultations, we offered the patient endovenous thermal ablation of the GSV and SSV of the left lower limb as a first stage. The rationale was that reflux elimination in the superficial veins might positively affect the venous return in the subfascial system. However, given the lack of a broad scientific evidence for such treatment of venous valvular aplasia, we opted to treat only one leg at first.

The patient agreed to the intervention, and endothermal laser ablation of the GSV and SSV was performed in one session. Ultrasound-guided puncture of the left GSV was performed at the medial proximal lower leg. The laser probe was inserted in the vein and advanced until 1 cm distal to the femoropopliteal junction. After introduction of saline solution alongside the vessel, the catheter was gradually retracted, applying a total dose of 80 J/cm to the treated distance of 40 cm. The left SSV did not feed into the VPOP but exhibited a proximal extension. The laser probe was, therefore, advanced to the distal upper leg until shortly before the subfascial progression of this femoropopliteal vein. After introduction of the saline solution, the laser was gradually retracted, applying a total dose of 60 J/cm to a treated distance of 21 cm. In addition, consistent use of compression stockings was strongly recommended at the first visit.

Postintervention duplex ultrasound revealed successful ablation of the left GSV and SSV. However, reflux in the VF (1300 ms) and VPOP (1100 ms) was still present. In the follow-up visits to 1 year after the intervention, the patient reported an alleviation of the pain, heaviness, and swelling in both legs. Subjectively, the improvement was greater in the left leg. The venous clinical severity score had decreased to 8 points in the left leg and remained at 9 points in the right leg. However, this minimal decrease was partly attributed to his compliance with the use of compression stockings, which accounted for 3 points.

DISCUSSION

Venous valve aplasia (or valvular rarefication) is a rare cause of chronic venous insufficiency, classified as primary etiology (Ep) in the CEAP classification.[5,6] The pathogenesis has not yet been elucidated. To date, the molecular regulation of embryologic venous valve evolvement and maintenance after formation is barely understood.[7-9] Venous valve development implies a complex organization of endothelial cells requiring reorientation perpendicular to the blood flow direction.[7] Several genes, including ephrin type B receptor 4, transcription factor forkhead box C2, and the gap junction protein, gap junction gamma 2, have been suspected to play a role in the development of valve aplasia, and it has been assumed that the disorder is inherited in an autosomal-dominant manner.[7-9]

Patients with venous valve aplasia will develop severe venous hypertension at a young age and, consequently, will experience pronounced leg edema, pain, and heaviness.[2-4] The symptoms will usually not be apparent at birth but will manifest during adolescence.[3,8]

The condition will often initially be misdiagnosed as post-thrombotic syndrome.[1] The primary diagnostic workup should include duplex ultrasound, which will typically demonstrate severe venous reflux in the superficial and deep venous system of both legs. If venous valve aplasia or rarefication is suspected, additional digital imaging or phlebography should be performed to exclude vascular malformations and proximal venous outflow obstruction. Measurement of the ambulatory foot vein pressure will provide evidence of severe ambulatory hypertension.[2] The suspected diagnosis can be confirmed by performing descending phlebography and/or intravenous ultrasound.[8] In the absence of venous valves, the deep veins will appear remarkably uniform. However, normal veins will exhibit an irregular shape indicating the presence of valvular sinus dilatations.[8] In contrast, when post-thrombotic valve destruction has occurred, the valves will appear scarred and shortened, with various amounts of intraluminal septae found.[10]

To date, no clinical studies have been reported on the treatment of patients with valvular aplasia, given the low prevalence of this disorder. Our literature research revealed only a few case reports and case series, with a total of 27 patients with congenital absence of the venous valves.[2-4]

In general, the management of venous valve aplasia consists of various treatment modalities individually adjusted to the patient's needs. Consistent application of compression therapy with bandages or compression stockings is of paramount importance to prevent the development of leg ulcers and reduce the severity of the symptoms.[1] Incompetent superficial veins can be treated with endovenous ablation or surgically. It has been shown that ablation of incompetent superficial veins can also lead to a significant decrease of reflux in the deep veins, an improvement in muscle pump function, and a decrease in venous hypertension.[1,2,11,12] Additionally, ablation can increase the effect of compression therapy, which mainly acts on the deep venous system.[13] Reconstructive valve surgery has been promoted by various investigators as the treatment of choice in the case of severe deep venous insufficiency.[3,14-19] However, evidence is sparse and no conclusions regarding the effectiveness of this intervention can be made owing to insufficient data.[20]

CONCLUSIONS

Venous valve aplasia should be considered in the diagnosis of young patients with severe symmetric combined (superficial and deep) venous insufficiency. The use of compression stockings represents the central element of conservative treatment. Additional ablation of superficial reflux can result in greater alleviation of the symptoms. However, further evidence is needed to guide the therapeutic measures for this type of chronic venous insufficiency.

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