EDITORIAL CIENTÍFICA

Venous Malformation is NOT a Hemangioma

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Term of "Hemangioma" is most frequently misused terminology for the venous malformation (VM), if not for entire congenital vascular malformation (CVM) altogether. But, genuine "Hemangioma" is NOT a CVM but it is a "vascular tumor".

Although VM has been often mistakenly called to "cavernous" and/or "capillary" hemangioma, VM is one of the CVMs as the outcome of the developmental arrest affecting the venous system as a predominant form and NOT an hemangioma.

A hemangioma is one of the vascular tumors developed after the birth mostly in the infantile/ neonatal period and naturally it is NOT a vascular malformation.

The term of hemangioma should represent only "infantile" hemangioma which includes "congenital" hemangioma as one of its variants and it should NOT be used for the CVMs.

Although the vascular malformation and vascular tumor belong to the "vascular anomaly" together, both conditions are fundamentally different not only in their anatomical, histological and pathophysiological findings but also in their clinical courses.

Hemangioma is a vascular tumor that originates from the endothelial cells with a distinctive growth cycle, a proliferation phase of early rapid growth followed by an involutional phase of slow regression. Hemangioma has such unique characteristic of "self-limited" growth as vascular tumor but vascular malformations are embryologic tissue remnant with "self-perpetuating" growth.

The absolute majority of the hemangioma appears through the neonatal period following the normal birth and it would grow soon after it is obvious clinically. It often takes an explosive growth in its initial/ proliferative phase followed by natural regression through involutional phase. They would regress to disappear before reaching the age of 5 to 7 years in its majority as a rule.

But the VM presents at birth as an inborn vascular defect like all others CVMs and would never disappear/ regress spontaneously through the rest of the life.

Indeed, its "extratruncular" type of the lesion will continue to grow slowly at a rate that is proportional to the growth rate of the body.

Therefore, differential diagnosis between vascular malformation and hemangioma is relatively easy, only with careful history and physical examination and simple test like Duplex ultrasonography when it is occasionally needed to confirm the clinical impression, if there is no doubt.

However, the diagnosis of the VM itself is a different story especially for its embryological characteristics: extratruncular type and truncular type.

The term of "extratruncular versus truncular" lesion is based on newly introduced embryological subclassification of modified Hamburg Classification (Table 1 A and B).

Hamburg Classification is a new version of the CVM classification replacing old classification mostly based on the name-based nosolgy only to add the confusion.

Extratruncular VM lesion is "prematured/ pretruncal embryonic lesion" as the outcome of the developmental arrest occurred in the earlier stage of embryonic life (the reticular stage) before the vascular system forms vascular trunks. It retains the characteristics of mesenchymal cells (angioblasts) as an embryonic tissue remnant of mesodermal origin: the evolutional potentials to grow when stimulated (e.g. trauma, surgery, hormone, pregnancy). It, therefore, accompanies the risk of a recurrence after suboptimum management.

It often presents clinically diffuse infiltrating condition to give mechanical impact to the surrounding tissues/organs in addition to its hemodynamic impact and to its belonging venous system.

Truncular VM lesion is "matured lesion" as the defect occurred during vascular trunk formation (truncal) stage, the "latter" stage of the fetal development following the reticular stage of vascular development. So, it has lost embryonic characteristics of the mesenchymal cells (angioblasts) with no risk of recurrence but it has much more serious hemodynamic impact to the involved vascular system (e.g. marginal vein).

It remains as fetal (truncal) vessel without a normal involution (e.g. sciatic vein, marginal vein), or as deffective vessel trunk formation: obstruction/dilatation of the formed vessel (e.g. vein web, venous aneurysm) or aplasia, hypoplasia and/or hyperplasia of the vessel development (e.g. agenesis/rudimentary deep vein).

This new classification system with standardized definition fulfills mandated condition to provide accurate anatomo-patho-physiologic information/status of the developmental arrest from different embryonic stages as the cause of various vascular defects.

Modern diagnostic and therapeutic technology provided appropriate ground for new contemporary classification as a new guideline for better understanding of this complex problem replacing old terminology (Table 1 A and B).

The majority of the VMs exists alone as an independent (predominent) lesion but infrequently it exists as a mixed condition with other CVMs (hemolymphatic malformation), lymphatic malformation (LM); capillary malformation (CM); aretrio-venous-shunting malformation (AVM), (e.g. Klippel Trenaunay Syndrome=VM+LM+CM; Parkes Weber Syndrome=VM+LM+AVM+CM).

Therefore, VM locates anywhere within the body in various numbers, shapes, extents/degrees and conditions either as one single type of predominent lesion or mixed condition with other CVMs.

Like all other CVMs, VM accompanies hemodynamic characteristics to affect involved venous system as well as embryonic characteristics

1A. Hamburg Classification[1] of Congenital Vascular Malformations (CVMs) - Types

- Predominantly arterial deffects
- Predominantly venous deffects
- Predominantly AV (arteriovenous) shunting deffects
- Predominantly lymphatic deffects
- Combined vascular deffects
- Predominantly capillary deffects

1B. Hamburg Classification of CVMs[2]: Forms - Embryological subtypes

1. Extratruncular forms
   - Infiltrating, diffuse
   - Limited, localized
2. Truncular forms
   - Stenosis or obstruction
     - Hypoplasia; Aplasia; Hyperplasia
     - Membrane; Congenital spur
   - Dilatation
     - Localized (aneurysm)
     - Diffuse (ectasia)

[1]The modified classification of the original classification which was established based on the consensus on CVM through the international workshop in Hamburg, Germany, 1988.

[2]Represents developmental arrest at the different stages of embryonic life: Earlier stage Extratruncular form; Later stage – Truncular form. And both forms may exist together.

Table 1 (A and B)

of "evolutional potentials" to grow, originated from mesenchymal cells when developmental arrest occurs in earlier stage of embryonic life.

This results in a wide range of clinical presentations of the VMs with unpredictable clinical course, erratic response to the treatment with high risk of recurrence, and confusing terminology with no information on the etiology, anatomy, and pathophysiology.

VM is, therefore, unique venous disorder of extreme variety with stigma of totally unpredictable behavior: "recurrence" is the trademark of the VM together with other CVMs.

Appropriate differential diagnosis of the VM from the "genuine" hemangioma should be done first especially in pediatric age group and a subsequent differentiation to identify its embryological sub-types-extratrucular and truncular lesions, should follow for proper implementation of different treatment criteria with different indications on various conditions involved to the VM.

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