Vascular anomalies

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Vascular anomalies: are congenital anomalies caused by abnormal growth of blood vessels, leading to masses originating and consisting of blood vessels with variable shapes. Vascular lesions in the head and neck region can result in significant cosmetic problems for the patient, and some may lead to even serious life threatening hemorrhage and other complications.

Vascular anomalies should be differentiated from congenital nevi. Vascular anomalies are formed of blood vessels filled with blood so their color is related to blood with different shades, while nevi are formed of melanin cells so their color is shades of melanin. This diffentiation is important as congenital nevi are premalignant and may lead to malignant melanoma but vascular anomalies are not premalignant.

Classification of vascular anomalies:

In the past, there has been confusion regarding the proper nomenclature for vascular lesions.

1-According to morphology:
(Salmon patch, strawberry hemangioma, port wine stain, cherry vascular anomalies)

2-According to the diameter of the blood vessel:
Thin small channel: capillaries wide cavernous
Mixed: cavernous and capillaries

3. In 1982, Mulliken and Glowacki "biologically classified" the vascular anomalies based on their clinical behavior and endothelial cell characteristics into two groups: hemangiomas and vascular malformations.

Now we should depend only on THE LAST BIOLOGICAL classification and ignore the previous classifications because in Mulliken biological classification the 2 sub-types of vascular anomalies differ regarding prognosis, outcome and management.

Group 1: Hemangiomas:

- The suffix **(–OMA)** in **(HAEM- ANGI- OMAS**) is due to the tumor like behavior of hemangiomas. **In fact they are not real neoplasms and they are also not premalignant lesions.**
- They are the most common tumors of the head and neck in infancy and childhood, comprising approximately 7% of all benign soft tissue tumors.
- The definition of hemangiomas is restricted to vascular anomalies caused by "endothelial proliferation".

Characteristics of endothelial cells in hemangiomas:

- Hemangiomas consist of young endothelial cells: these cells are: (Plump, active cells with high mitotic activity; they have high number of mitotic figures indicating division of endothelial cells.
- They have receptors to mediate cellular proliferation. In between the endothelial cells there are mast cells.
- These cells are considered as embryonic cells with short doubling time.

HEMANGIOMAS HAVE 2 PHASES:

Hemangiomas start de novo or as tiny lesions at the age of 3-4 weeks. This is contrary to the vascular malformations which appear at birth.

- **Proliferative phase: relatively rapid early growth until approximately 6 to 12 months of age.**
- **Involution phase or regression phase: follows the proliferative phase and lasts till 5 to 9 years of age. It is characterized by a decrease in size and fading of the red color (mottling)**

In the proliferative phase mast cells will increase in number playing a role in neoangiogenesis, so the lesion will expand and grow rapidly till the age of one year, then the lesion will stabilize, then starts to involute characterized by a decrease in size and fading of color (mottling).

Clinical picture of hemangioma

1. Natural history: This is important for diagnosis of hemangiomas

- **Appear or start as small lesions at the age of 3-4 weeks.**
- **Grow rapidly to reach their maximum size at the age of one year.**
- **They usually involute and disappear either completely or remain as a thin fibro-fatty tissue.**
- **The process of involution is normally slow and will not be completed until the age of 5 to 9 years.**

2. They are the most common tumors of infancy and childhood, comprising approximately 7% of all benign soft tissue tumors.
3. Female to male is 3:1.
4. More common in pre-mature babies.
5. **80% are solitary, 20% are multiple.**
6. Most of them (60%) are in the head and neck, less in trunk and less in the extremities.

Treatment of hemangiomas

- **As they resolve spontaneously, they are usually managed by expectant observation: follow up to check for involution and to check for possible complications.**
- **If it is hemangioma you reassure the family, it will involute and resolve spontaneously.**
- **Treatment is indicated when they are complicated.**
- **First line of treatment is by systemic steroids or beta blockers (propranolol) which induce involution of the lesion.**
- **Other methods include LASER or surgery.**

In summery, hemangioma is one of the 2 divisions of vascular anomalies, they form blood vessels, multiply, proliferate, involute and disappear spontaneously, treatment is by observation unless they are complicated; in this case the first line of treatment is by systemic steroids or by beta blockers (propranolol)
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Complications of hemangiomas:

1) Obstruction: hemangioma can grow in the eyelid obstructing the vision leading to amblyopia (lazy eye). They may also obstruct airway or auditory canal.
2) Bleeding.
3) Large hemangioms may entrap platelets leading to thrombocytopenia, this is called Kassbach-Merit syndrome.
4) Skeletal distortion.
5) Congestive heart failure due to multiple hemangiomas.
6) Ulceration and infection.

Group 2: Vascular malformation:

1. They are structural abnormalities resulting from errors in the morphogenesis of embryonic vessels between weeks 4-10 weeks of gestation.
2. Almost always sporadic.
3. They appear at birth and their growth is parallel to the growth of the child.
4. They are formed of mature endothelial cells which have normal turnover rate throughout their natural history. These cells have no receptors and no mast cells between them.
5. Vascular malformations can be one of tow types: either high flow or low flow (capillary, venous, lymph or combined).
6. Never goes spontaneously as hemangiomas and they do not respond to steroids.
7. It may need treatment if complicated or for cosmetic reasons.
8. Female: male is 1:1

Complications:

1-Erosion of bones leading to fractures.
2-Stealing blood from a limb leading to atrophy of distal parts.
3-Entrapment of platelets.
4-Bleeding.

Treatment:

1-Surgery
2-Laser
3-Embolization: by injecting material embolizinng the feeding artery.

Port-wine stains are capillary vascular malformation not (hemangiomas).
They are usually evident at birth.
They are facial lesions restricted to one or more of the three trigeminal sensory areas (ophthalmic, maxillary and mandibular branches).
They are flat and sharply demarcated, grow proportionally with the child.
Their surface is studded with nodules.

Port-wine stains can be part of Sturge-Weber syndrome:

1-Vascular malformations of the face with involvement of chorroid plexus and meninges.
2- The stain mainly involving the ophthalmic division.
3- Local or generalized seizures.
4- Ipsilateral glaucoma.

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