Lateral Marginal Vein: Have We Understood its Significance?

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Abstract

The lateral marginal vein is one form of truncular venous malformation seen commonly in Klippel–Trenaunay syndrome. It is characterized by avulvulosis and presents with features of chronic venous insufficiency. As there can be associated intravascular thrombosis, there is an increased risk of venous thromboembolism and pulmonary embolism. In children, it can cause vascular bone syndrome. Surgical excision is indicated when possible to prevent complications.

Keywords: Klippel–Trenaunay syndrome, lateral marginal vein, vascular malformation

INTRODUCTION

The lateral marginal vein (LMV), has recently drawn the attention of vascular surgeons in India, thanks to the understanding of the anomaly brought by doyens of “vascular malformations (VM)” like Prof. Byung-Boong Lee. He states in his lectures that the “Marginal vein (MV) is not another varicose vein, but a VM, besides being one of the most important components of Klippel–Trenaunay syndrome [KTS].”[1-4]

Understanding the embryology and treatment options will enable the surgeon to explain the importance of having the LMV treated.

EMBRYOLOGY

The MV is a fetal remnant vein that has failed to involute or regress in a normal manner, hence persists subcutaneously. This is a type of truncular VM, which, when present in the lower limb is termed – “marginal/lateral (LMV)/sciatic embryonic vein.” As a persistent fetal/embryonic vein, the LMV is always “valveless.” It is this nature of the vein that contributes to severe reflux, stasis, venous hypertension, and pulmonary embolism (PE). The intravascular thrombosis risk is high because of the lack of media/smooth muscle in the wall of the LMV. Recent evidence shows that LMV when combined with coagulopathy (as seen in extratruncular VM) can cause life-threatening PE.[1,2,5-7]

Although the LMV is considered to be superficial, it can be termed a misnomer as it frequently penetrates the deep fascia and involves the deep muscles of the leg.[1,8]

INVESTIGATION AND INTERPRETATION

MVs can be detected by ultrasound. Of importance during the scan is to note the structure of the deep venous system and the hemodynamics within, as this will determine the therapeutic options that would be offered.

If the deep veins are not patent or hypoplastic, the LMV maybe the only drainage for the lower limb and hence treating the LMV can be detrimental to the limb. Cases of venous gangrene following treatment of LMV have been reported in such patients.[1,2,9]

Based on the duplex findings, the MV can be classified into five types described by Weber [Figure 1].[8,10]

A computed tomography or magnetic resonance imaging may be needed to get a clear outline of the deep venous system. A direct puncture venogram would show the LMV and not the deep veins leading one to think that there is aplasia of the deep veins. Hence, a traditional venography should be combined with a venous duplex to avoid this error. Whole-body blood pool/transarterial lung perfusion/radionuclide lymphoscintigraphy have also been used in the evaluation of KTS.[1,7,9,11]

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D-dimer to assess hypercoagulability helps decide the need for anticoagulation, especially in the presence of coexisting VMs. The dosage of anticoagulants is as per existing guidelines.\[^{9}\]

**Treatment Options**

Several treatment modalities have been tried – foam sclerotherapy, coil embolization, endoablation, and surgical excision (segmental/staged excision).\[^{1,2,6,7}\]

In the pediatric cohort with normal or mild hypoplasia of deep veins, it is best that the LMV is surgically excised to prevent vascular bone syndrome. This will prevent/reduce the limb length discrepancy, and if this has set in, compensation can occur as the child grows. If there is aplasia of the deep veins, the LMV should not be removed. This surgical principle is applied in treating adults too.\[^{8}\]

**Surgical Excision Technique**

Surgical excision is to be done with a tourniquet and Esmarch bandage application to reduce the blood loss, which can be significant due to the fragile nature of the vein from a lack of media, presence of multiple abnormally sized perforators, and presence of dysplastic lymphatic tissue around the vein. The dissection should be gentle and meticulous. Stripping of the vein is not an option for all the reasons already mentioned. A semi-closed technique is the safest suggested [Figure 2]. Surgical excision may need to be staged in patients with hypoplastic deep vein system to let the deep veins get used to the increased flow. Open surgery is associated with no recurrence [Figures 3 and 4].\[^{2,3,6}\]

All patients with abnormal D-dimer must receive venous thromboembolism (VTE) prophylaxis before excision.

Foam sclerotherapy and endoablation are associated with a risk of fatal PE and skin ulceration due to the subcutaneous nature of the vein. These procedures are also large quantities of foam and high energy for thermal ablation due to the sheer size of the MV leading to higher morbidity.

Coil embolization to occlude the vein requires a large number of coils and has been known to lead to PE due to the coils dislodging from a valveless vein.

**Prognosis and Follow-up**

We have 88 patients on follow-up with KTS, most of whom are managed with custom-made compression garments. Semi-closed excision of the LMV has been done in three cases with no recurrence and clinically significant reduction in edema. These patients, however, do need to continue manual decongestive therapy on a long-term basis for the lymphatic component along with compression therapy.

**Summary**

- The LMV is not an ordinary varicose vein but is a truncular venous malformation

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**Conflicts of interest**

There are no conflicts of interest.
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