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Hemangioma of Oral Cavity- A Review

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Abstract

Hemangiomas are vascular lesions presenting as proliferations of vascular channels. They are tumor like hamartomas. They are common in children frequently regress on it own or sometimes needs proper mangement of the same or else leads to complications. It can be present as intraosseous, intramuscular, soft tissues lesions of oral cavity. The current paper aims to highlight the various aspects of hemangioma.

Keywords: Hemangioma, oral cavity,Vascular lesion, Capillary hemangioma, Cavernous Hemangioma, Hamartomas.

Introduction

Hemangiomas can be defined as a tumor like malformation composed of seemingly disorganized masses of endothelial lined vessels that are filled with blood[1].Pindborg and Hjorting-Hansen (1974) define hemangioma as a benign neoplasm of vascular origin. Lucas (1976) states that they are hamartomatous lesions with growth resulting form the opening of malformed vessels, rather than true proliferation of cells[1].Stout and Lattes define hemangioma as a vascular tumor which may be of two types:[1]Capillary hemangiomas is comprised of haphazard arrangement of capillaries, Cavernous hemangiomas if vessels are widely dilated. Lucas (1976) stated that the hemangiomas occur in the head and neck with a greater frequency than any other part of the body[1].

Classification

The term 'hemangioma' has been commonly used to describe a large number of vasoformative tumors. The classification of these entities have been complex and not entirely consistent over time.In 1982, Mullikan and Glowacki described the classification scheme that is most accepted today. This scheme divides vasoformative tumor into 2 broad groups: **Hemangiomas and Vascular Malformations**. The vascular malformations can be subdivided into arterial, venous, capillary and lymphatic malformations.[2]

Table-1: Types of Hemangiomas

Vasoformative Tumors	New Nomenclature	Old Nomenclature
Hemangiomas	1.Capillary Hemangioma	Strawberry Hemangiomas
	2.Cavernous Hemangioma	Juvenile Hemangioma
		Parotid Hemangioma
	3.Mixed Hemangioma	
Vascular Malformations	Venous Malformation	Cavernous Hemangioma
	Intramuscular-Venous	Hemangiomatosis Intramuscular Hemangioma

	Malformation	Capillary Hemangioma
	Capillary Malformation	Port Wine Stain
	Arteriovenous Malformation	Arteriovenous Hemangioma
		Arterial Angioma
		Arteriovenous Aneurysms
		Cirsoidangioma
		Red Angioma
		Serpentine Aneurysm
	Lymphatic Malformation	Capillary Lymphangioma
		Cavernous Lymphangioma
		Lymphangioma Cystic Hygroma

The International Society for the Study of Vascular Anamolies (ISSVA), in 1996, approved a modified classification system.[3] The diseases were subdivided into:

A.**Tumors:** Hemangioma, Pyogenic Granuloma, Rapidly Involuting Congenital Hemangioma, Non involuting Congenital Hemangioma, Hemangiopericytoma, Tufted Angioma and Kaposiform Hemangioendothelioma.

B. **Vascular Malformations:**

Another Classification is that proposed by Watson and McCarthy [4] based upon a series of 1,308 blood vessel tumors and is as follows:

1. Capillary Hemangiomas
2. Cavernous Hemangioma
3. Angioblastic or Hypertrophic Hemangioma
4. Racemose Hemangioma
5. Diffuse Systemic Hemangioma
6. Metastasizing Hemangioma
7. Nevus Vinosus or Port Wine Stain and
8. Hereditary Hemmorrhagic Telangiectesia. [4]

Pathophysiology [5]

The classic sequence of events usually falls in 3 stages:

1. The Undifferentiated Capillary Network Stage
2. The Retiform Developmental Stage
3. The Final Developmental Stage

Undifferentiated Developmental Stage: The primitive mesenchyme primordia is nourished by an interlacing system of blood spaces without distinguishable arterial and venous channels.

Retiform Developmental Stage: It begins at about 48 days of embryonic development.Seperate venous and arterial stems appear on either side of the capillary network in the retiform developmental stage.

Final Developmental Stage: It begins at 2 months after development and involves the gradual replacement of the immature plexiform network by the mature vascular channels.

- The more common capillary hemangioma represents an arrest in the development of the mesenchyme primordia in the undiffrentiated capillary network stage.
- As differentiation progresses, primitive vessels penetrate deeper into the subcutaneous layer, the muscle or the bone tissue and give rise to **Capillary Hemangiomas.**
- Termination of development in the retiform developmental stage may produce venous, arterial, or capillary malformations because this stage is characterized by an established venous, arterial and capillary system.
- In the final developmental stage, the maturation of the venous and lymphatic system predominates. Aberrations in this mature stage of development result in venous malformations and lymphangiomas.

Other Hypothesis

1. Proliferating hemangiomas have been shown to have estradiol-17 beta-receptors in the cytoplasm and

corticosteroid treatment has been theorized to block these receptors.[5]

2. A number of growth factors including vascular endothelial growth factors (VEGF), basic fibroblast growth factor (bFGF), transforming growth factor(FGF) and interleukin 6 (IL-6) have been demonstrated as regulators of angiogenesis.[5]
3. Another theory suggest that endothelial cells of hemangiomas are derived from a distant population of endothelial precursors carried by vascular pathways to receptive environment. Potential sources include the bone marrow and the placenta.[5]
4. Recently, a comparison of the transcriptome of the human placenta and infantile hemangiomas supported a placental origin of the tumors. [6]

Epidemiology

Hemangiomas of oral cavity are not common pathological entity, however, among hemangiomas, the head and neck are frequent site of occurrence. Fifty percent of the venous malformations occurs in head and neck.[7]In the oral cavity, the bones and the muscles are affected as well as the mucosa and the skin. The incidence of intraosseous hemangiomas varies from 0.5-1.0% of all intraosseous neoplasms.[8]

Site: The most common bones affected are vertebral coulumn and calveria. The most commonly affected facial bones are mandible, the maxilla and nasal bones. Mandible is reported to be affected more often than maxilla (2:1).[9]

Intramuscular hemangiomas in the oral region are most commonly seen in the masseter, comprising 5% of all intramuscular hemangiomas. Tongue hemangiomas frequently extend deeply between the intrinsic muscles of the tongue. The lip mucosa is another common site for hemangiomas in children.[10]

- Race: It affects 12% of whites,but it rarely occurs in darker individuals.
- Sex: Hemangiomas are 3-5 times more common in females than in males.
- Age: Hemangiomas occurs in infants and children.[11]

Clinical Features

A). Capillary Hemangiomas: These are by a pale, well demarcated, flat area, most visible on agitation.Prodromal lesion may appear as pale halo area of telangiectasis or as very fine telangiectasia similar to port wine stain. Elevation occurs during 1st year of life and increases from the age of 3-8 months, with some growth continuing into second year of life.Stable interval period of 6-12 months often follows the growth period. A slow spontaneous involution begins in center of the lesion in most of the cases.

Involution often begins as darkening of color followed by appearance of numerous gray or pallid regions and fibrous septae within lesion.Most lesions get involuted by the time the patient reaches 7 years of age.Patient's sex, size of lesion, location and number of lesions do not influence the speed of resolution. Involution may continue into late teenage years also. [12-16]

B).Cavernous Hemangiomas: Cavernous hemangiomas are composed of large, irregular, deep dermal and subcutaneous blood filled channels that impart a purplish discoloration to the overlying skin. Typically soft, poorly defined, and readily blanch with compression, giving them a charecterstic "bag of worms" feel.The lesion may expand and darken, when agitated, or when placed in a dependent position.Often a capillary component overlies a cavernous component, and it may be difficult to distinguish these component histologically.Cavernous and mixed hemangiomas demonstrate the same patterns of proliferation as those of capillary lesions.However, involution is often incomplete, depending on the location

and the presence of associated arterovenous malformations. [12-16]

Oral Manifestations

The hemangioma of the oral soft tissue is similar to the hemangioma of the skin and appears as a flat or raised lesion of the mucosa, usually deep red or bluish red and seldom well circumscribed.[5]

The affected area might show bleeding around tooth necks, regional hyperthermia, pulsation, audible bruit, spontaneous bleeding, tooth mobility, pulsing teeth, local gigantism, painful or non tender. The most common sites of occurrence are the lips, tongue, buccal mucosa and palate.[5]

The tumor often is traumatized and undergoes ulceration and secondary infection. Paresthesia seldom occurs.[17] Certain tiny vascular formations of lip vessels called “microcherry”, “glomerulus” and “venous lake” have been described by Gius and his associates as lesions encountered with increasing frequency in the later decades of life and occurring with greater frequency in patients with gastric and duodenal ulcers.[5]

The intramuscular hemangioma is one special form of hemangioma which is quite rare in the head and neck region. It arises within the normal skeletal muscle, comprises less than 1% of all hemangiomas and is important chiefly because of the problem in differential diagnosis in treatment of the lesion. Intramuscular hemangiomas in the oral region are most commonly seen in the masseter, comprising 5% of all intramuscular hemangiomas.[5]

Stanley (1849) was the first to report intra-osseous hemangioma. Worth and Stoneman (1979) state that the vertebrae are the most commonly involved bones, although skull is also a frequent site.[18]

Radiographic Appearance

Hemangiomas have no pathognomonic radiologic features.It may appear cyst like either as unilocular or multilocular with septae (soap bubble) , Cyst like trabaculated

Showing sparse or spoke like trabaculation, cancellous or linearly trabaculated (Tube like appearance) with radiating bony spicules seen tangentially.[18]

Associated radiographic findings which are present in some cases include, ill defined and irregular margins often as multiloculated appearance because of residual bone trapped around the vessels. Small radiolucent locules may resembles enlarged marrow spaces surrounded by coarse, dense and well defined trabeculae. These internal trabeculae may form a honeycomb pattern.[18]

The trabaculae may be radiating like the spokes of a wheel or sunburst appearance, erosion of lamina dura, displacement and resorption of teeth may be noted.[1]

Investigations

- **Angiography:** Most definitive aid for the diagnosis of hemangioma.The angiographic appearance of intraosseous lesions are less well defined than that of soft tissue lesions.[19]
- **Ultrasonography:** To determine whether a lesion is angiomatous in nature, but it cannot diffrentiate between lymphangioma and hemangioma.
- **Contrast-enhanced MRI:** To differentiate a hemangioma from lymphangioma in the oral cavity.[20] MRI appears to be highly reliable for lesions of either soft tissue or bone.
- **Orthopantograms (OPG) :** Reveals a central vascular malformation of the bone and usually has a honeycombed appearance or cystic radiolucencies.[19] Intraosseous vascular malformations show a nonspecific reticulated or honeycombed pattern that is well demarcated from normal bone.

A sunburst effect, created by spicules radiating from the center, is often present.

- **CT Scan:** It often shows an expansile process with a high density amorphous mass that can be falsely diagnosed as fibrous dysplasia.
  - Aspiration of intraosseous lesions, that are diagnosed as oral hemangiomas readily produce frank blood.

Histologic Findings

Hemangiomas are classified as capillary and cavernous hemangiomas, depending on the size of vascular channels.A hemangioma may comprise numerous large dilated vascular channels lined by endothelial cells without a muscular coat; such lesions are referred to as cavernous hemangiomas. Rarely, cavernous hemangiomas may show a media muscularis. Cellular or capillary-type hemangiomas show significant endothelial proliferation, and the vascular lumina are very small. Histopathologically, vasoformative tumors share many similar microscopic features, and overlap between hemangiomas and vascular malformations exists.Salient histopathologic findings of vasoformative tumors that distinguish them are as follows [5]:

Hemangiomas (Proliferative Phase):

Endothelial cell hyperplasia forming syncytial masses.

Thickened (multilaminated) endothelial basement membrane.

Ready incorporation of tritiated thymidine in endothelial cells.

Presence of large numbers of mast cells.

Hemangiomas (Involuting Phase)

Less mitotic activity.

Little or no uptake of tritiated thymidine in endothelial cells.

Foci of fibrofatty infilteration.

Normal Mast cell counts

Vascular Malformations

No endothelial cell proliferation.

Contain large vascular channels lined by endothelium.

Unilamellar basement membrane.

Does not incorporate tritiated thymidine in endothelial cells.

Syndromes Associated with Hemangiomas[5]

1. Rendu-Osler-Weber Syndrome: Autosomal dominant inheritence, multiple telangiectesias, occasional GI tract involvement, occasional CNS involvement.
2. Sturge-Weber-Dimitri Syndrome: Non-inherited and nonfamilial, portwine stain, leptomeningeal angiomas.
3. Kasabach-Merritt Syndromes: Thrombocytopenic purpura associated with hemangioma, consumptive coagulopathy, microangiopathic hemolysis, intralesional fibrinolysis.
4. Maffucci Syndrome: Hemangiomas of the mucous membranes, dyschondroplasia.
5. Von Hippel-Lindau Syndrome: Genetic transmission variable; hemangiomas of the cerebellum or the retina, cysts of the viscera.
6. Klippel-Trenaunay-Weber Syndrome: Port-wine stain, angiomatosis of the extremities.

Complication of Hemangiomas

The hemangiomas may exhibit ulceration with or without secondary infection and hemorrhage. Hemangiomas of neck and laryngeal region can obstruct the airway, while the lesions of ocular region can lead to dimness of vision, strabismus, or astigmatism.

Differential Diagnosis

It is most important in the differential diagnosis to distinguish capillary hemangioma from angiosarcoma. Capillary hemangiomas, whether deep or superficial, do not develop the freely anastomosing sinusoidal pattern encountered in the most well differentiated angiosarcomas, or they possess nuclear pleomorphism and

hyperchromatism. In addition, the location of the lesion is important. Most superficial angiosarcomas are located in the scalp of elderly patients and angiosarcoma of the deep soft tissue is quite rare; therefore, a vascular tumor of skeletal muscle is statistically more likely to be benign than malignant.

Treatment

The therapy of hemangioma of the oral mucosa depends upon several factors, including the age of the patient, size of the lesion and the general clinical character.

- Most hemangiomas involute till teenage, so no treatment needed.[21]
- Patients who require treatment can undergo conventional surgery, laser surgery, or cryosurgery.[21]
- Larger lesions that extend into muscles are more difficult to eradicate surgically, and sclerosing agents such as 1% sodium tetradecyl sulfate may be administered by intralesional injection.[21]
- These agents result in postoperative pain, and the patient must be managed with a moderate-level analgesic such as oxycodone or aspirin with codeine.[21]
- Cutaneous port-wine stains can be treated by subcutaneous tattooing or by argon laser.[21]

Conclusion

Vascular lesions presenting as proliferations of vascular channels are tumor like hamartomas when they arise in childhood; in adults (particularly elderly persons), benign vascular proliferations are generally varicosities. Most of the hemangiomas involute with time but sometimes, it needs intervention.

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