

Vascular anomalies of orofacial region- A review

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ABSTRACT

Vascular anomalies well-known as birthmarks are benign lesions of blood vessels or vascular elements are often considered errors of vascular morphogenesis. Numerous terms have been used to describe vascular anomalies on the basis of physical appearances leading to misdiagnosis followed by complications in the management of the lesion. Lack of complete understanding of the anomaly result in misdiagnosis. Main aim of this paper is to discuss the vascular anomalies of the head and neck region in detail which will enable the dental practitioner to arrive at a better diagnosis.

Key words: vascular anomalies, classification, lasers, radiographic features

From being a good luck charm to the kiss of an angel bestowed by the heavenly stars, these marks of life are sometimes the cause of disfigurement. Be it clusters of veins, arteries or lymph channels, often give rise to these marks which are noticed within or under the skin as soft mass or stain that are compressible, non-pulsating with bluish-red tinge. They differ from each other in having different clinical, histological and imaging features and hence differ in management protocols. Although painless, these lesions can result in significant cosmetic disfigurements and lead to life threatening hemorrhages after a simple surgical procedure such as extraction.

Due to the usage of perplexing colloquial terms based only on clinical outlook, practitioner often misdiagnoses the anomaly which will bring about reduced outcome of treatment advised. Therefore thorough knowledge and understanding of each type of vascular anomaly plays an eminent role in the diagnosis. In this review we discuss about the newer terminologies, current classification,

pathogenesis, salient clinical and immunohistochemical features of individual anomaly, different imaging modalities used and the management of lesion so as to avoid further bewilderment in diagnosis and treatment protocol.

Classification

Vascular anomalies are either congenital or appear within first year of life [1]. These are benign lesions which can appear anywhere in the body but often evident in the head and neck region with 5-10% prevalence rates [2, 3]. Dormant lesion can be triggered by certain factors such as trauma, infection, hormonal alterations and progressive increase in age. Abnormal vascular morphogenesis, increased mitotic activity as well as cellular hyperproliferation are thought to play a role in its pathogenesis [1, 4]. Several classifications have been put forward to categorize these lesions. In 1982, Mulliken and Glowacki classified the vascular anomalies based on the endothelial cells and clinical characteristics into vascular

tumors and vascular malformations. In 1996, the International Society for the Study of Vascular Anomalies

(ISSVA) used fundamentals of Mulliken and Glowacki’s classification system along with immunohistochemical aspects. In 1999, Waner and Suen modified the ISSVA classification to clinically useful classification system [1-6]. (Table 1)

Table 1: classification of vascular anomalies [1]

Vascular Tumors	Infantile hemangiomas		
	Congenital hemangiomas (RICH, NICH)		
	Tufted angioma (with or without Kasabach-Meritt Syndrome)		
	Kaposiform hemangioendothelioma (with or without Kasabach-Meritt Syndrome)		
	Spindle cell hemangioendothelioma		
	Dermatologic acquired vascular tumors (Pyogenic granuloma, targeoid hemangioma, glomeruloid hemangioma, microvenular hemangioma, etc)		
Vascular Malformations	Slow-flow vascular malformations	Capillary malformations (CM)	Port-wine stain
			Telangiectasia
			Angiokeratoma
		Venous malformation (VM)	Common sporadic VM
			Bean syndrome
			Familial cutaneous and mucosal venous malformation(VMCM)
	Glomuvenous malformation(GVM)		
	Lympatic malformation (LM)	Mafucci syndrome	
	Fast-flow malformations	Arterial malformation (AM)	
		Arteriovenous fistula (AVF)	
		Arteriovenous malformation (AVM)	
	Complex-combined vascular malformations	CVM,CLM,LVM,CLVM,AVM-LM,CM-AVM	

VASCULAR TUMORS:

Hemangioma:

Hemangiomas are the most commonly occurring tumor in infants mostly in females with incidence of 10% [1, 4, 7]. People with light colored skin, premature birth and low birth weight are considered to be at higher risk. Hemangiomas generally occur sporadically however, may perhaps be familial expressed as autosomal dominant inheritance pattern, with loss of heterozygosity on chromosome 5q31-33.5 comprising of rapidly dividing endothelial cells [8, 9].

Pathogenesis has been put forward based on following two theories: First theory suggests, disrupted placental tissue give rise to hemangiomas endothelial cells which is found embedded in fetal soft tissue during gestation/birth. The tissue specific markers are: GLUT-1, CD14 (Lewis antigen) CD32 (FcGammaR2) and Merosin [10,11].

Second theory evolved on sighting the endothelial progenitor and stem cells in the circulation of patients with hemangiomas. Abnormal levels of MMP-9 and proangiogenic factors (VEGF, b-FGF and TGF-β1) play a role in pathogenesis [12].

When hemangiomas are visualized in utero and perceived at birth they are termed as congenital hemangiomas (CH) having 20% incidence rates. They do not proliferate after birth. Based on the involution they are subcategorized as Rapidly Involuting Congenital Hemangiomas (RICH) And Non- Involuting Congenital Hemangiomas (NICH). Infantile hemangiomas (IH) present shortly after birth [13-15]. Hemangiomas manifest in 3 developmental phases: proliferation- rapid growth phase lasts for 3 months of life. Followed by a slow or no growth phase termed quiescence, lasts from 9-12 months. Finally there is graying of overlying skin and deeper component shrinking knows as involution phase [16].

Superficial Hemangiomas are red, nodular with no subcutaneous component. Whereas, Deep hemangiomas have bluish tinge or telangiectasia. Compound hemangioma consists of superficial and deep components. Focal Hemangiomas are localized, unilocular lesions while, segmental hemangiomas are diffuse plaque-like. If multifocal hemangiomas present with lesions >5 in number examination of visceral organs is recommended to rule out syndromes associated like, PHACE and LUMBAR syndrome [17].

Diagnosis is best made by history and physical examination. MRI with contrast or Doppler Ultrasound is used to confirm the diagnosis [18-20]. Currently established treatment ideology is “close observation” reinstated traditional “watch and wait” method with following approaches i.e., accurate diagnosis, early management of complications, decrease in complications and increased morale with positive psychological state [21]. Medical management includes first line steroid therapy, lasers, bleomycin a5 is an effective substitute for vincristine and interferon resistant hemangiomas. Surgical resection is considered in cases of accessible hemangiomas [22-24].

Tufted Angioma (TA) and Kaposiform Hemangioendothelioma (KH)

Being presented as vascular tufts of capillaries (TA) and components of vascular and lymphatic channel (KH), these rare vasoproliferative tumors display at or shortly after birth. They are sometimes associated with secondary destructive osseous changes. A syndrome associated with is Kasabach-Meritt syndrome. Immunostaining patterns of monoclonal antibody D2-40 help to differentiate TA from KH. Imaging modalities are same as vasoproliferative neoplasms. Management is similar to those for infantile hemangioma. When there is aggressive local behavior wide local excision and supportive treatment the mainstay of therapy [25, 26].

Hemangioendothelioma

Being considered a benign reactive vasoproliferation that present at any age throughout the body, was first described in 1986 as a rare vascular neoplasm. Immunohistochemistry is positive for CD31 and factor VIII antigen and negative for CD34 [5]. Epithelial hemangioendothelioma is rare, slowly progressive, distinct

pathological tumors occur anywhere in the body. Most common in the skeleton and is an accidental finding during normal investigative procedures. Other rare types of hemangioendotheliomas are Composite, Retiform, Polymorphous, Dabska Tumor and Lymphangioendotheliomas. Dermatologically acquired vascular tumors include Pyogenic Granuloma, Targeoid Hemangioma, Glomeruloid Hemangioma, Microvenular Hemangioma etc [18,19].

Angiosarcoma:

Is a rare aggressive neoplasm, has female predominance and average age of onset is 3.7 years. Cross sectional imaging shows an aggressive heterogenous mass with pooling of contrast and multiple synchronous or metastatic lesions in the liver [5, 13].

VASCULAR MALFORMATIONS:

Vascular malformations are congenital morphogenic anomalies composed of inappropriately connected vasculature. Lesions infiltrate normal tissues, do not regress and continue to expand with time. They can be subdivided into 2 categories, slow/low flow consists of combinations of capillary, venous and lymphatic components and fast/high flow malformations contain arterial component in combination with other vascular structures [2,4,19].

Slow flow malformations:

Venous malformations:

Previously known as “venous angioma” and “cavernous hemangiomas”, these are most commonly seen vascular malformations. They are generally sporadic, sometimes familial with association of chromosome 9p [18,27]. Studies show loss of function mutation on angiopoietin receptor gene TIE2/TEK, and up regulation of TGF- β and β -FGF factors as well as presence of progesterone receptors.

Often congenital, these consist of abnormally formed and dilated superficial or deep veins, with thin walls due to lack of smooth muscle and appear as a bluish patch/mark. VM can be localized/generalized and can occur anywhere in the body which are compressible and swell with increased hydrostatic pressure. Elevated D-dimers is marker of the disease [4]. They can be found

intraosseously which differentiates it from hemangioma. Complications could be pain, swelling with formation of phleboliths and coagulopathies. MRI is most useful diagnostic. Treatment includes sclerotherapy with ethanol and sotradecol, laser therapy (ND: YAG, PDL, Gentle YAG or KTP) and surgery [27-29].

Capillary malformations:

Hemodynamically inactive superficial sporadic lesions affecting the skin and/or mucosal capillary network are found anywhere in the body, mostly in cervicofacial region [1]. They can be categorized into medial form consisting of stork bites on nape of the neck, angel kisses on forehead which gradually lighten with time and disappear [30]. Lateral forms commonly known as port wine stains. Other examples of CM's are nevus flammeus, telangectasias, and spider nevus.

Pathogenesis is indefinite. Genome wide linkage analysis has identified a locus on chromosome 5q association with disease [4]. Congenital CM's are flat, red or purple patches with irregular borders. Port wine stains are usually seen along the distribution of trigeminal nerve. Lesions progress with time and become darker, elevated and nodular. Diagnosis is by physical examination. In case of associated symptoms MRI may be performed. FDPL Laser is the main treatment modality. Surgical excision is an option in lesions amenable to lasers [31-33].

Lymphatic malformations:

LM's are sporadic lesions composed of lymphatic vessels with inappropriate communications, lined by endothelial cells and filled with lymphatic fluid. Earlier known as "cystic hygroma" and lymphangioma" terms are no longer used. They are mostly congenital or appear by 2 years of life, are non-tender and soft can occur in any part of the body. Local infections may cause LM to swell, protrude and become painful. They can be classified as, Macrocytic (>2cm³) which are present beneath the skin, Microcytic (2cm³<) have small vesicles over the swelling involving skin or mucosa and Mixed [2].

Etiology is unclear. Receptors involved in the formation of lymphatic vascular channels like, VEGFR3 and Prox-1 may play a role in development of disease. LM can also occur as a result of trauma or infection. Complications include dysphagia, odynophagia, impaired

speech, severe obstruction. Involvement of skeletal framework can cause osseous dystrophy leading to dental or extremity abnormalities [34]. Diagnosis is based on clinical presentation and imaging modalities like MRI, Ultrasonography and Doppler. Treatment modalities for larger lesions include aspiration of contents followed by injection of sclerosing agent. Laser ablation and surgical treatment could also be used. NdYAG and diode laser photocoagulation can be used to treat microcystic LM [35].

High flow vascular malformations:

Arteriovenous Malformation and Arteriovenous Fistula:

AVM and AVF are congenital and acquired malformations respectively. They are hemodynamically active malformations with rapid flow consist of capillary beds shunting blood from arterial system to the venous system. AVM consist of the core (nidus), multiple feeding arteries and widened draining veins which are pulsatile, either superficial or deep [5,36]. Whereas AVF lacks the nidus. Even though they are present at birth, they are not detected until puberty, sometimes lesion presents after trauma. A defect in vascular stabilization is thought to be the cause of these malformations. Defects in TGF-beta signaling and a genetic two-hit hypothesis theories help explain the pathogenesis. Expansion in puberty is due to the presence of progesterone receptors [37]. The characteristic features will be palpable warmth, pulse, or thrill due to high vascular flow. Common locations are intracranial, intraosseous and subcutaneous fat. They are infiltrative causing destruction of local tissues and are life threatening secondary to massive bleeding. They can be associated with hereditary hemorrhagic telangiectasia syndrome [5].

Imaging is mainstay in diagnosing the extent of AVM and AVF which includes, MRI, MRS, CTA and color Doppler ultrasound. Angiography should precede the treatment for complete mapping of the vessels. Endovascular embolization and surgical resection are main treatment modalities which includes palliative, complication control and curative as major protocols. Treatment of AVM is challenging because of high recurrence rate [1,4,37,38].

CONCLUSION

Though vascular anomalies do not bring about any

functional oriented problems, it does pose a source of psychological stigma to the patient and their families. Approach in management of such anomalies differs from lesion to lesion. Identifying the anomaly with proper nomenclature is the utmost importance in the diagnosis. Lesion can be diagnosed on basis of physical examination, histochemical markers and other imaging modalities. Treatment of vascular anomalies is complex and involves multidisciplinary approach to prevent further complications and rendering a positive psychological approach to patients.

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