# Hemangioma and vascular malformations

**Hemangiomas**

- Benign tumors of infancy
- Rapid growth phase
- Endothelial cell proliferation,
- Followed by gradual involution.

**Vascular malformations**

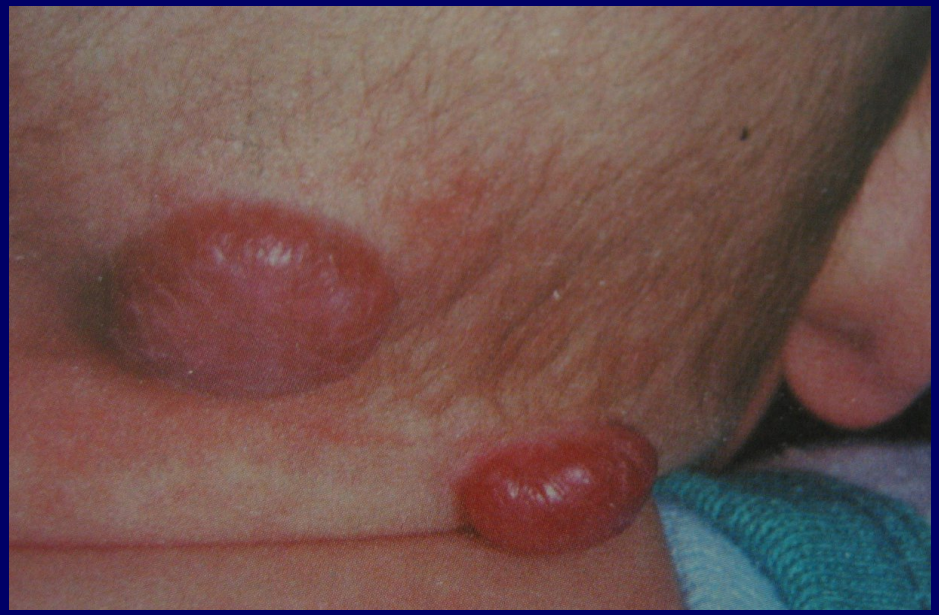
- Structural anomalies of blood vessels
- No endothelial cell proliferation
- Present at birth
- Persists for life

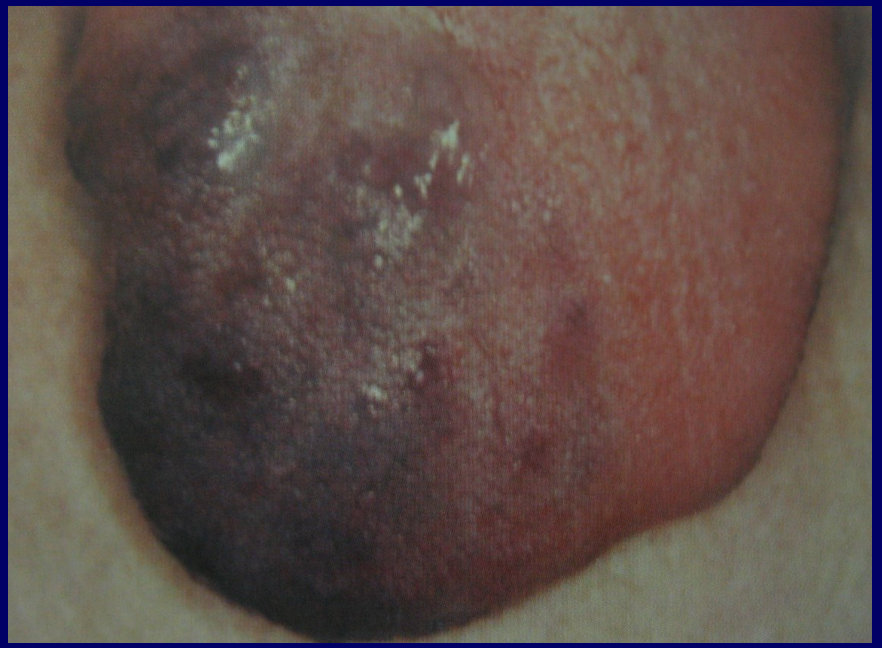
# Categories

- Type of vessel
  - Capillary
  - Venous
  - Arterial
- Hemodynamic features
  - Low flow
  - High flow

# Hemangioma

- Most common tumors of infancy
- More common in females (3:1)
- In head and neck
- Rapid development during the first few weeks faster than the growth of the infants.
- Superficial tumors of skin – raised and bosselated – bright red color – strawberry hemangiomas
- Firm to rubbery on palpation.



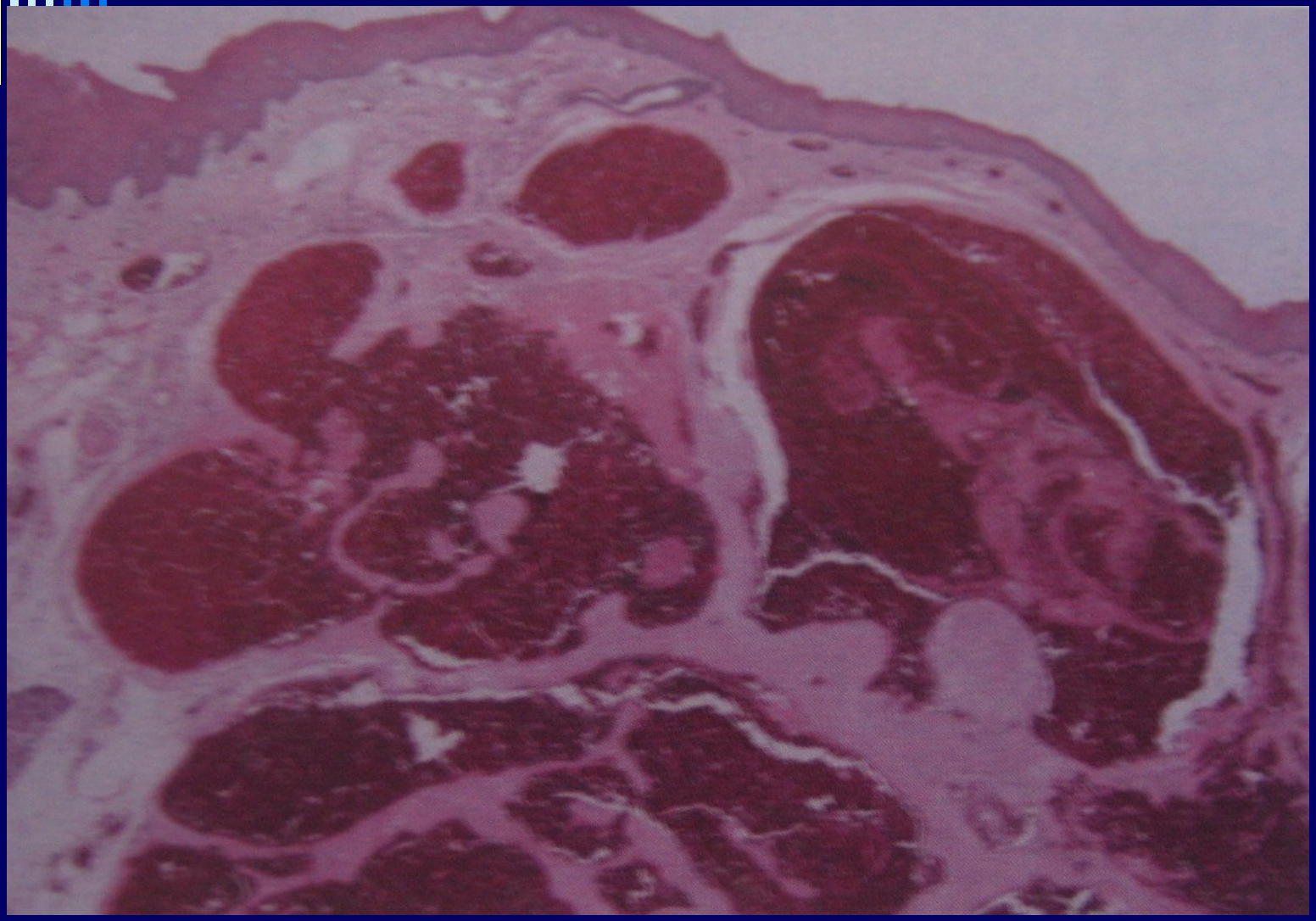


# Hemangioma

- Proliferative phase lasts for 6-10 months.
- Slows in growth and begins to involute.
- Color changes to dull purple hue
- By 5 years of age, most disappear
- Can heal to normal skin, can show atrophy, scarring, wrinkling or telangiectasia.

# H/P- Hemangiomas

- Early hemangiomas – numerous plump endothelial cells, indistinct vascular lumina.
  - Also known as Juvenile or cellular H
- As the lesion matures, endothelial cells become flattened and small, capillary-sized vascular spaces
- Hemangiomas undergo involution and vascular spaces become more dilated (Cavernous) and widely spaced.



# Complications

- Secondary infection
- Peri-ocular tumors – altered vision
- Multiple cutaneous hemangiomas – increased risk for concomitant visceral hemangiomas
- In neck or larynx – airway obstruction
- Kasabach-merritt syndrome – serious coagulopathy
  - Large extensive hemangiomas
  - Thrombocytopenia and hemorrhage

# Vascular malformations

- Present at birth, persist throughout life
- Portwine stains – capillary malformations in newborns
- Common in face
- Seen along with sturge-weber syndrome
- Pink to purple macules, grows with the patient, darkens with age and becomes nodular.

# Low flow venous malformations

- Small isolated ectasia to complex growths
- Involves multiple tissues
- Present at birth
- Blue color and easily compressible
- Grow proportionately with the patient

# Arteriovenous malformations

- High flow lesions
- Persistent direct arterial and venous communication
- Present from birth
- Fast vascular flow thru the lesion – palpable thrill/bruit
- Overlying skin is warmer to touch
- May have pain, bleeding and skin ulceration.

# Intrabony vascular malformations

- Could be venous or arteriovenous malformations
- Seen between 10-20 years of age
- Common in females
- Radiolucency within the bone
- Angiogram is helpful to confirm the vascular nature of the lesion.
- Risk of severe bleeding – spontaneous or during surgical manipulation.

# H/P – Vascular malformations

- Do not show active endothelial cell proliferation
- Channels resemble the cell of origin
  - Capillary – similar to capillary H
  - Venous – dilated spaces like venous H
- Arterio venous malformations shows thick walled arteries and veins along with capillary vessels.

# Ttt and prognosis

- Hemangiomas involute with age – follow up
- Prognosis depends on the organ of involvement
- Sclerotherapy and excision

# Sturge-weber angiomatosis

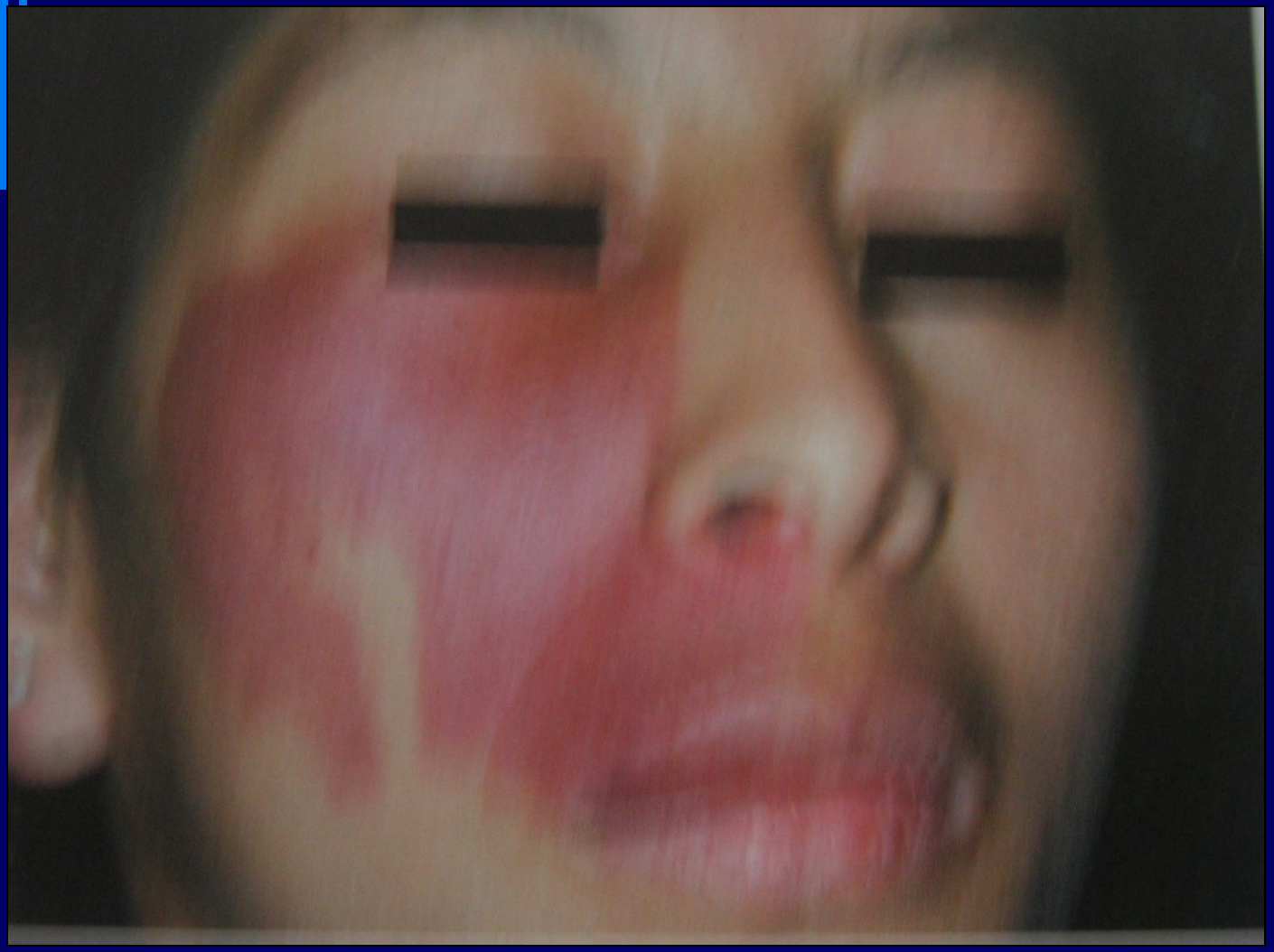
- Also called encephalo-trigeminal angiomatosis
- Rare, non-hereditary developmental condition
- Hamartomous vascular proliferation involving brain and face.
- Caused by persistence of vascular plexus around the neural tube, which normally regresses by ninth week.

# Clinical features

- Dermal capillary vascular malformation of the face – Portwine stain or Nevus flammeus – deep purple color.
- Unilateral distribution along one or more segments of trigeminal nerve.
- Involvement of ophthalmic nerve – Risk of full condition
- Leptomeningeal angiomas, convulsive disorders, retardation or hemiplegia
- Radiographs – Tramline calcifications on affected side.

# Features

- Ocular involvement
- Intra oral – affects the ipsilateral mucosa
  - Gingival hyperplasia
- H/P
  - Excessive numbers of dilated blood vessels
  - Gingival lesions can be similar to pyogenic granulomas
- Ttt and prognosis
  - Depends on the severity of the disorder



# Lymphangioma

- Benign, hamartomatous tumor of lymphatic vessels
- Developmental malformation arising from sequestrations of lymphatic tissue which do not communicate normally with the rest of lymphatic system

# Types

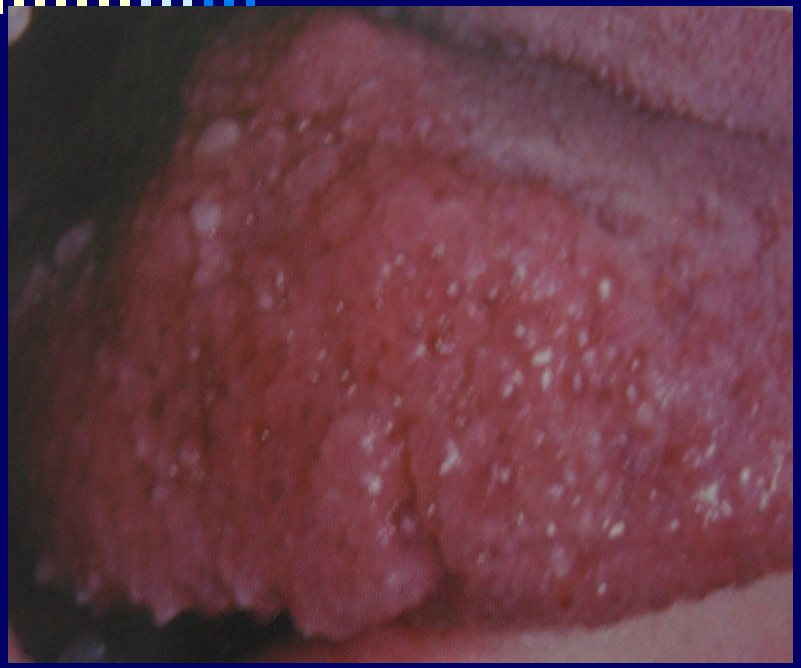
- Three types
  - Lymphangioma simplex – capillary lymphangioma – small, capillary sized vessels
  - Cavernous lymphangioma – large, dilated channels
  - Cystic lymphangioma – large, macroscopic cystic spaces
- All three types can be seen within the same lesion.
- Subtypes could be same pathology but the size of vessels is determined by the surrounding connective tissue.

# Clinical features

- Marked predilection for head and neck
- 90% develop by 2 years of age.
- Cervical L
  - More common in posterior triangle of neck
  - Soft, fluctuant masses
  - Rapid enlargement after upper respiratory tract infection

# Clinical features

- Oral L frequently occur on the anterior two thirds of the tongue
- Often result in microglossia
- Tumor is superficial in location – pebbly surface with cluster of translucent vesicles
- Called appearance of frog eggs or tapioca pudding.
- Secondary hemorrhage – purple vesicles



# Histopathology

- Lymphatic vessels diffusely infiltrating adjacent soft tissues and may demonstrate lymphoid aggregates.
- Lining endothelium is typically thin
- Spaces contain a proteinaceous fluid
- Intraoral – lymphatic vessels located just beneath the epithelial surface and often replaces the connective tissue papillae

# Hemangiopericytoma

- Rare neoplasm derived from pericytes – cells that encircle the endothelial cells of capillaries.
- In lower extremities
- Primarily in adults
- Slow growing, painless mass
- Can occur in nasal cavity and para nasal sinuses – nasal obstruction

# H/P

- Fairly well circumscribed
- Tightly packed cells surrounding endothelium lined vascular channels
- Cells are haphazardly arranged with round to ovoid nuclei and indistinct borders.
- Blood vessels with irregular branching – stag horn or antler like appearance
- Sinonasal – more of spindle cells
- Ttt – surgical excision