Disontogenetic Neoplasms

GENERAL FEATURES

D A P
UNIBA

DISONTOGENESIS

Persistance of embryonic transient tissues

- **Amartia:** irregular quantitative mingling of tissues that are normally present in an organ
- **Choristia:** germs of organ or tissue are found at an anomalous site (ectopic germs)
- **Unused embryonic germs:** they still have the potential to develop into different tissues

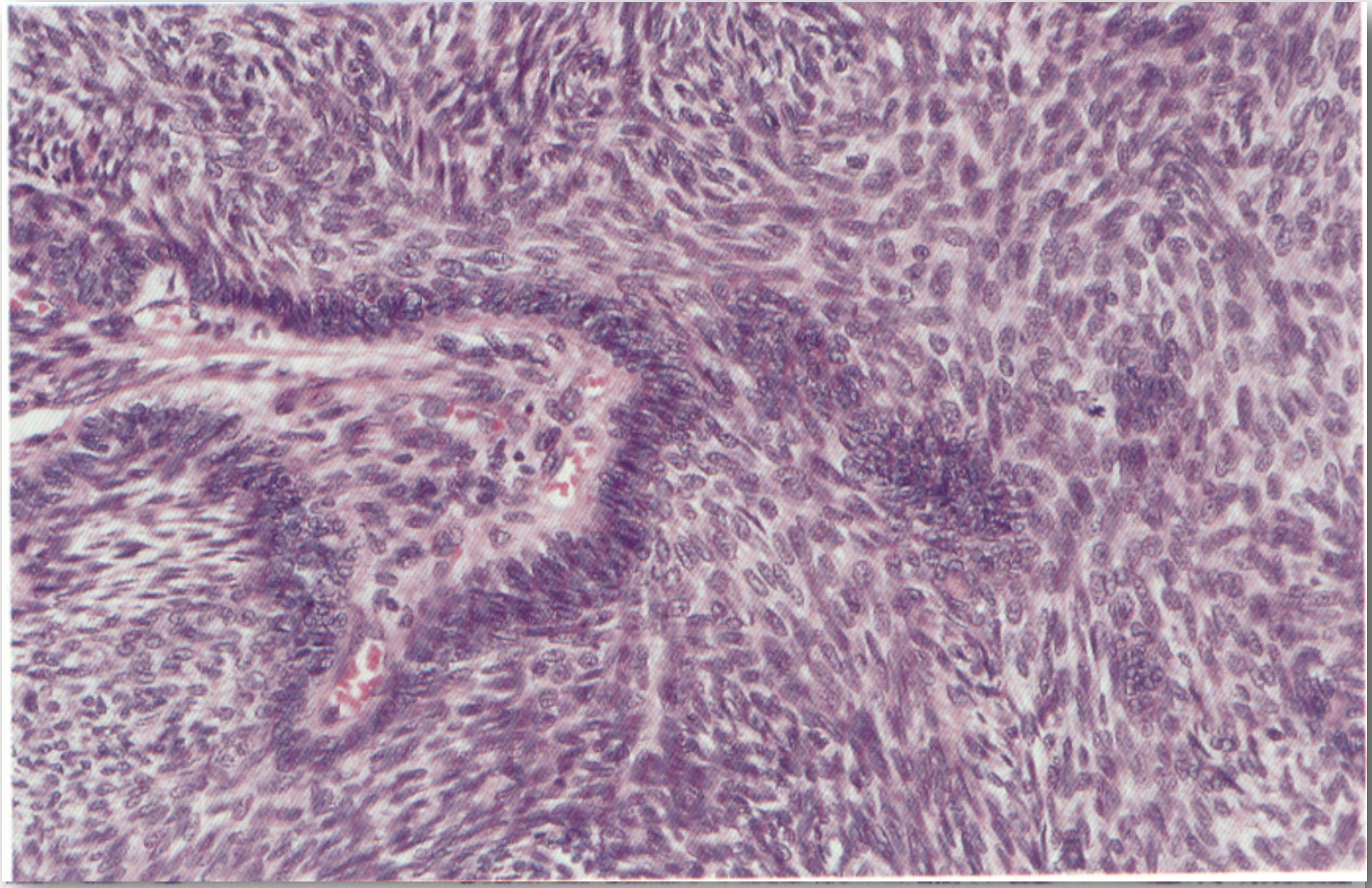
DISONTOGENETIC TUMORS

- *Neoplasms made of one or more tissues that are not fully mature and, possibly, located at an ectopic site*
- They can develop from:
  - Transient embryonic tissue
  - Amartia
  - Coristia
  - Unused embryonic germs

DYSONTOGENETIC TUMORS

Tumours from abnormal embryonic persistance

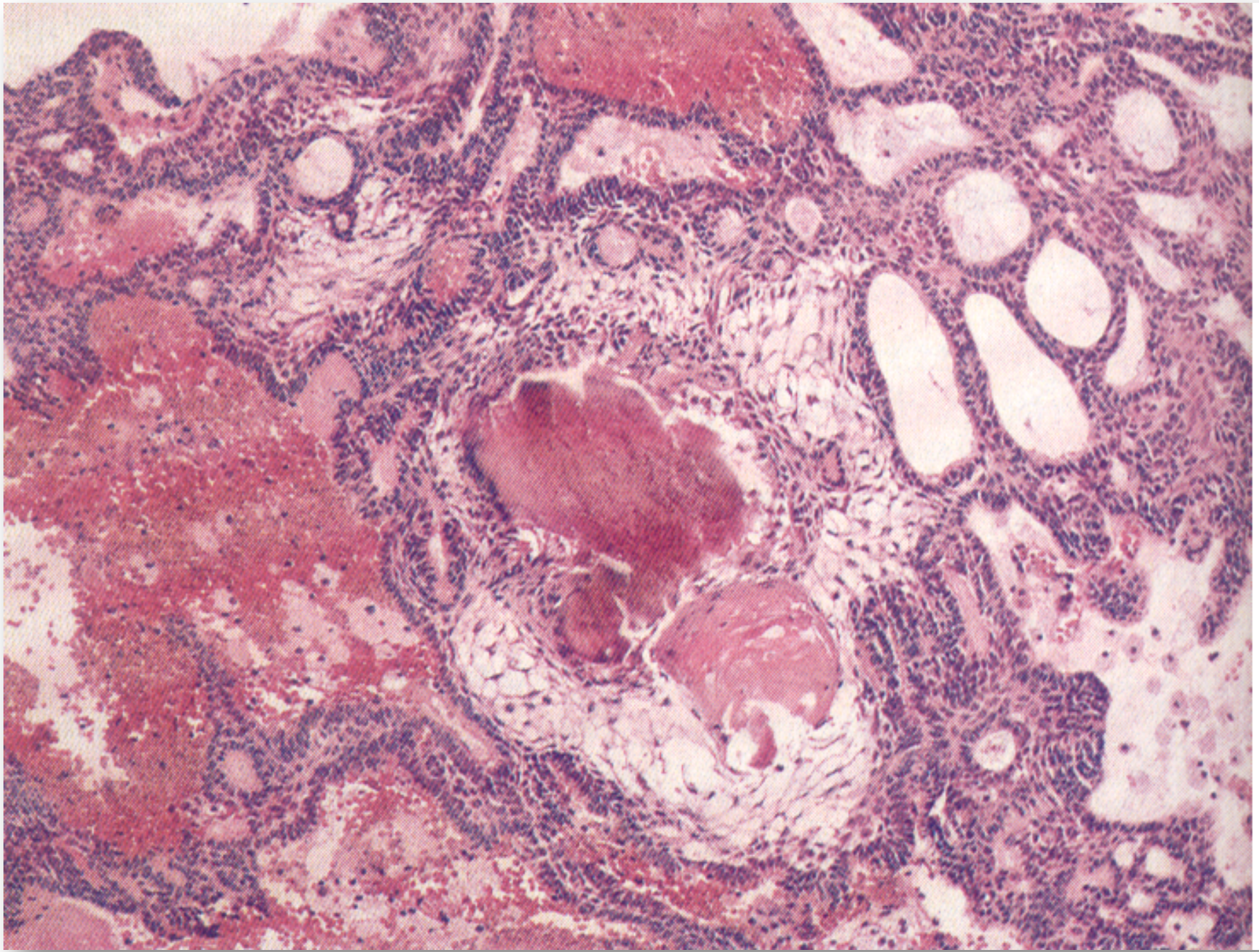
- **Adamantinoma o ameloblastoma**
  - Sex: Male, 3-4th decade
  - Paradental residues of Malassez (tooth enamel)
  - Molar region of the mandible
  - Does not contain enamel and dentin
  - Locally invasive
  - Bunches of polygonal cells (ameloblasts) (plexiform or follicular)
  - uni/multi-cystic



DYSONTOGENETIC TUMOURS

Tumors from abnormal embryonic persistance

- **Craniopharyngioma,** Erdheim's tumor or hypophyseal adamantinoma
  - Epithelial residues in the cranio-pharyngeal duct (Rathke's pouch),
  - Derived from buccal ectoderm
  - Rare benign neoplasm
  - 1st-2nd decade of life
  - Suprasellar: hypothalamic compression, III ventricle (hydrocephalus), optic and oculomotor nerves.
  - Macroscopic aspect: variable dimensions , smooth or lumpy surface, solid or cystic.
  - Microscopic aspect: cords or anastomosed epithelial islets enclosed in a stroma with few cells, palisading, necrosis, calcium deposits



DISONTOGENETIC TUMOURS

Median cysts or fistulas of the neck, aberrant thyroid, lingual goiters ecc.

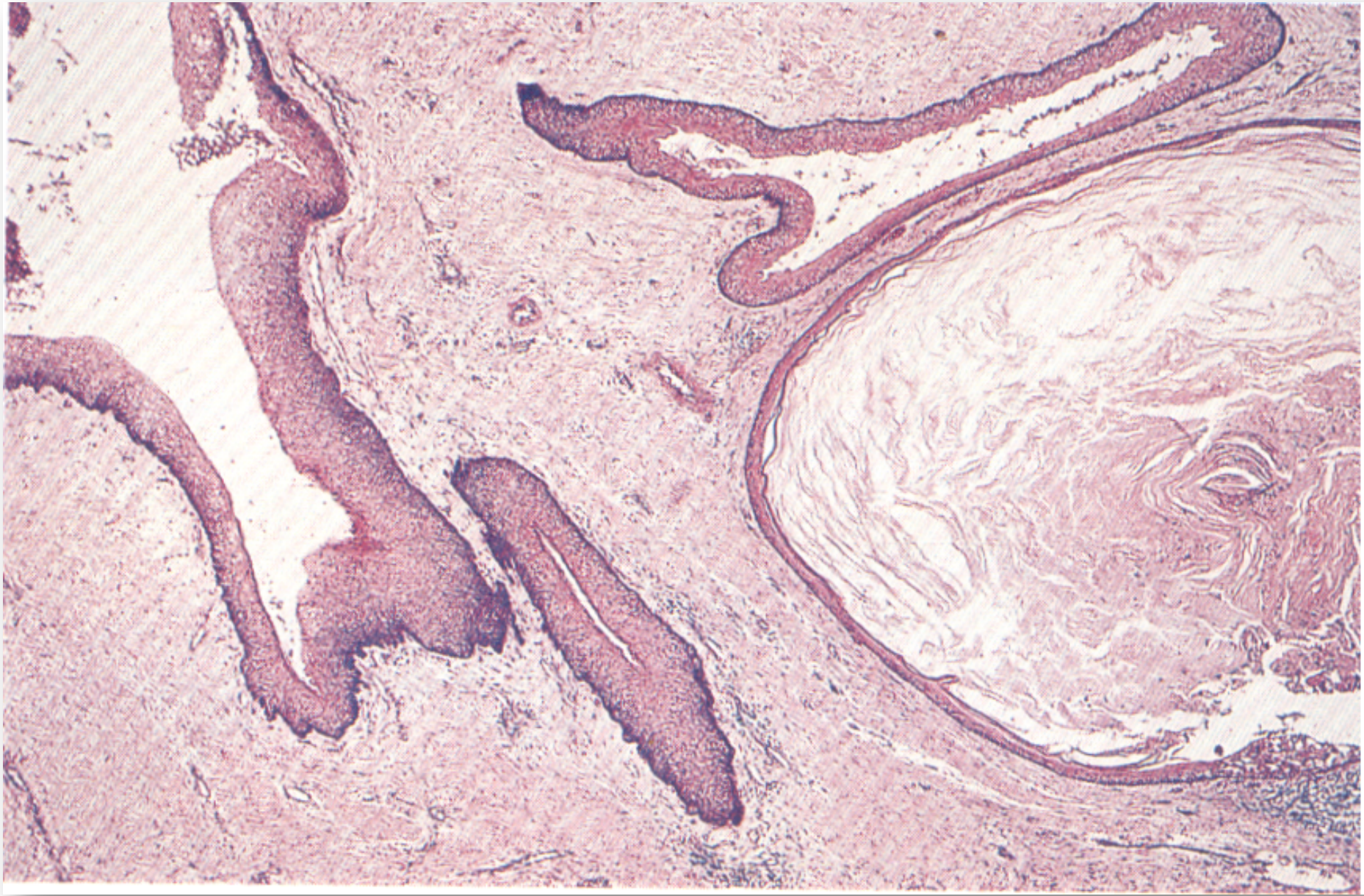
- Residues of thyroglossal duct
- Any point on neck midline (hyoid bone)
- First infancy
- Slow growth (1-2 cm)
- Covered by simple cylindrical epithelium, islets of thyroid follicles

Lateral cysts or fistulas of the neck

- Residues of epithelium of pharyngeal pouches
- Lateral region of the neck (sterno-cleido-mastoid)
- Early onset, evident after 1° decade
- Slow growth
- Covered by squamous or respiratory epithelium

Epidermoid cysts

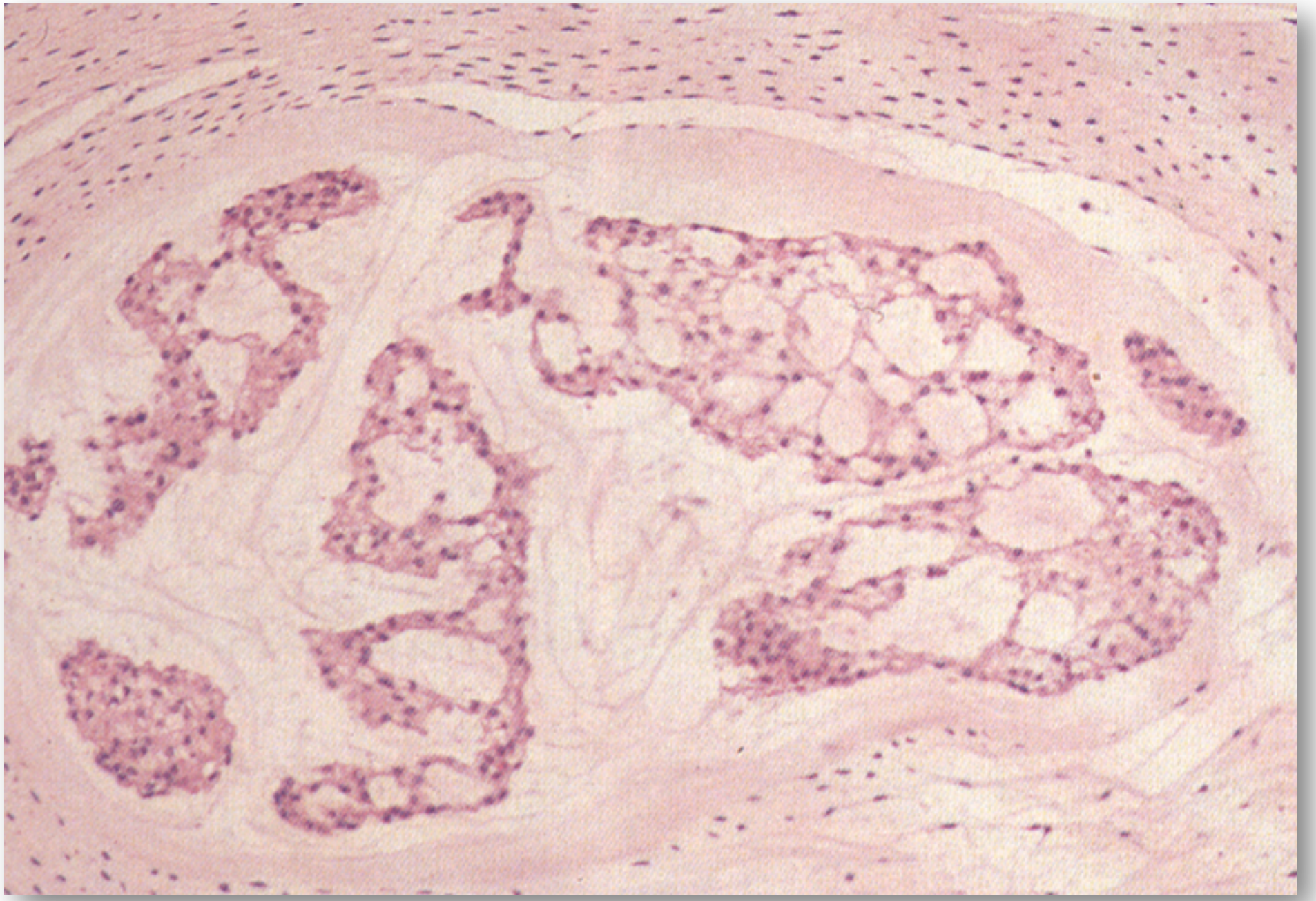
- Epidermic germ embedded in the skin
- Flat epithelial coating, sebaceous and squamous content



DISONTOGENETIC TUMOURS

Chordomas

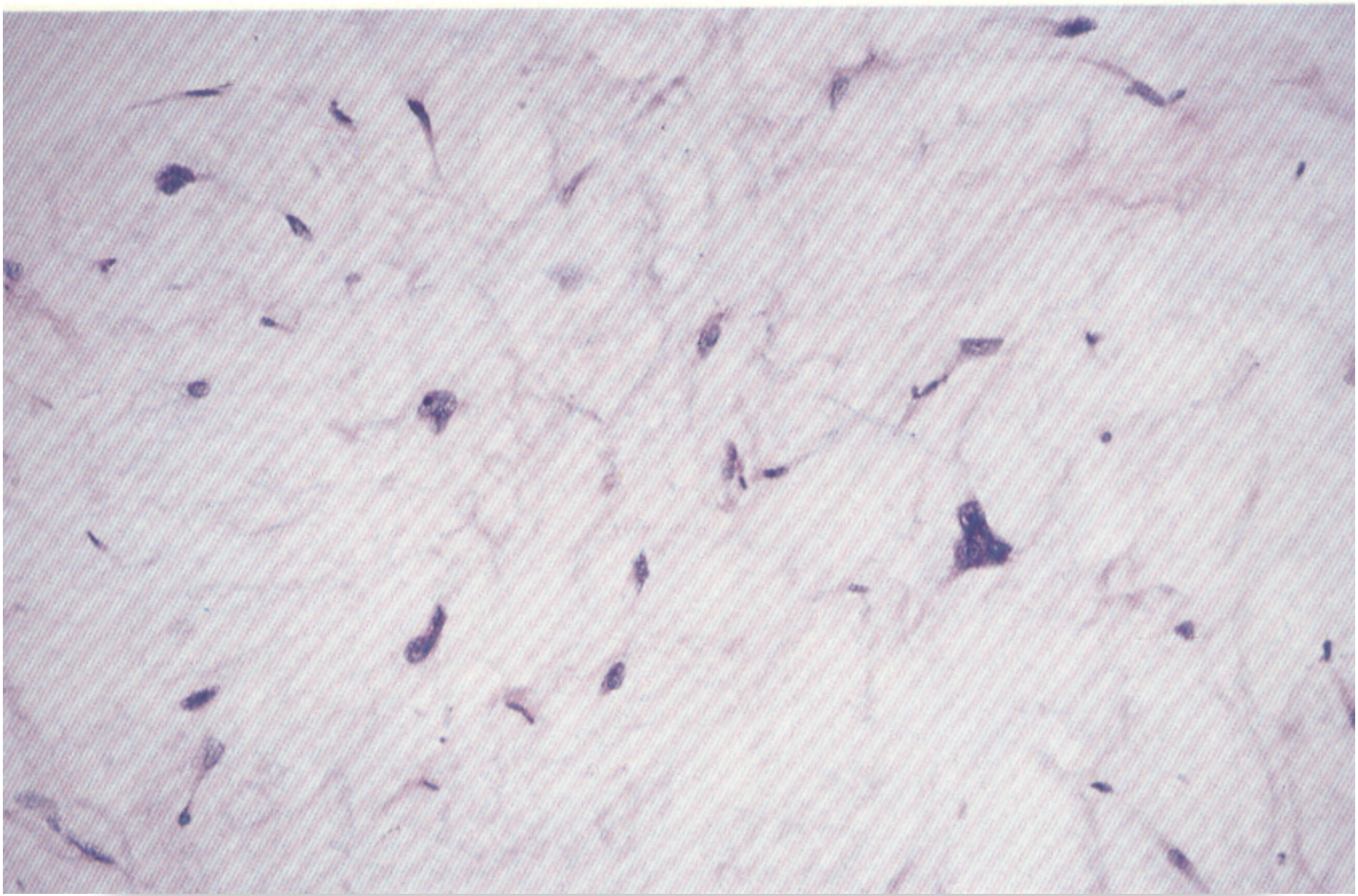
- Residues of notochord (transitory mesodermal organ)
- Cranial base (sphenoid) or sacro-coccygeal region
- Rare tumor
- Age: 30-60 , Sex: Female
- Locally infiltrating and destructive
- Slow growth and recurrent (remarkable dimensions)
- Sometimes metastasis (10 %)
- Macroscopic appearance: tender mass, traslucent, grey-pinkish
- Microscopic appearance : cords of big “vegetal” cells, with light cytoplasm ,rich of vacuoles (glycogen) and with a small nucleus, enclosed in fundamental amorphous substance



DISONTOGENETIC TUMOURS

Myxomas

- Residues of mucous, embryonic (mesenchymal) tissues
- Fusiform and stellate cells enclosed in abundant myxoid matrix
- Skin , muscle, heart, deep/internal organs



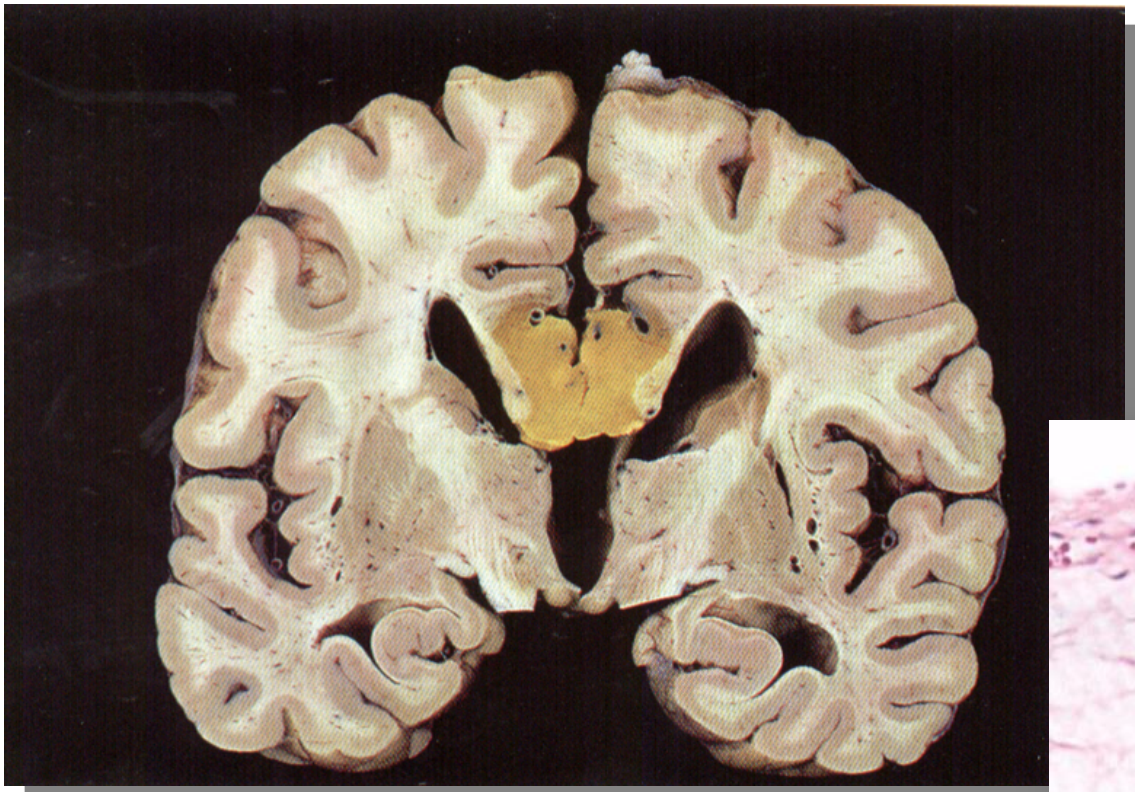
DISONTOGENETIC TUMOURS

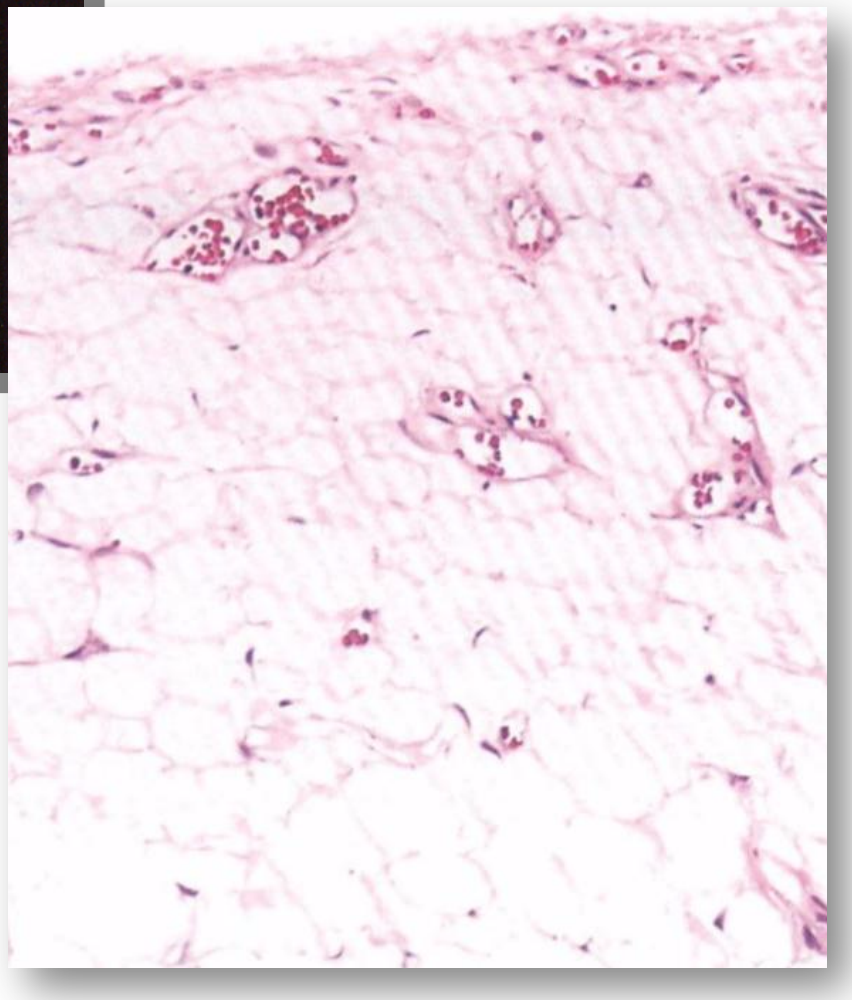
Tumors on Amartia (Amartomas)

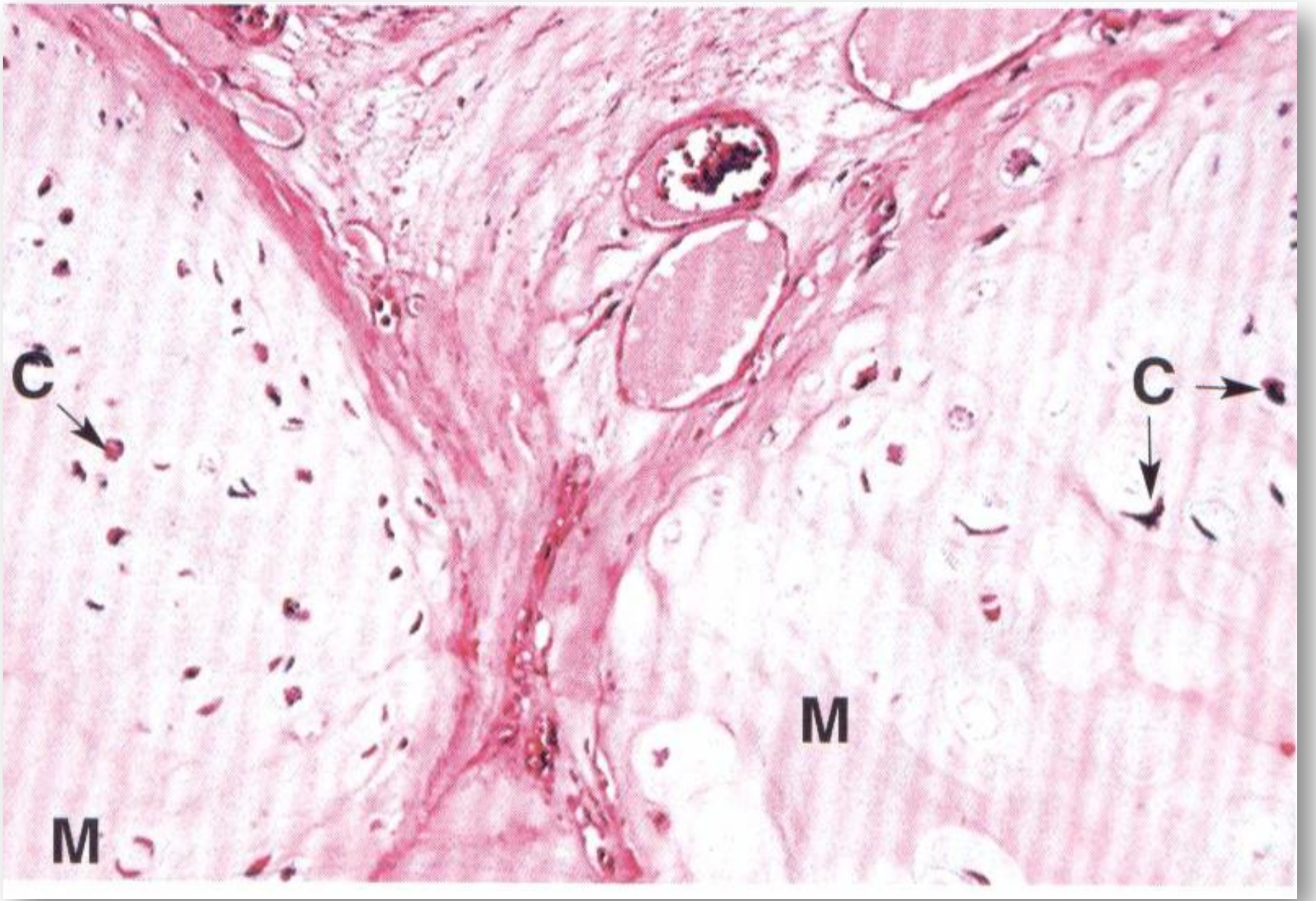
- **Lipoma**
  - Localized excess of adipose tissue
  - Unique or multiple, symmetrical, variable dimensions
  - Armpits, shoulders, buttocks ecc.

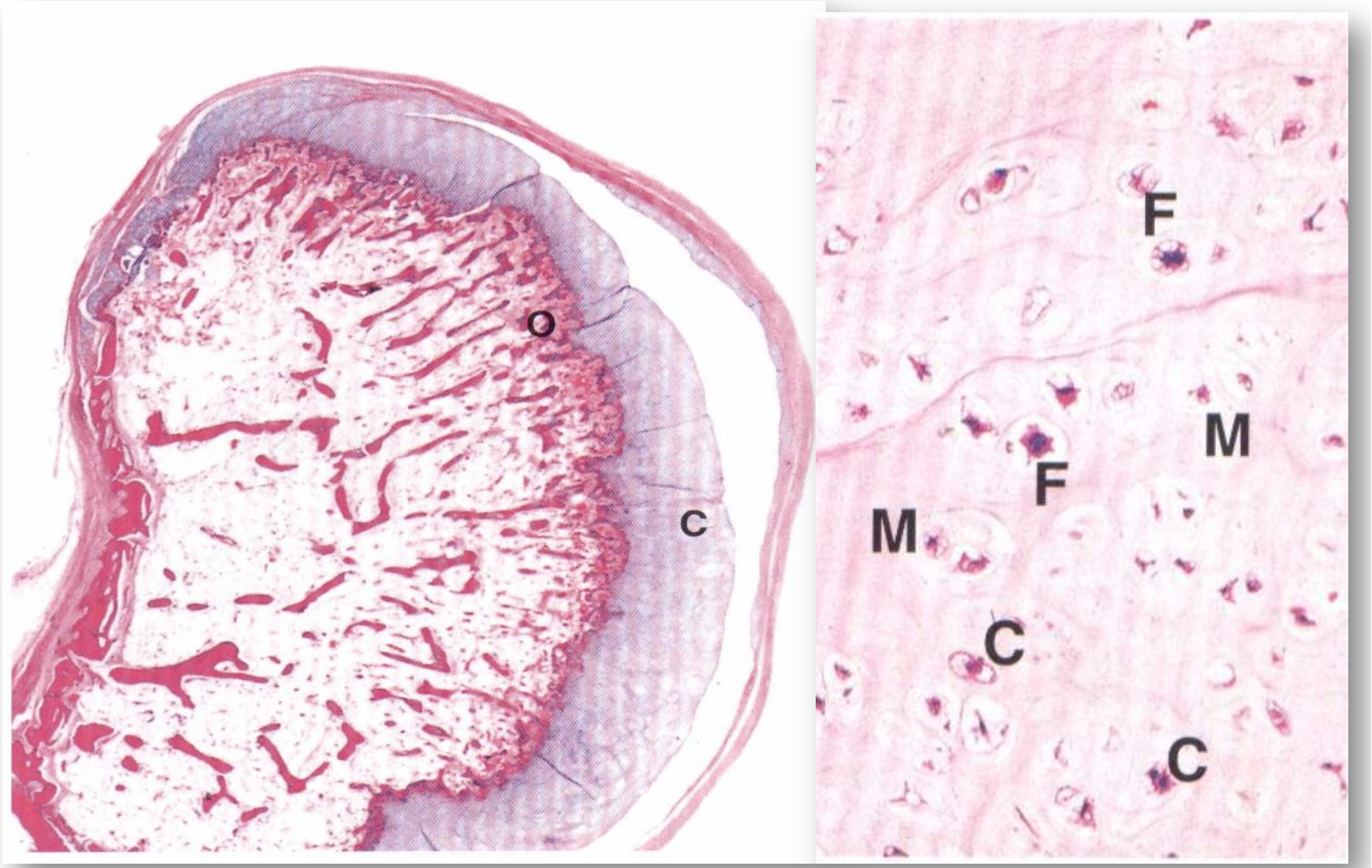
Condroma

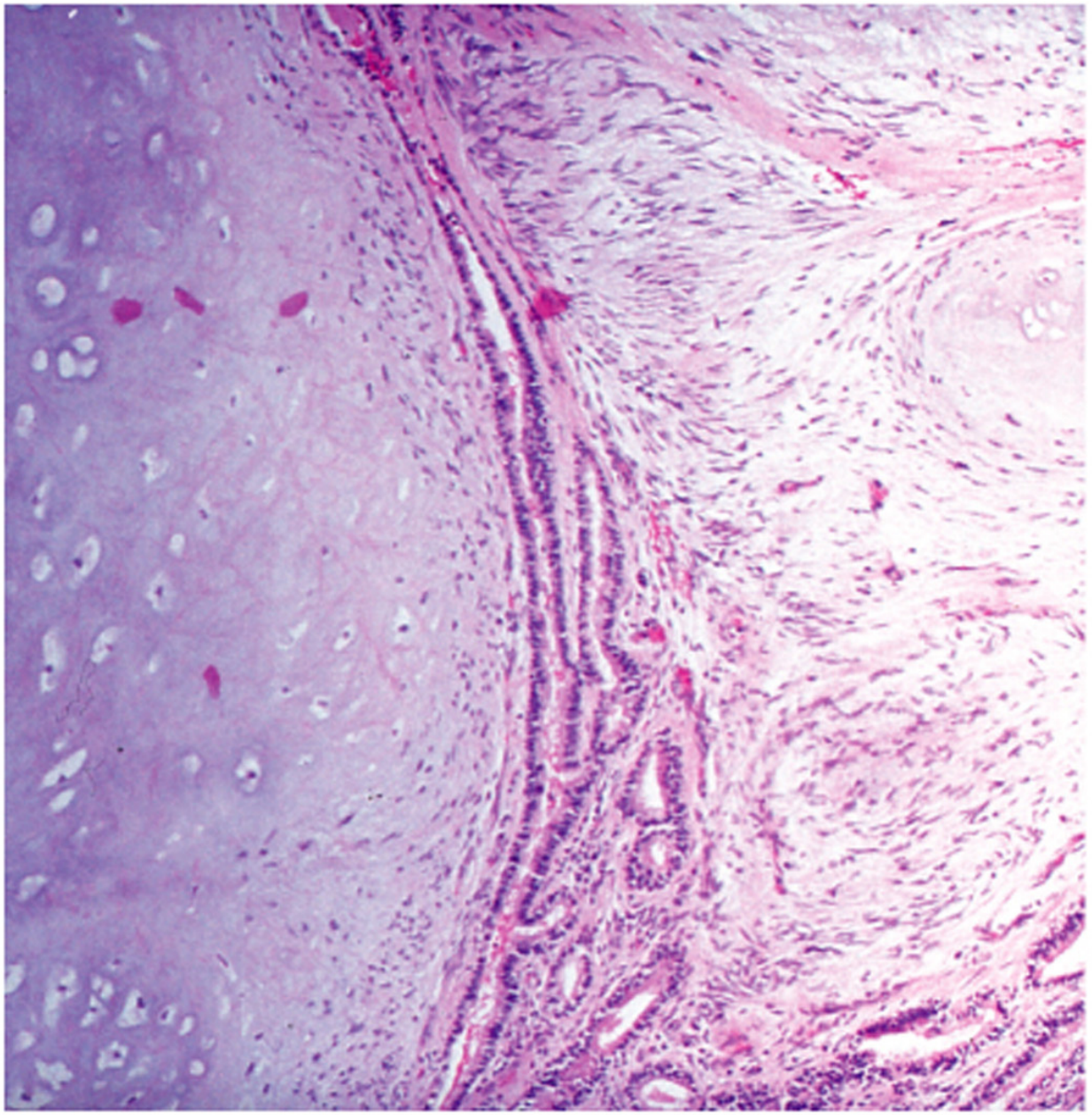
  - Accumulation of cartilaginous tissue on congenital predisposition
  - multiple and simmetrical, flat and short bones






C
C
M
M


O
C
F
M
F
M
C
C

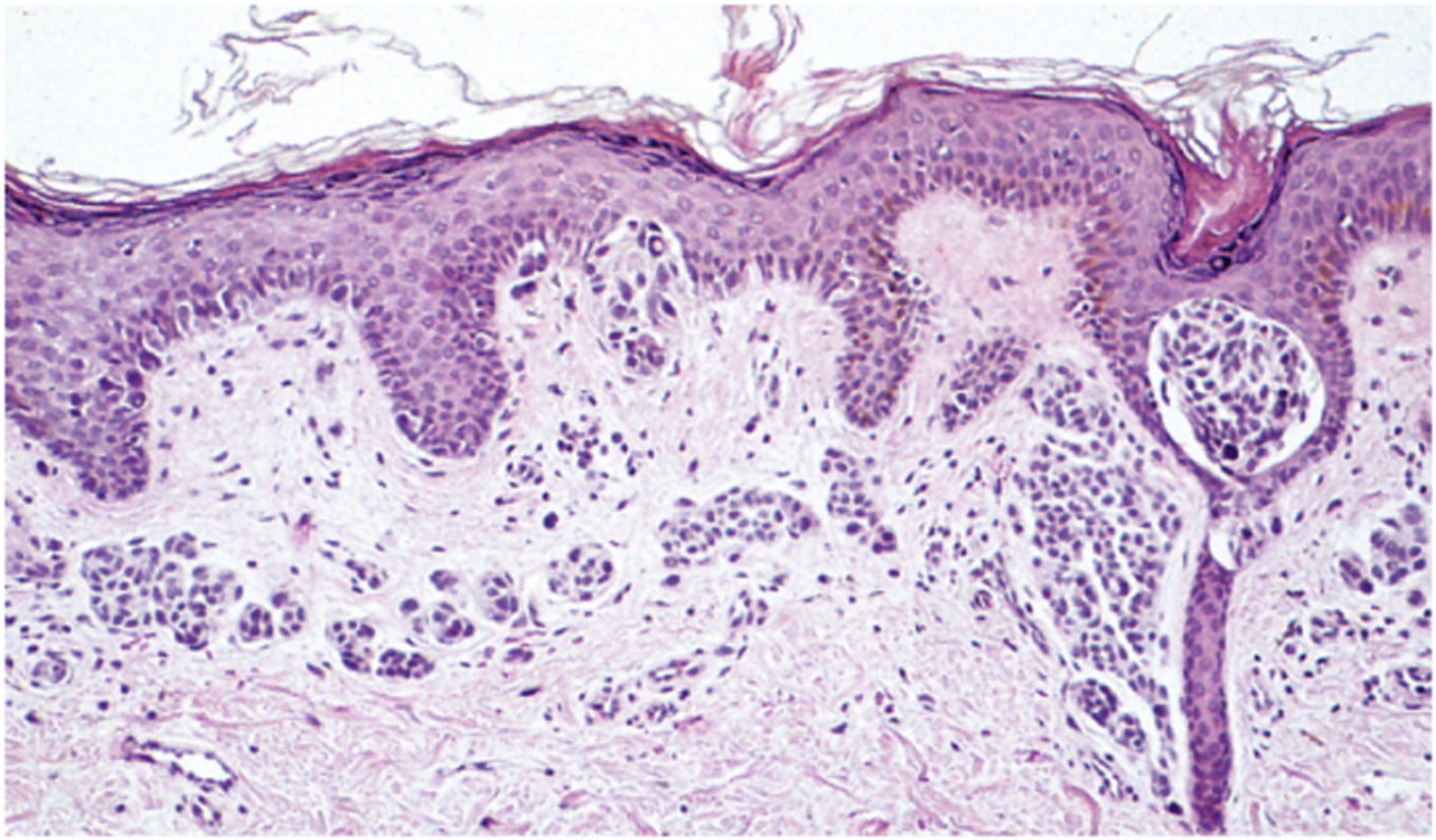


Rubin, Patologia

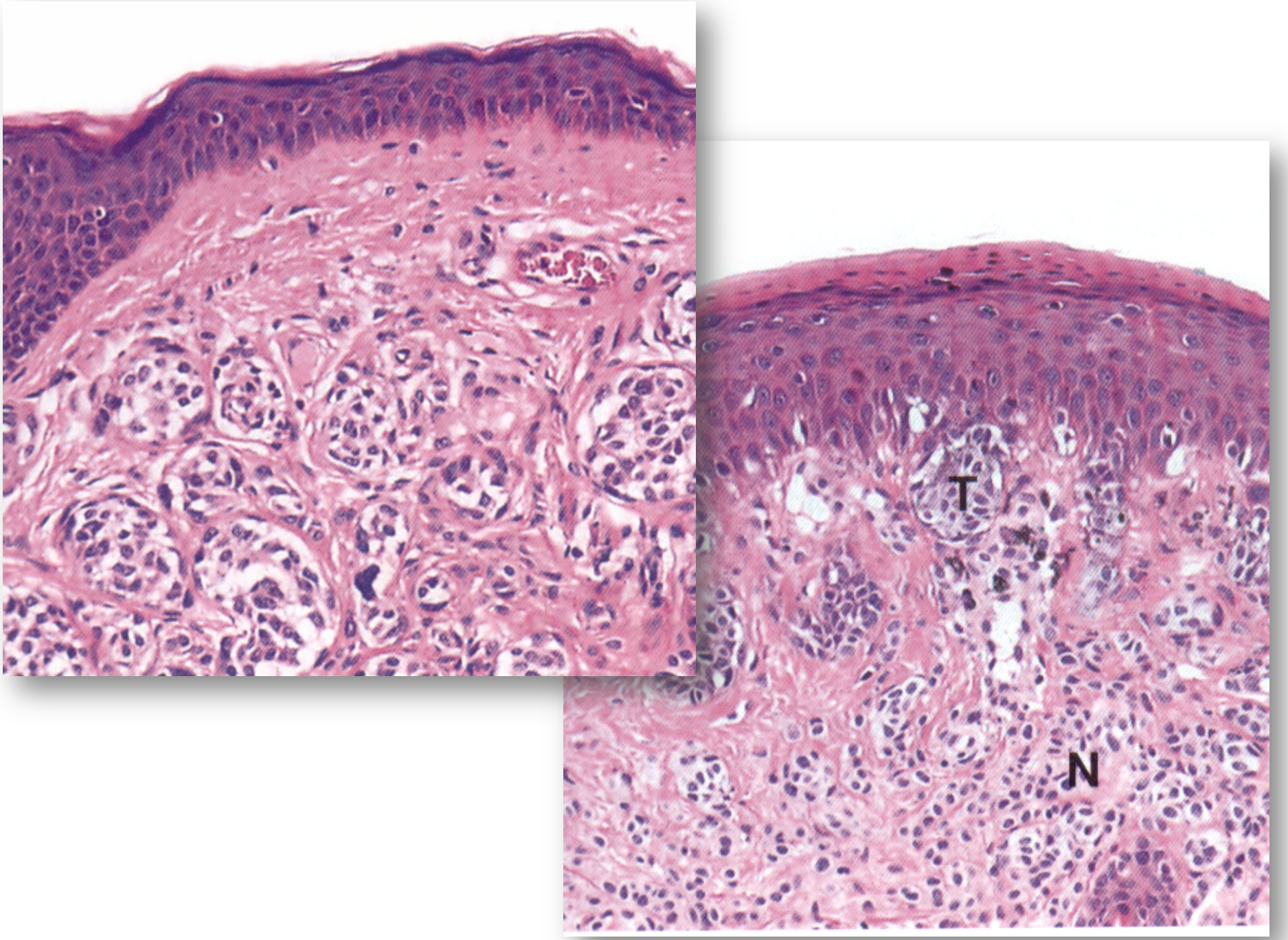
DISONTOGENETIC TUMOURS

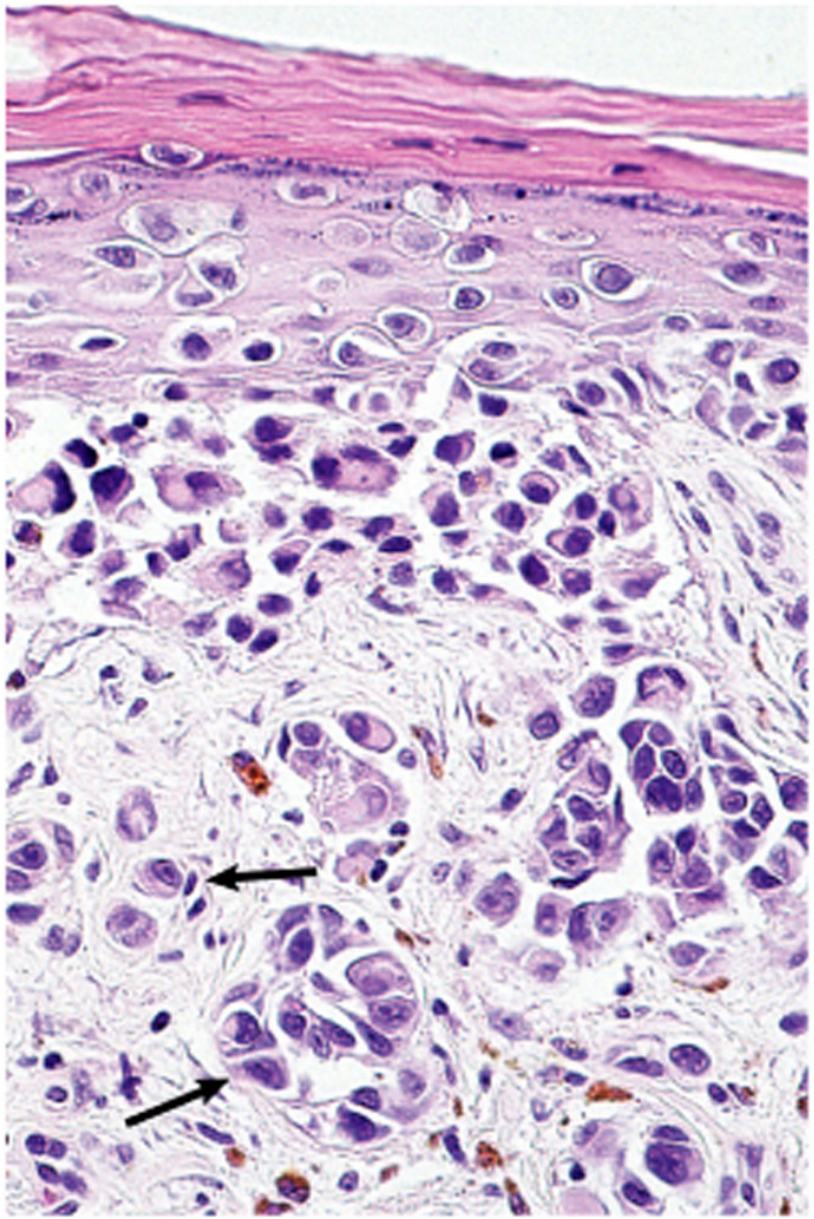
Nevi (neurogenic amartia)

- Small local accumulations of melanocytes
- At the border between epidermis and dermis
- Superficial or deep dermis
- Single or multiple
- Flat or prominent
- Smooth or warty
- Skin and all melanin–containing tissues (uvea, iris, conjunctiva, pia mater and internal organs)

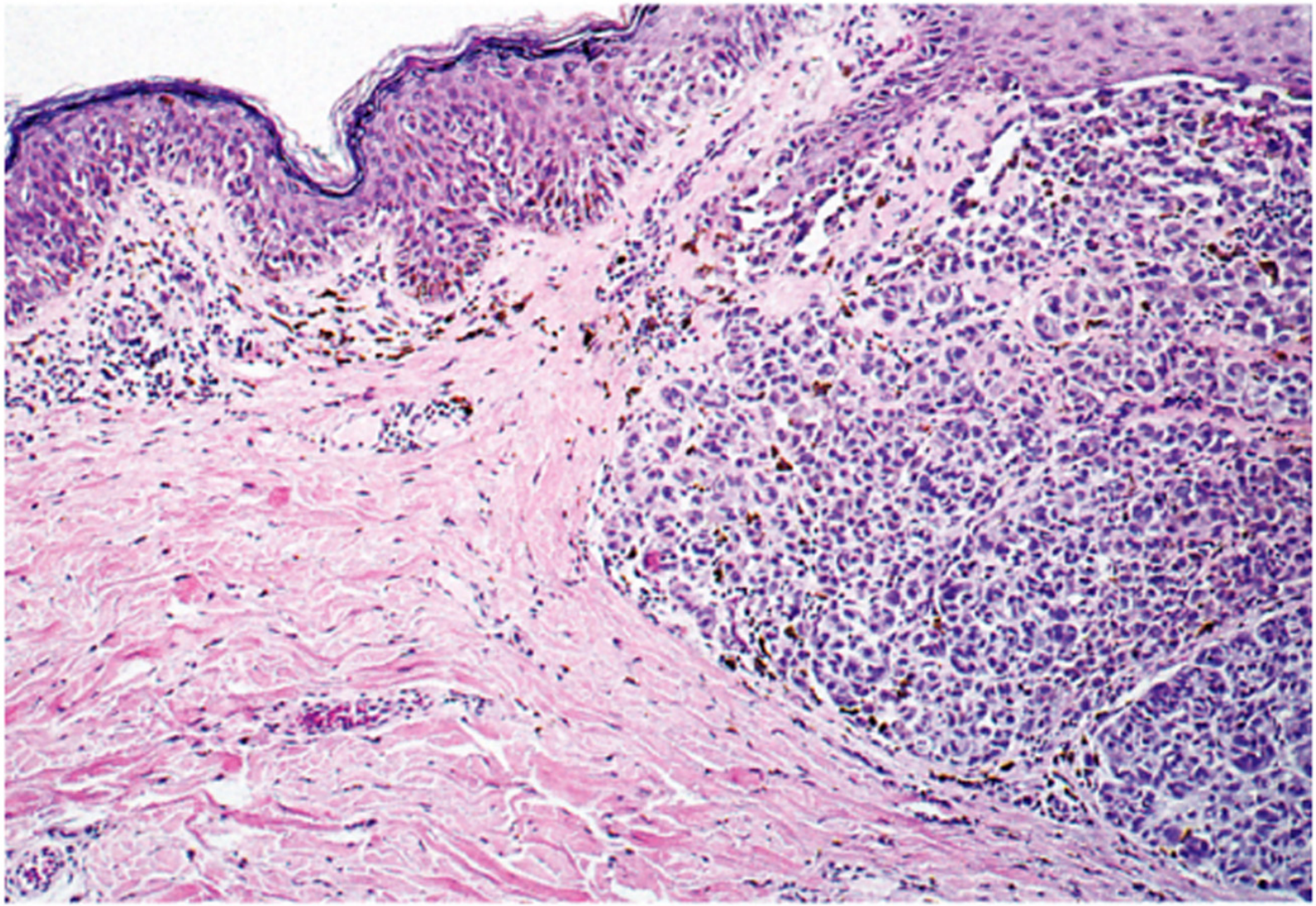


Rubin, Patologia


T
N



Rubin, Patologia

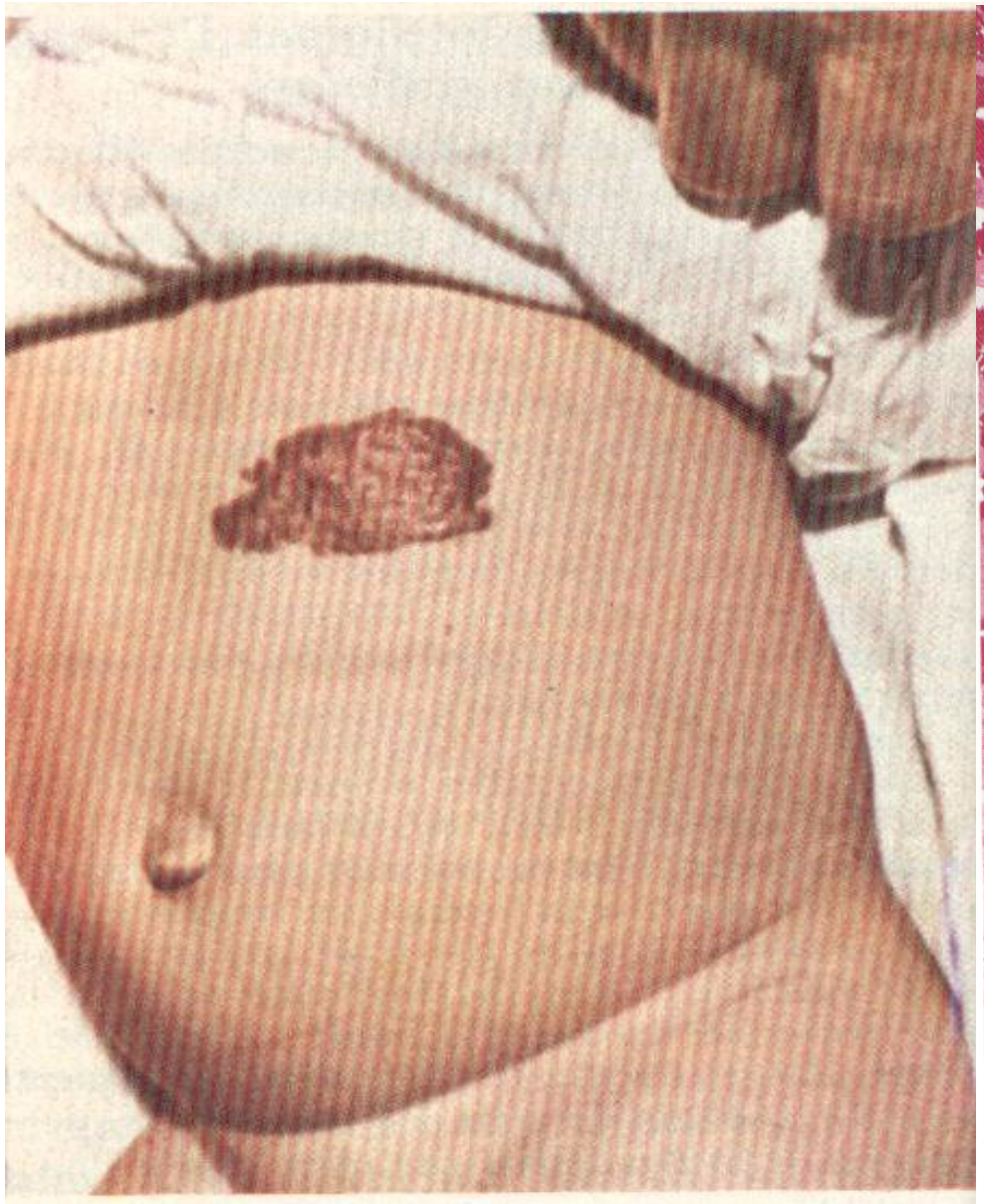


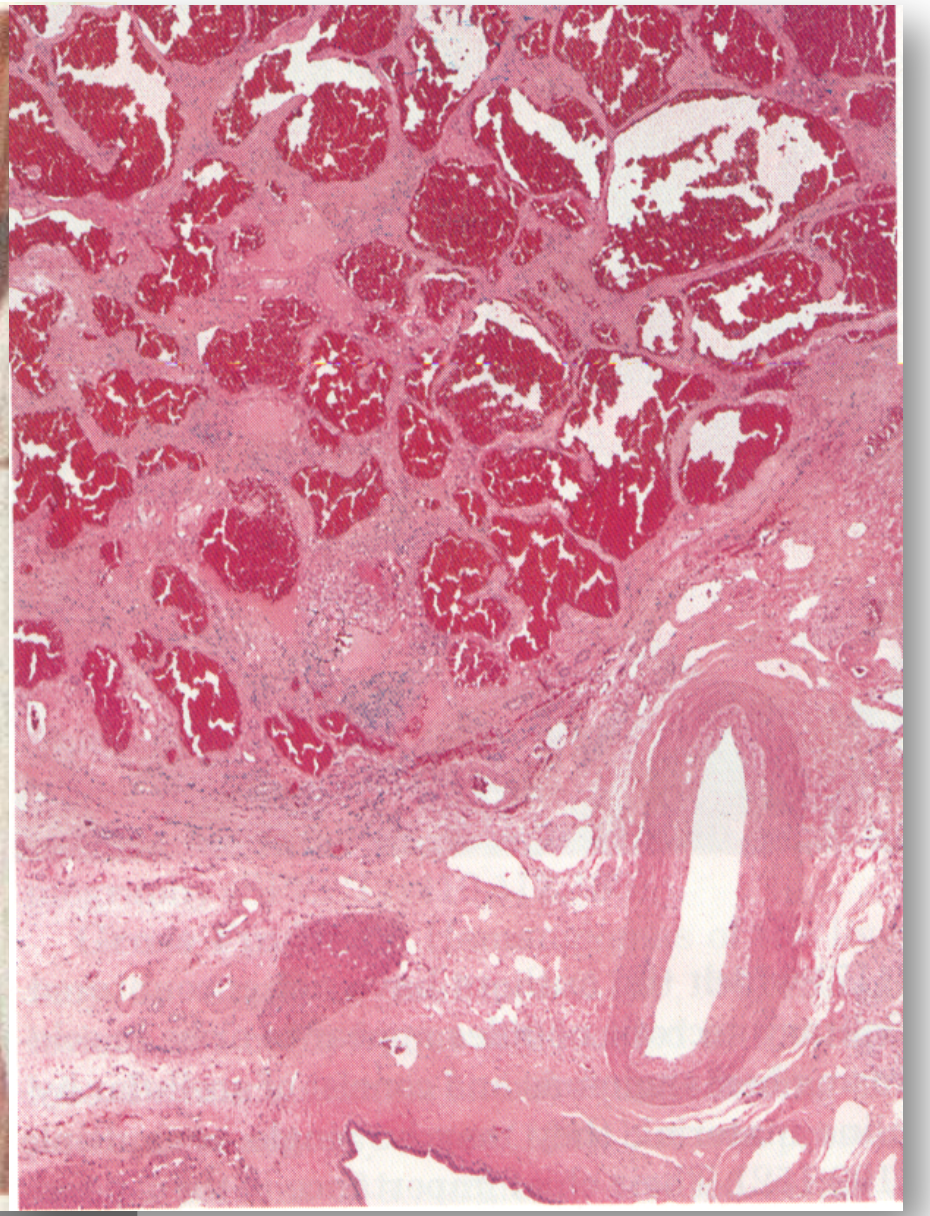
Rubin, Patologia

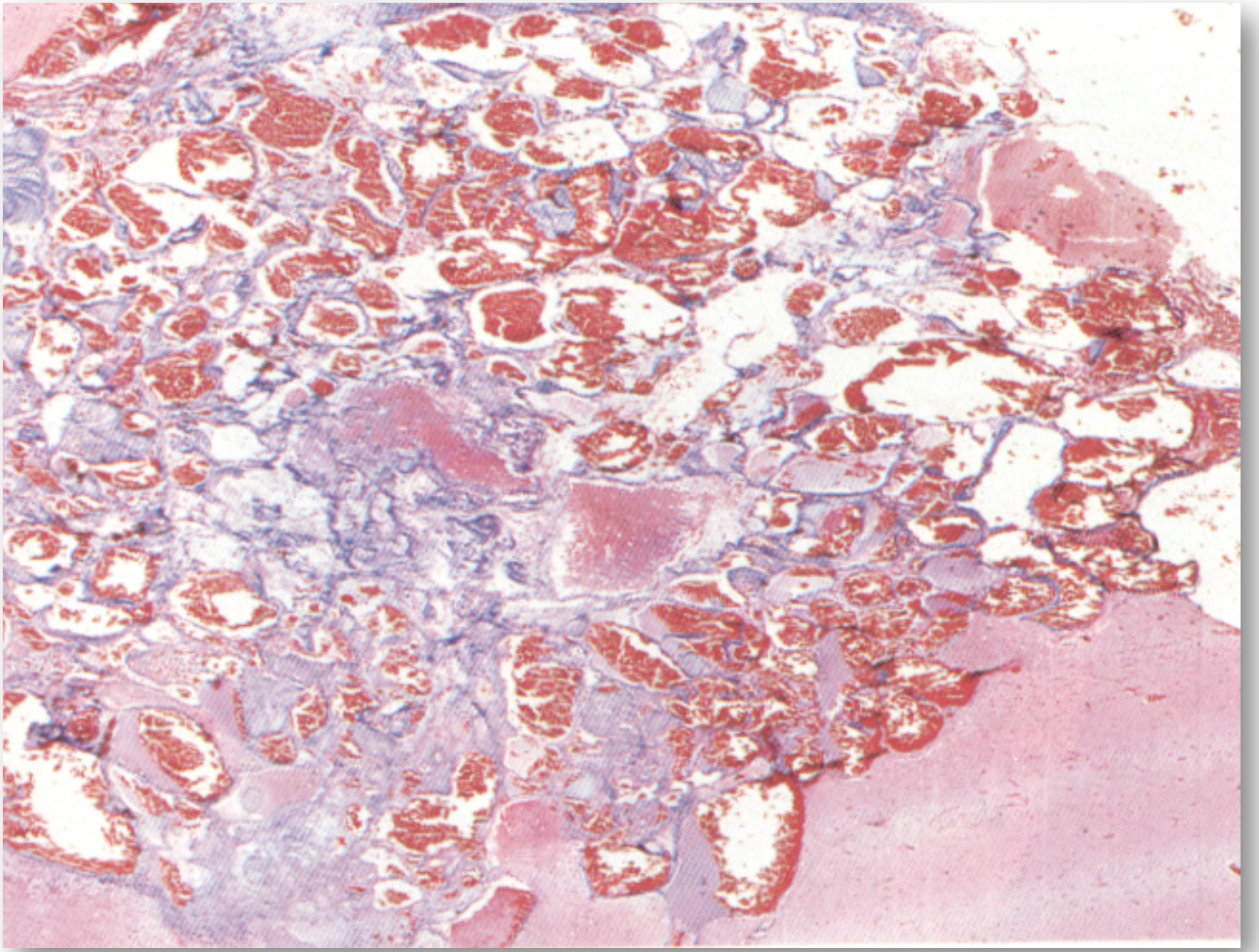
DISONTOGENETIC TUMOURS

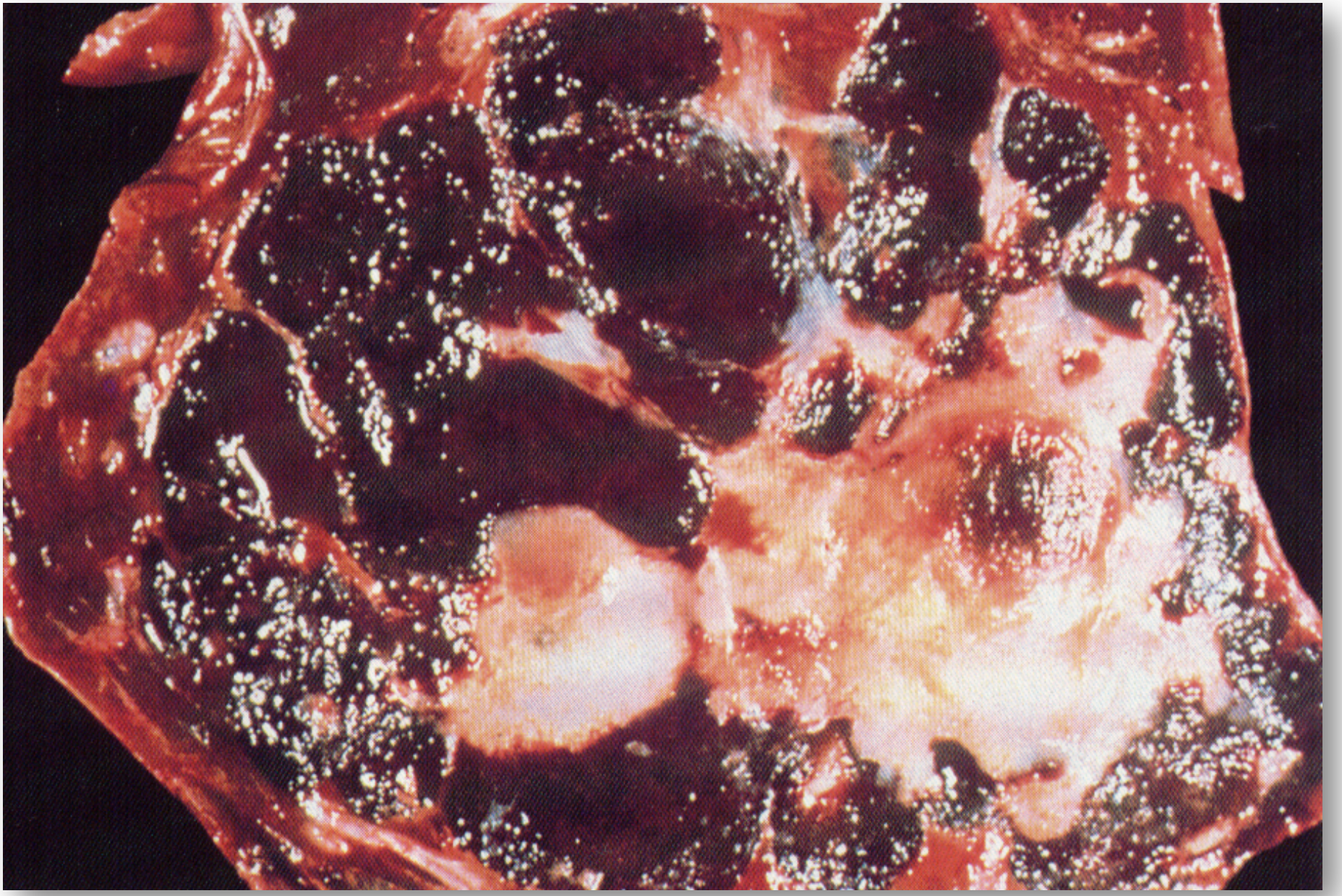
Vascular Tumors (haemangioma, lymphangioma)

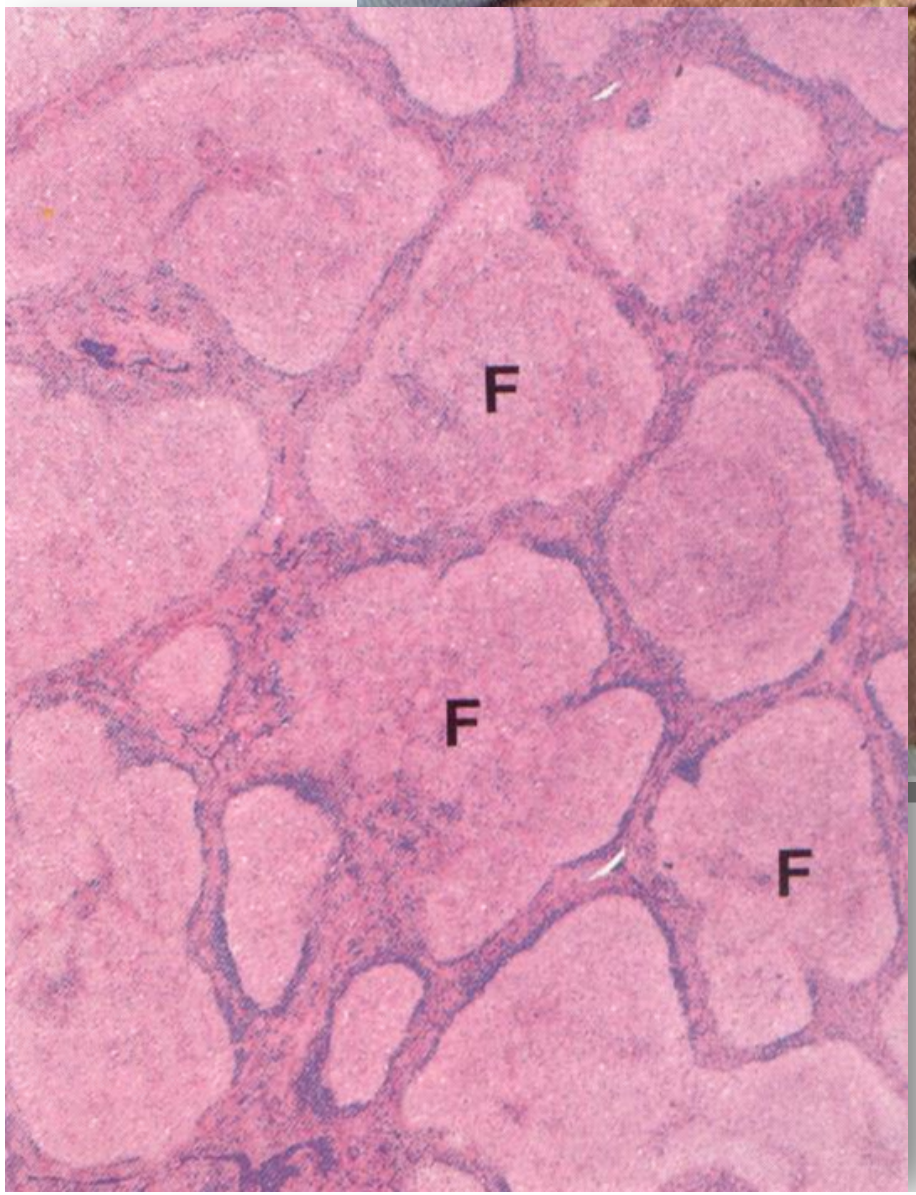
- Localized malformations
- Sometimes multiple
- Abnormal vascular proliferation
- **Skin or deep organs**










F
F
F

DISONTOGENETIC TUMOURS

Odontoma (complex or compound) associated to Gardner's syndrome

- > thickness of mandible and maxilla
- **Radiological appearance:** dense opacity

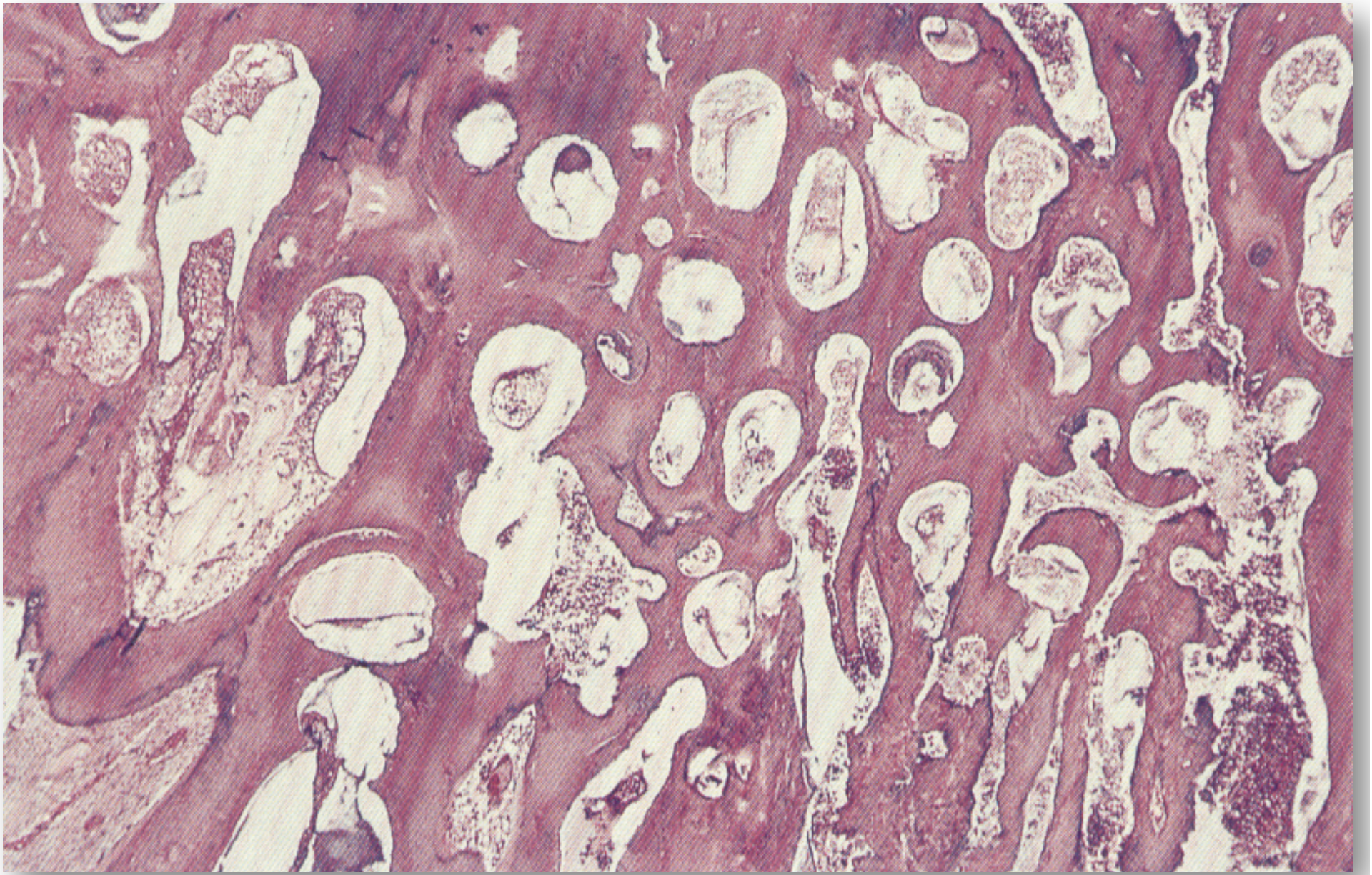
- **Complex (irregular masses of hard and soft dental tissues)**

  - enamel + dentin

  - enamel + cementum

- **Compound**

  - denticles separated by fibrous tissue



DISONTOGENETIC TUMOURS

Tumors on coristia (coristoma)

- ***Simple tissue malformations with limited local proliferative activity, in organs devoid of such components***

  Pleuric/pulmonary chondroma

  - displacement of bronchial cartilaginous tissue

DYSONTOGENETIC TUMORS

Tumors on unused embrional germs

- Mixed Tumors:
  - mesenchymal
  - organoid
  - teratoid

DYSONTOGENETIC TUMORS

Mixed tumors

- **Mesenchymal**

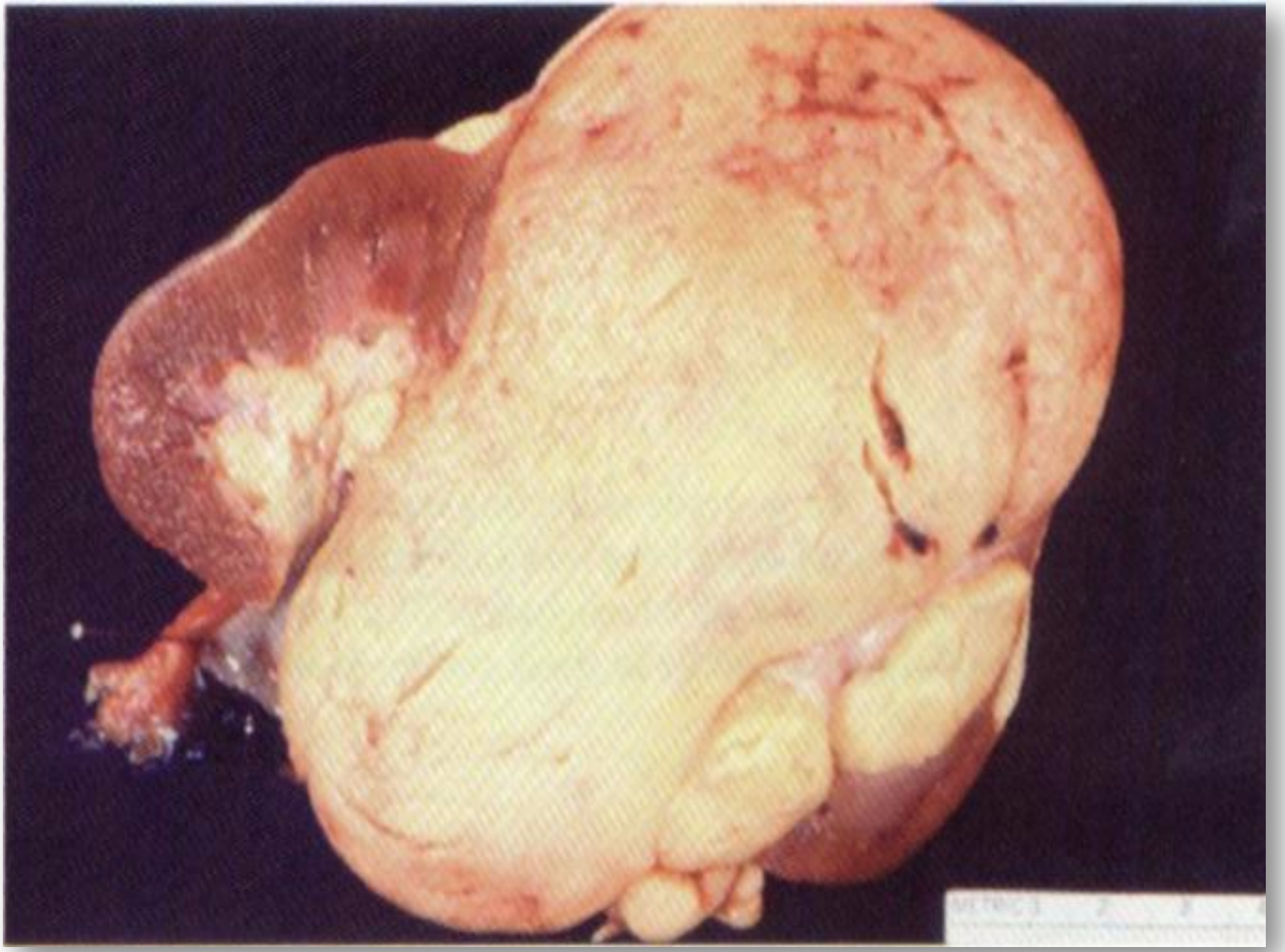
  Different mesenchymal tissues, ***irregularly*** mixed

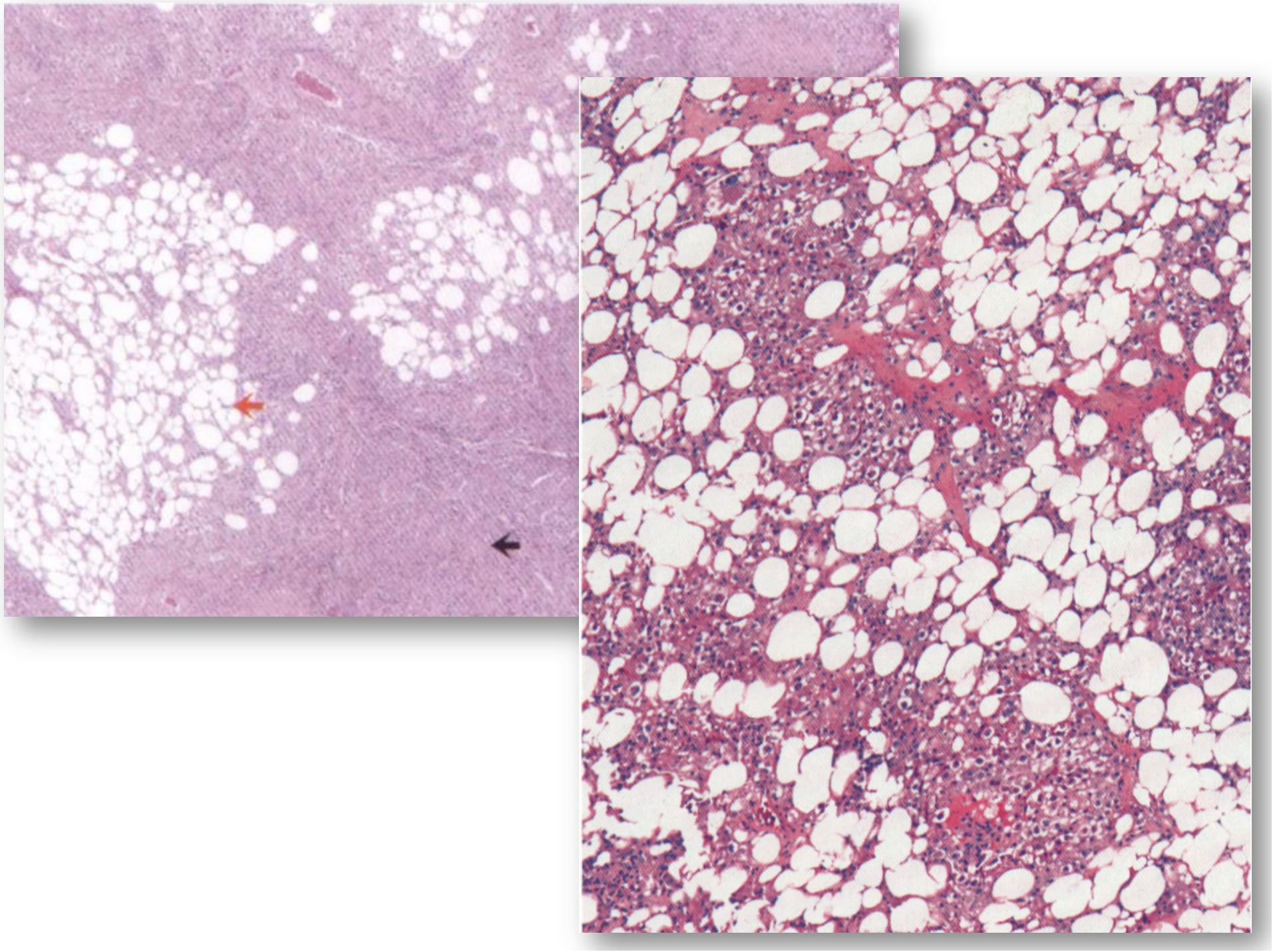
  -fibrolipoma, fibro-chondro-osteoma, angiomyolipoma…

- **Organoid**

  connective or epithelial tissues ,organized in organoid formations, they mimic structures that are similar to normal constituents of the organ in which they develop

  - Benign tumors , accessory enclosed in the maternal tissue
  - Exception : T. Wilms, rapid growth, destructive, malignant

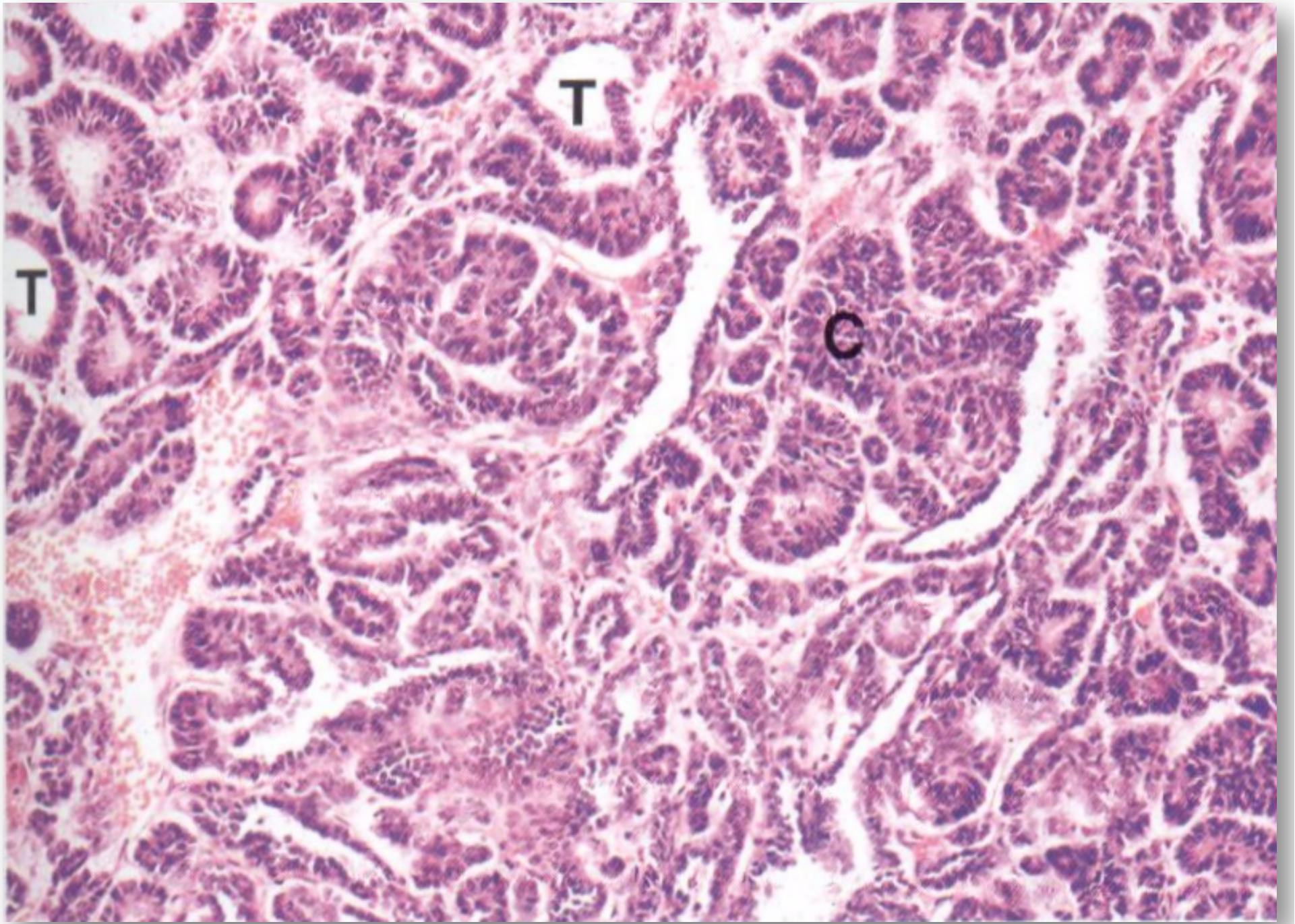




WILMS' TUMOUR

Frequent tumour

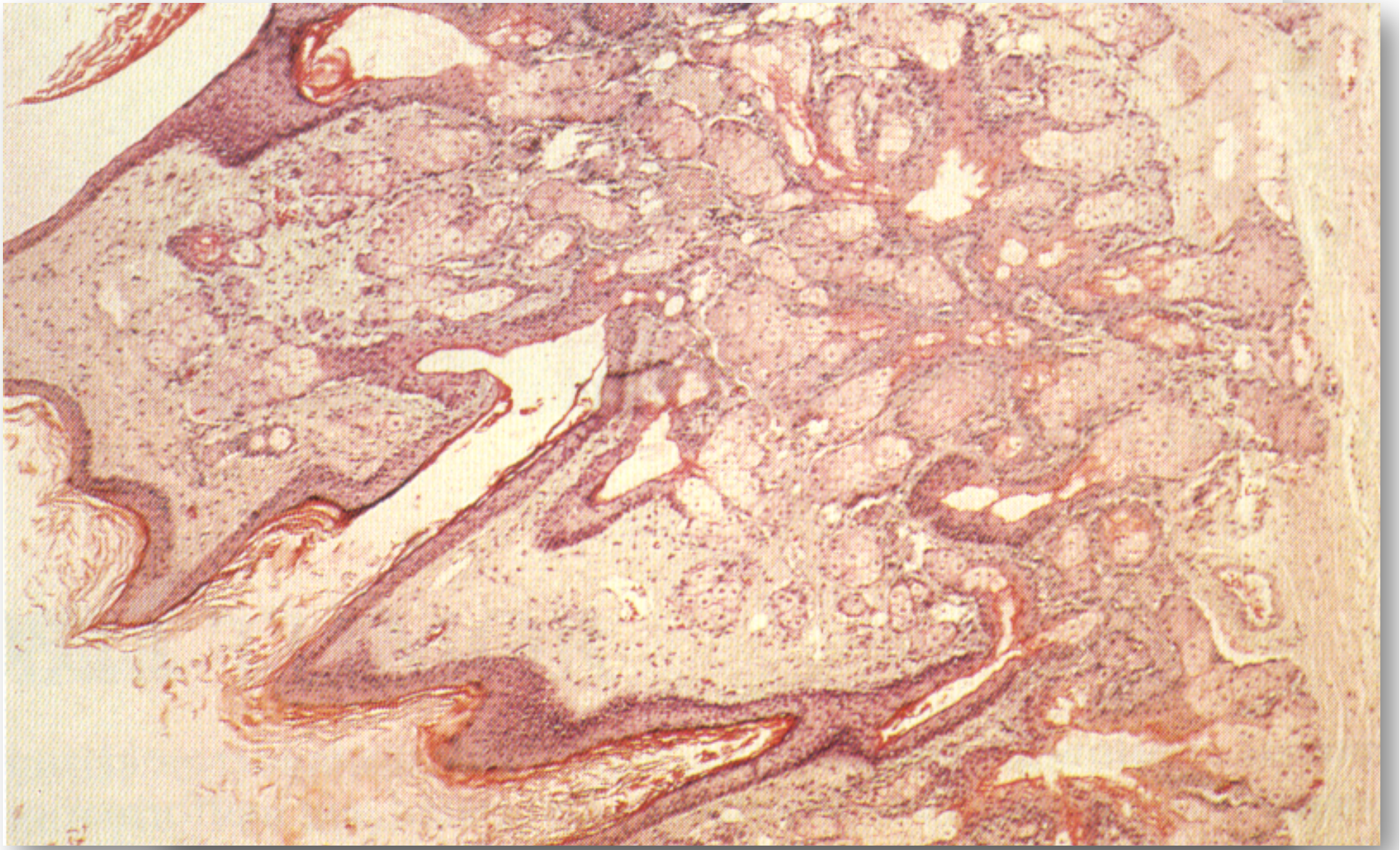
- Present at birth
- Affects both males and females
- Malformative syndrome (microcephalia, spina bifida, meningocele, aniridia, genito-urinary abnormalities, macroglossia..)
- Monolateral
  - Macroscopic appearance: globose shape (12 cm), gross encephaloid aspect, grey-reddish in colour
  - Microscopic appearance: epithelial + mesenchymal tissues
  - Cords or clusters of small and dark cells , sometimes with tubular differentiation + different tissues: fibroblastic , mixoid, adipose, cartilaginous, bony, striated muscle)

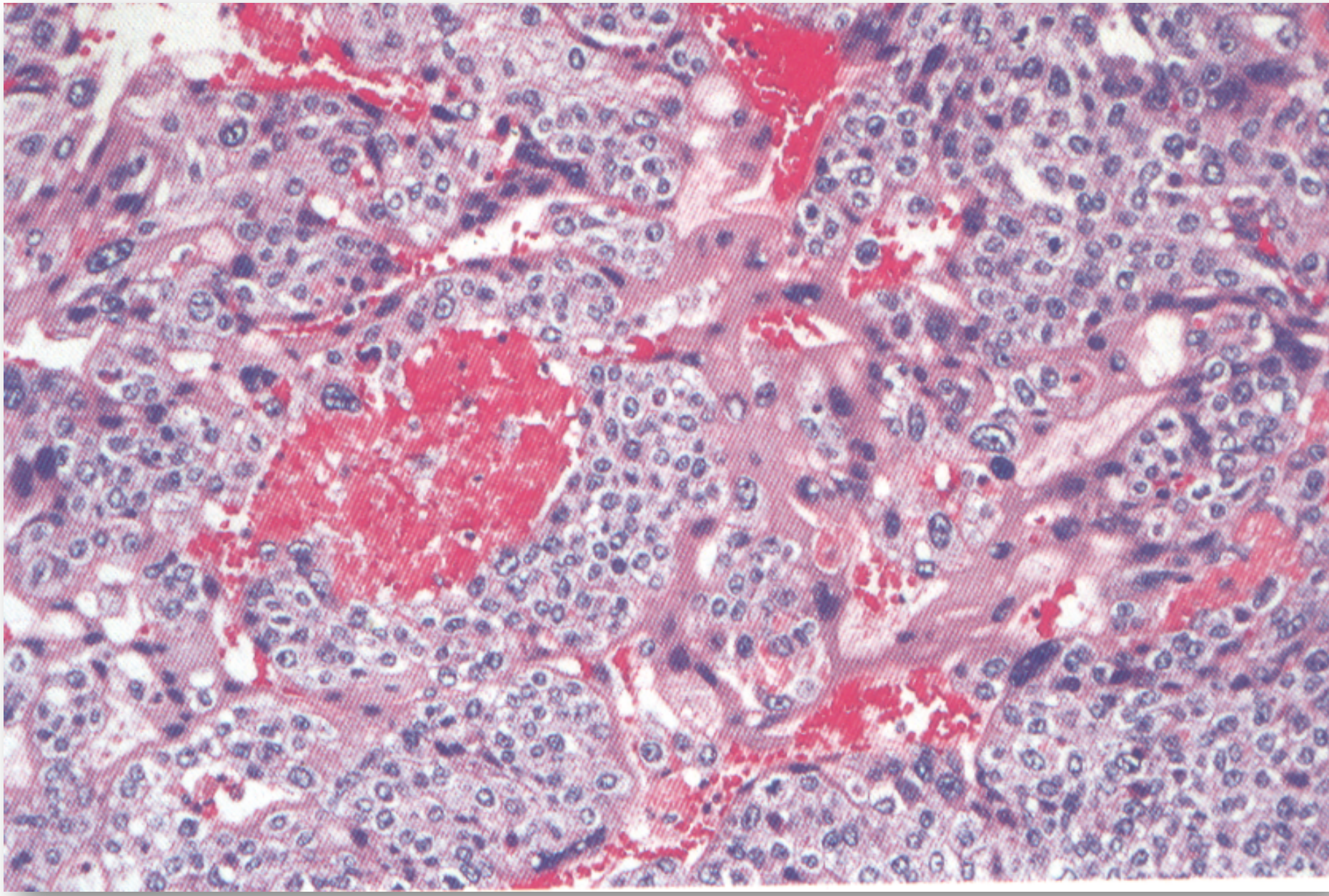

T
T
C

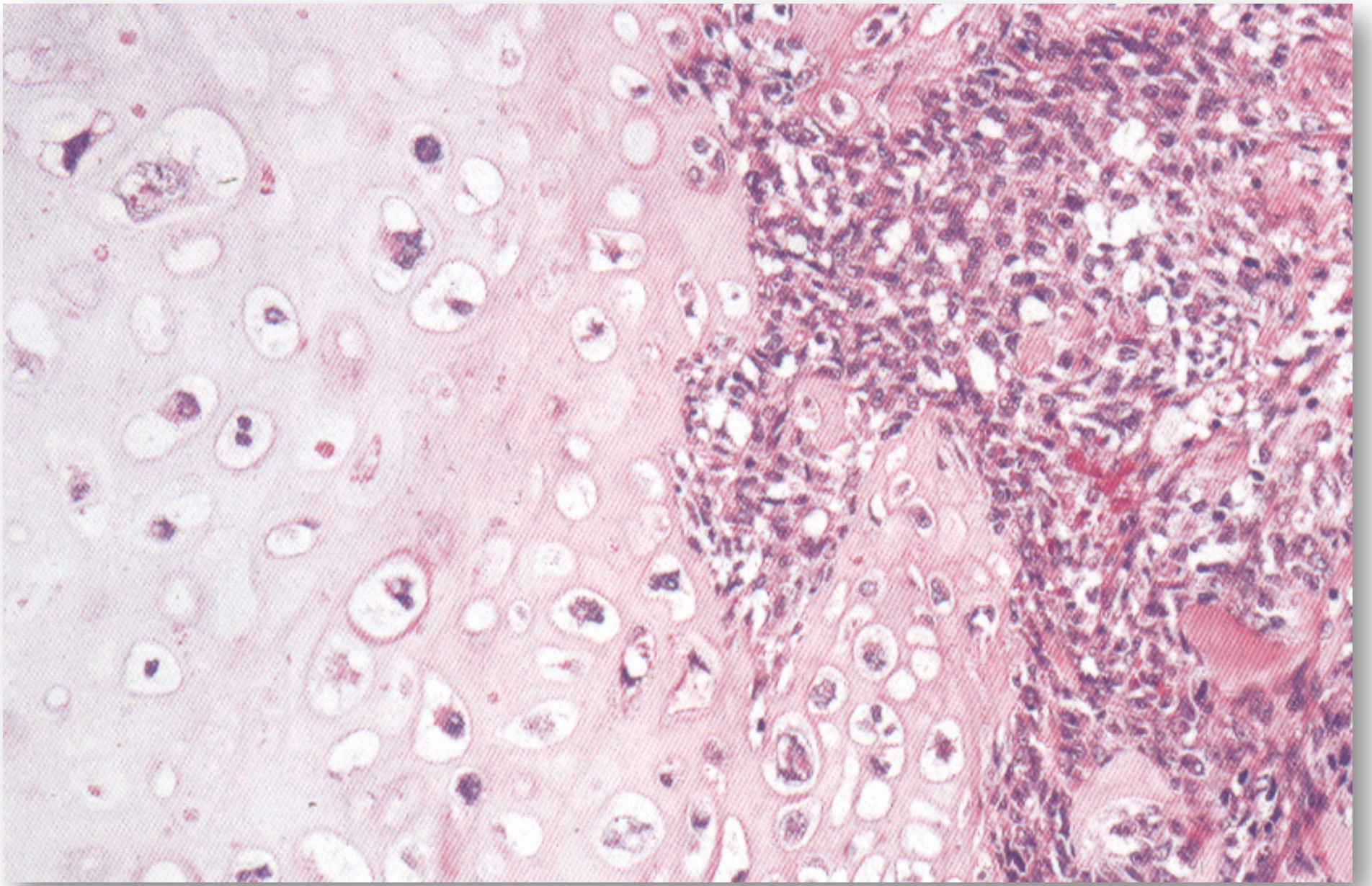
DISONTOGENETIC TUMOURS

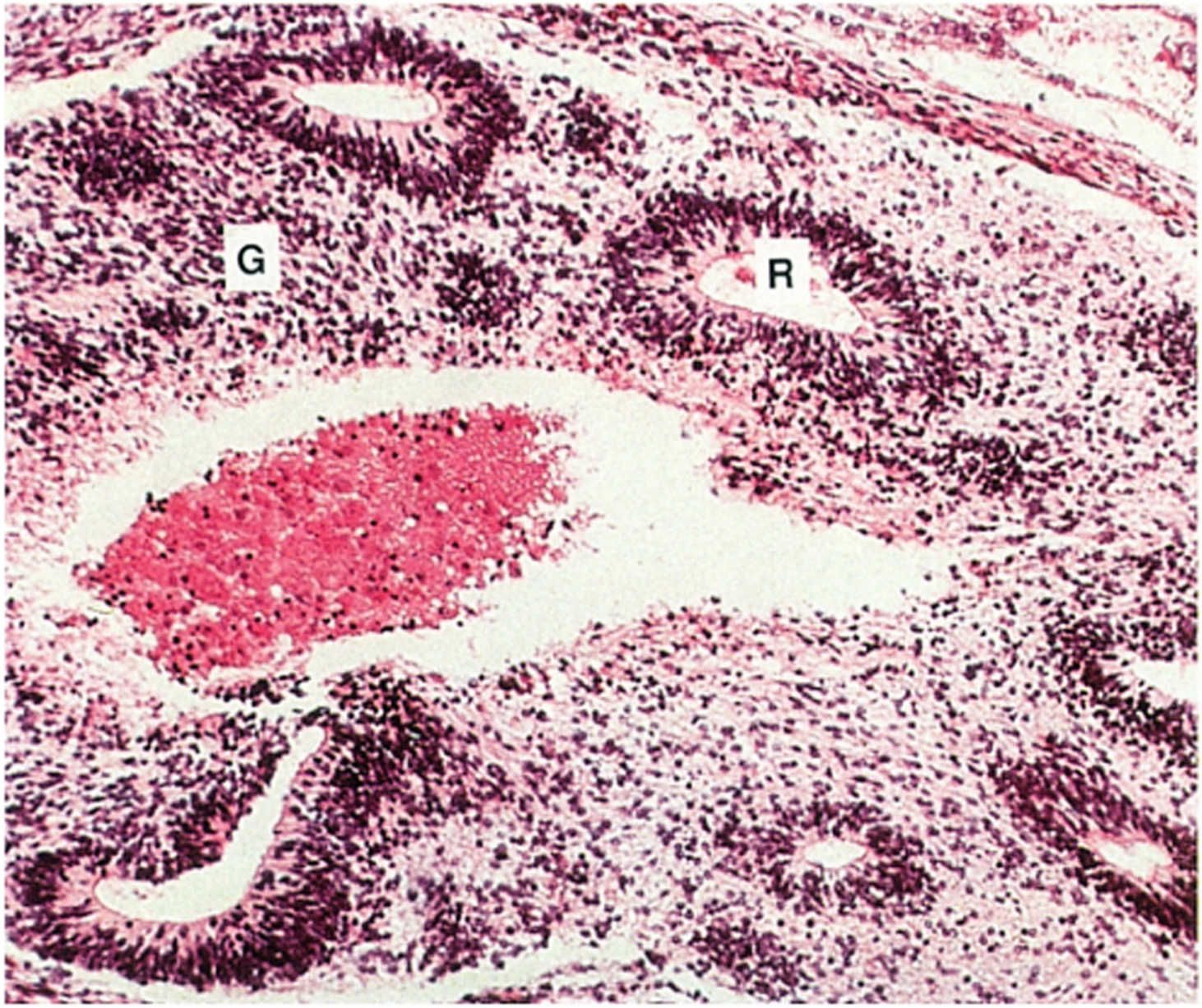
Teratoid mixed tumours

- Made of tissues of different origin
- Arise from totipotent cells (ovary, testis, embryonal residues on midline)
- Contains mature or immature tissues of one to three embryonal layers
  - ***Mature Teratoma*** (benign): differentiated tissues , corrispondent age of the carrier (dermoid cyst)
  - ***Immature Teratoma*** (aggressive): made of a random mixture of immature tissues (coriocarcinoma)









Rubin, Patologia