

A REVIEW ON THE HISTOLOGICAL TYPES OF HEMANGIOMA

ABSTRACT

Even after the approved classification of congenital vascular tumours/malformations which was first published by Mulliken and Glowacki, in the year 1982, there is still a significant amount of confusion to categorize hemangiomas and vascular malformations. Hemangiomas are considered to be true, benign neoplasms arising from endothelial cells and must be clearly differentiated from localized defects of vascular morphogenesis, i.e., vascular malformations.

Keywords: hemangioma, capillary, spindle cell variant

INTRODUCTION

Hemangiomas are benign tumours which arise from proliferation of endothelial cells surrounding blood filled cavities. Hemangiomas remain to be the most commonly occurring benign tumours of infancy and childhood. Mostly they present from birth and start to increase in size initially, but majority of such tumours, ultimately regress spontaneously. Incidence of this group of tumour is seen more in females as compared to males. Eighty percent of hemangiomas occur as a single lesion, but 20% of the affected individuals **will** have multiple **tumours**. **Hemangiomas** of the small bowel, lungs, liver and mucous membranes can occur in inherited Osler-Weber-Rendu disease.

There are many types of hemangiomas, and they can occur throughout the body, including the skin, muscle, bone, and internal organs though skin is the most common site for hemangioma.

They are a group of vascular lesions with different clinico-pathological subtypes, with their clinical behavior varying with the stage of the **tumour** as well. As such, they can and do have a varied clinical, imaging and pathological appearance according to the location of the **tumour** and also the stage at which the patient is seen.

DISCUSSION:

When considering the diagnosis of hemangioma, there should be definite elicitation of endothelial cell hyperplasia, in order to avoid mimickers like vascular malformations. Malformations of blood vessels are caused by derangement in embryogenesis and vascular genesis and thus the morphological defects would be localized.^(1,2) Even by the nomenclature,

hemangioma with “oma” as its suffix mean proliferation of cells causing a tumour which is out of context to be used in malformations.⁽³⁾

Considering the age of onset, in infancy and childhood, hemangioma is considered to be the most commonly occurring benign tumour of the soft tissue origin. Girls are affected in a higher frequency, with preponderance to light skin colour, twins, premature infants and those born to women of higher age order during their pregnancy. These are typically grouped under two terminologies as congenital hemangiomas and **infantile hemangiomas**.⁽⁴⁾ Location wise, head & neck appears to be most common (60%), the next common being the torso (25%) and the extremities (18%).⁽⁵⁾

Vascular anomalies on the other hand, are congenital lesions of abnormal vascular development. Vascular lesions were majorly classified in 1982 by Mulliken and Glowacki into vascular tumours which grow by cellular hyperplasia and vascular anomalies which lack proliferating capacity.⁽⁶⁾

A hemangioma presents on the skin and mucosa, morphologically having a typical appearance of macules that are paler with fine, thread-like telangiectasia on the surface of the lesions.

Hemangioma may also appear as a soft mass, smooth or lobulated and sessile or pedunculated and may vary in size from few millimeters to several centimeters.

According to Enzinger and Weiss, hemangiomas are broadly, histologically classified into capillary, cavernous and miscellaneous forms such as verrucous, venous and arteriovenous hemangiomas.

Capillary **hemangiomas** include salmon patch, port wine stain, strawberry hemangioma.

Strawberry hemangioma lesions can appear from birth or can be noticed within a few weeks of infancy. They develop into a raised dimpled lesion. The lesions grow in the site as the child grows and then they will start regressing in size. This process can take up to 10 years.^(7,8)

Salmon patches are pink or red, flat, irregularly shaped patches that appear on the child face or the back of the neck. They are always present at birth.

Port wine stain or nevus flammeus appears at birth. They ordinarily persist throughout life. They appear anywhere on the body particularly on the neck and upper trunk. Early stains are usually flat and pink in appearance as the child matures the color may deepen to a dark red or purplish in color.

Capillary hemangiomas are further sub classified as juvenile hemangioma, pyogenic granuloma and epithelioid hemangioma. Capillary or telangiectatic hemangioma microscopically consist of

numerous tubules lined by endothelium and surrounded by cellular intercapillary tissue of varying thickness and that of cavernous hemangioma consist of blood channels which are irregular and more widely dilated. There are different types of capillary hemangiomas with different histological findings.

Pathologically, a capillary hemangioma progresses from dense endothelial cell proliferation, denoting the early phase of disease, to a clearly formed mass of capillaries, arranged as lobules in the later phases of disease. There is a resemblance to pyogenic granuloma in the later stages of capillary hemangioma, when the former is devoid of inflammatory processes.

Pyogenic granulomas are nothing but capillary hemangiomas that present as fast growing, red, pedunculated lesions on the skin, gingival or oral mucosa. Histologically there is vascular proliferation, edema and inflammation noticed. The epidermis is thinned out and sometimes ulcerated associated with acanthosis and hyperkeratosis. The most striking feature is the so called vascular (capillary) lobule which is a central branching vessel, surrounded by hypercellular proliferation of newly formed endothelial and perithelial cells. ^(9,10)

Microscopically, dilated vessels, containing papillary proliferation of plump endothelial cells without atypia is characteristic of *Masson's hemangioma* which often occurs on fingers or as hemorrhoids. Intravascular papillary endothelial hyperplasia (Masson's hemangioma) may be associated with preexisting pyogenic granuloma or hemangioma or in vascular malformation of blue rubber bleb nevus syndrome. It may also be secondary to trauma or be superimposed on cavernous hemangioma that mimics other benign or malignant vascular proliferations such as angiosarcoma or Kaposi sarcoma.

Spindle cell hemangiomas are slowly growing, vascular tumours, appearing as solitary or multiple, dermal or subcutaneous nodules with preferences for the distal extremities such as hands and feet. They occur in middle aged adults commonly but can appear at any age and even have been reported to be presented at birth.

There is one variant of hemangioma with distinct predilection to distal extremities which is spindle cell hemangioma. Microscopically the lesions shows thin-walled vessels lined by flattened endothelial cells and between these vascular spaces were spindle, round to epithelioid cells which appeared vacuolated, within loose fibrous connective tissue. These features are suggestive of a spindle cell hemangioma. CD34 immunohistochemical study was also done to mark vascular distribution. The presence of thrombi and plump endothelial cells are features that differentiate spindle cell hemangioma histologically from Kaposi sarcoma and intravascular papillary endothelial hyperplasia.

There is a specific type of hemangioma called the infantile hemangioma, which arise from hematopoietic progenitor cells.

Hemangiomas may mimic other lesions clinically, radiographically, just like they do histopathologically.

The differential diagnosis of hemangioma includes pyogenic granuloma, chronic inflammatory gingival hyperplasia, epulis granulomatosa and even squamous cell carcinoma.

Hemangiomas can also be termed as superficial, deep or compound, according to their extent of lesion.

A superficial hemangioma is red and nodular with no subcutaneous component while deep **hemangioma** presents as a protrusion with an overlying bluish tint or telangiectasia and compound hemangiomas have both superficial and deep components.

Abnormal levels of matrix metalloproteinases (MMP) and proangiogenic factors such as vascular endothelial growth factor (VEGF), basic fibroblastic growth factor (b-FGF) and transforming growth factor beta1 (TGF-beta1) play important role in **hemangioma** pathogenesis.⁽¹¹⁾

Although different types of hemangiomas show different clinical and histopathological findings, hemangiomas are benign (noncancerous) conditions.

Hemangiomas usually fade gradually overtime and may not require treatment. The treatment is based primarily for cosmetic reasons. However, hemangiomas that may be disfiguring or that which are located at sites causing impairment (eyelids, **airway**) **require** early medical treatment intervention by beta blockers, corticosteroids and laser therapy.

Only 10 to 20% require surgical intervention because of their size, location and their behavior. Different interventional procedures used in treating hemangiomas include microembolization, radiation, cryotherapy, sclerosing agents and surgical excision.

CONCLUSION:

Any subtype of hemangioma requires early detection and diagnosis to avoid any complications at a later date. There are many treatment options for hemangioma including both medical and interventional, where in surgical excision is the of the gold standard treatment in warranted cases.

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