1 History, Terminology, and Classifications of Vascular Anomalies

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1.1 Introduction

The objective of this chapter is to present a ready reckoner for the understanding of the history, incidence and a review of all noted classifications. This may provide a comprehensive understanding of this group of vascular-related entities.

Majority of the vascular anomalies are seen in the head and neck region. Even though the incidence of this anomaly could be construed as a rare disease entity, with only 5% of overall affliction, the lack of knowledgeable management has disfigured many. A comprehensive understanding of this benign yet complex life-changing entity is essential. A historical perspective, pathophysiological evolution, and the current knowledge of management modalities are essential for rendering clinical care in this subspecialty care.

The core points to consider in vascular lesions management -

1. Majority are benign entities and not "cancers." The high-flow vascular malformations can be treated effectively too.
2. Lesion eradication should not be a piecemeal methodology.
3. There is no one treatment modality that would fit every vascular anomaly.
4. Biopsy of the lesions may not be necessary; malformations are more common. Biopsy only if you understand the malignant behavior.
5. Vascular anomalies have a" dynamic lifecycle."
6. Between vascular tumors and malformations- tumors are more common, but you may encounter "malformation" more often as they persist and need varied care.
7. Vascular anomalies are a "group" of different entities of varied etiopathological which may not share the same treatment approaches. The grouping is for logistical convenience.
8. Airway has to be a consideration in managing head and neck anomalies, especially with syndromes and segmental presentations.
9. No single susceptible population group has been identified.
10. Patients usually present with multiple earlier investigations and interventions.
11. That is a great potential for molecular and genetic research in this field-so consented patients could save tissue samples for laboratories discovering targeted therapies.

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1.2 History

The first scientifically documented "surgery under an ether anesthetic agent" by Morton in 1846 done in Boston, Massachusetts USA, was for excision of a "low flow" vascular malformation. Rudolph Virchow, in 1863 first categorized vascular anomalies by microscopic architectural patterns. From earlier use words "Port wine stain," "Angiomas," etc. Virchow's attempted an acceptable Cellular classification as angioma simplex, angioma cavernosum and angioma racemosum. His trainee Wagner further studied lymphatic malformations and contributed to the understanding of their challenging clinical management. The further biological classification was proposed in 1982 by Mulliken and Glowacki [1]. The international collaborative group ISSVA-International Society for the Study of Vascular Anomalies is a way forward to advance constant review of the current and updated science in this field. In this rapidly changing understanding of the molecular basis of disease, we may need a constant redefinition of our comprehension and learning.

Classifications for vascular lesions are complex. It serves as a means of communication between the clinicians. Today, the ISSVA is an umbrella association for the classification of vascular lesions. At the broadest level, the ISSVA classifies vascular lesions as Vascular Tumors and Vascular Malformations (Table 1.1) [2]. Vascular malformations are structural abnormality that is always present at birth but may not become apparent until later in life [3]. They include capillary (arteriole and venule), venous, arteriovenous, lymphatic, and a combination of these vessels. Vascular tumors have increased mitotic activity and will appear soon after birth, with approximately 30% already apparent at the time of birth [1, 3]. Vascular tumors are divided by the ISSVA as benign, locally aggressive or borderline, and malignant. Benign tumors include hemangiomas. Locally aggressive or borderline tumors include Kaposi Sarcoma and hemangioendothelioma. Malignant tumors include angiosarcomas and epithelioid hemangioendothelioma. Intraosseous vascular lesions have not been classified by the ISSVA. Table 1.1 is a summary of individual authors who proposed vascular anomaly

Table 1.1 Various classifications of vascular anomalies

Year	Author	Basis of classification
1863	Virchow RLK	Microscopic channel architecture
1877	Wegener	Histomorphic subclassification of Virchow's classification
1973	Degni and coworkers	Site of origin of the detect
1974	Malan	Embryologic site of origin of the defect
1982	Mulliken JB and Glowacki J	Endothelial characters
1983	Burrows and colleagues	Angiographic flow patterns
1988	International Society for the Study of Vascular Anomalies (ISSVA), Hamburg	Anatomopathologic classification of vascular defects (Hamburg classification)
1989	Belov	Etiologic and pathophysiologic classification system
1992	ISSVA, Colorado	Cellular features, vascular flow characteristics, and clinical behavior
1993	Jackson and associates	Flow rate
1996	ISSVA, Rome	Modified ISSVA classification
2011	S C Nair	Anatomical presentation
2014	ISSVA, Melbourne	Modified ISSVA classification

classification and the basis of their effort to differentiate various presentations.

Head and Neck Vascular anomalies can cause significant morbidity. There are various misguided attempts to treat with the "one size fits all" approaches. Chemotherapy, Interferons, chemical agents with no long-term outcome reviews have disfigured many an individual seeking care.

We have attempted to give you a list of classifications more for a perspective and the efforts which have gone to well-intentioned methods to care in Table 1.1 above.

The international Society proposed its fast classification in 1988 in Hamburg. In multiple meetings have happened in Hamburg, Germany, Colorado, USA, and Rome, Italy etc. an ongoing. Occasionally the names of the meetings have been associated with the classifications (Hamburg classification). Multiple societies like the oculoplastic association have its own classification of vascular anomalies of the orbit.

1.3 Incidence

The overall incidence of vascular lesions is around 5% [4]. Among tumors, infantile hemangiomas have the same percentage of incidence. Among vascular lesions, venous malformations are commonly similar to lymphatic malformations with an incidence of 1: 5000 to 1:10000. More than 50% of them are present in the head and neck anatomical area.

1.4 Terminology and Classifications

Terminology of Vascular lesions evolved from descriptive to the more scientific basis for nomenclature, the former possibly arising from maternal explanations of the lesions. It posted 1982 after Mulliken and Glowacki et al. [1] attributed biologic differences within the lesion that they were described as separate entities. A neoplastic Hemangioma was found to be distinctly different from the developmental VM.

1.4.1 Historical Terminology on Appearances

Descriptive terms and historical nosology of vascular anomalies offers an array of overlapping descriptive and histopathologic terms (Figs. 1.1, 1.2, and 1.3)

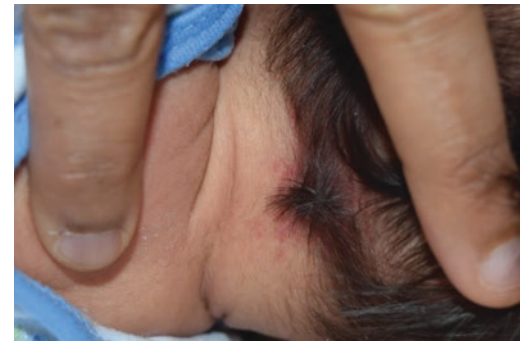


Fig. 1.1 Superficial vascular lesion was seen on the neck of infant. Historically was also referred to as "stork bite"

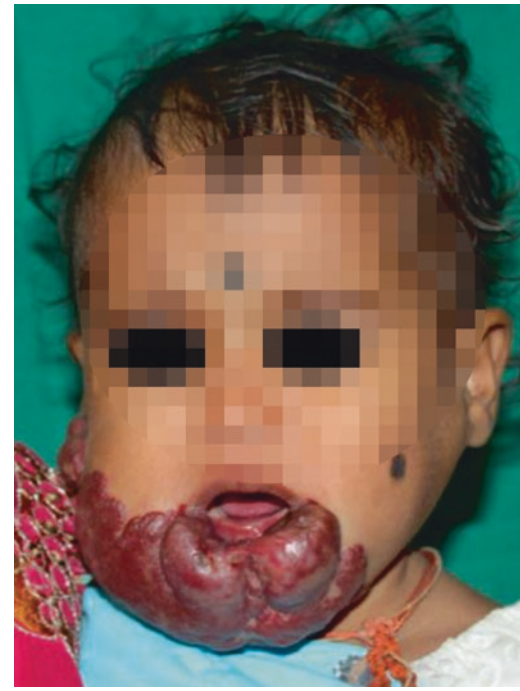


Fig. 1.2 Capillary malformation of lower lip, chin, and right cheek

- Port wine, as illustrated below it can be used for "capillary hemangiomas" historically. It can be as small and superficial.

1.4.2 Histopathologic Terms Used Historically

- Capillary
- Cavernous
- Arterial
- Mixed

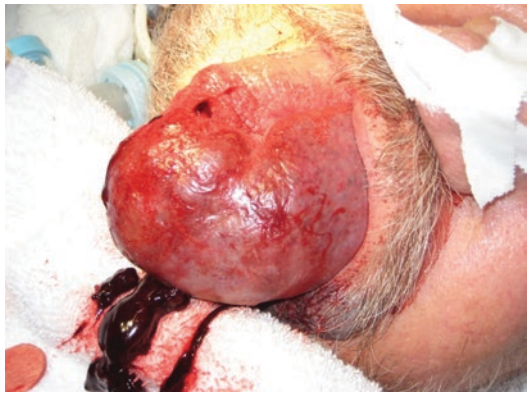


Fig. 1.3 Strawberry lesion or a "capillary-cavernous hemangioma" was commonly used for lesions like present on the tongue as illustrated in Fig. 1.3. Such terminology added to the confusion as shown in the tongue vascular lesion, which is a low flow vascular lesion, with sequela of trauma

Biologic behaviour-based terminology [1]—This is the current ISSVA terminology simplified and widely accepted. The classification is elaborated below.
Vascular Tumors
Vascular malformations

1.5 Mulliken and Glowacki's Biological Classification (1982)

The ISSVA classification has been modified and based on the biologic classification by Mulliken and Glowacki [1] and further with the mutational knowledge.

Hemangiomas
- Proliferating phase.
- Involuting phase.

Malformations
- Capillary
- Venous
- Arterial
- Lymphatic
- Fistulas

- Hemangioma (Fig. 1.4)

1.6 Vascular Malformations (Figs. 1.5 and 1.6)

1.6.1 Classifications

Here we have listed all relevant classifications espoused by various groups. Each has their merits. The current International Society for the study of Vascular Anomalies classification system. There have been various editions over the years.

Here is a link to the entire extensive classification

- https://www.issva.org/classification
- https://www.issva.org/UserFiles/file/ISSVA-Classification-2018.pdf

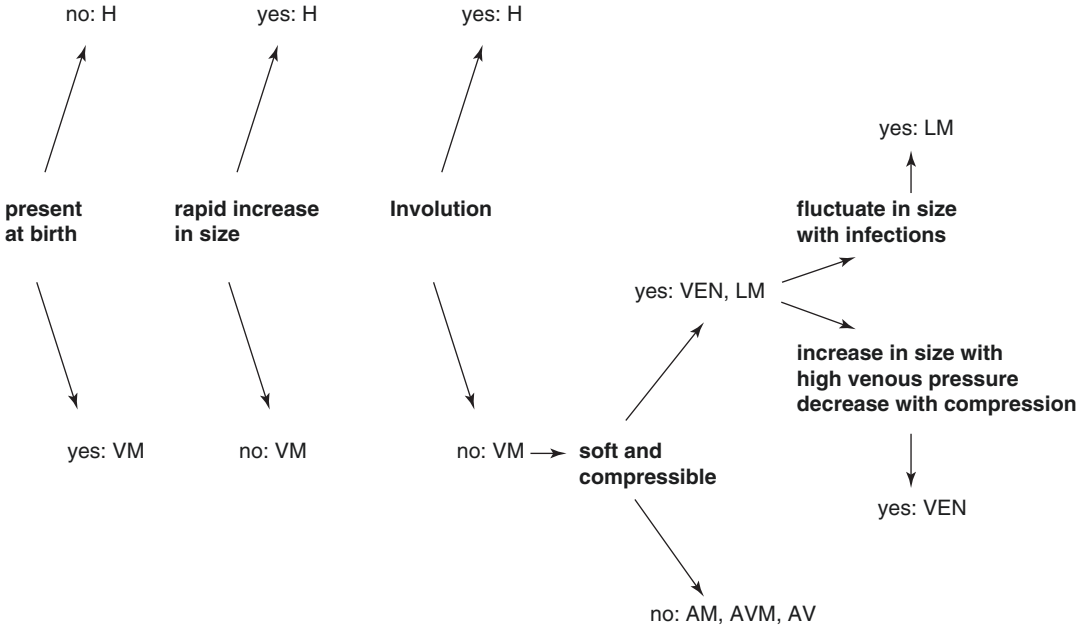
International Society for the study of Vascular Anomalies classification system (ISSVA, last revision May 2018)
Vascular tumors
Benign
Locally aggressive or borderline
Malignant
Vascular malformations
Simple
Capillary malformations
Lymphatic malformations
Venous malformations
Arteriovenous malformations
Arteriovenous fistula
Combined (2 or more VMs in one lesion)
CVM, CLM, LVM, CLVM, CAVM, CLAVM, others

(C-capillary, V-venous, L-lymphatic, AV-arteriovenous)
Anomalies of major named vessels
Vascular malformations associated with other anomalies

Based on the anatomic location, Hemangiomas were described by Waner and Suen [3] as:

Classification by Waner and Suen:
• Superficial hemangioma
• Deep hemangioma
• Combined hemangioma

M.Ethunandan et al., [5] bjoms 44(2006) described a flow chart that correlated the various clinical signs, which helped differentiate a Vascular malformation from a Hemangioma.



***H* hemangioma; *VM* vascular malformation; *VEN* venous malformation; *LM* lymphatic malformation; *AM* arterial malformation; *AVM* arteriovenous malformation; *AV* AV-fistula.**

Further advances in the understanding of the vascular dynamics allowed changes in the classification, and this was first proposed by Ian Jackson et al., allowing major breakthroughs in treatment planning and management of vascular lesions.

Box 1 Classification of vascular anomalies by vascular dynamics according to Jackson

I. Hemangiomas
II. Vascular malformations
(a) Low-flow (VM)
(b) High-flow (AVM)
III. LMs

From Jackson IT, Carreno R, Potparic Z, et al. Hemangiomas, vascular malformations, and lymphovenous malformations: classification and methods of treatment. Plast Reconstr Surg 1993;91:1217

1.7 CNS and Non-CNS List of Vascular Lesions

Central nervous system vascular malformations are rare abnormalities of blood vessels in your brain or spinal cord and their membranes. They distinctly differ in their appearance and management. A classification differentiating the CNS VM from its non-CNS variant is presented.

Table I Classification of CNS vascular malformations

Arteriovenous shunts
Classical AV Malformations
Pial AV fistulas
Dural AV shunts
Galenic shunts
Cavernous malformations
Capillary telangiectasias
Venous malformations
Developmental venous
Anomalies
Mixed malformations

Table II Classification of non-CNS vascular malformations

Haemangiomas (proliferative lesions or tumours)
Vascular malformations (non-proliferative)
High flow
• Arteriovenous malformations
Low flow:
• Capillary malformations
• Venous malformations
• Lymphatic malformations
• Mixed malformations

The severity of Artery venous malformation was scored by Schobinger into four stages based on their clinical appearance and behavior. From lesions exhibiting minimal symptoms to the severe malformations that resulted in hi-output cardiac failure consequent to the decompensatory phenomenon. Priority in therapeutic management decisions evolved.

1.8 Schobinger Classification

Schobinger clinical staging system of arteriovenous malformations

Stage I	Quiescence	Cutaneous blush, skin warmth, arteriovenous shunt on Doppler ultrasound
Stage II	Expansion	Darkening blush, lesion shows pulsation, thrill and bruit
Stage III	Destruction	Steal, distal ischemia, pain, dystrophic skin changes, ulceration, necrosis, soft tissue and bony changes
Stage IV	Decompensation	High-output cardiac failure

Based on Eighth Meeting of the International Workshop on Vascular Malformations. Amsterdam, Netherlands, 1990

Adapted from Kohout MP, Hansen M, Pribaz JJ, et al. Arteriovenous malformations of the head and neck: natural history and management. Plast Reconstr Surg 1998;102:643–54

1.8.1 Jackson's Classification

IAN JACKSON IN 1992 further expanded on the earlier classification. Based on this, Hemangiomas were described on their location and VM on the dynamics of blood flow.

Hemangiomas
Superficial (capillary hemangioma) (Fig. 1.2)
Deep (cavernous hemangioma)
Compound (capillary-cavernous hemangioma)
Vascular malformations
Simple lesions
Low flow lesions
Capillary malformations (capillary hemangioma, port wine stain)
Venous malformation (cavernous hemangioma)
Lymphatic malformation (lymphangioma, cystic hygroma)
High-flow lesions
Arterial malformation
Combined lesions
Arteriovenous malformations
Lymph venous malformations (Fig. 1.6)
Other combinations

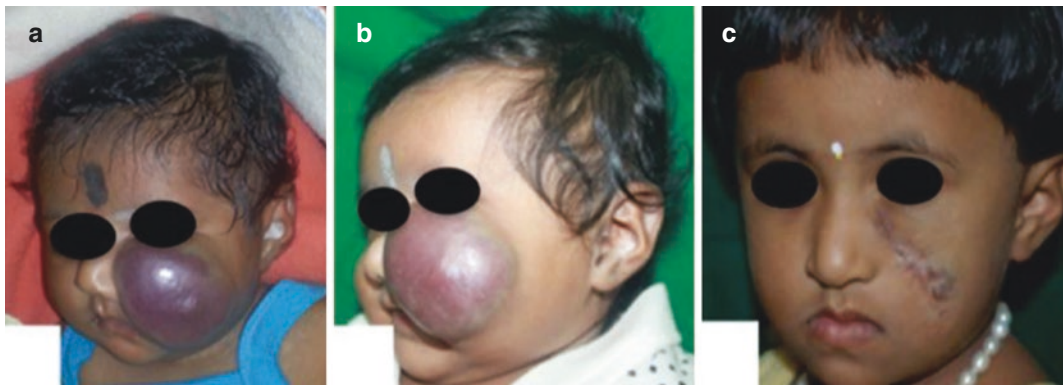


Fig. 1.4 Non-involuting congenital Hemangioma (NICH) of the left cheek ((**a**) initial therapy with propranolol 10 days post-birth, (**b**) 6 months following propranolol therapy, (**c**) excision of lesion at 2 years age)

1.8.2 Surgical Management Based Classification for Head and Neck Lesions

More recently Sanjiv Nair et al. [4] classified both Hemangiomas and VM based on their anatomic location. This helped understand the depth of the lesion and allowed a surgical treatment plan .The classification has been effectively used in the surgical ablation of the lesions, as will be described in Chap. 7.

Type I	Mucosal/cutaneous
Type II	Subcutaneous/juxta muscular
Type III	Glandular
Type IV	Skeletal (intra bony)
Type V	Deep visceral (temporal, infra temporal, parapharyngeal)

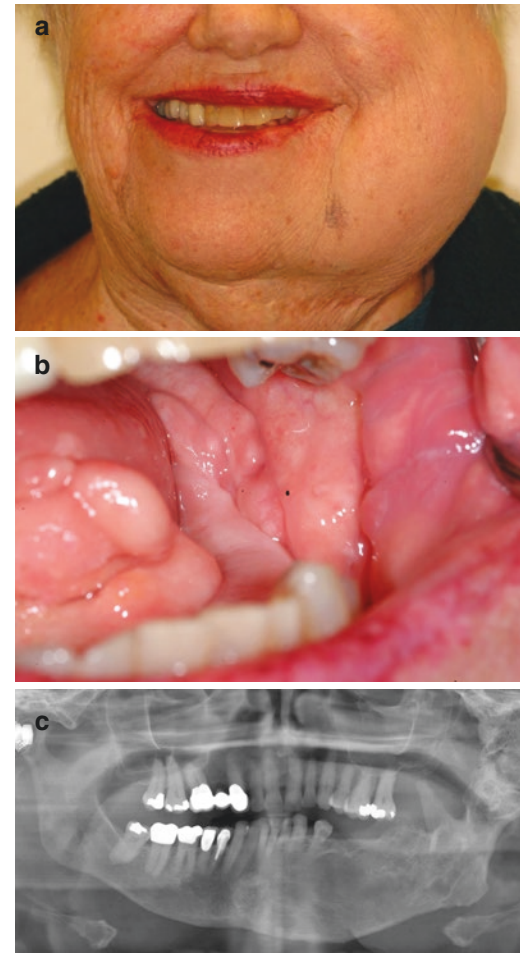


Fig. 1.5 Left facial low flow lymphovenous malformation of an intraosseous type of the left lower facial skeleton. (**a**) Noted left lower facial asymmetry with no superficial vessels. (**b**) Mucosal area of the intraoral picture with tissue expansion and no surface ulceration or superficial vessels. (**c**) Left mandibular and condylar hypertrophy of the skeleton involved. Courtesy: O Ross Bernie DDS PhD

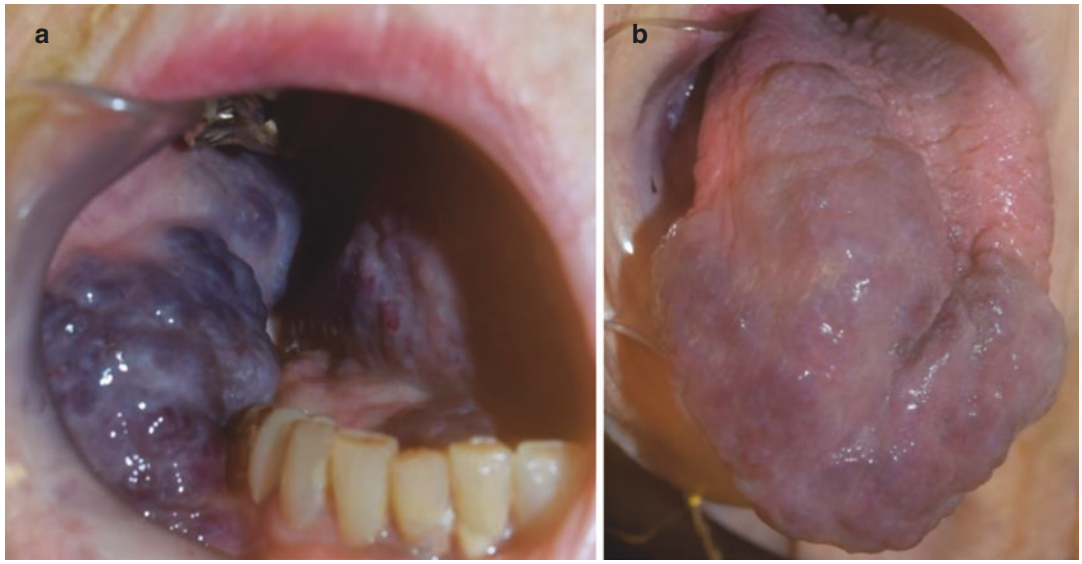


Fig. 1.6 The appearance is easy to be remarked as "berry" or "purple" which caused the historic confusion in nomenclature (**a**) Notice extensive dark purple lesions of right buccal mucosa extending to tongue and floor of the mouth. (**b**) Shows extensive growth and discoloration of anterior ventral tongue

1.9 Conclusion

The evolution of various classification has been based on the increased understanding of both clinical and biologic behavior of VA. Earlier confusion in terminology has been largely dispelled with a better understanding of the clinical features aided by advances in imaging techniques.

Vascular lesions have a varied clinical and radiographical presentations. A surgical plan and approach based on Nair's Clinical Classifications and imaging data can minimize complications. The radiologist, pathologist, and surgeon must consider clinical, radiographic, and histological data. A classification system for intraosseous vascular lesions needs to be established and is a work in progress.

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