

LECTURE

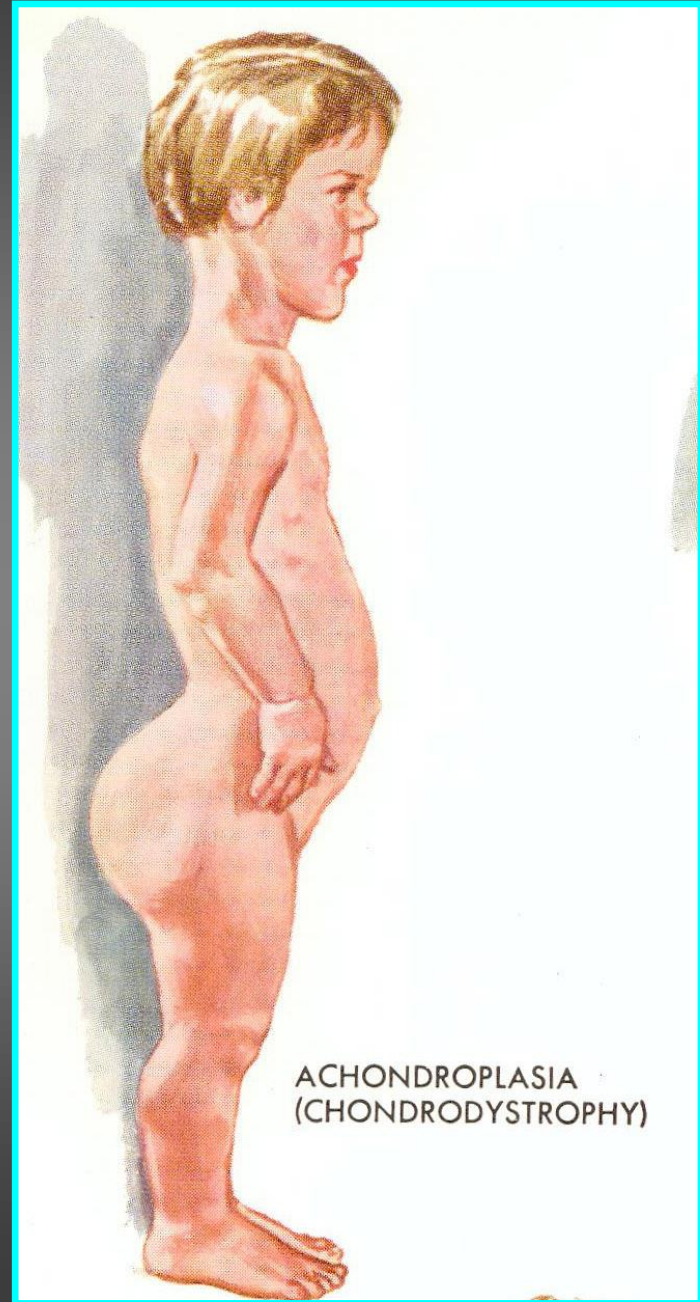


X-RAY OF
ROENTGEN'S WIFE
HAND

CONRAD WILHELM ROENTGEN

BONES + MUSCLES + JOINTS

ACHONDROPLASIA



ACHONDROPLASIA
(CHONDRODYSTROPHY)

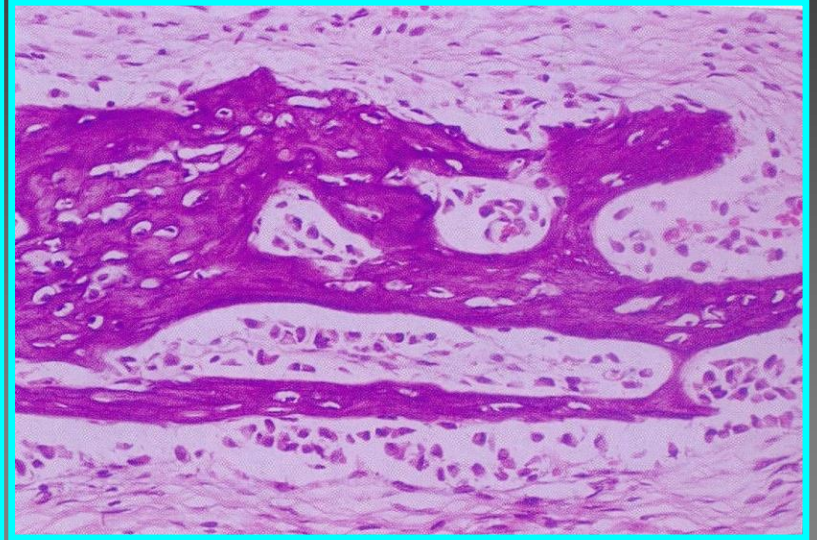
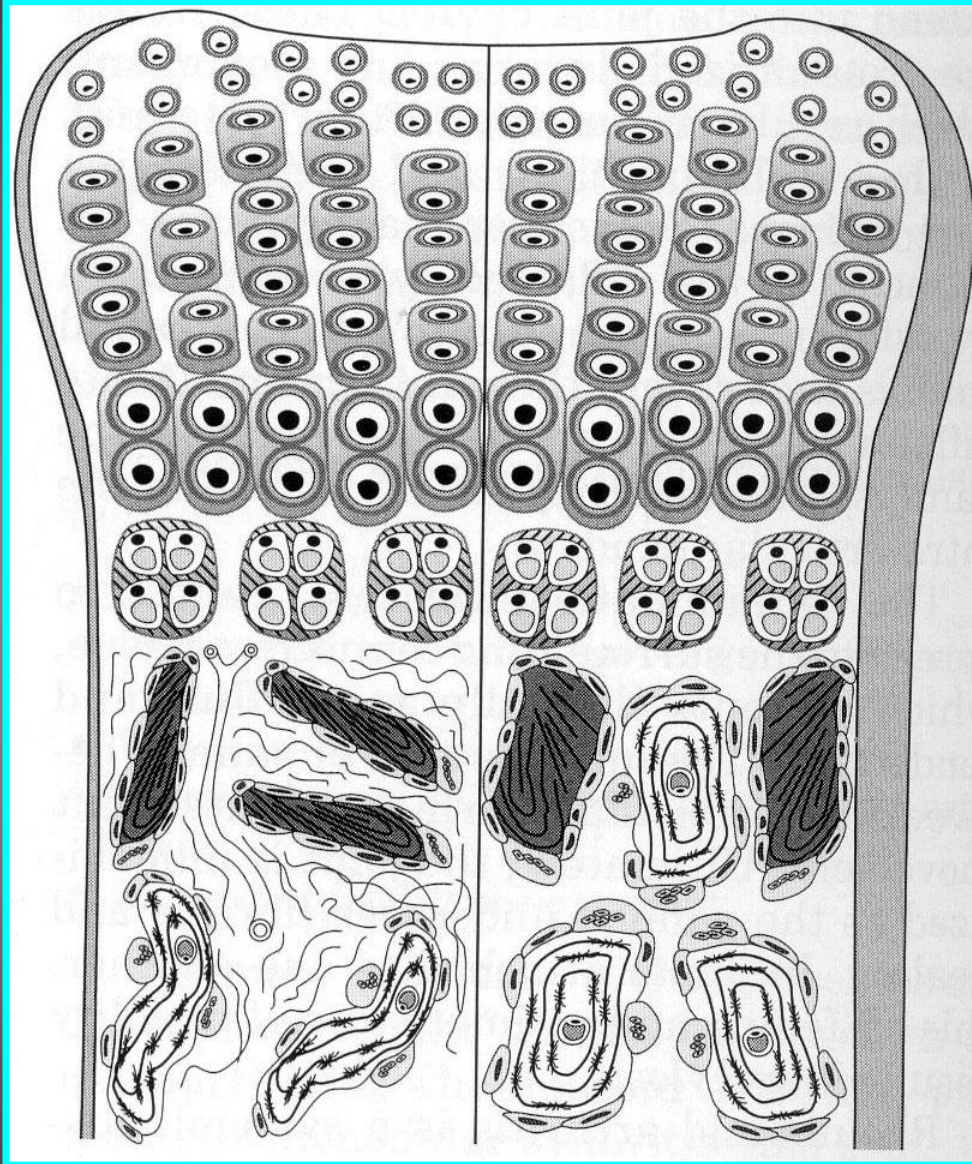
- **OSTEOPETROSIS, „STONE BONE”,**
also known as **MARBLE BONE**
DISEASE and **Albers-Schonberg disease**
is an extremely rare inherited disorder
whereby the bones harden, becoming
denser, in contrast to more prevalent
conditions like osteoporosis, in which the
bones become less dense and more brittle,
or osteomalacia, in which the bones soften.

OSTEOPETROSIS

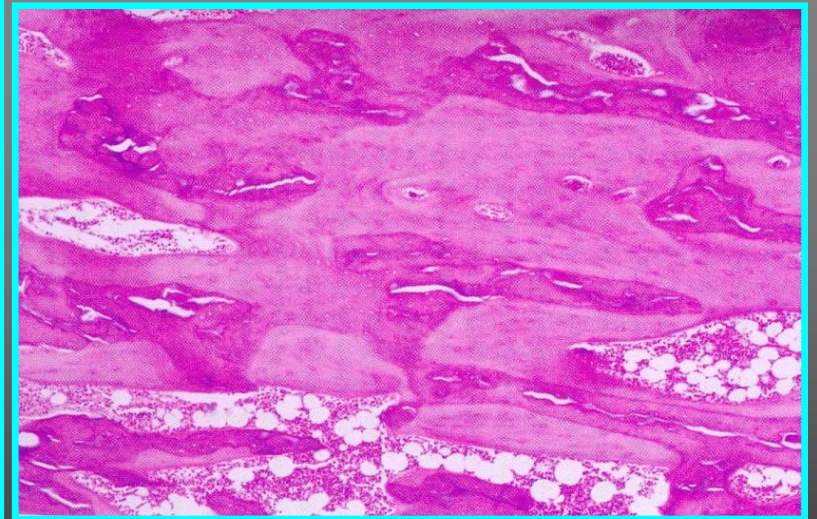


- **OSTEOGENESIS IMPERFECTA (OI) is SKELETAL DISEASE commonly known as the "brittle bone" disorder, a genetic disorder characterized by bones that break easily, often from little or no apparent cause. A person with OSTEOGENESIS IMPERFECTA may break a rib while coughing or a leg by rolling over in their sleep.**

PATHOLOGY OF BONES AND CARTILAGES



OSTEOGENESIS IMPERFECTA

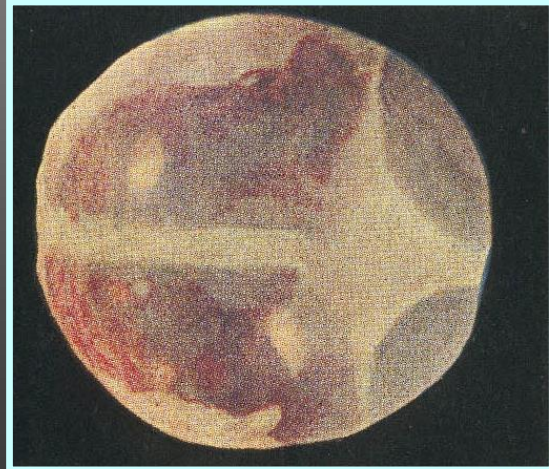
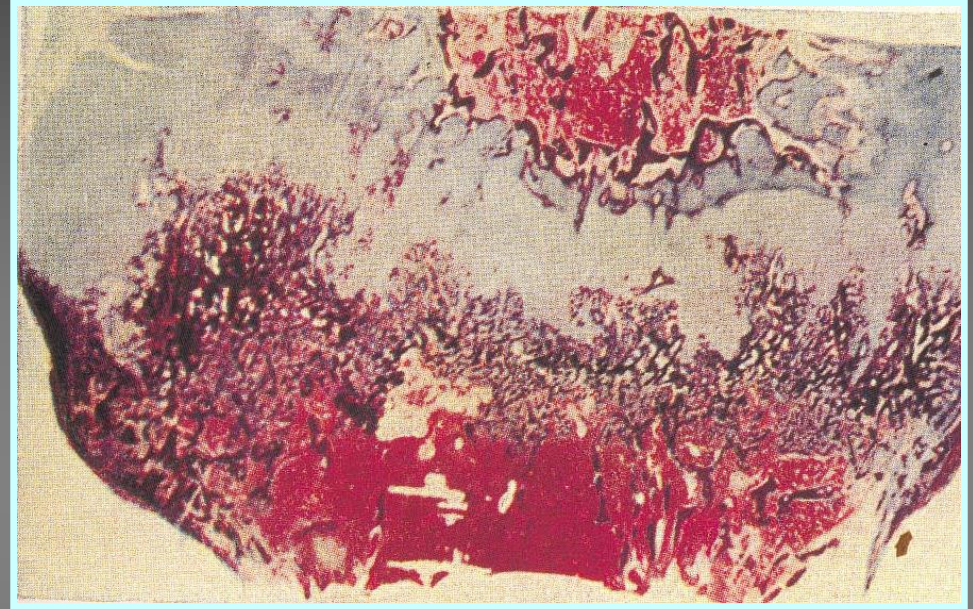
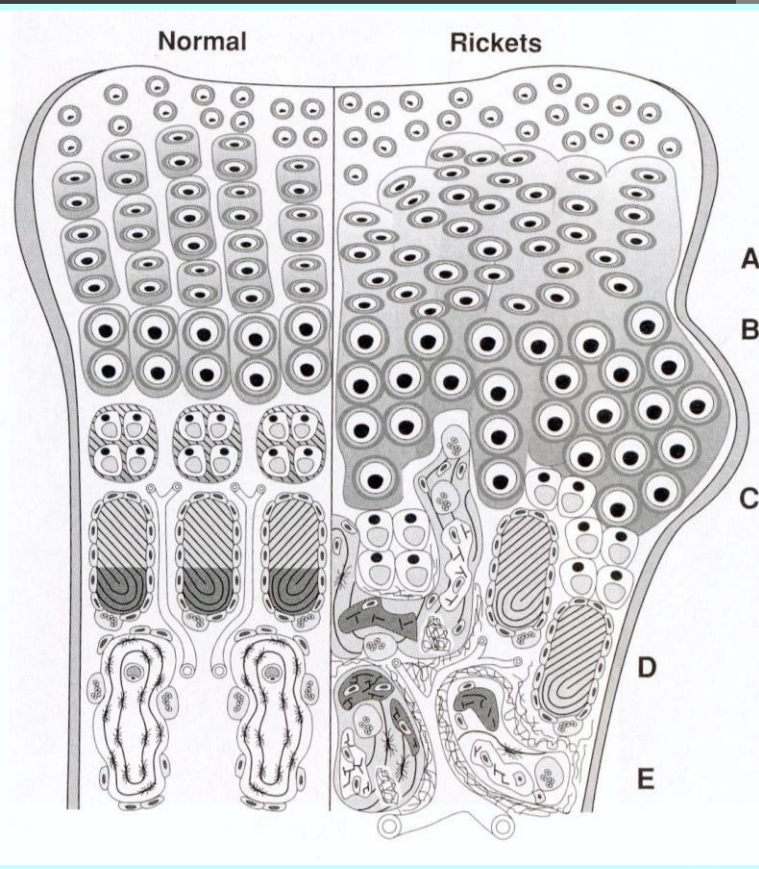


OSTEOPETROSIS

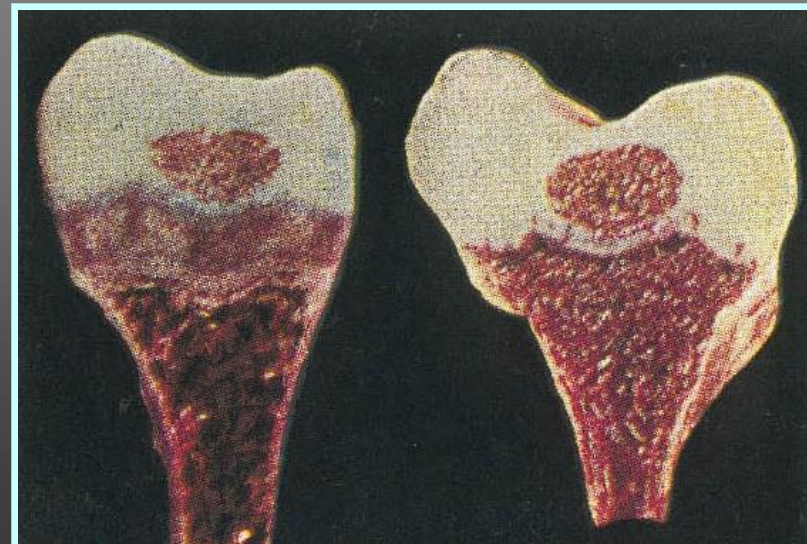
**OSTEOGENESIS IMPERFECTA AND
OSTEOPETROSIS**

PATHOLOGY OF BONES AND CARTILAGES

RICKETS - RACHITIS



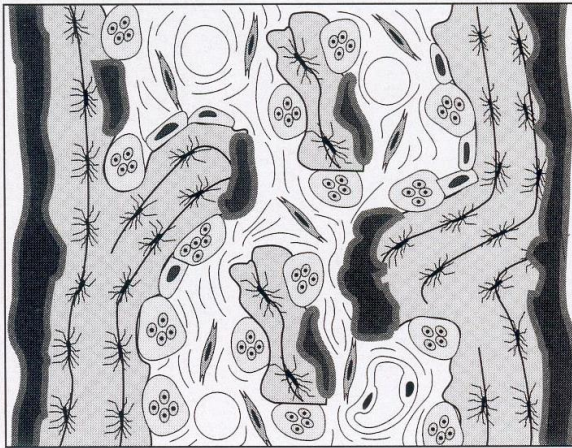
THE EXCESS OF
UNCALCIFIED
TISSUE,
OSTEOID, IN
AREAS OF BONE
GROWTH



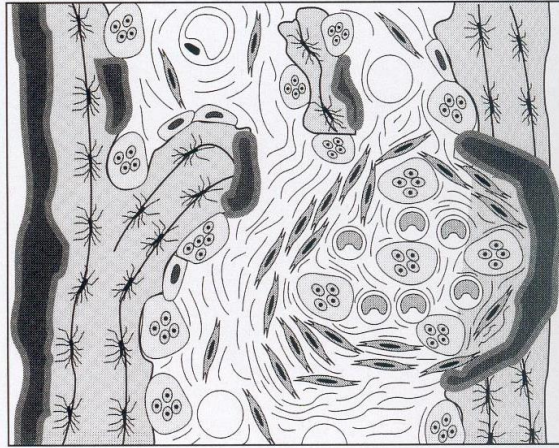
PATHOLOGY OF BONES AND CARTILAGES

HYPERPARATHYROIDISM

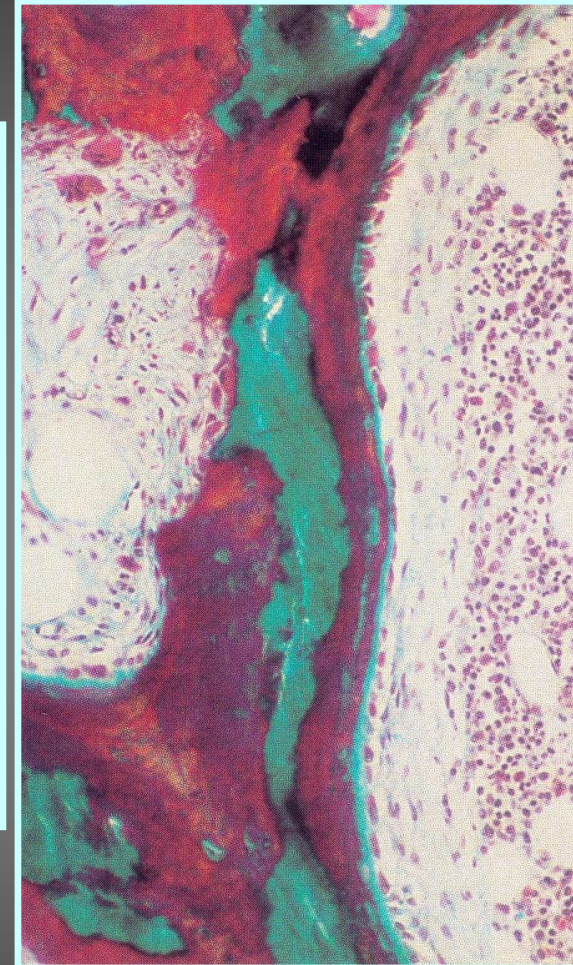
Diagram 17.5. Osteitis fibrosa caused by hyperparathyroidism. A. The outer cortex is ragged, and the bone surfaces contain an increased amount of osteoid. Osteoclasts are present in increased numbers, and the marrow shows fibrosis. **B.** Brown tumor of hyperparathyroidism. This destructive expansive bone lesion is composed of osteoclasts, macrophages, and fibroblasts.



A



B

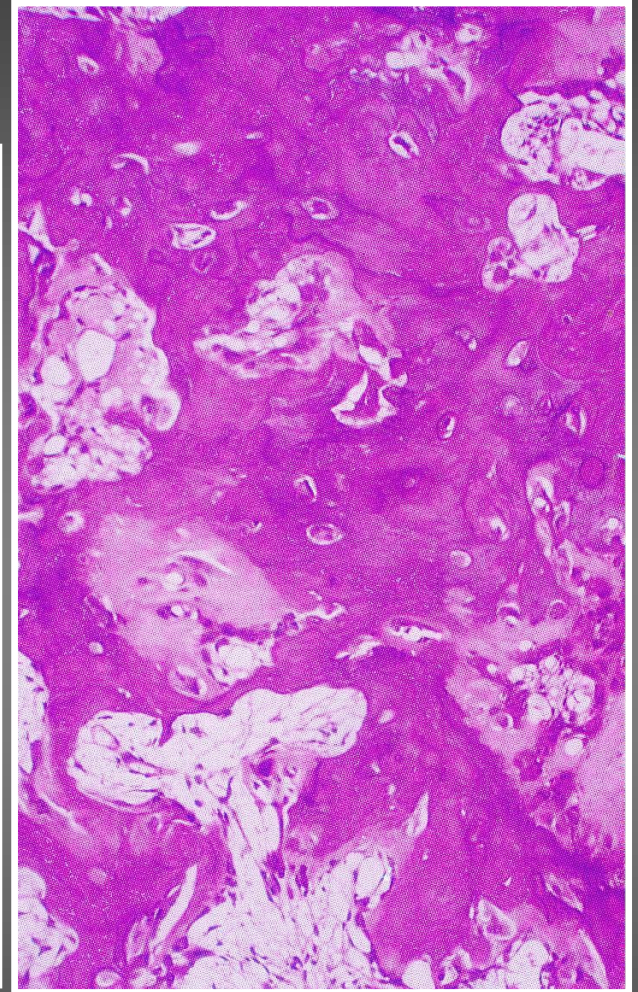
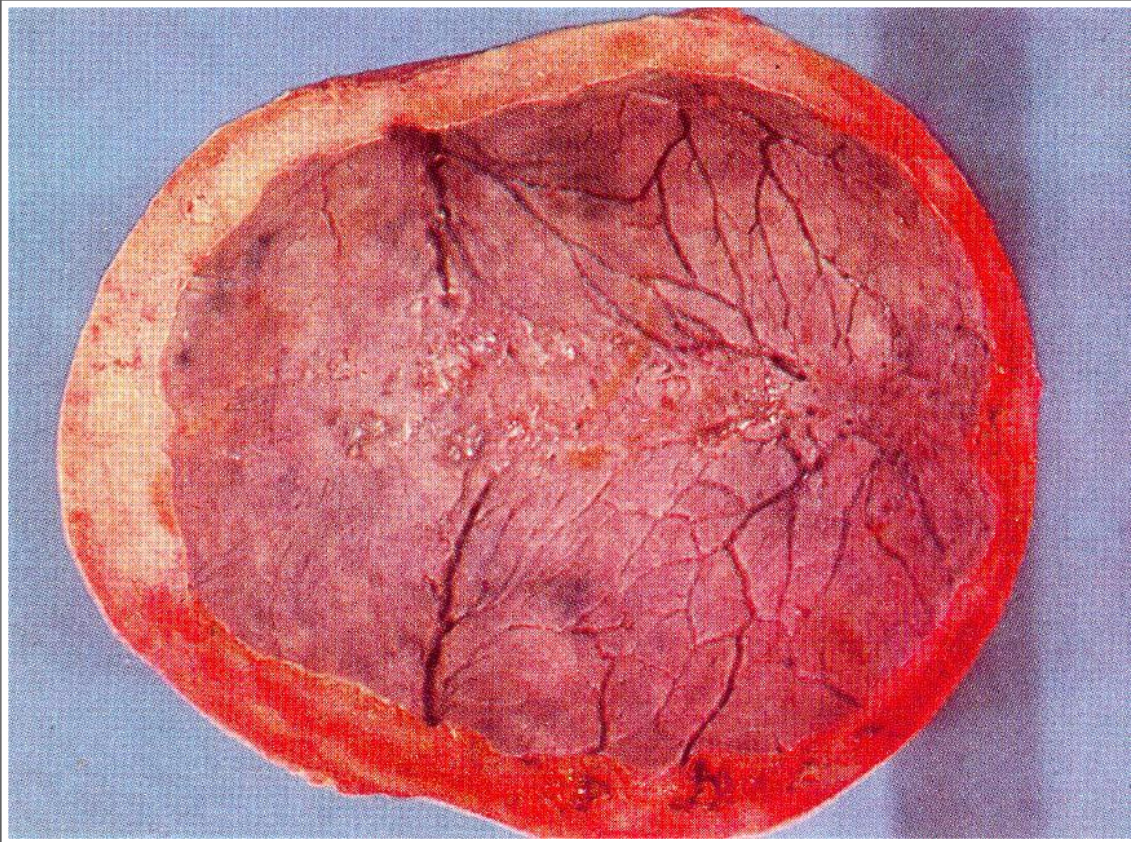


BROWN TUMORS

- **BROWN TUMORS** are tumors of bone that arise in settings of excess osteoclast activity, such as hyperparathyroidism, and consist of fibrous tissue, woven bone and supporting vasculature, but no matrix. They are radiolucent on x-ray.
- The osteoclasts consume the trabecular bone that osteoblasts lay down and this front of reparative bone deposition followed by addition resorption can expand beyond the usual shape of the bone, involving the periosteum and causing bone pain.
- The characteristic brown coloration results from hemosiderin deposition into the osteolytic cysts. Also characteristic of giant cell tumors of the bone.

PATHOLOGY OF BONES AND CARTILAGES

OSTEITIS DEFORMANS- MORBUS PAGET ETIOLOGY - UNKNOWN



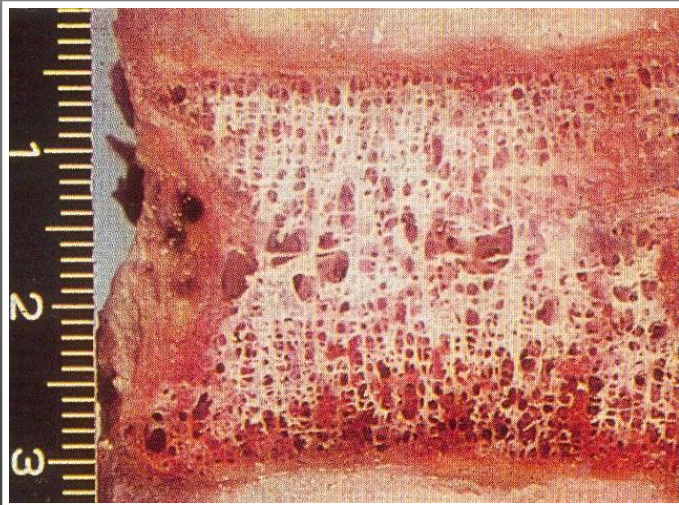
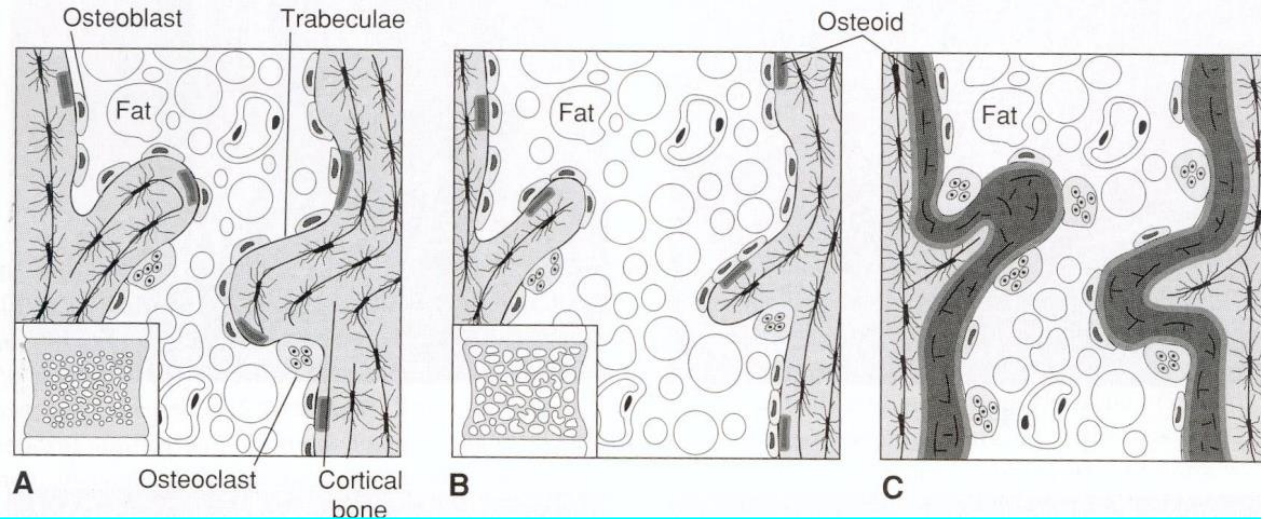
MIDDLE-AGED PATIENTS AND OLDER. 3 STAGES: 1. RESORPTION, 2. OSTEOGENESIS, 3. SCLEROSIS

LATE CONSEQUENCE: SARCOMA

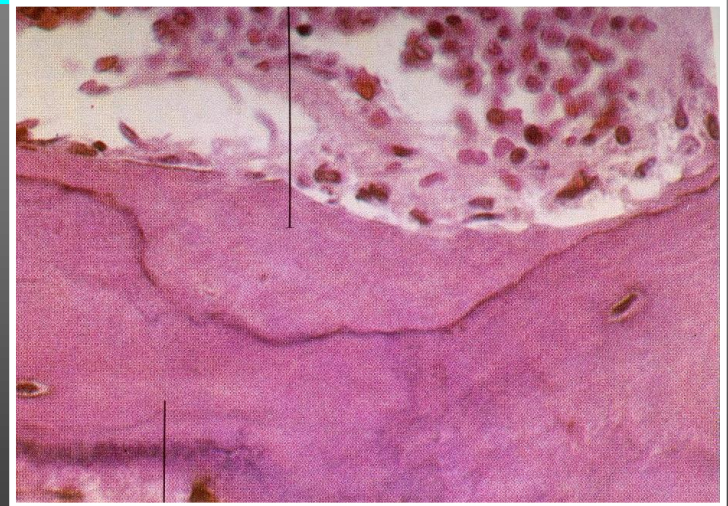
- **PAGET'S DISEASE OF BONE (OFTEN JUST PAGET'S DISEASE) OR OSTEITIS DEFORMANS**, is a chronic disorder that typically results in enlarged and deformed bones. The disease is named after Sir James Paget, the British surgeon who first described it in 1877. The excessive breakdown and formation of bone tissue that occurs with Paget's disease can cause bone to weaken, resulting in bone pain, arthritis, deformities, and fractures.
- Paget's disease is rarely diagnosed in people less than 40 years of age. Men are more commonly affected than women.

PATHOLOGY OF BONES AND CARTILAGES

Diagram 17.3. Metabolic bone diseases. **A. Normal bone.** The cortex and trabeculae of spongiosa are relatively thick. Osteoblasts and osteoclasts are present on the surfaces of trabeculae and endosteal cortical bone in a ratio of 10:1. **B. Osteoporosis.** Cortex and trabeculae are thinner but well mineralized. Osteoblasts and osteoclasts are present in normal numbers. **C. Osteomalacia.** The cortical bone and trabeculae appear of normal thickness, but are composed mostly of osteoid.



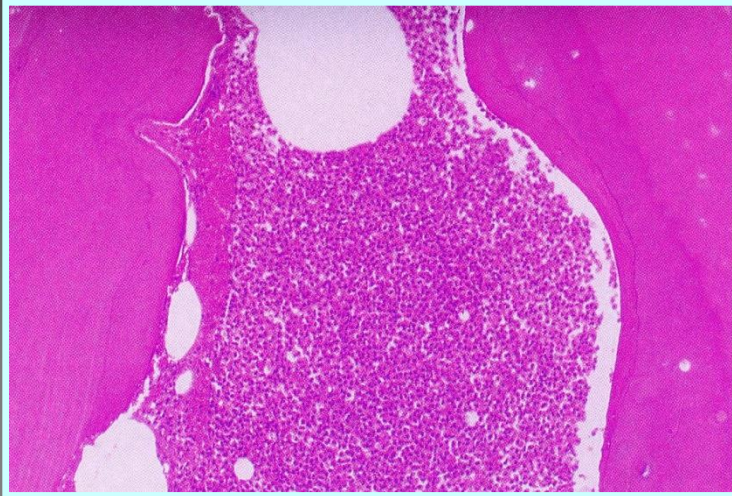
OSTEOPOROSIS



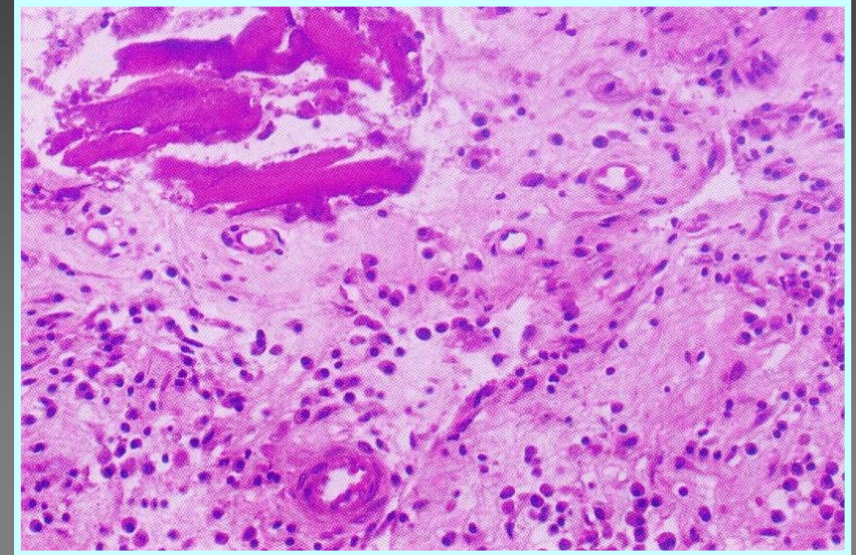
OSTEOMALACIA

PATHOLOGY OF BONES AND CARTILAGES

OSTEOMYELITIS



PURULENT OSTEOMYELITIS



OSTEOMYELITIS CHRONICA FIBROSA



NECROTIC AREAS IN BONE

BACTERIAL OSTEOMYELITIS

CONSEQUENCES:

- A. SEPTICOPYAEMIA
- B. SECONDARY AMYLOIDOSIS

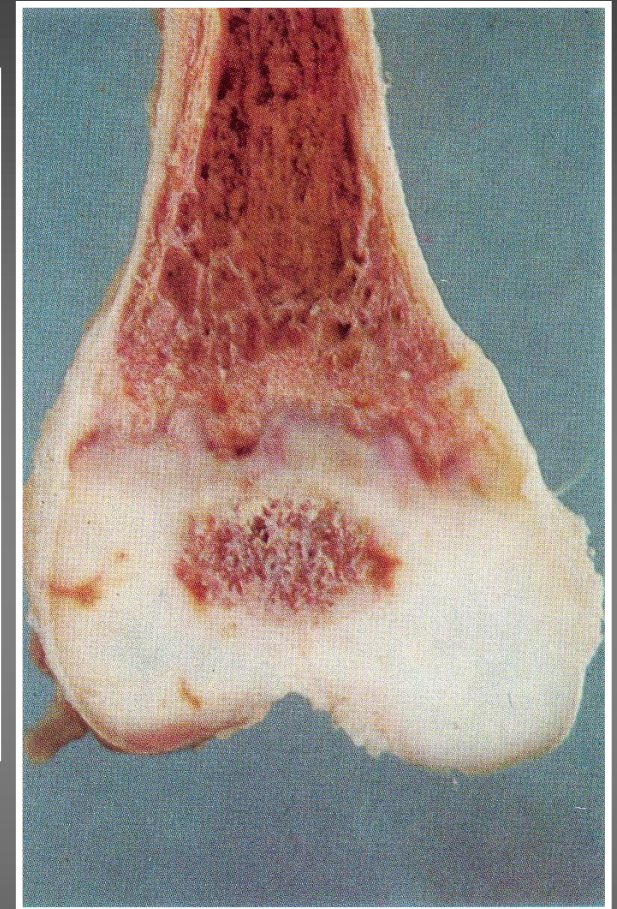
PATHOLOGY OF BONE AND CARTILAGES

OSTEOMYELITIS



TUBERCULOUS OSTEOMYELITIS

SECONDARY TBC, MAINLY
HEMATOGENOUS, CHRONIC, TREATMENT
OFTEN FAILS, IN VERTEBRAL COLUMN

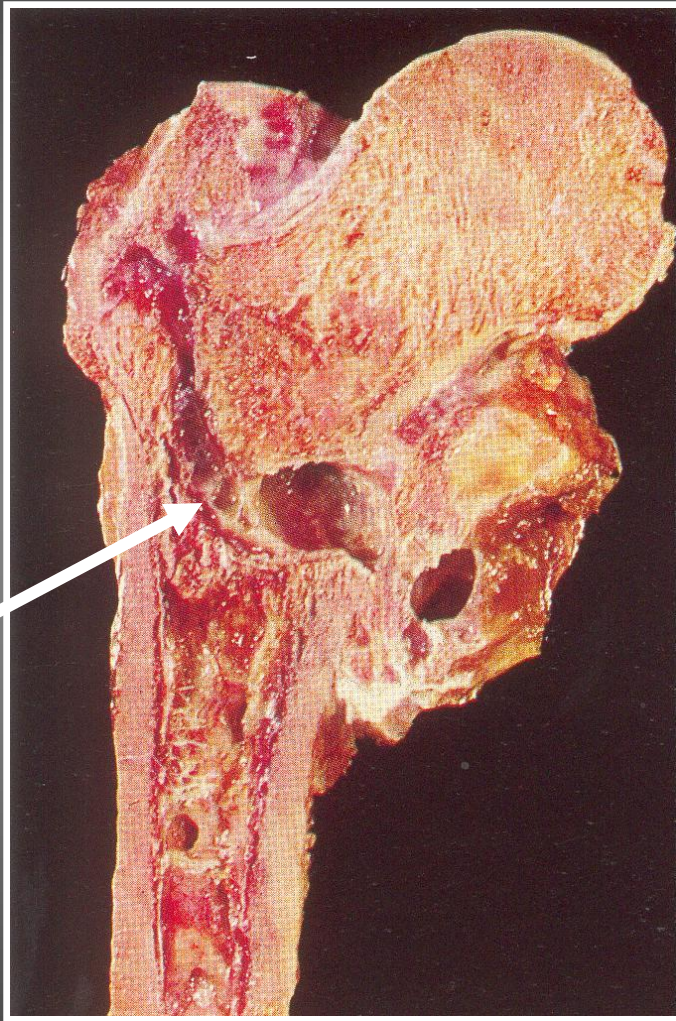


LUETIC OSTEONCHONDRITIS; LUES

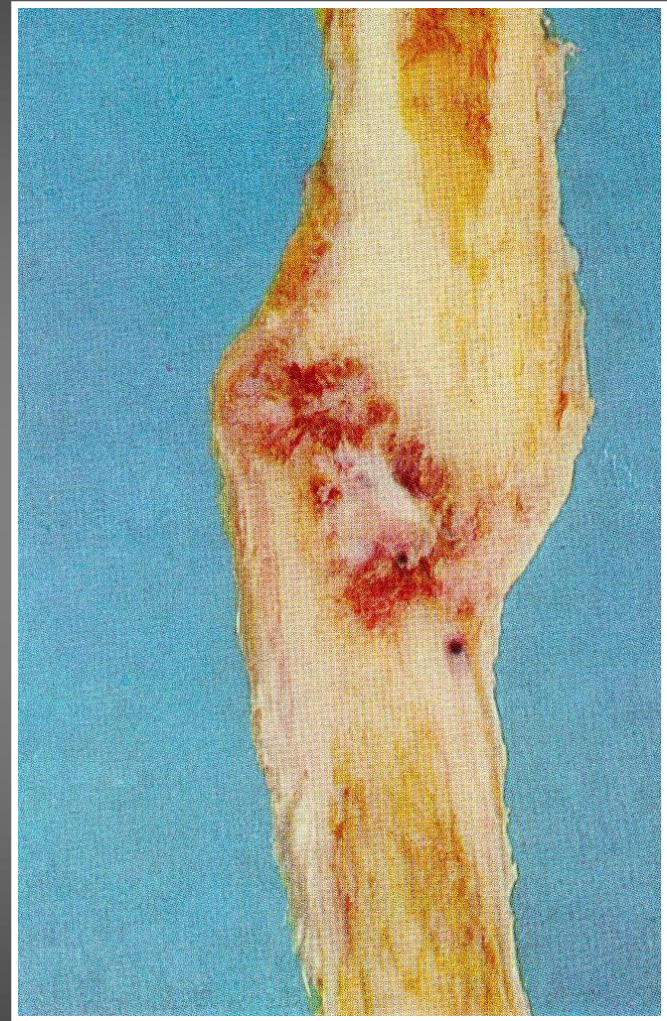
CONGENITAL LUES; LESS OFTEN
ACQUIRED AS
OSTEOCHONDRITIS,
PERIOSTITIS OR GUMMAS IN
MEDULLA

PATHOLOGY OF BONES AND CARTILAGES

FRACTURE HEALING



TROCHANTERIAN FRACTURE

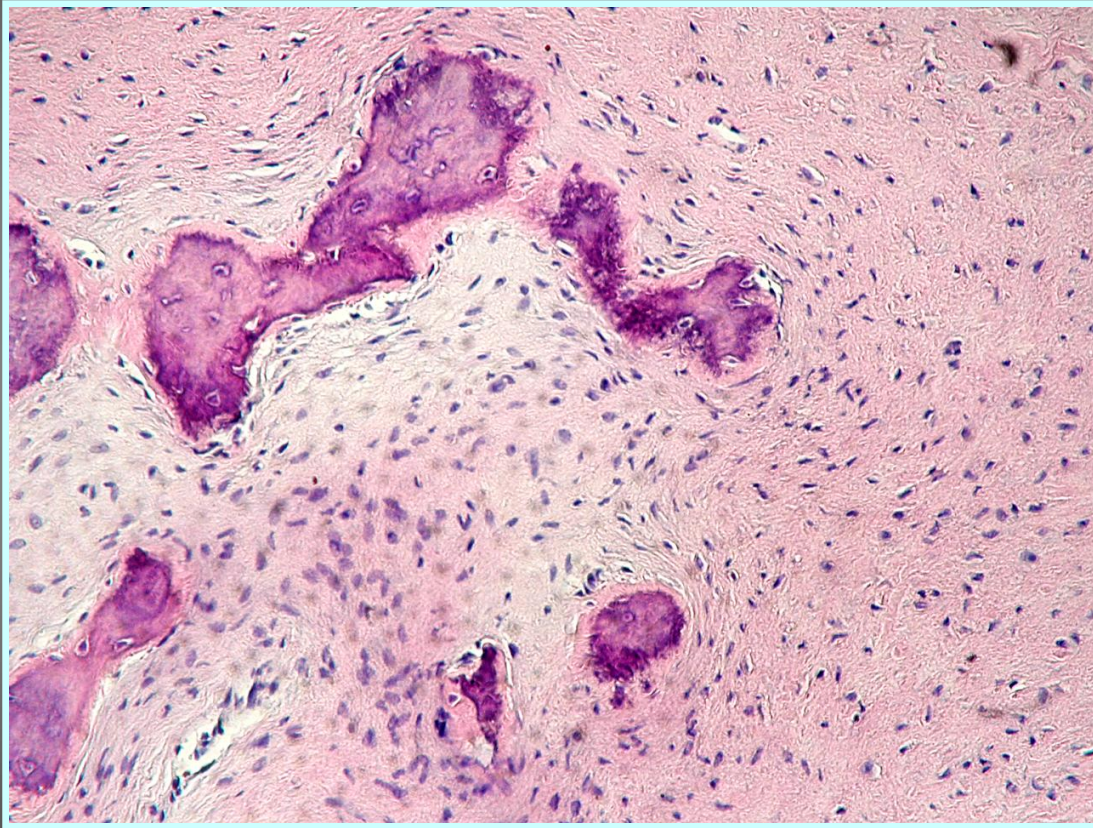


**PSEUDOARTHROS AFTER THE
FRACTURE OF THE LONG BONE**

- **Pseudarthrosis, Pseudoarthrosis**
- a pathological entity characterized by nonosseous union of bone fragments of a fractured bone due to inadequate immobilization leading to existence of the 'false joint' that gives the condition its name.

PATHOLOGY OF BONES AND CARTILAGES – TUMOR-LIKE CONDITIONS

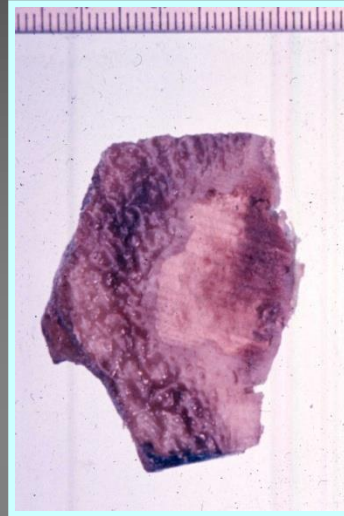
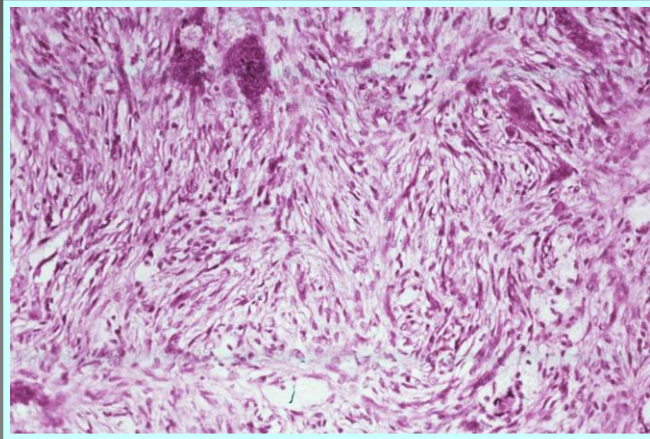
FIBROUS DYSPLASIA – ETIOLOGY UNKNOWN



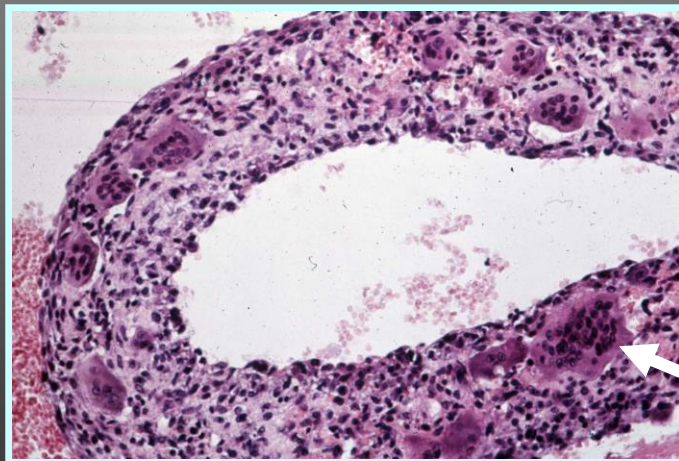
THE REPLACEMENT OF BONE TISSUE BY CONNECTIVE TISSUE. THREE FORMS: a. LOCALIZED, b. MULTIFOCAL
c. ALBRIGHT DISEASE.

PATHOLOGY OF BONES AND CARTILAGES – TUMOR-LIKE CONDITIONS

FIBROUS DYSPLASIA



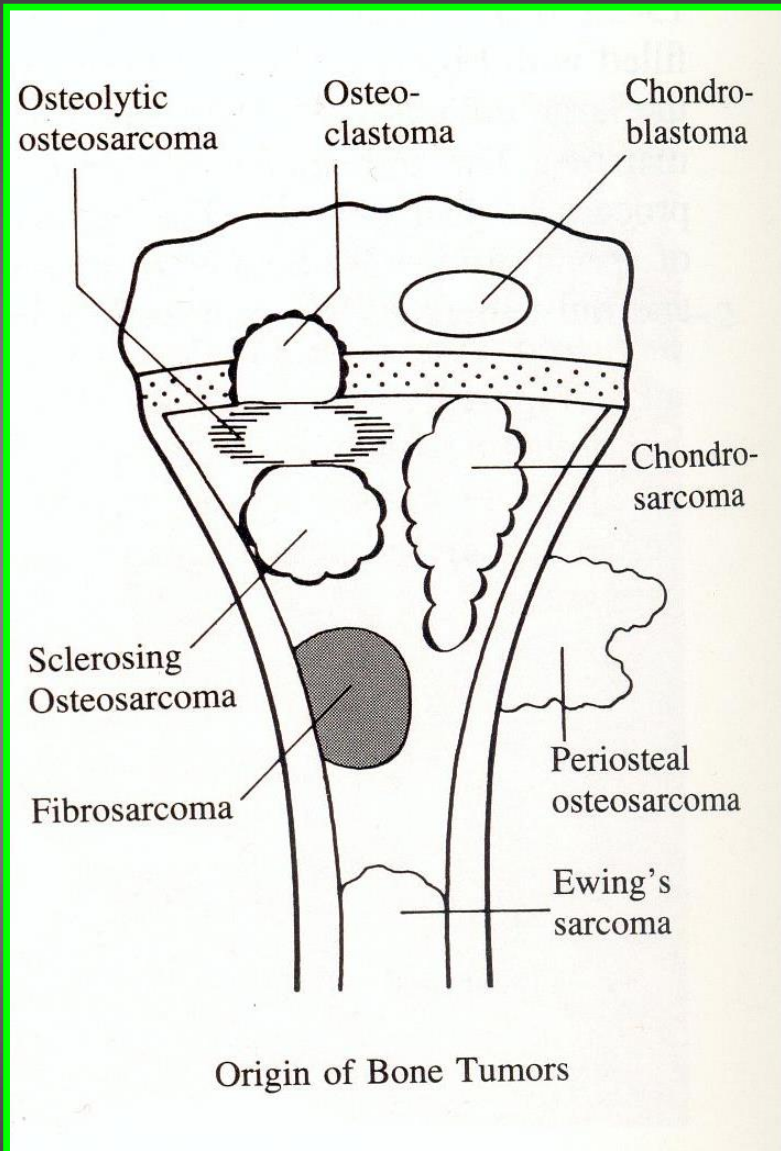
THE REPLACEMENT OF BONE TISSUE BY CONNECTIVE TISSUE. 3 FORMS: a. LOCALIZED
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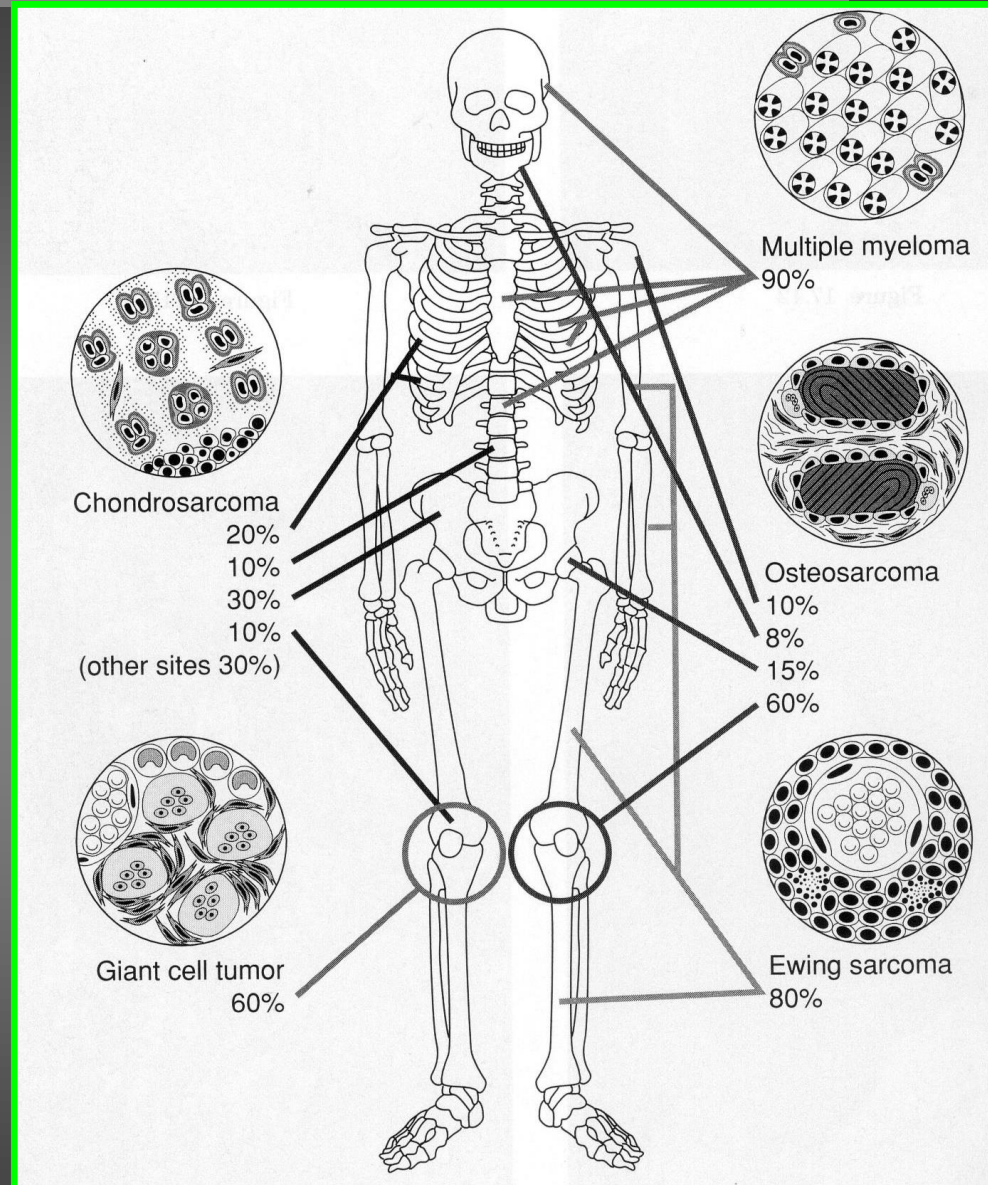
ANEURYSMAL BONE CYST

POST TRAUMATIC +
OSTEOCLASTS

PATHOLOGY OF BONE AND CARTILAGES - TUMORS



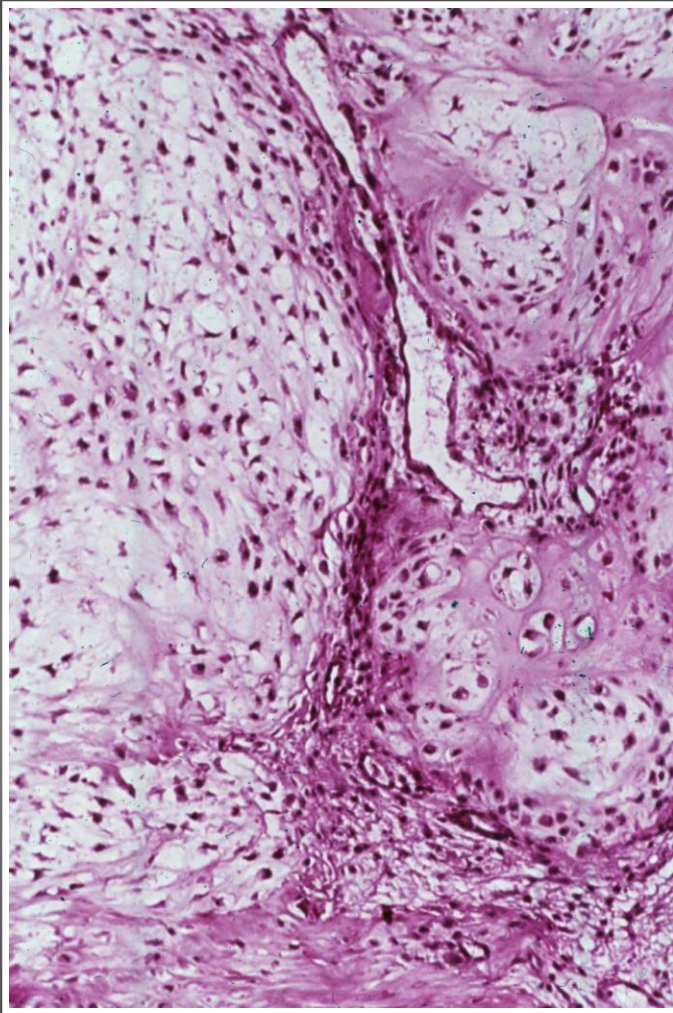
HISTOGENESIS OF BONE TUMORS



THE FREQUENCY OF BONE TUMORS IN DIFFERENT PARTS OF BONE SYSTEM

PATHOLOGY OF BONE AND CARTILAGES - TUMORS

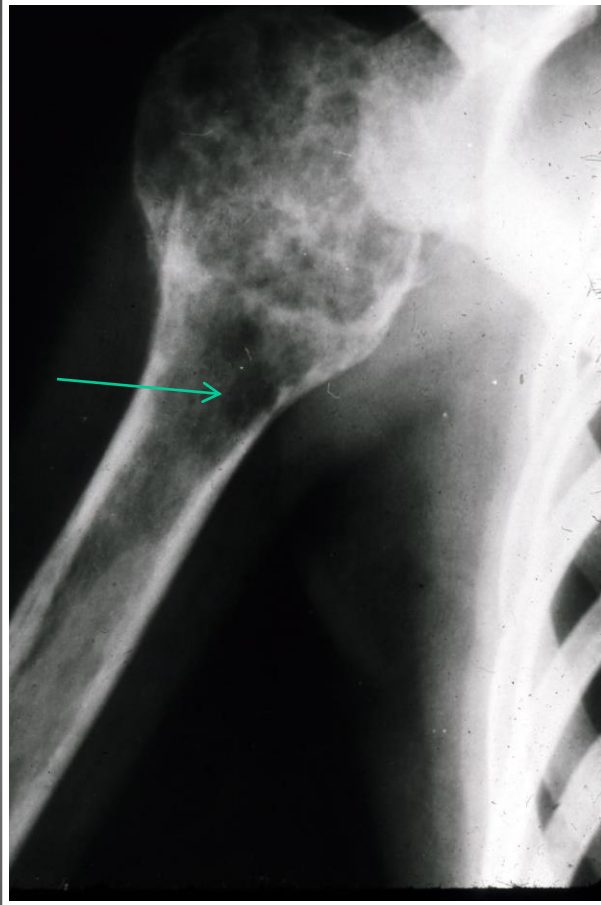
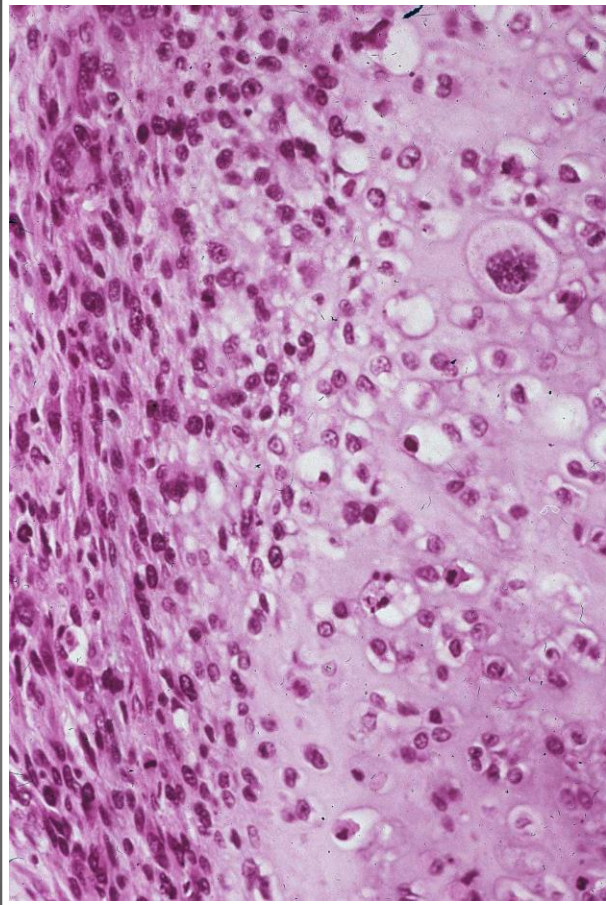
CHONDROMA



CHONDROMA MOST OFTEN SOLITARY TUMOR, LESS OFTEN MULTIPLE (**OLLIER DISEASE**)

PATHOLOGY OF BONES AND CARTILAGES - TUMORS

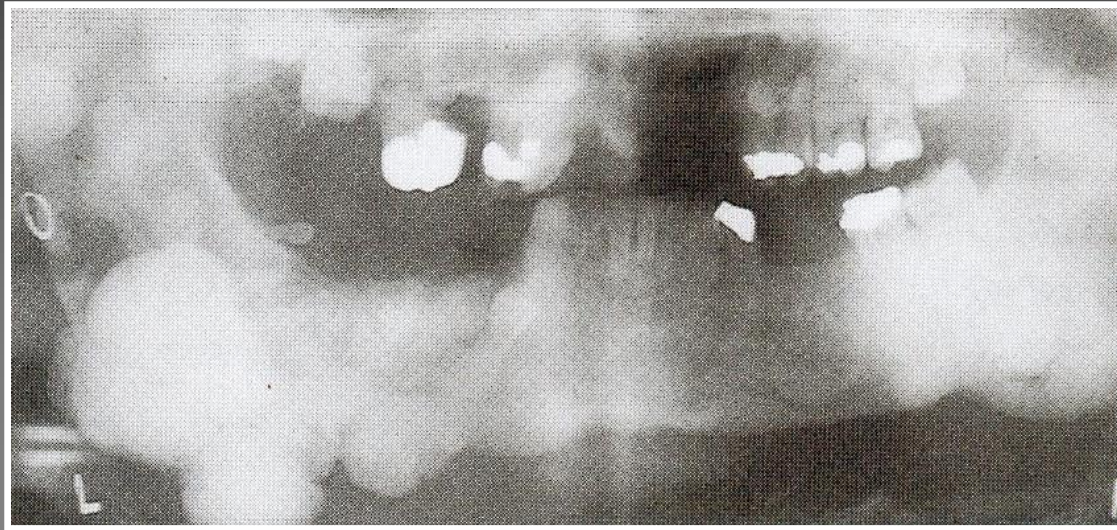
CHONDROSARCOMA



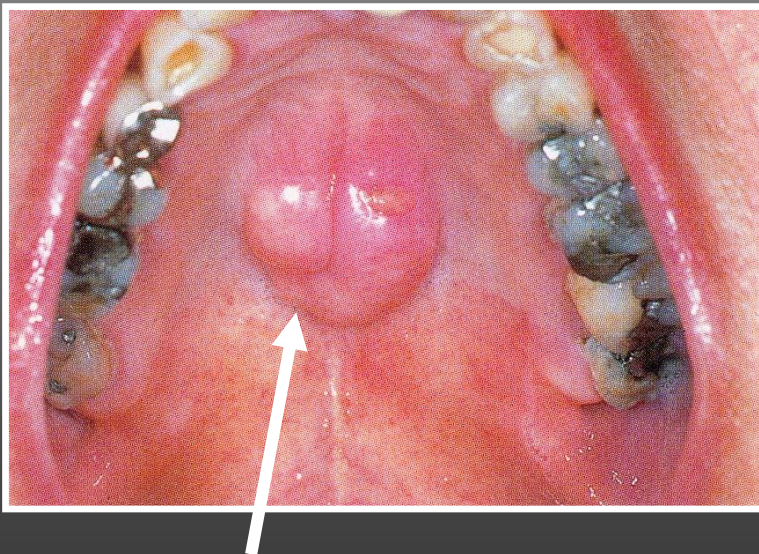
APPROXIMATELY 20% OF PRIMARY MALIGNANT TUMORS IN BONES, 1/5 DEVELOPS FROM CHONDROMA, MORE OFTEN IN MEN. SLOW GROWTH – METASTASES ARE VERY COMMON IN LUNGS

PATHOLOGY OF BONES AND CARTILAGES - TUMORS

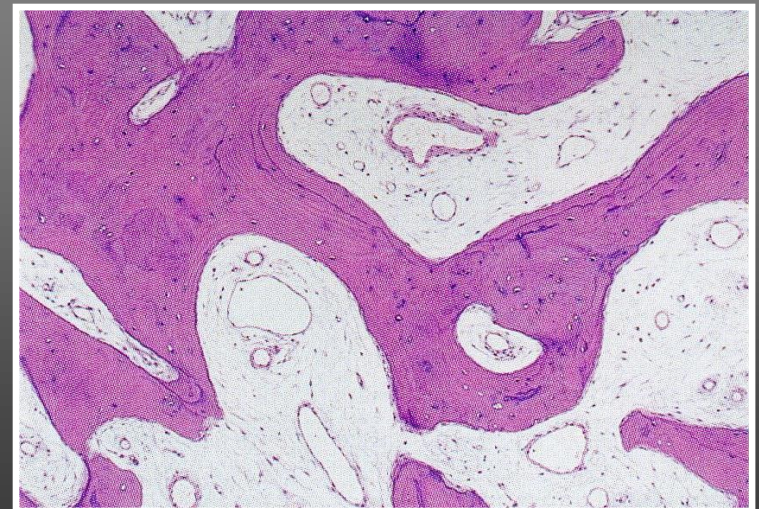
OSTEOMA



A CASE OF GARDNER SYNDROME: OSTEOMAS, INTESTINAL POLYPS, ADDITIONAL TEETH

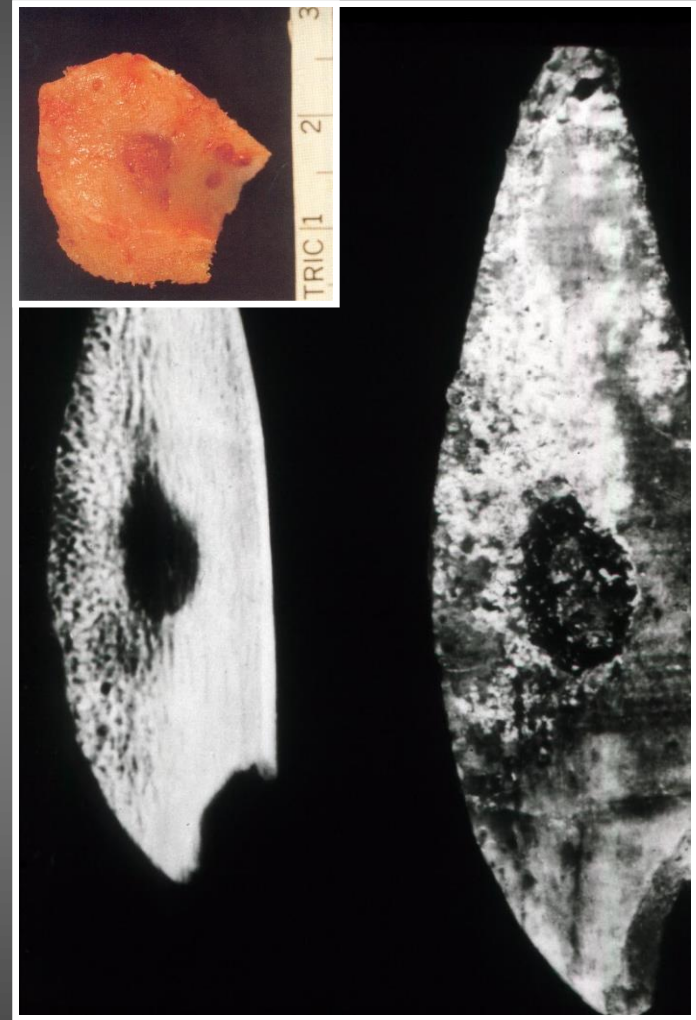
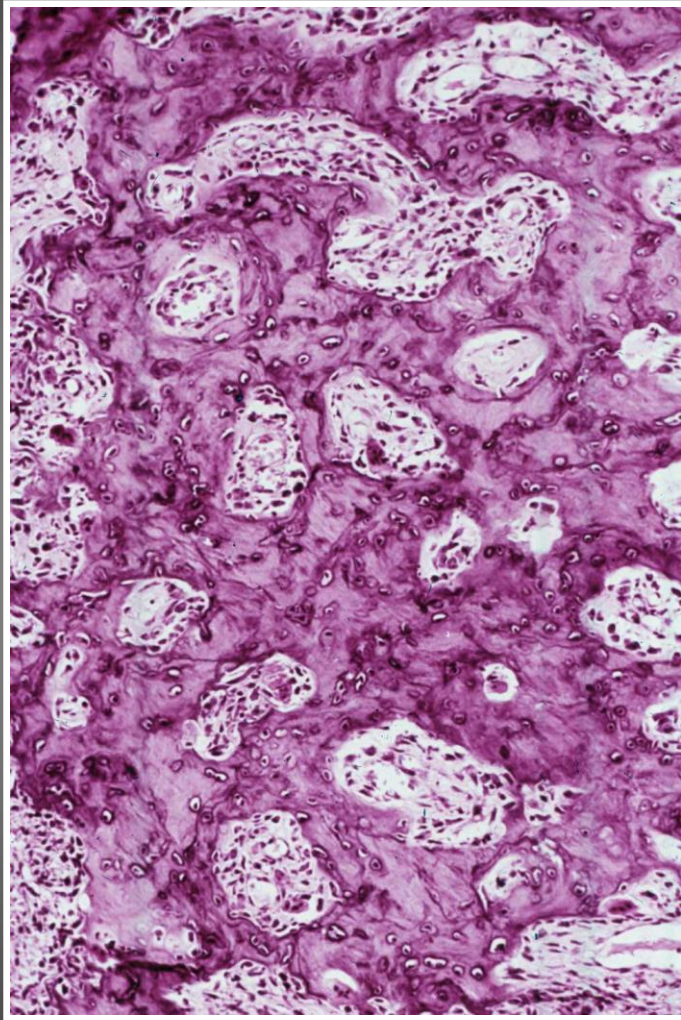


OSTEOMA



PATHOLOGY OF BONES AND CARTILAGES - TUMORS

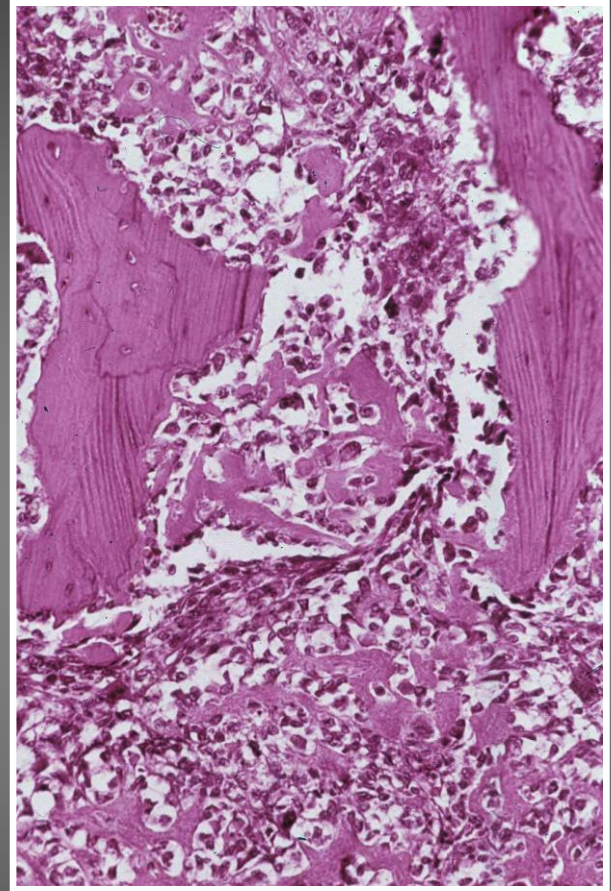
OSTEOID OSTEOMA



SMALL IN SIZE AND BENIGN. PATIENTS ARE MOST OFTEN IN THE 1st OR 2nd DECADE OF LIFE. NIGHT PAINS ARE FREQUENT.

PATHOLOGY OF BONES AND CARTILAGES - TUMORS

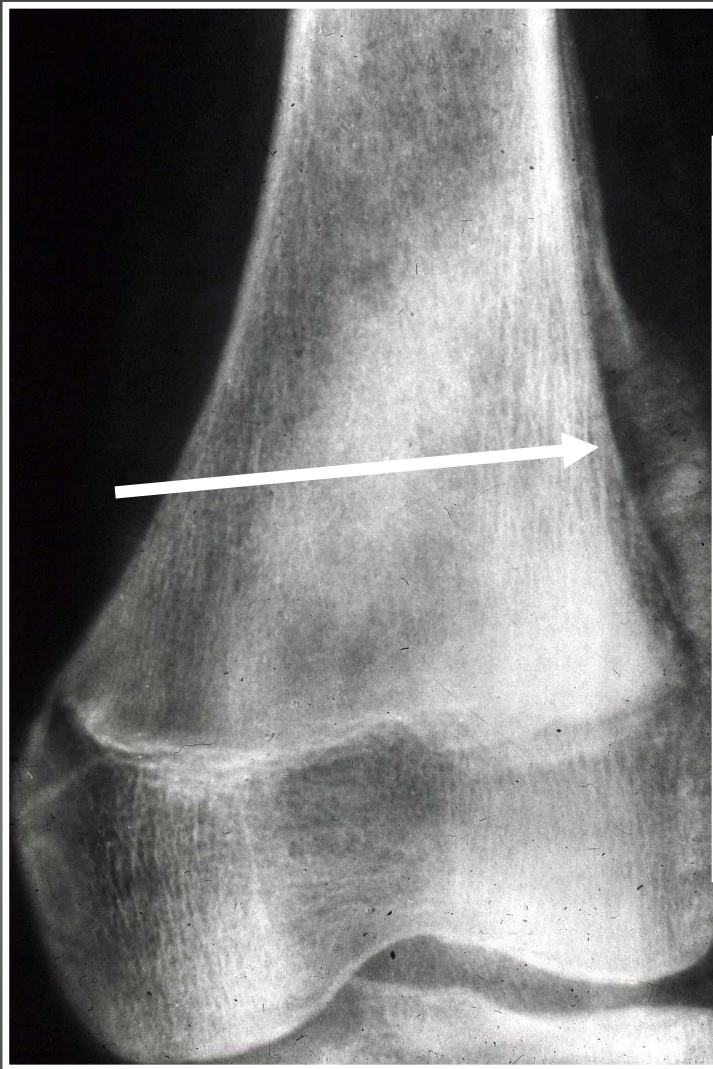
OSTEOSARCOMA



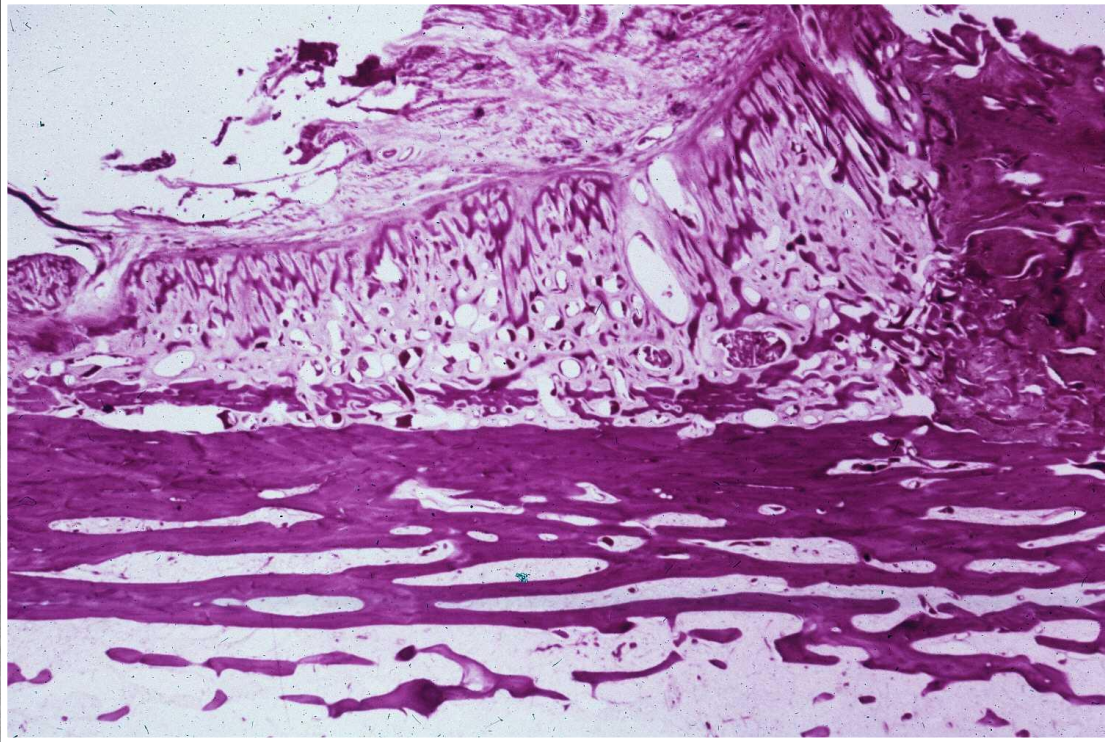
OSTEOID-PRODUCING TUMOR, MAINLY IN MEN BEFORE 20.

KNEE IS THE MOST OFTEN SITE OF THIS SARCOMA

PATHOLOGY OF BONES AND CARTILAGES -TUMORS



CODMAN TRIANGLE



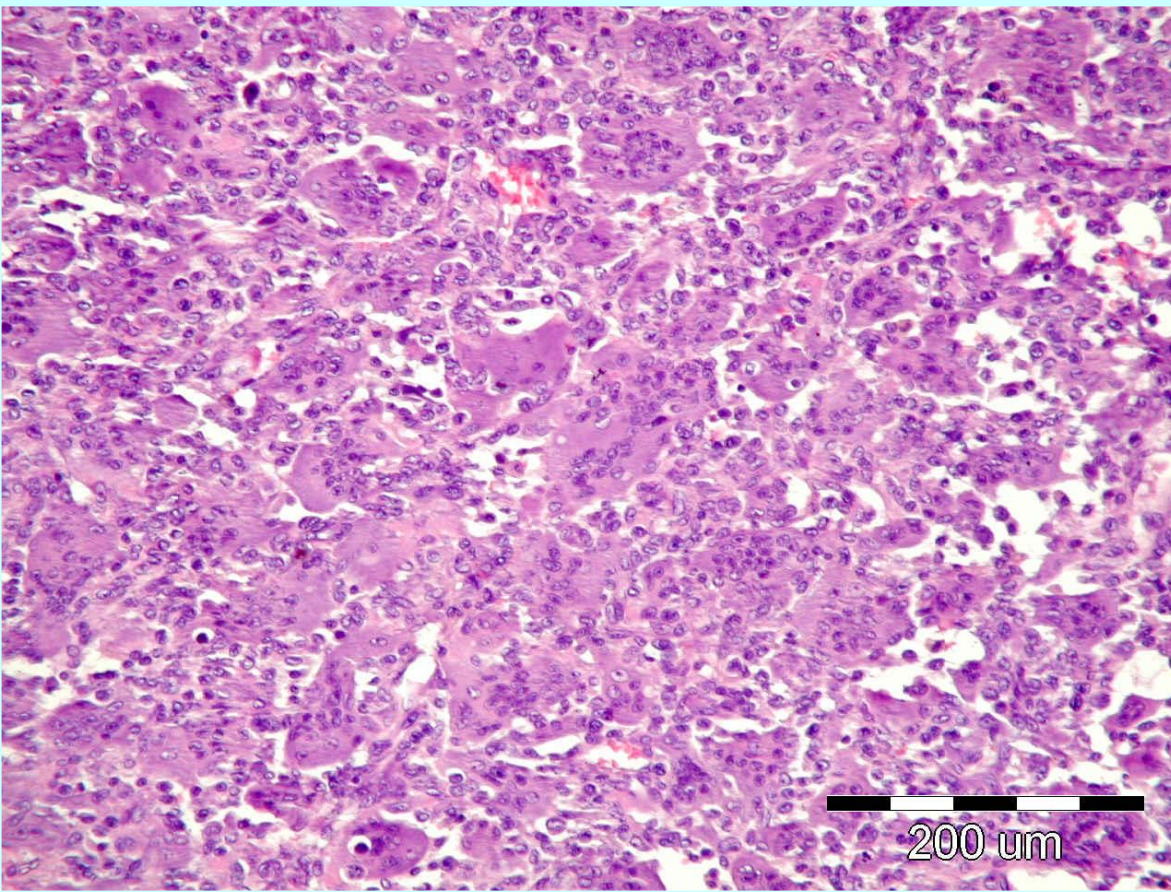
CODMAN TRIANGLE IN OSTEOSARCOMA

A TYPICAL FEATURE: DEMARCATION LINE (→) AND THE GAPS IN PERIOSTEUM IN MALIGNANT TUMORS OF BONES

- **Codman triangle** is a term used to describe the triangular area of new subperiosteal bone that is created when a lesion, often a tumour, raises the periosteum away from the bone

PATHOLOGY OF BONES AND CARTILAGES - TUMORS

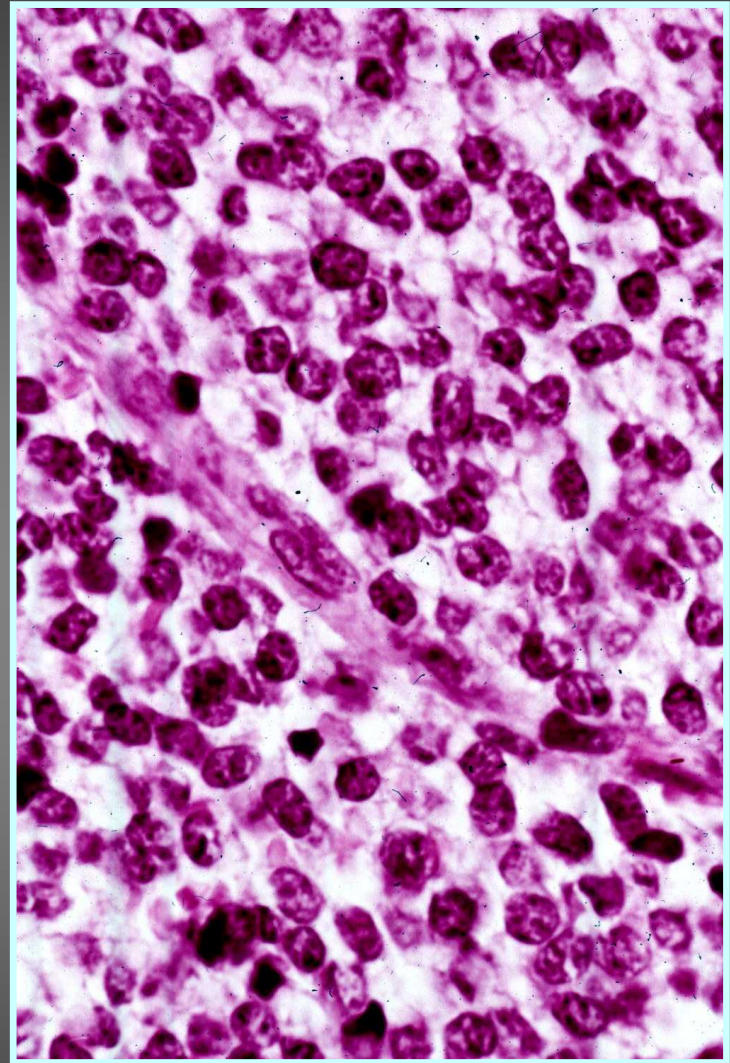
GIANT-CELL TUMOR - OSTEOCLASTOMA



APPROX. 5% OF BONE TUMORS: MAINLY BETWEEN 20 AND 55. LOWER LIMBS (KNEE). HISTOGENESIS IS UNKNOWN. COMMON PATHOLOGICAL FRACTURES

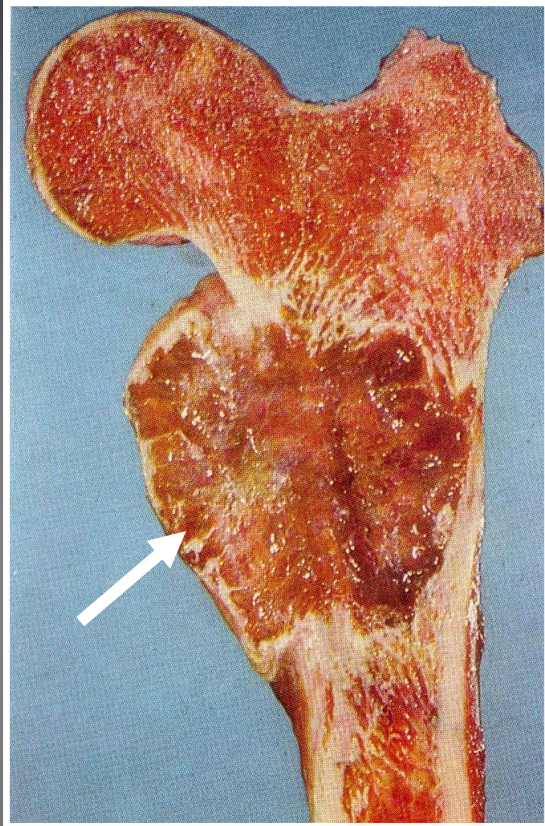
PATHOLOGY OF BONES AND CARTILAGES - TUMORS

SARCOMA EWING

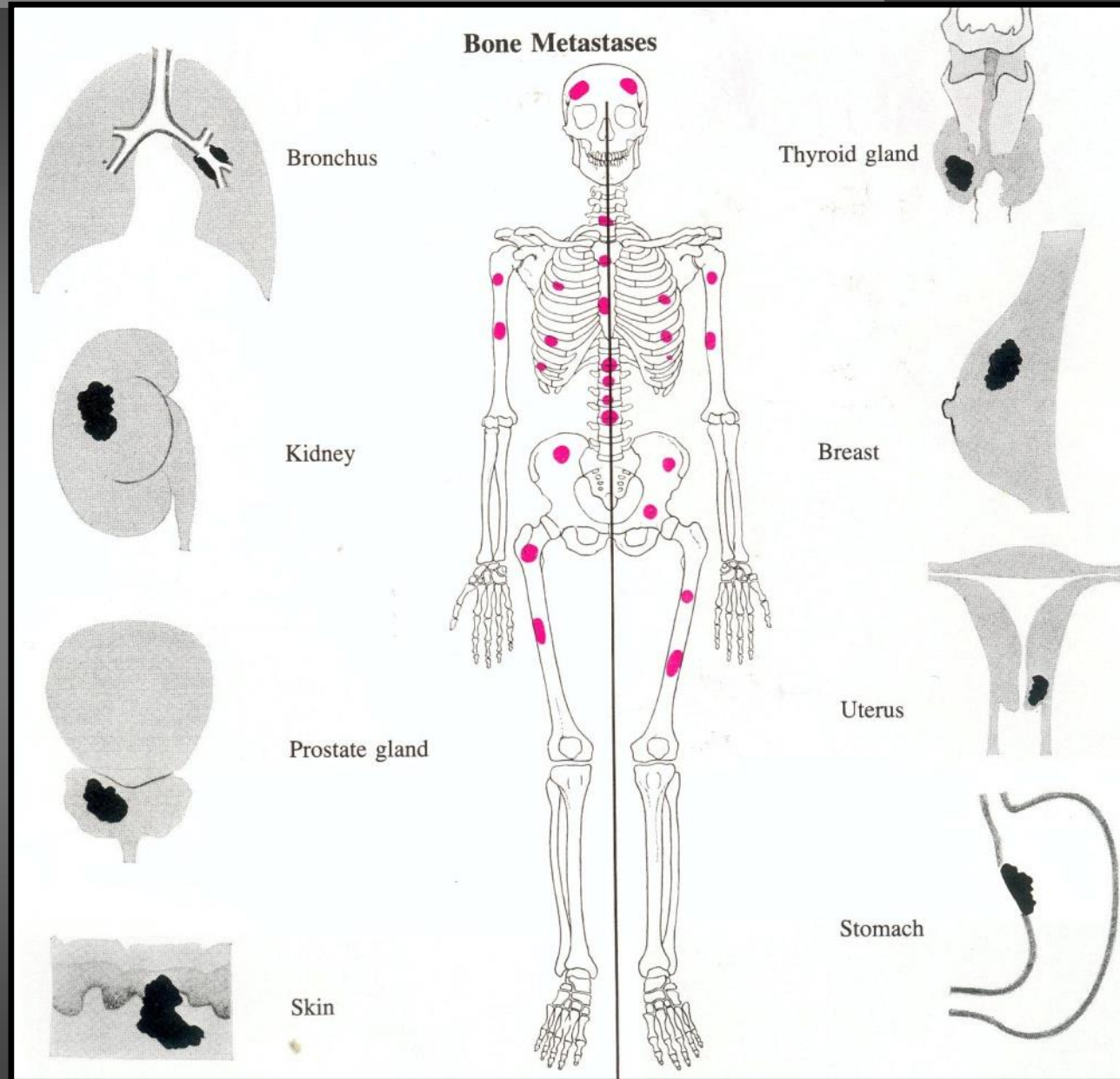


HIGHLY MALIGNANT; HISTOGENESIS IS UNKNOWN; RAPID GROWTH; UP TO 30 YEARS;
CELLS CONTAIN GLYCOGEN

PATHOLOGY OF BONES AND CARTILAGES - TUMORS



**OSTEOLYTIC
METASTASIS OF THE
LUNG CANCER TO
FEMUR**



WHERE THEY METASTASIZE TO

JOINTS - GANGLION

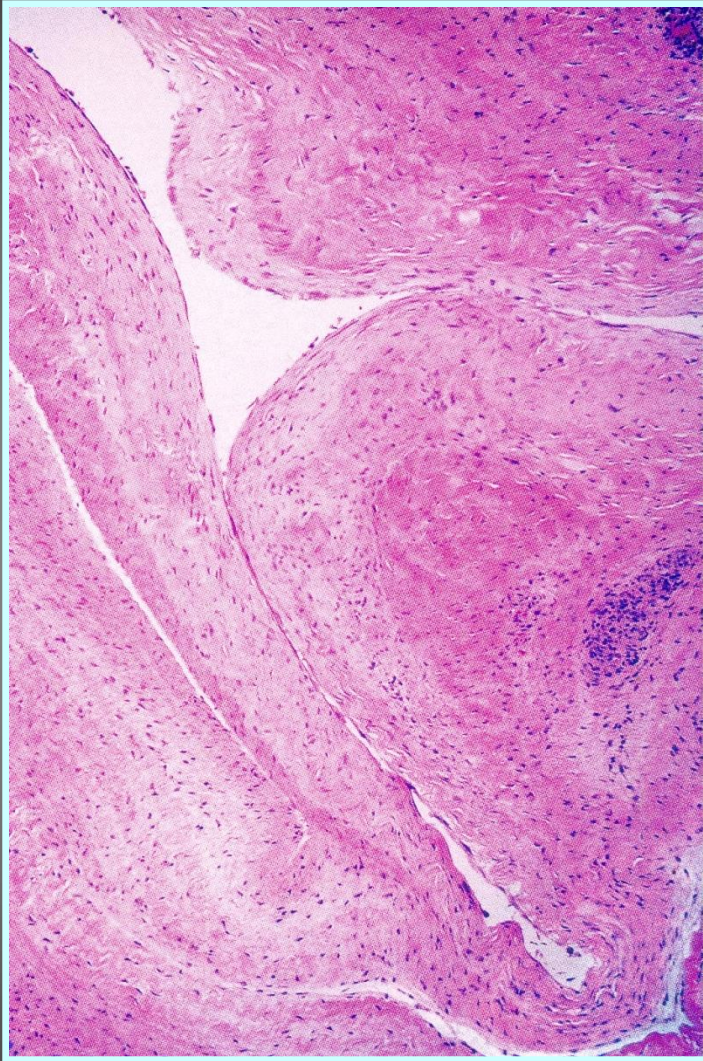
- Common tumor-like lesion arising from soft tissue, caused by mucoid degeneration of joint capsule, tendon or tendon sheath
- Small cyst-like mass (no epithelial lining) near joint capsule or tendon sheath
- Common site is wrist, also hand and foot, rarely in intratendinous region
- May cause pain, weakness, bone changes, partial disability of joint
- May be due to injury or overuse of joint
- May be multilocular; **Fluid is similar to synovial**

GANGLION

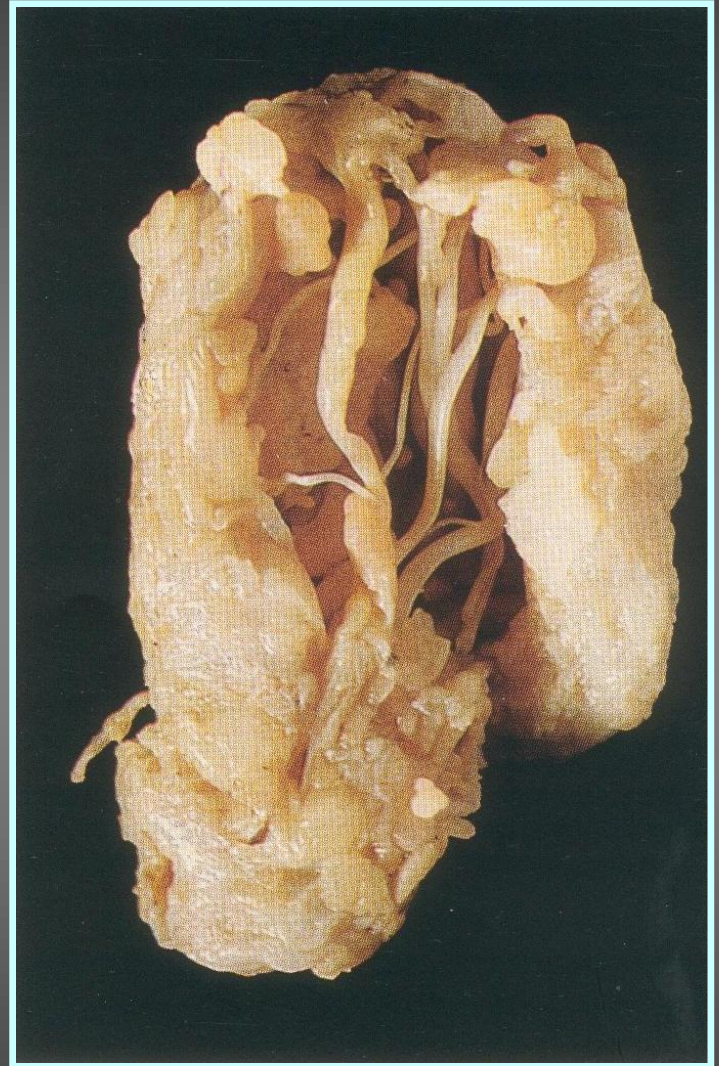


Usually does not communicate with joint space; rarely is intraosseous (medial malleolus of tibia)

PATHOLOGY OF JOINTS



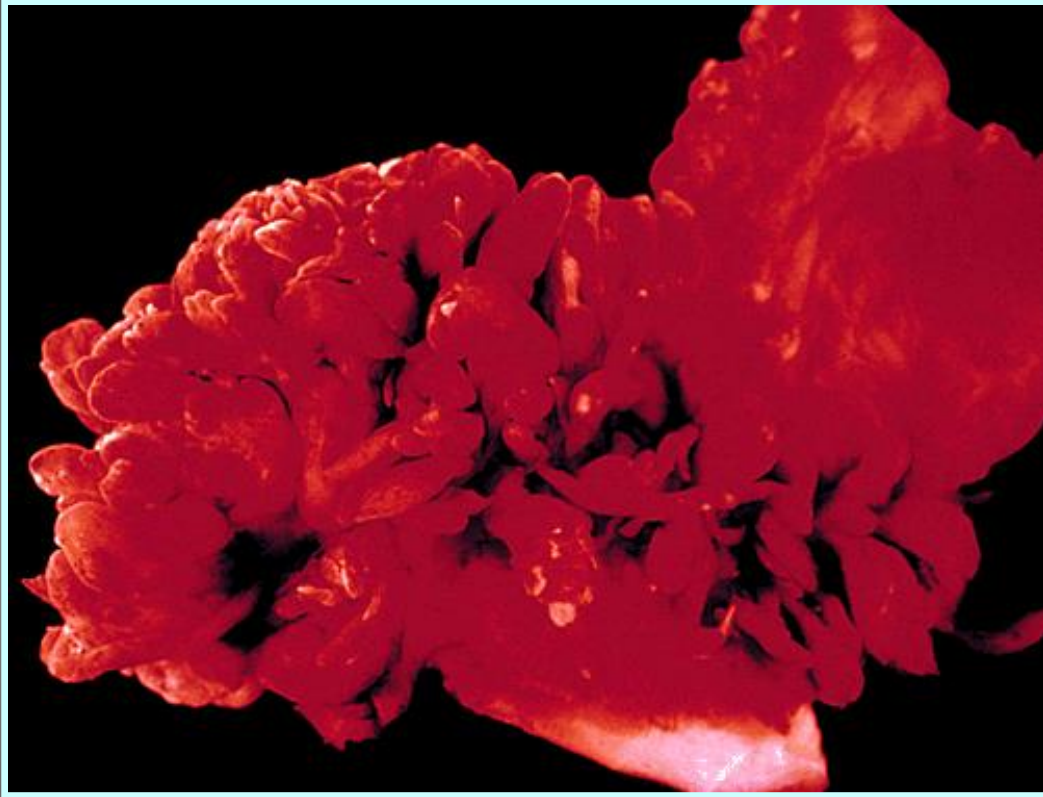
GANGLION



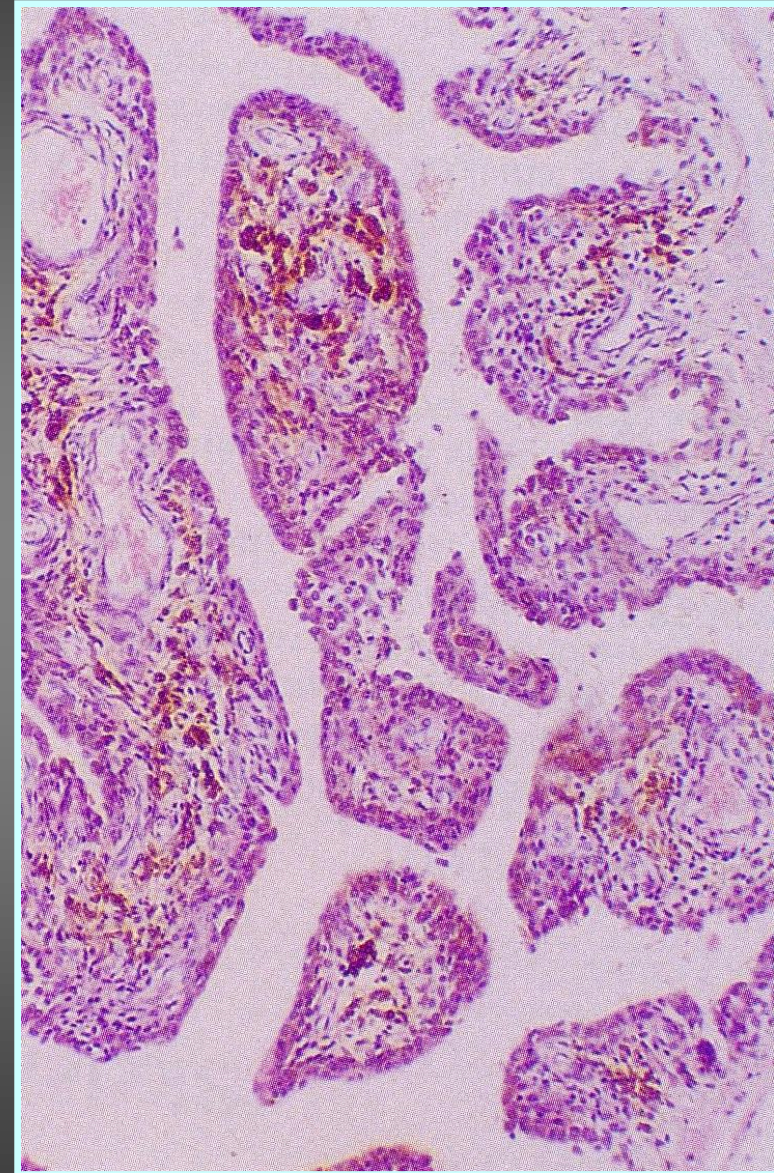
BURSITIS

PATHOLOGY OF JOINTS

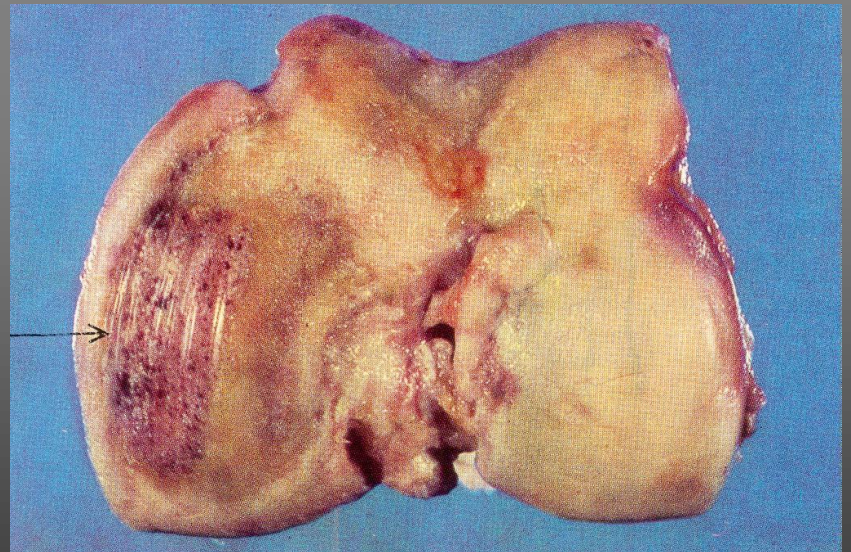
PIGMENTED VILLONODULAR SYNOVITIS



PROLIFERATIVE INFLAMMATION AFFECTING
SYNOVIAL TISSUE. ETIOLOGY IS UNKNOWN.



PATHOLOGY OF JOINTS



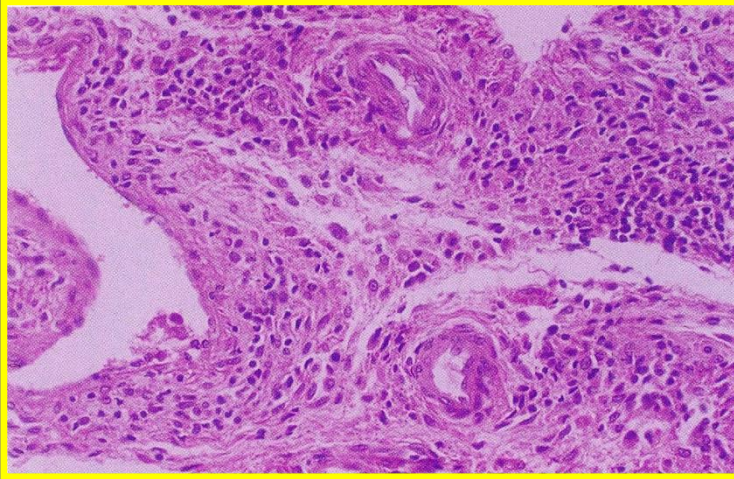
(OSTEO)ARTHRITIS DEFORMANS – EARLY AND LATE PHASE

- **Definition:** A progressive, degenerative joint disease, the most common form of arthritis, especially in older persons. The disease is thought to result not from the aging process but from biochemical changes and biomechanical stresses affecting articular cartilage. In the foreign literature it is often called osteoarthritis deformans.

**Synonym(s): Arthritis, Degenerative /
Osteoarthritis / Osteoarthritis Deformans /
Arthritides, Degenerative**

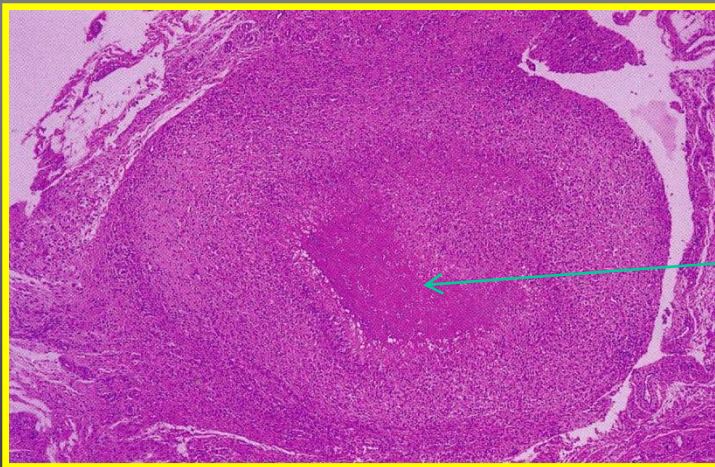
PATHOLOGY OF JOINTS

RHEUMATOID ARTHRITIS



LYMPHOCYTES AND PLASMA
CELLS IN SYNOVIA

ETIOLOGY IS UNCLEAR.
LEADS TO
DEFORMATIONS OF
JOINTS, FIBROSIS AND
REGRESSIVE LESIONS.



RHEUMATOID NODULE IN SYNOVIA

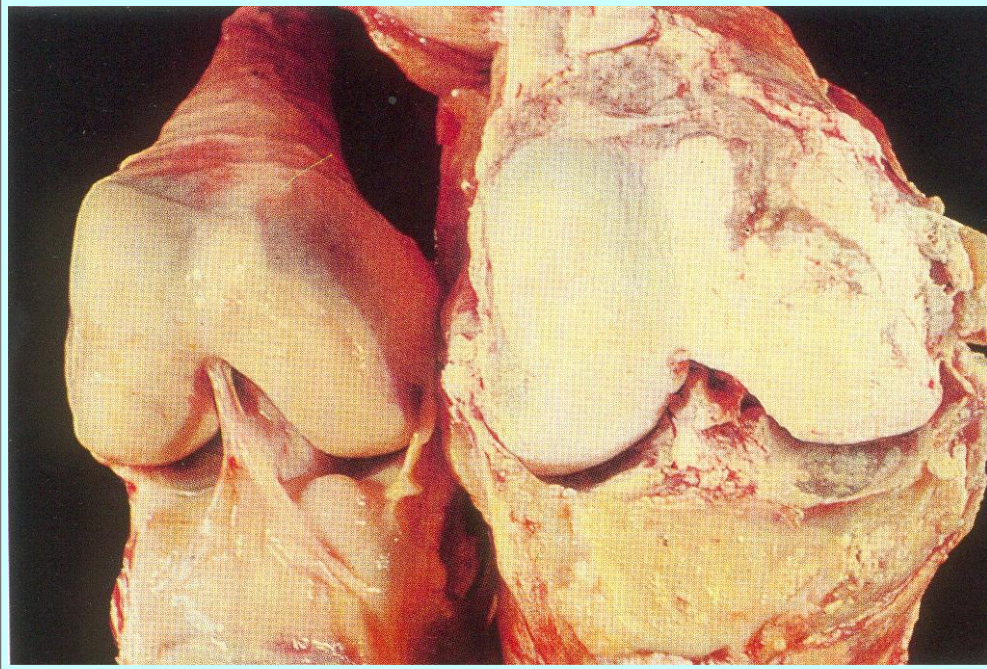


ANKYLOSIS OF
INTERPHALANGEAL
JOINT

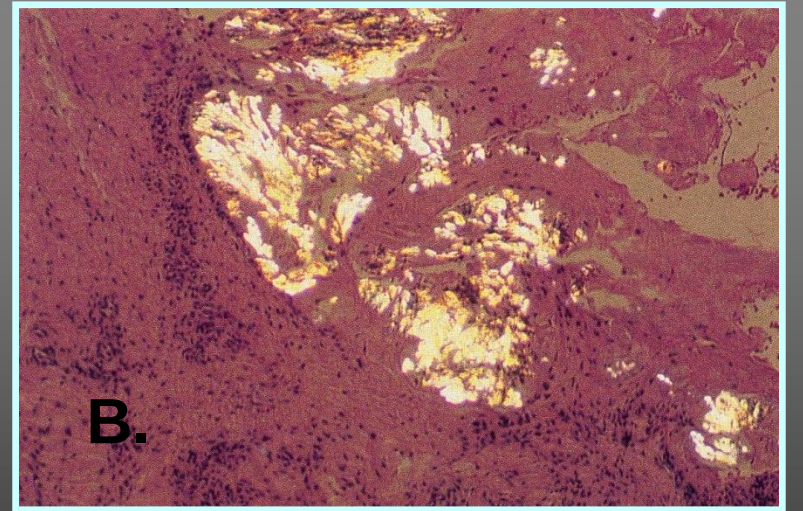
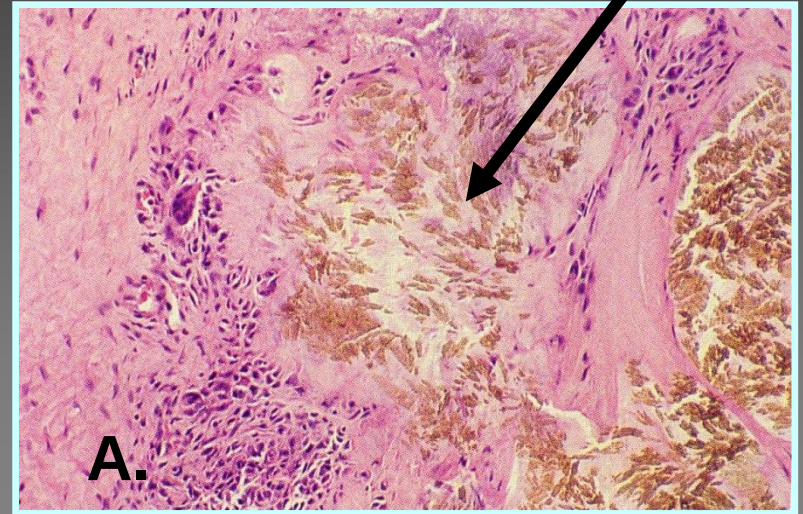
- **ANKYLOSIS**, or **ANCHYLOSIS** (from Greek ἀγκύλος, bent, crooked) is a stiffness of a joint due to abnormal adhesion and rigidity of the bones of the joint, which may be the result of injury or disease

PATHOLOGY OF JOINTS

URIC ACID DIATHESIS

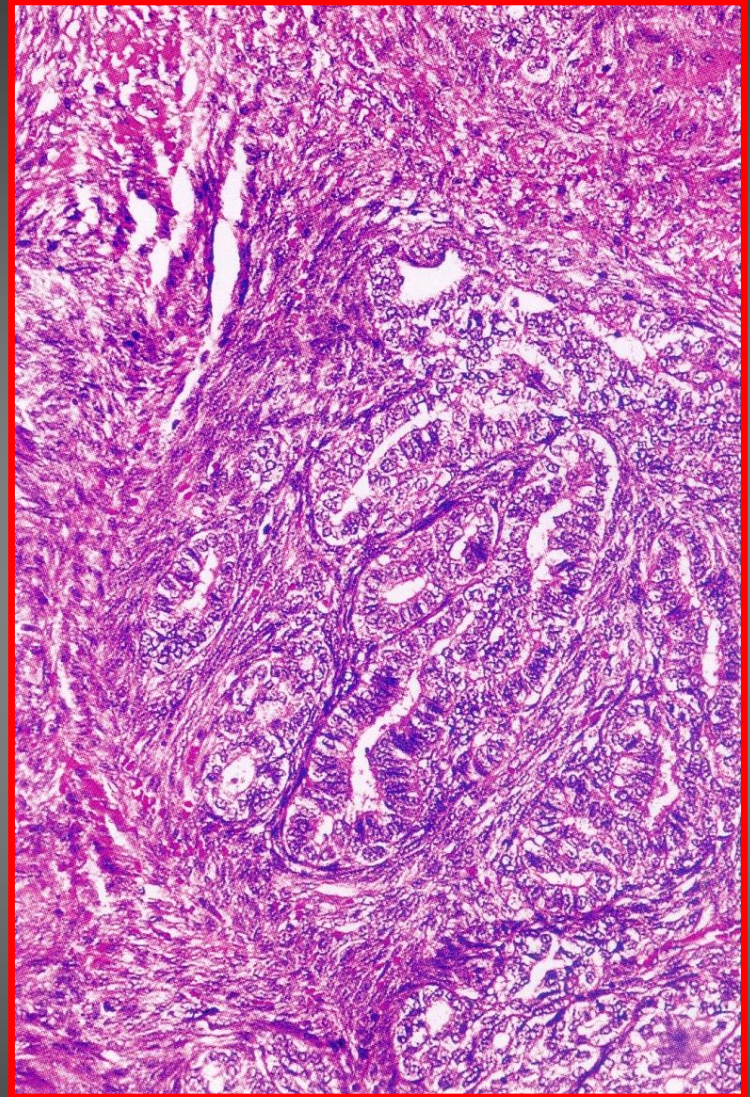
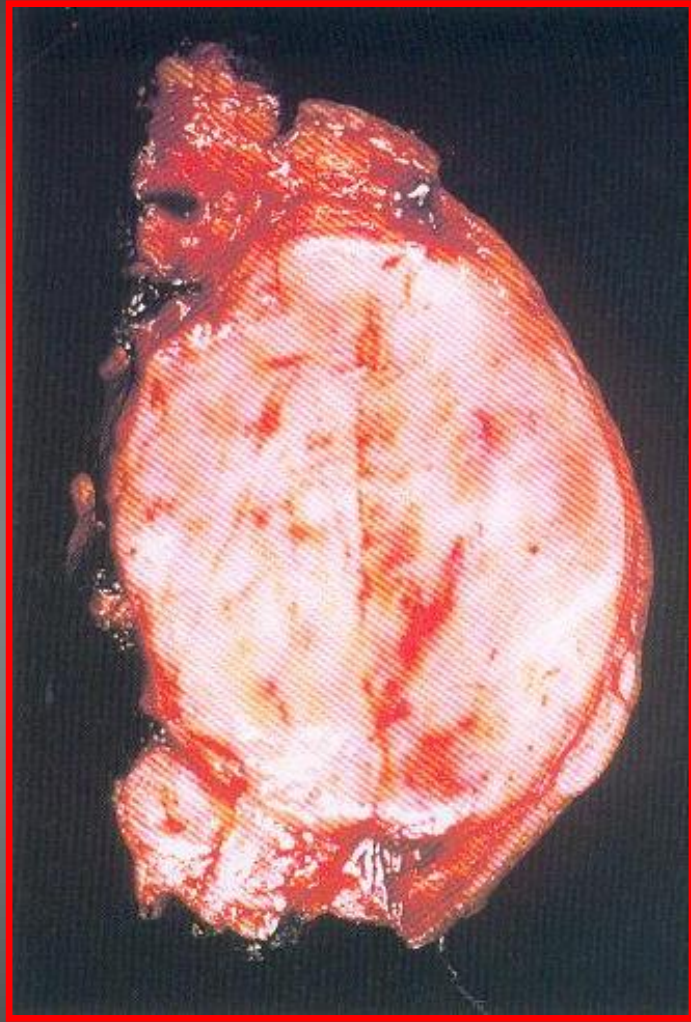


URATIC ARTHRITIS



URIC ACID DEPOSITS IN SYNOVIA
A. H&E, B. IN POLARIZING MICROSCOPE

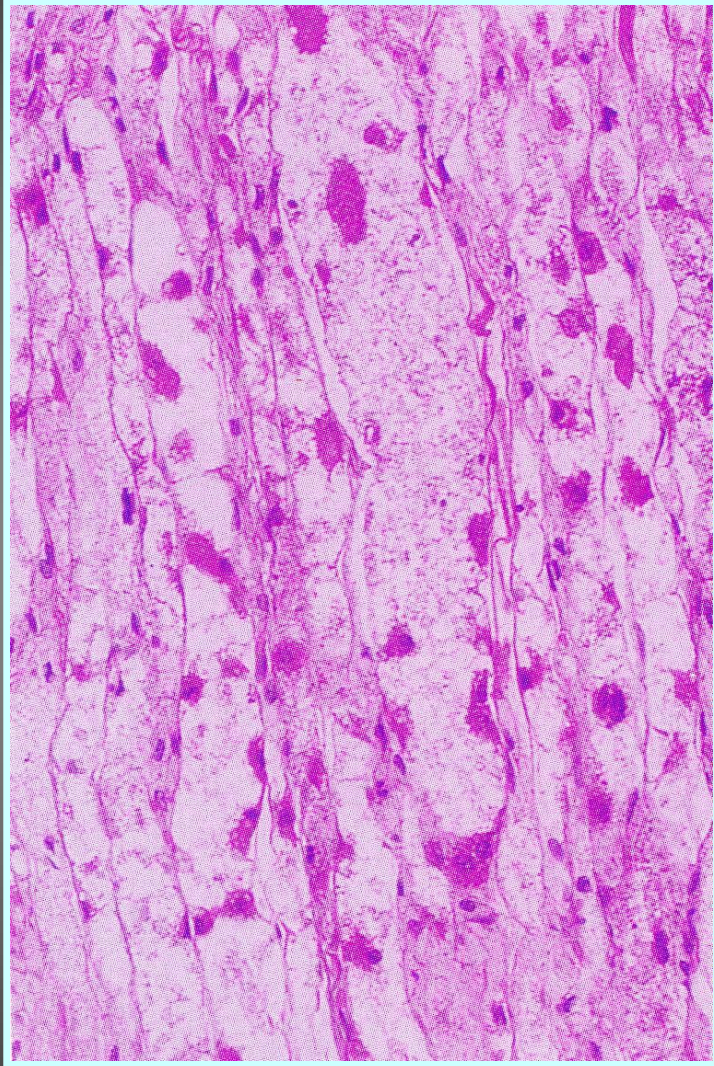
PATHOLOGY OF JOINTS



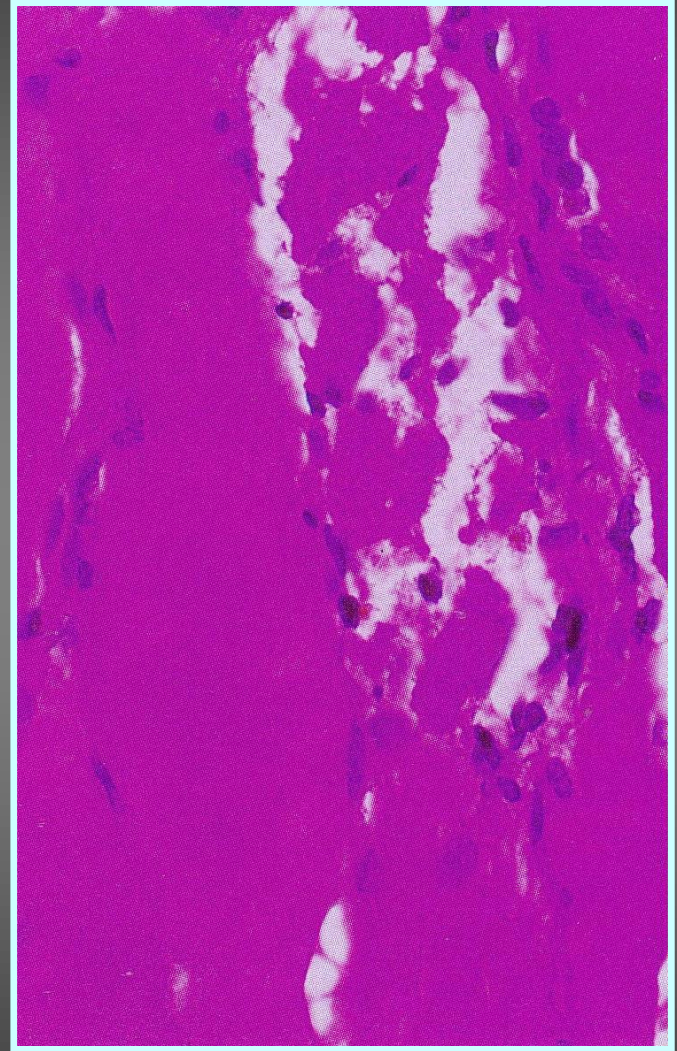
BIPHASIC SYNOVIAL SARCOMA

RARE TUMOR ORIGINATING FROM SYNOVIA; MAINLY IN THE LARGER JOINTS;
HIGHLY MALIGNANT

PATHOLOGY OF MUSCLES



GLYCOGENOSIS IN A MUSCLE

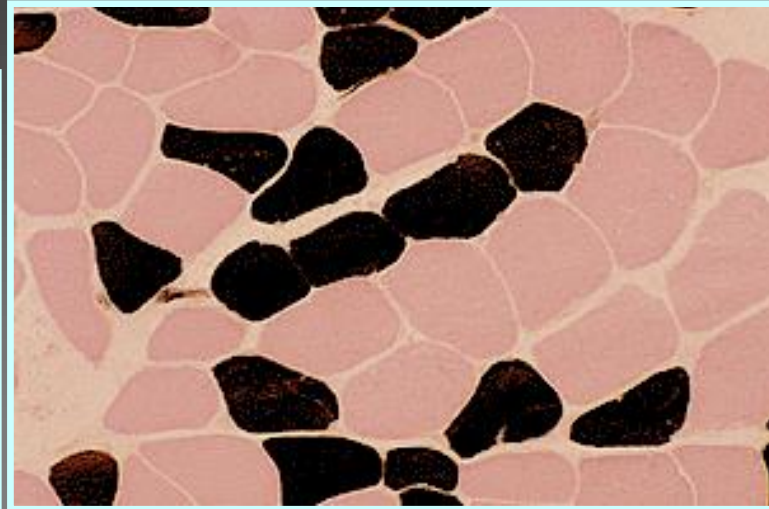
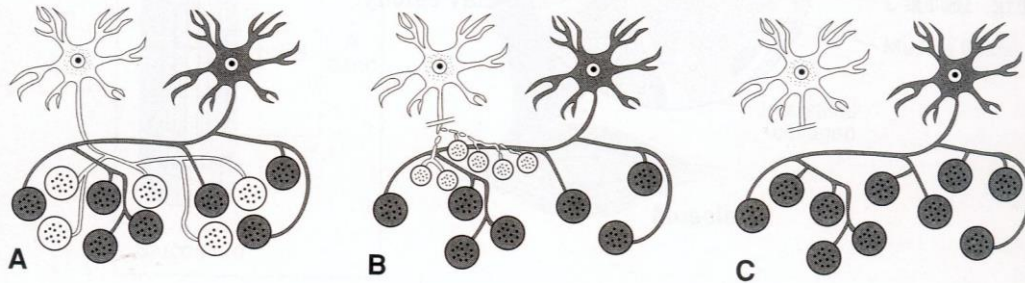


NECROSIS - MYOLYSIS

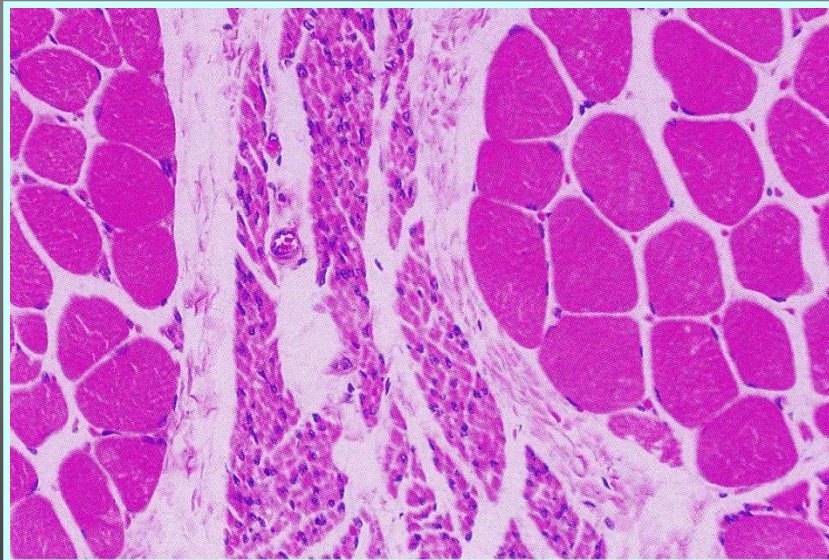
PATHOLOGY OF MUSCLES

NEURITIC ATROPHY OF MUSCLES

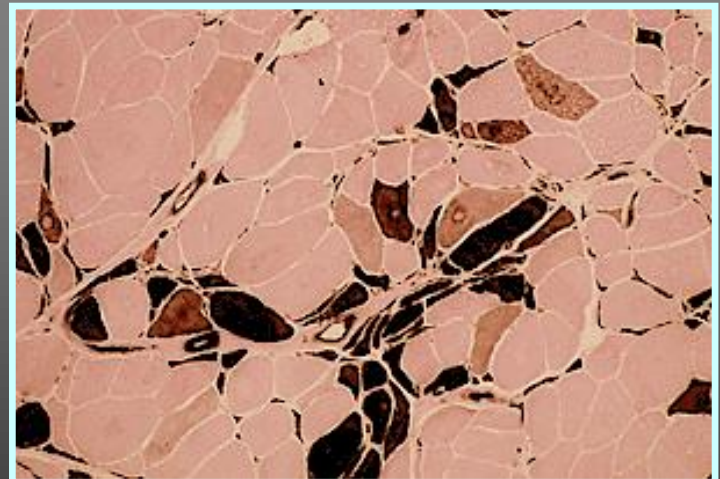
Diagram 18.2. Denervation of muscle due to nerve injury with subsequent reinnervation. A. The normal muscle is composed of a mixture of type I and type II fibers. **B.** Transection of nerve causes atrophy of denervated muscle cells. **C.** Reinnervation leads to fiber type grouping because the nerve sprouts reach muscle fiber in groups and thus make them all of the same type—either fast or slow.



NORMAL MUSCLE. ATPase REACTION. FIBRES ARE IN TWO COLORS



NEURITIC ATROPHY - WERDNIIG HOFFMANN DISEASE (PROGRESSIVE SPINAL AMYOTROPHY)



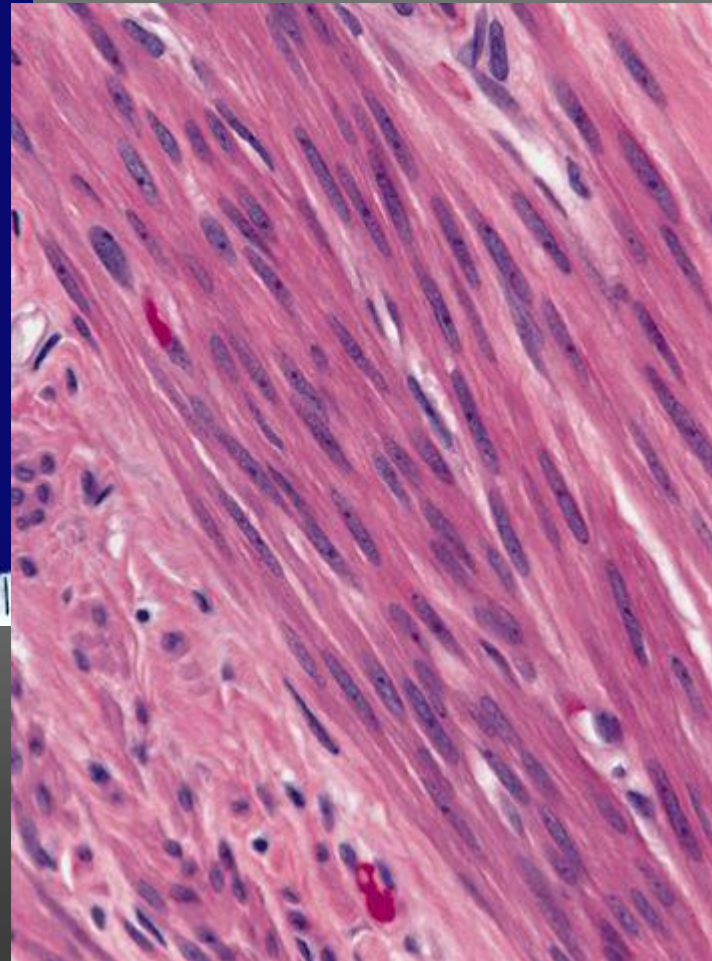
NEURITIC ATROPHY; POLYGONAL FIBRES ARE SEEN

- **WERDNIG-HOFFMAN DISEASE** (also known as "Severe infantile spinal muscular atrophy", or "spinal muscular atrophy type I") is an autosomal recessive neuromuscular disease. It is the most severe form of spinal muscular atrophy, which is one of a number of neuromuscular diseases classified as a type of muscular dystrophy.
- **Werdnig-Hoffman** affects the lower motor neurons only.

LEIOMYOMA

- Bland smooth muscle tumor without mitotic figures
- Skin and subcutis; also deep soft tissue, uterus (most common neoplasm in women)
- Patients with multiple cutaneous leiomyomas may have autosomal dominant disorder

LEIOMYOMA



LEIOMYOSARCOMA

- Smooth muscle tumor with atypia plus either mitotic activity, tumor cell necrosis or size > 10 cm
- 10% of adult soft tissue sarcomas
- Skin / subcutis; better survival than retroperitoneal tumors
- Retroperitoneum: third most common retroperitoneal sarcoma after liposarcoma and MFH; usually women, 5 year survival is only 29%
- Immunocompromised patients: associated with EBV in HIV patients; may be multifocal
-

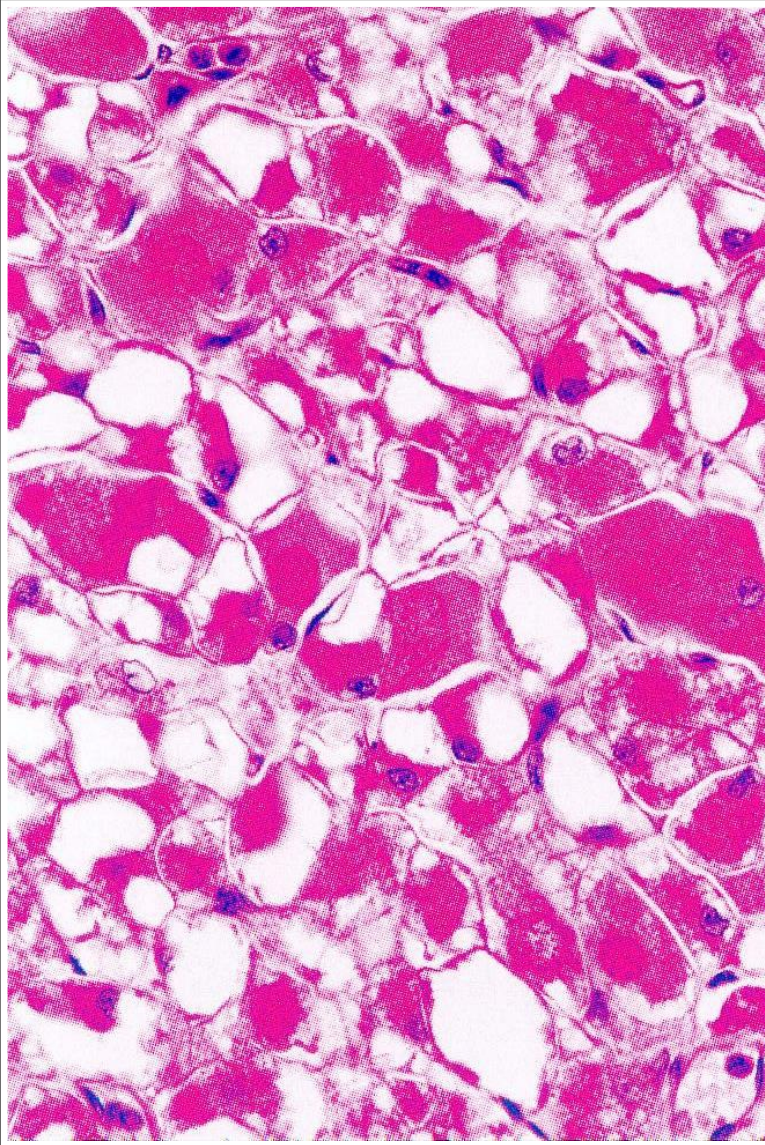
LEIOMYOSARCOMA



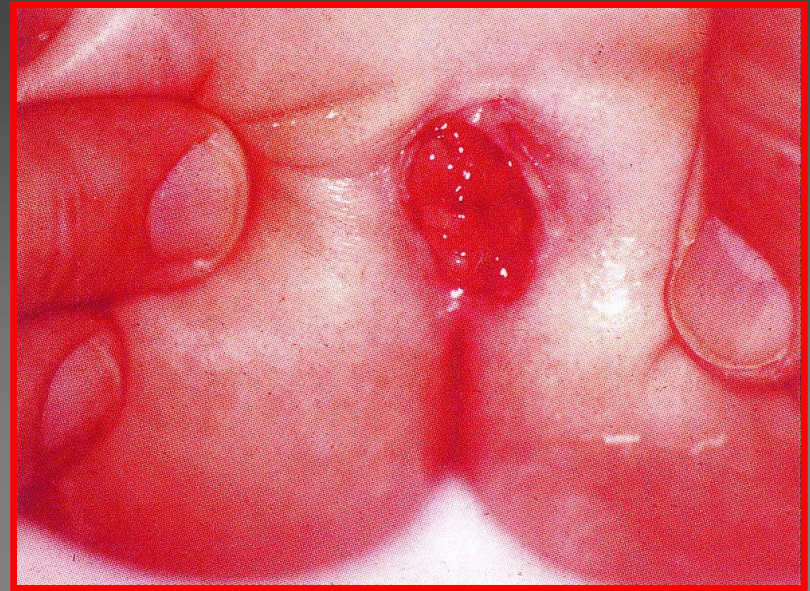
RHABDOMYOMA

- Benign tumor of mature skeletal muscle
- Extracardiac rhabdomyomas are divided into fetal, adult and genital histologic types (eMedicine)
- Extracardiac tumors are not associated with tuberous sclerosis
- Some cases may be due to degeneration and regeneration, and not be neoplastic

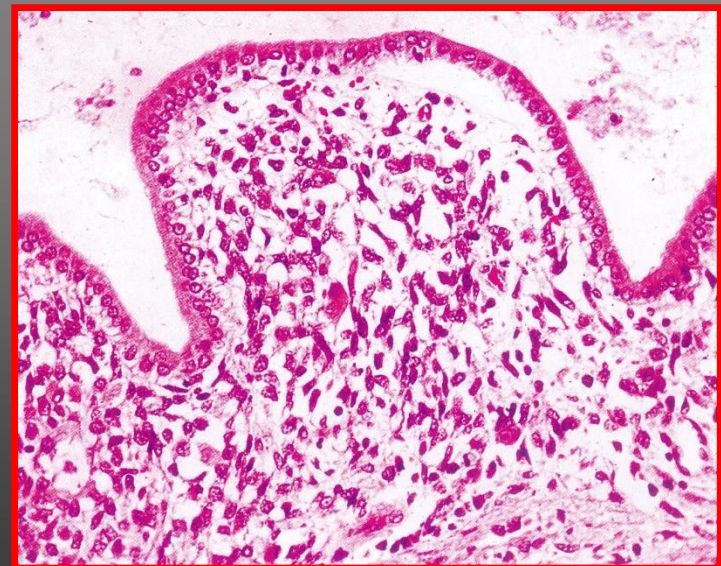
PATHOLOGY OF MUSCLES - TUMORS



RHABDOMYOMA (RARE CASE)



BOTRYOID SARCOMA



RHABDOMYOSARCOMA

- Primitive malignant soft tissue sarcoma with skeletal muscle phenotype by H&E, immunohistochemistry or EM
- Subtypes: alveolar, anaplastic, embryonal, pleomorphic, sclerosing (Mod Pathol 2001;14:506), although mixtures are common
- Note: some alveolar and embryonal tumors have similar gene expression

RHABDOMYOSARCOMA

- **Most common soft tissue sarcoma of childhood/adolescence (5 - 8% of solid pediatric tumors, 50% of pediatric soft tissue sarcomas)**
- **50% at 0 - 9 years**
- **Children 2 - 6 years usually have head, neck or GU tumors**
- **Teenagers usually have paratesticular, trunk or abdominal tumors**
- **Relatively rare in adults, who often have pleomorphic and NOS subtypes**
- **Slight male predominance (M/F: 1.3:1)**
- **Head and neck tumor are more often embryonal types**

EMBRYONAL RHABDOMYOSARCOMA

- Most common subtype of rhabdomyosarcoma in the pediatric and adolescent setting
- Anaplasia is associated with TP53 mutations, p53 protein overexpression and worse overall prognosis (Cancer 2014;120:1068)
- Displays a wide spectrum of morphologic features, including cases with spindled morphology

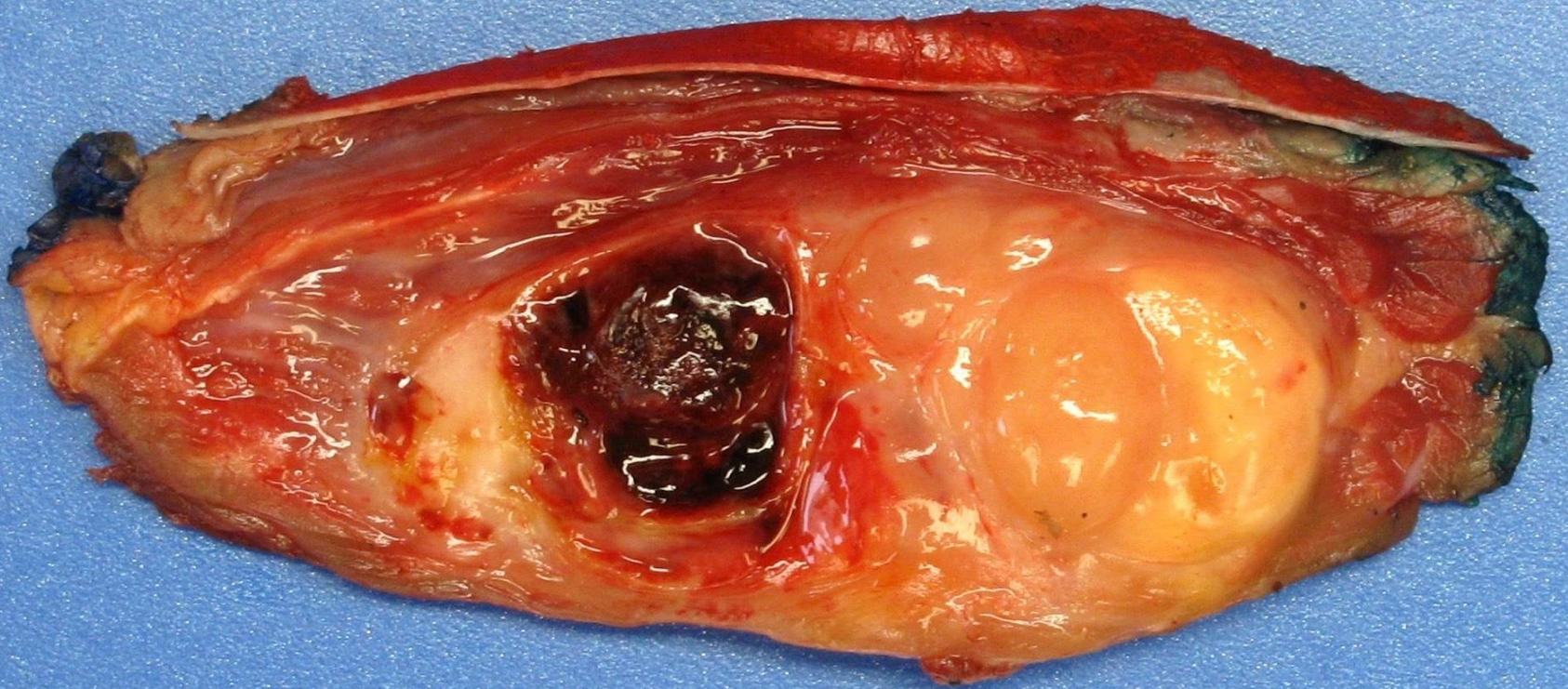
EMBRYONAL RHABDOMYOSARCOMA - orbital



PLEOMORPHIC RHABDOMYOSARCOMA

- **High grade sarcoma composed of undifferentiated cells (Fletcher: WHO Classification of Tumours of Soft Tissue and Bone, 4th Edition, 2013)**
- **Exceedingly rare category of rhabdomyosarcoma (RMS) in adults**
- **Not well characterized in the pediatric population; many of these cases can be considered RMS with diffuse anaplasia**

PLEOMORPHIC RHABDOMYOSARCOMA



Thigh mass (7 cm)

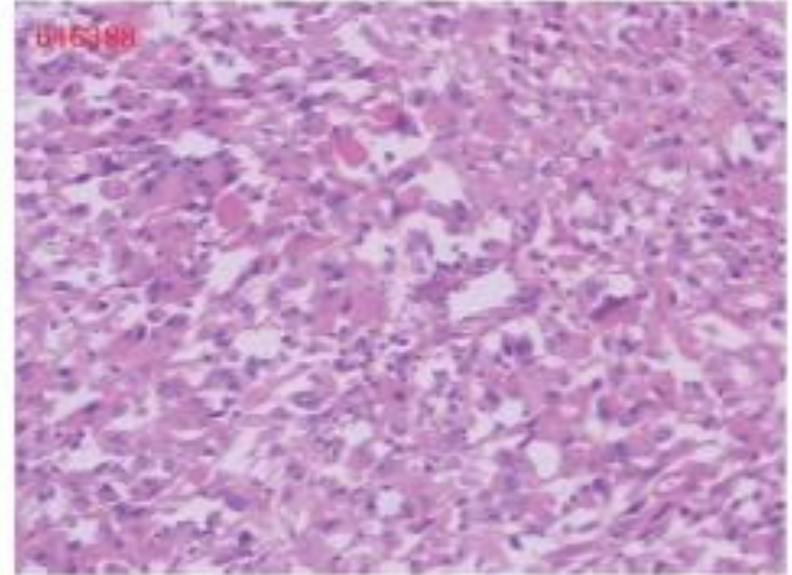
PLEOMORPHIC RHABDOMYOSARCOMA

C – myoD1, D -desmin

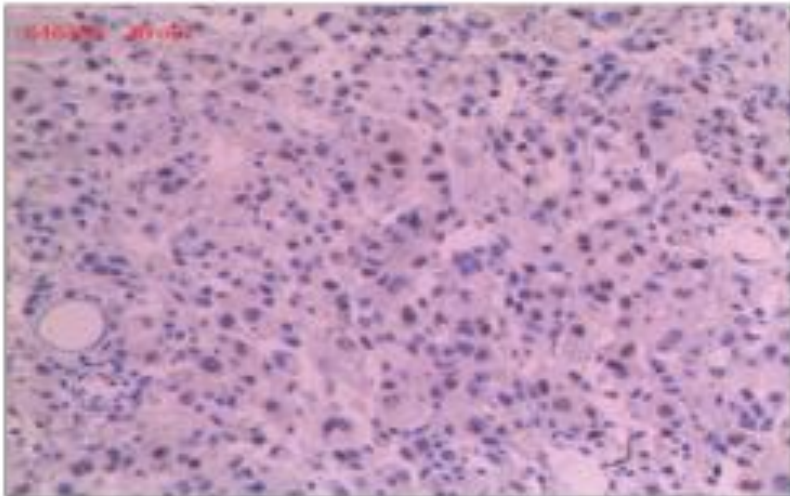
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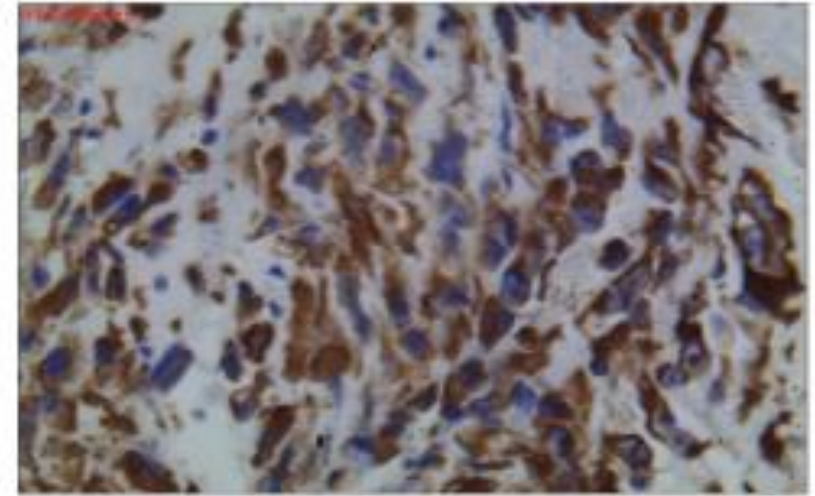
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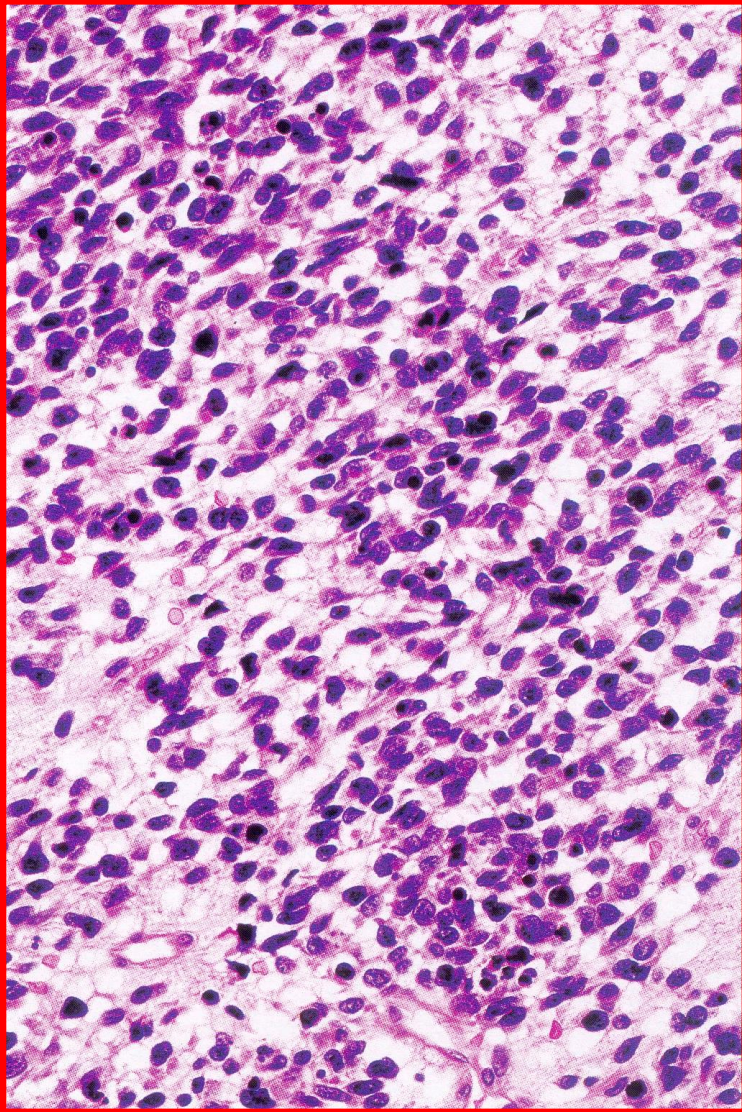
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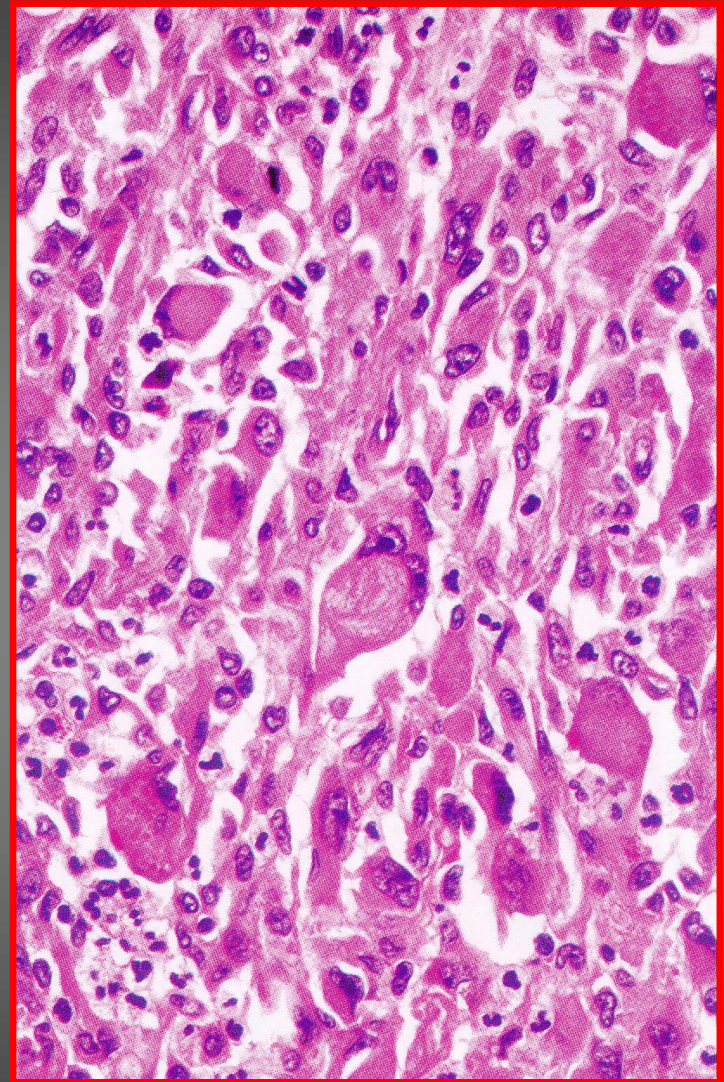
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PATHOLOGY OF MUSCLES - TUMORS



**EMBRYONAL
RHABDOMYOSARCOMA**



**PLEOMORPHIC
RHABDOMYOSARCOMA (IN ADULTS)**

THANK YOU

