Editorial

Venous malformation: treatment needs a bird’s-eye view

A better understanding of congenital vascular malformations (CVMs) has evolved throughout the last two decades and is still evolving to address and correct many earlier misconceptions about this group of unique vascular lesions. Old terminology of these lesions based on old data has slowly fallen out of favour over the years and has given way to new terminology and classification of CVMs over the last two decades. There are still a few lingering issues regarding these vascular lesions that continue to be a source of confusion to clinicians involved in the diagnosis and treatment of CVMs.

One area of confusion is the classification of an extratruncular venous malformation (VM) as a capillary or cavernous haemangioma. There is indeed a genuine 'haemangioma' with entirely different aetiology, biology and clinical characteristics.

Haemangiomas and VMs are fundamentally distinct, not only in their anatomical, histological and pathophysiological findings, but also in their clinical courses. A haemangioma is not a congenital vascular lesion, but rather a vascular tumour that is derived from endothelial cells and develops in the postnatal period. In contrast, the VM is an embryological tissue remnant and is one type of CVM.

The haemangioma has a unique characteristic of 'self-limited' growth along with a distinctive growth cycle characterized by a proliferation phase of early rapid growth, followed by an involutorial phase of slow and gradual regression. The extratruncular VM, however, has an embryological characteristic of 'self-perpetuating' growth that is a result of a birth defect that arises during the 'early' stage of embryogenesis.

Indeed, 'extratruncular' VM lesions retain their mesenchymal cell characteristics and potential for growth. These lesions are able to proliferate when challenged by a growth stimulus, both internal stimuli (e.g. female hormones, menarche and pregnancy) and external stimuli (e.g. trauma, injury, surgery, etc.).

Therefore, the VM will not involute and regress like a haemangioma, but will persist through the life of the patient with the potential for a dreadful 'recurrence' which is the major clinical issue associated with an ill-planned surgical resection in particular. Indeed, over the last century we clinicians and our patients have paid a high price for neglecting this critical embryological aspect of the VM. Now, the idea Eivazi B et al. have proposed in their interesting manuscript 'Phleboliths from venous malformations of the head and neck' might fall into this same situation, where the biology of the VM was ignored. Certainly it sounds very appealing that direct surgical removal of the phleboliths from within the VM lesion would relieve symptoms and that anticoagulation treatment of the VM would prevent both recurrent phlebolith formation and phlebolith progression.

The Achilles heel of this approach is the fact that the authors neglected the nature of the VM from a macro-view. Phlebolith formation is a natural outcome of intravascular coagulation induced by a 'slow' blood flow through the infiltrating, extratruncular VM lesions. Such slow, sluggish blood flow is due to the natural anatomy and peculiar structure of the VM, such that the phlebolith is a hallmark of the VM, and will remain so as long as the VM continues to exist.

Simple removal of the phlebolith alone is, therefore, a mere stopgap measurement at best and is associated with very limited success in providing even temporary relief of the symptoms. Indeed, the VM lesion will continue to produce multiple phleboliths as its natural response. It is futile to attempt to remove one stone after another with such a high risk of bleeding which is often uncontrollable. Therefore, unless the VM lesion itself is eradicated, the phlebolith will continue to persist as the source of pain with no doubt.

In addition, in the absolute majority of cases, the coagulopathy associated with the VM is not the 'primary' cause of the phlebolith formation. Phleboliths are the result of a 'consumptive coagulopathy' following the intravascular coagulation and stone formation due to sluggish flow and trapped blood within the VM lesions. Therefore, it is not as effective as those with primary coagulopathy, and also definitely not logical while allowing its primary cause to persist intact. Hence, systemic anticoagulation to treat the intravascular coagulopathy to prevent phlebolith formation and its progression within the VM is not always practical.

Furthermore, there is always the possibility of the presence of another type of CVM lesion co-existing
with the VM (e.g. lymphatic malformation, LM), making the condition much more complicated. This is especially true for patients in the paediatric age group.

VM combined with another type of CVM is now classified as a haemolympathic malformation (HLM) (e.g. Klippel–Trenaunay and Parkes–Weber syndromes). The VM in this situation should be treated from a macro-view standpoint as part of a multicomponent vascular malformation as a whole.

The approach to treatment without an appropriate bird’s-eye view, limiting the focus of treatment to only the VM component, will invite another unwelcome problem causing more harm than good to other co-existing CVMs. For example, overzealous management of extratruncular or truncular VM often results in the worsening of a co-existing extratruncular LM (e.g. increasing lymph leakage). 10

Lastly, another commonly ignored component of VM management is the ‘truncular’ VM lesion, occurring alone or combined with other CVMs. Truncular VM lesions generally present as aplastic, hypoclastic or hyperplastic, normally-developed vessels (e.g. rudimentary femoral vein and absence of iliac vein). They also present as defective veins with various conditions such as a vein web, aneurysm or ectasia. Occasionally embryonic and fetal vessels persist due to failure of involution (e.g. lateral/marginal embryonic vein) resulting in complicated haemodynamic effects on the deep venous system.

Truncular VMs are vascular defects that arise during a ‘later’ stage of embryogenesis where the ability to grow and proliferate is lost. These lesions are associated with profound haemodynamic effects on the venous system depending on the location and extent. This type of lesion is directly involved with the vascular trunks resulting in various anomalous conditions of the affected vessels.

Only with an appropriate bird’s-eye view of the VM as one of the many different types of CVM can management of the VM be carried out with minimal risk of complications and morbidity.

References