PLASTIC & RECONSTRUCTIVE SURGERY

LEC 4

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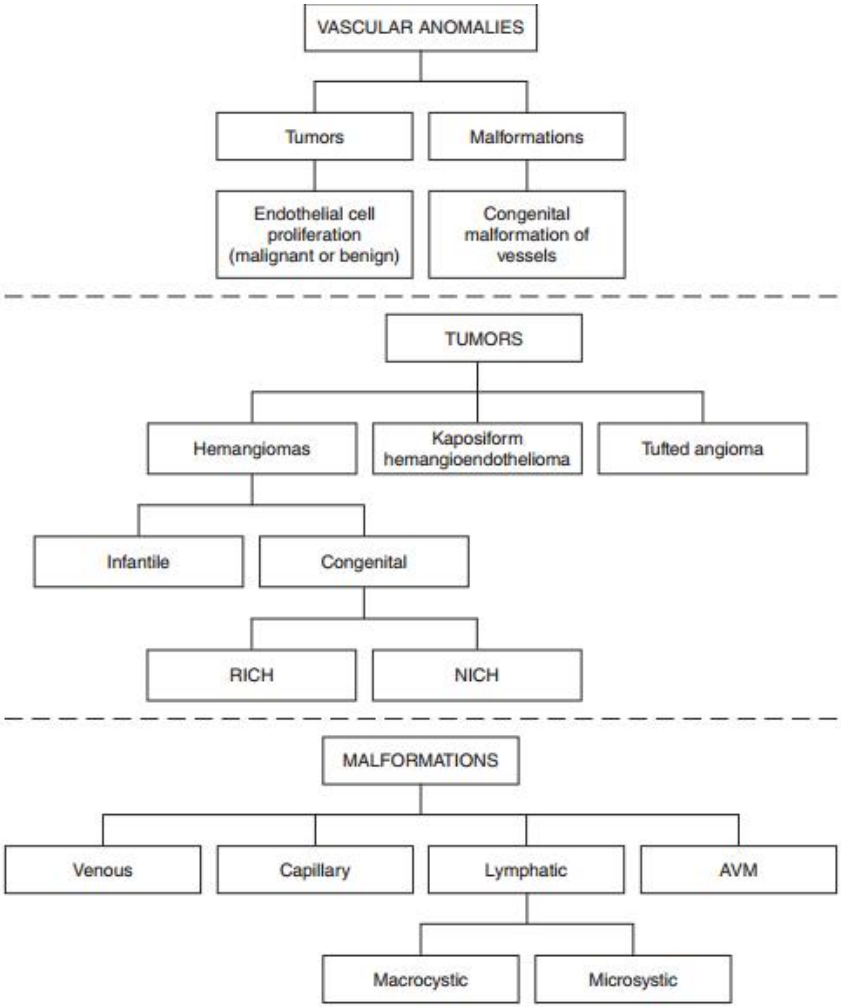
VASCULAR ANOMALIES

CLASSIFICATION:



INFANTILE HEMANGIOMA

Benign proliferation of endothelial cells which is present after birth. Most common tumor of infancy

- Incidence: 1:10 infants
- 10% of white infants and 2% of black infants.
- More common in females (3:1).
- More frequent in premature low-birth-weight infants: 23%
- **Most common location:** Head/neck region

Diagnosis

A- Mainly based on history and physical examination findings: Lesion presents after birth as cutaneous mark, such as a pale area, macular stain, telangieaatic macule, or ecchymotic spot or scratch and continues to enlarge in infancy and become bright red, tense. IH is characterized by a three-stage life cycle, consisting of the proliferating phase, involuting phase, and involuted phase

1. Proliferating phase: In typical hemangiomas, the majority of proliferation occur during a rapid growth phase in the first 6 to 8 months with a cessarion of growth by 1 year of age. the tumor is typically in its most florid crimson presentation

2. Involuting phase: the florid crimson color of tumor fades to a dull purplish hue, with increased pallor of the skin and decreased turgor of the tumor. It lasts anywhere from *2* to 10 years. In many children the involuting phase results in virtually normal skin, but in a number ofcases children with hemangiomas will exhibit residual telangiectasias, pallor, atrophy, textural changes, and sometimes residual fibrofatty tissue.

3. Involuted phase: Loose fibrofatty tissue replaces previous parenchymal tissue. Regression is complete in *50%* of children by *5* years and in 70% ofclill.dren by 7 years, with continued improvement up to 10 to 12 years of age.

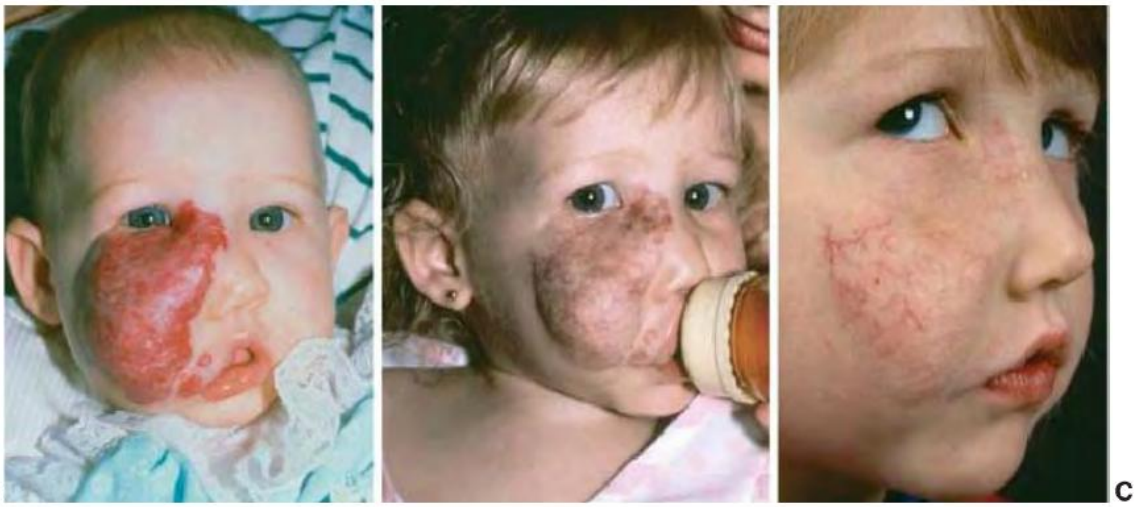


FIGURE 21.1. Hemangioma. This girl with a right facial hemangioma demonstrates the three-stage life cycle of IH, consisting of the proliferating phase ((A) age 3 months), involuting phase ((B) age 18 months), and involuted phase ((C) age 7 years).

B- Ultrasound (US). Difficult to distinguish hemangioma from arteriovenous malformation (AVM) as both are high-flow lesions. US shows shunting pattern of flow.
C- MRI with contrast (gold standard test): Especially important if suspect visceral hemangiomas

Associated anomalies/syndromes and rare presentations

1. Spina bifida occulta: Associated with lumbar hemangioma
2. PHACES: Posterior fossa anomalies, Hemangiomas, Cardiac anomalies, Eye abnormalities, Sternal cleft
3. Kasabach–Merritt syndrome: Hemangioma + thrombocytopenia (Platelet count <10,000, normal PT/PTT)

4. Cutaneous visceral hemangiomas: Multiple hemangiomas (>5) should elicit concern for visceral hemangiomas:

- **a.** Congestive heart failure
- **b.** Hepatomegaly (intrahepatic hemangiomas)
- **c.** Anemia

Treatment

1. Observation—appropriate in most cases because total involution occurs in 50% of hemangiomas by 5 years, in70% by 7 years, and in .90% by 9 years.

- **a.** Reassurance of parents and patients is critical
- **b.** Take serial photographs to monitor progress

2- Local Wound Care for ulcerated hemangioma

3- Medical Management: Pharmacologic therapy is indicated for hemangiomas that threaten function or result in local complications.

- Corticosteroids: Steroids can be administered intralesionally or topically for small, well-localized tumors *or* orally for large and/or aggressive hemangiomas that may impair function, cause severe disfigurement, or are life-threatening
- Propranolol
- Interferon Alpha
- Vincristine

4- Lasers: Pulsed dye laser (PDL) can be effective in treating relatively flat, superficial hemangiomas

5-Surgery:
Depending on circumstances, surgery is typically delayed until the child is of school age and beginning to experience psychological consequences.
Urgent surgery is performed during proliferative phase if lesion threatens important structures or function.

- **Visual obstruction:** During the first year of life for a period of 1 week can result in deprivation amblyopia or anisometropia
- **Nasolaryngeal obstruction:** Hemangiomas in a "beard distribution" proliferating in subglottic airway potentially life-threatening; requires aggressive treatment with propranolol, steroids, surgery, or laser
- **Auditory canal obstruction:** Can result in mild to moderate conductive hearing loss

Complication:

Bleeding: Usually responds to pressure and rarely requires surgical ligation
Ulceration: Most common complication, in ,5% of patients
Infection: Rare but more common with intraoral or perianal hemangiomas
Kasabach-Merritt syndrome: Profound thrombocytopenia with kaposiform hemangioendothelioma
High-output heart failure: Associated with very large liver/visceral hemangiomas, usually seen with hemangiomatosis
Skeletal distortion: Bones are typically deformed by pressure from hemangioma; bony hypertrophy is rarely seen.
Emotional/psychological distress: Children become aware of their deformity around age 5; treatment should be more aggressive at this time.

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VASCULAR MALFORMATIONS

Unlike hemangiomas: Present at birth, grow proportionately with the child, do not regress and equal M:F ratio.

Diagnosis:

1. Clinical history and physical examination
2. Imaging
a. US with Doppler: Differentiates slow-flow from fast-flow lesions
b. MRI with contrast: Gold standard test. It Provides details on the anatomic distribution of the lesion and differentiate the types of vascular malformation
c. Arteriography: Invasive study

Capillary malformations (port wine stain):

Incidence: 0.3% of newborns. Most common location is on the face
3:1 female:male ratio
They usually present at birth as pink or red intradermal discolorations that may involve small areas or involve an entire limb or face

Common associated syndromes: **Sturge–Weber syndrome:** facial CM in the trigeminal nerve {ophthalmic (Vl)} distribution, ipsilateral leptomeningeal, and ocular vascular anomalies and seizures.

Treatment options
a. Observation
i. Lesions do not regress
ii. Can progress to "cobblestone" appearance
b. Pulsed dye laser : 70% to 80% of patients respond with decrease in pigmentation of the lesion..
c. Surgical excision. Must be used for management of soft-tissue/skeletal hypertrophy to address contour deformity.

LYMPHATIC MALFORMATION:

1. Slow-flow lesion
2. Appearance
a. Anomalous lymphatic channels filled with lymphatic tissue which may be clustered into vesicles
b. Further classified as microcystic versus macrocystic

3. Most common cause of macroglossia, macrochelia in children. Can also cause facial asymmetry, distortion of surrounding tissue, soft tissue/skeletal hypertrophy.

4. Treatment options

a. Observation

Intralesional bleeding can be treated with NSAIDS for pain control and rest.

Antibiotics indicated for cellulitis/other infections.

b. Sclerotherapy (mainstay treatment)

c. Surgical resection

Venous malformations:

diagnosis:

Blue or purple lesions with spongy texture

Swell in dependent position

Deflate when elevated

Aching in extremity lesions

Can be hormone sensitive and enlarge during puberty and with pregnancy

Treatment:

Many lesions amenable to sclerotherapy

Extremity aching managed with compression garments, NSAIDs, analgesics

Laser treatment: Nd:YAG or argon laser

Surgical resection

Complications

•Episodes of thrombosis

•Localized intravascular coagulation that could evolve into disseminated intravascular coagulation

Arteriovenous malformation

DIAGNOSIS

Pulsatile high-flw lesion

Anatomy and hemodynamics defined by angiography

MRI useful in determining extent of lesion

Clinical aspect:

Stage 1 (quiescent): Warm pink to bluish stain

Stage 2 (expansion): Thrill and dilated venous network formation

Stage 3 (destruction): Cutaneous ulcers, necrosis, frequent bleeding

Stage 4 (decompensation): Cardiac decompensation

Treatment:

1-Preoperative medical management of any underlying coagulation defect secondary to thrombotic consumption

2-Preoperative embolization followed by surgical resection **within 72 hours**
3-**Wide local excision because recurrence rates are very high** and reconstruction with flaps often necessary

Complication:

Consumptive coagulopathy
Congestive heart failure
Local destruction of normal anatomy
Surgical bleeding.

Congenital Melanocytic Navi

Incidence:

Lesion must be present at birth to be classified as congenital.
Equal prevalence in males and females
Occurrence in all races
One percent incidence in newborns, with greater incidence in blacks (1.8%).

Classification of Congenital Nevi

Small (1.5 cm^2)
Medium (1.5 cm^2--20 cm^2)
Giant (>20 cm^2)

Clinical feature:

CMN may appear initially as a hairless, pale brown flat lesion at birth, which evolves with time to develop variegation and hyperpigmentation. Dark, coarse hair may develop during the first 1 to 2 years of life. By 10 years of age, the lesions often develop a verrucous texture and become more elevated, with hypertrichosis and hyperkeratosis. Nodule formation typically represents benign neurotization in the nevus. CMN may be associated with multiple smaller satellite lesions dispersed over the trunk, extremities, or head and neck.
The most common anatomic location for a giant CMN is the posterior trunk, followed in frequency by the extremities and head and neck.Giant nevi may be found in specific anatomic patterns, such as the "bathing trunk" and "glove-stocking" distributions.

COMPLICATIONS:

1-Malignant Transformation

2- Neurocutaneous Melanosis

Treatment:

Intervention, if performed, should be done early in life, as the risk of malignant transformation is greatest in the first decade of life (starting intervention at 6 months ofage, with completion of staged surgeries before school age).

Treatment options for CMN include nonexcisional and excisional methods:
Nonexcisional methods: Chemical peels, lasers, curettage, and dermabrasion have been reported as treatment for CMN.
Excisional methods: include primary excision and closure, serial excision, skin grafting, tissue expansion and skin substitutes. To address the malignant potential, only complete excision of the nevus c:an be recommended as a solution.

Differential Diagnosis

The following lesions can sometimes be confused with CNN:
Café au lait spot.
Nevus spilus
Epidermal nevus:
Common acquired nevus
Atypical (dysplastic) nevus
Blue nevus:.
Becker's nevus
Mongolian spot
Nevus of Ota