

# OSPE

## Musculoskeletal Block



You must know features, Diagnosis and Definition of all cases.

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## 1- Duchenne Muscular Dystrophy (DMD)

### **Describe:**

- 1- weakness muscle.
- 2- stiffness.
- 3- deformity.

### **Definition:**

- DMD is the most severe and common type of muscular dystrophy.
- DMD is characterized by the wasting away of muscles.
- DMD affects mostly males at a rate of 1 in 3,500 births
- Diagnosis in boys usually occurs between 16 months and 8 years.
- Death from DMD usually occurs by age of 30.

### **Q: Explain the cause of DMD?**

**A: DMD is an X-linked recessive disorders caused by mutation of dystrophin gene on location [Xp21]**

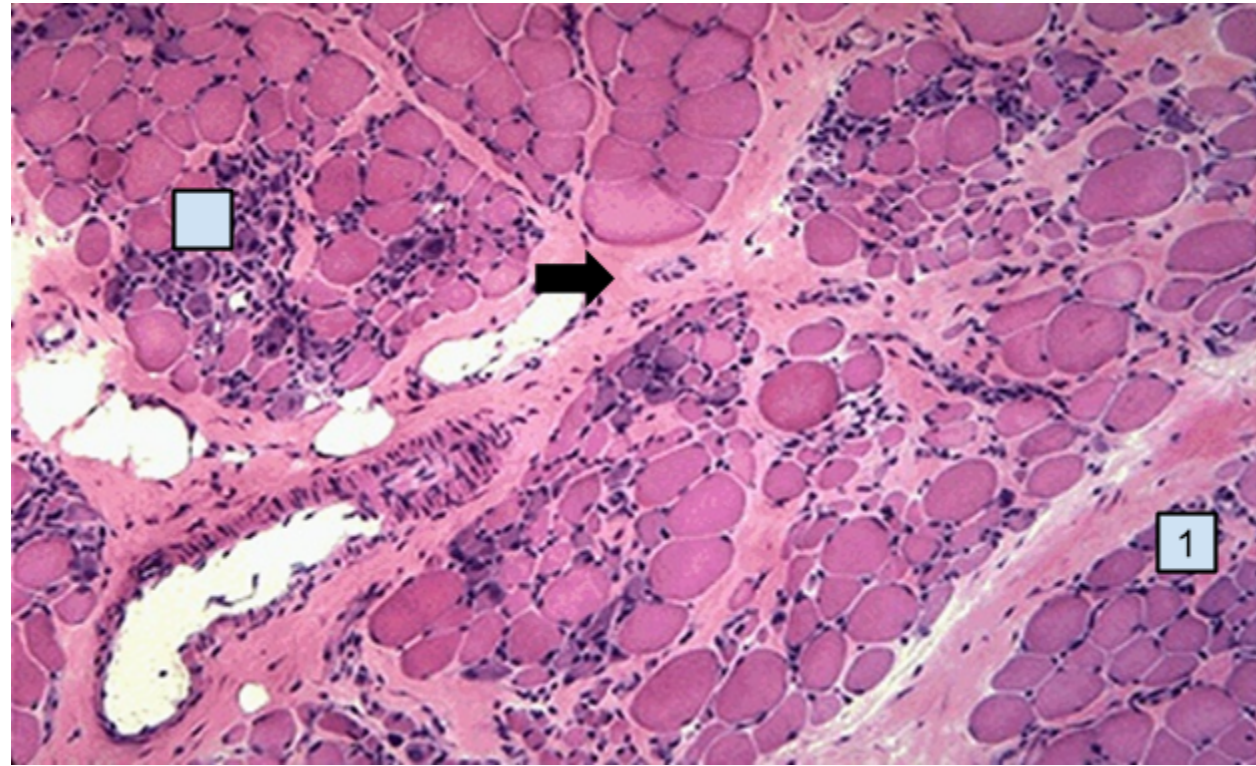
### **Case:**

A 3 year old boy presented to his pediatrician with complaint of his parents from difficulty in walking , poor balance , and frequent falls .Laboratory investigation shows elevated creatine kinase. Muscle biopsy show absence of dystrophin by western blot analysis

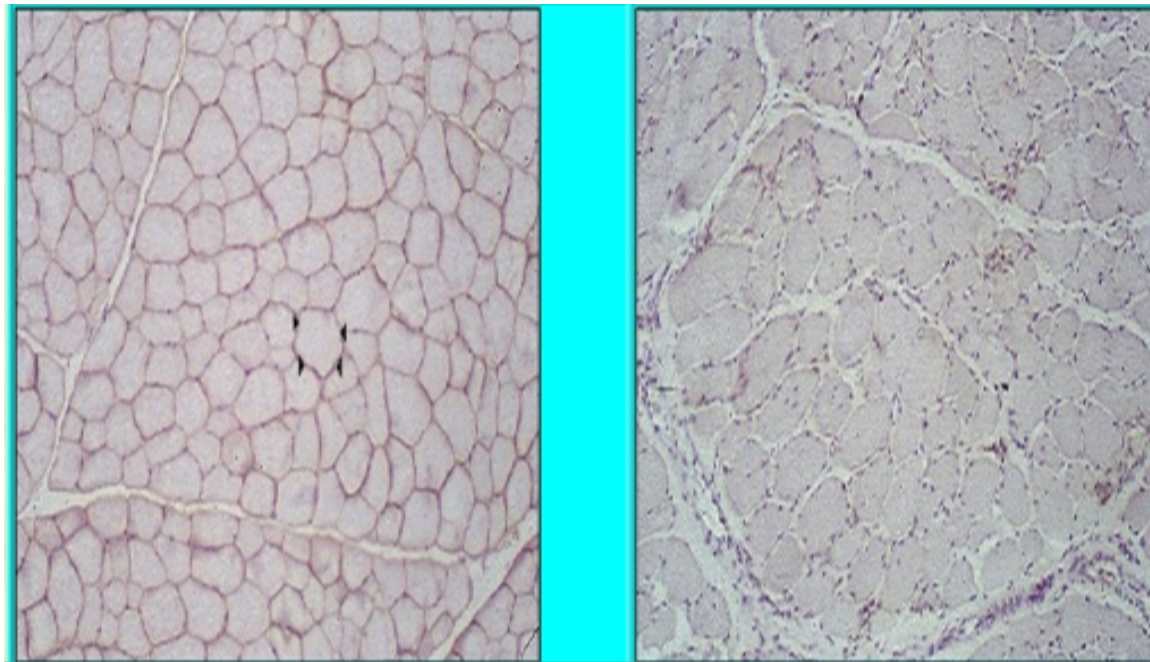


## Histology:

- 1- Bluish **regenerating** muscle fibers.
  - 2- Increase of connective tissue (endomysial increased).
  - 3- Variation in muscle fibers size.
- H & E Stain used in this picture.







R: Normal Ms

L: DMD

**R:** Positive brownish staining indicates normal muscle fibers.

**L:** Negative brownish staining which indicate lack an absence of dystrophin around the muscle fibers.

Stain : Dystrophin Stain. [Dys1 Stain]

## 2- Dermatomyositis

### **2 features:**

- 1-Violet red skin rash around the nose and eyes.
- 2-Wrinkled skin [indicates a muscle disease].
- 3-Inflammation.

### **Definition:**

- Is an inflammatory myopathy characterized by inflammation of muscle tissue and a skin rash.
- Can occur in any individual with peak age patterns at:
  - 5-15 years of age
  - 40-60 years of age.
- Occurs more frequently in women.



## CASE:

A 52-year-old woman presents with 6-month history of progressive muscle weakness and a skin rash. Physical examination is remarkable for a diffuse purple/red discoloration of the skin over her cheeks, nose, and eyelids. Examination confirms proximal muscle weakness. Laboratory findings show an increase in creatine kinase (10 times the normal).

## Signs :

- A. Two enzymes are elevated: Creatine Kinase & NA.
- B. Proximal muscle weakness.

## Histology:

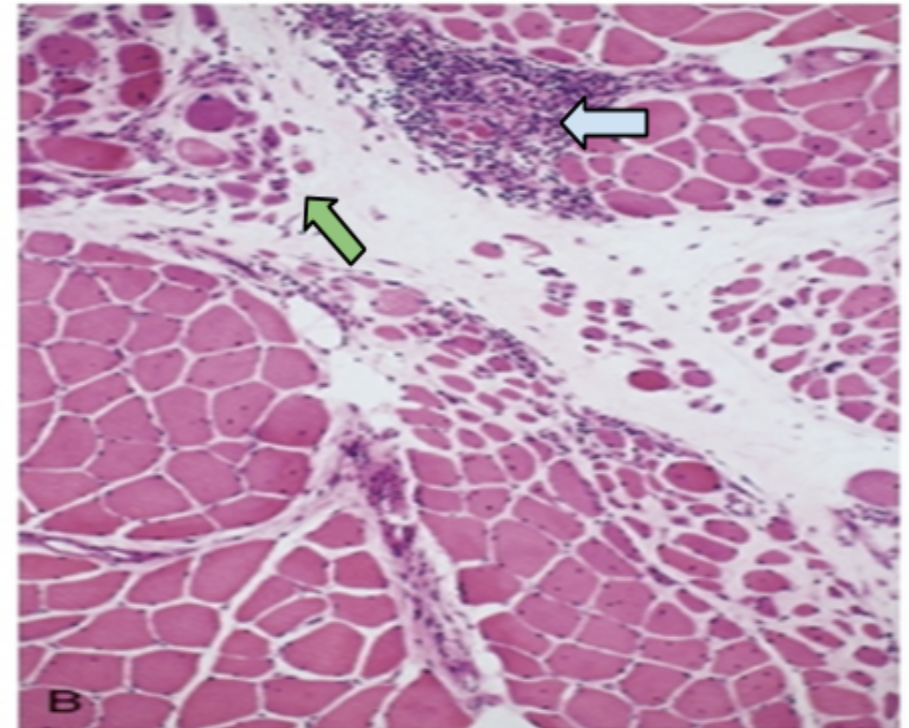
1- Inflammatory Cells (Lymphocytes).

2- Perifascicular atrophy (muscle fibers atrophy).

[The muscle fibers around the margins of the fascicular undergoes atrophy]

Fascicular is the bundle that contain the muscle fibers.

the



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### 3- Myasthenia Gravis

**Main feature:** Ptosis of the left eyelid.

[Ptosis is the dropping of the upper eyelid]

**Q:** What is the possible diagnosis, and its cause?

**A:** Myasthenia Gravis, it is an autoimmune disease which causes a generation of antibodies that blocks “postsynaptic Ach receptors”.



#### **Several Types of Myasthenia Gravis:**

- 1- Neonatal Myasthenia Gravis: A transient condition in 10% to 15% of infants born to mothers with MG.
- 2- Congenital Myasthenia.
- 3- Juvenile Myasthenia: Onset is around 10 years of age.
- 4- Ocular Myasthenia.
- 5- Generalized Autoimmune Myasthenia.

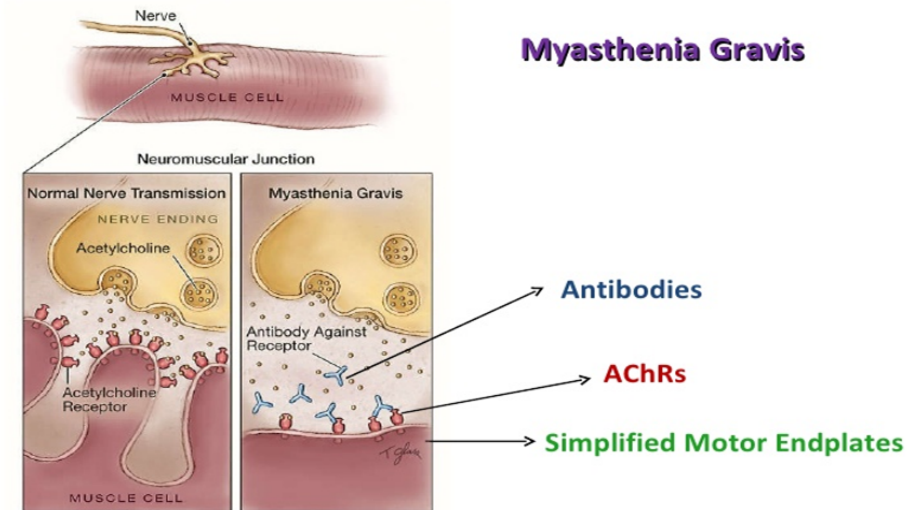
#### Acquired autoimmune disorder:

fundamental defect is a decrease in the number of available AChRs at the postsynaptic muscle membrane  
simplification of the postsynaptic folds and widening of the synaptic cleft .

#### Clinically characterized by:

Weakness of skeletal muscles  
Fatigability on exertion.

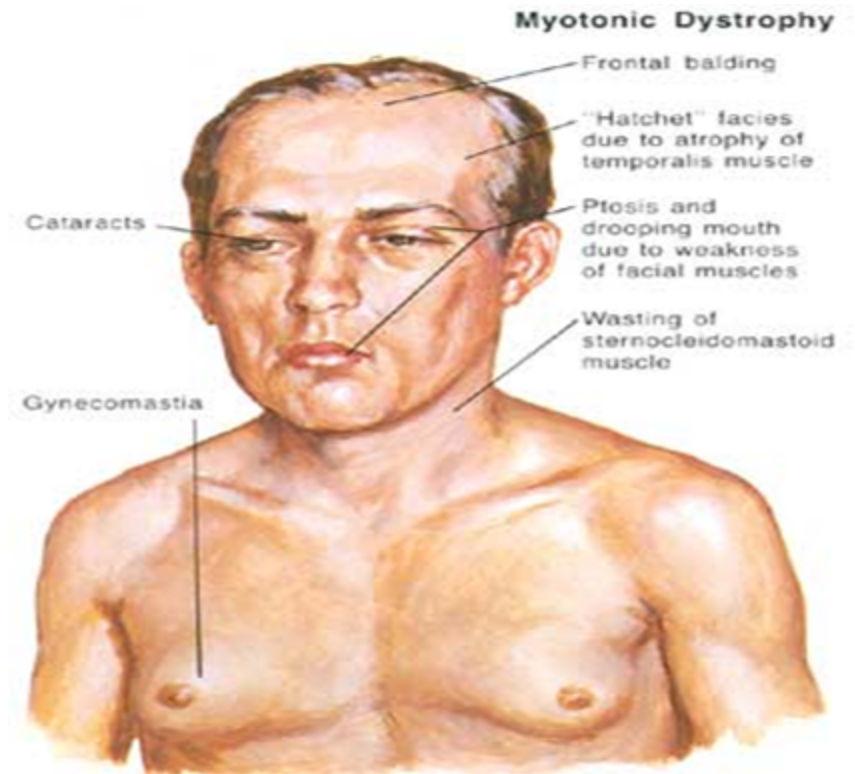
The neuromuscular abnormalities in MG are brought about by an autoimmune response mediated by specific anti-AChR antibodies  
These antibodies reduce the available AChR's at neuromuscular junctions  
the thymus is abnormal in approximately 75% of patients with MG  
In 65% of patients the thymus is hyperplastic.





## 4- Myotonic dystrophy

Also known as dystrophia myotonica  
Composed of 2 clinical disorders with overlapping phenotypes & distinct molecular genetic defects:  
1. DM1- the classic disease.  
2. DM2- proximal myotonic myopathy.  
It is autosomal dominant disease.





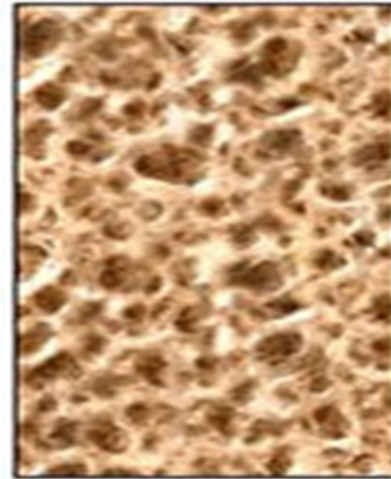
## 5-Osteoporosis

A disease characterized by low bone mass and microarchitectural deterioration of the bone tissue.  
Leading to: enhanced bone fragility and increase in fracture risk.

1-In Left : Normal structure of bone.

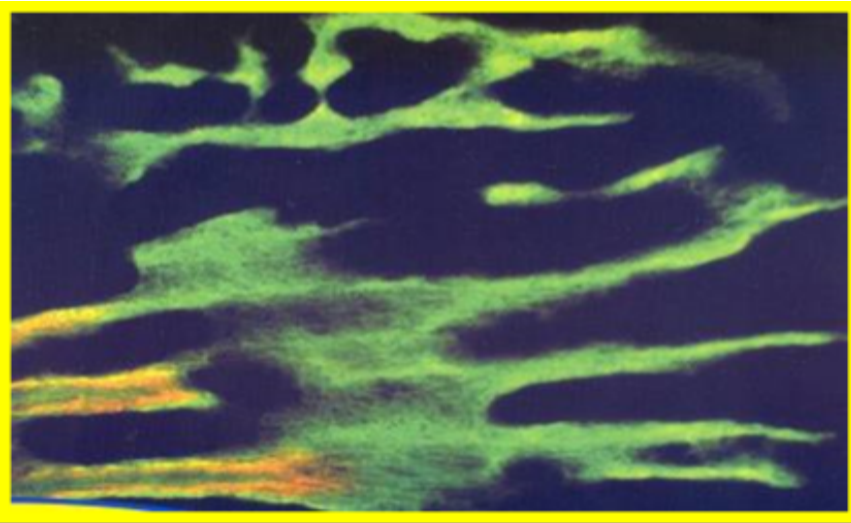
2-In Right: Dysregulatory cells (Osteoporosis).

**Normal bone**

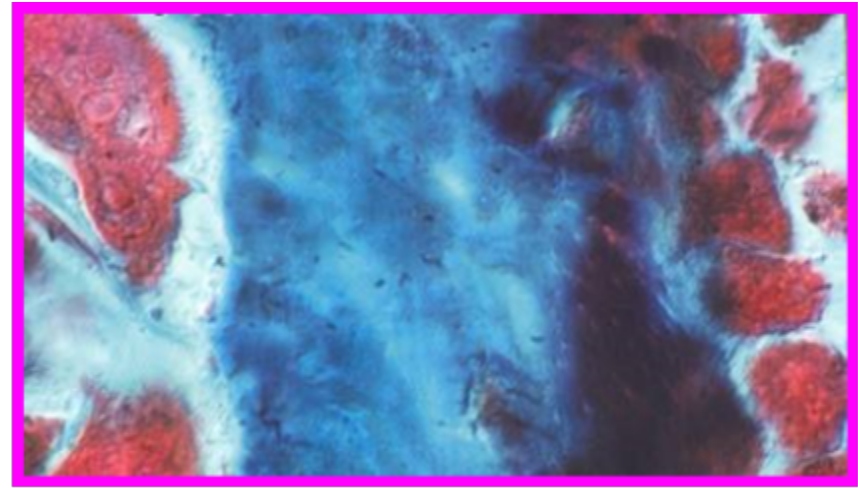


**Bone with  
Osteoporosis**

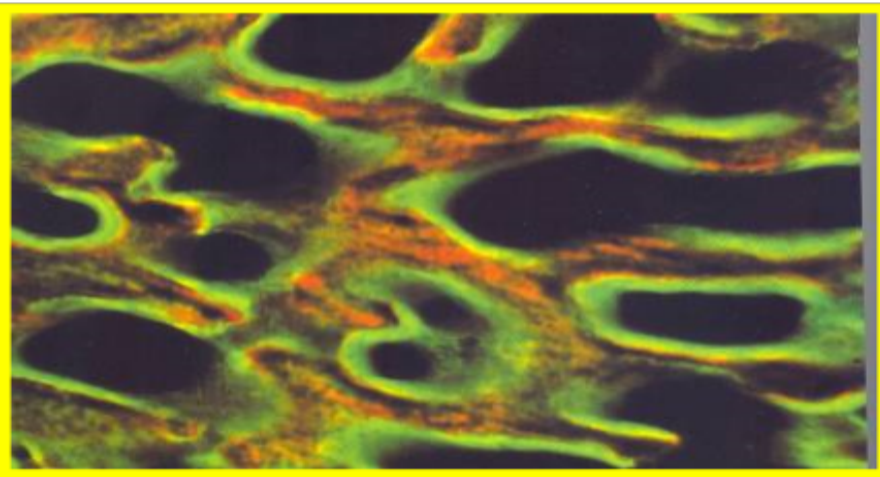




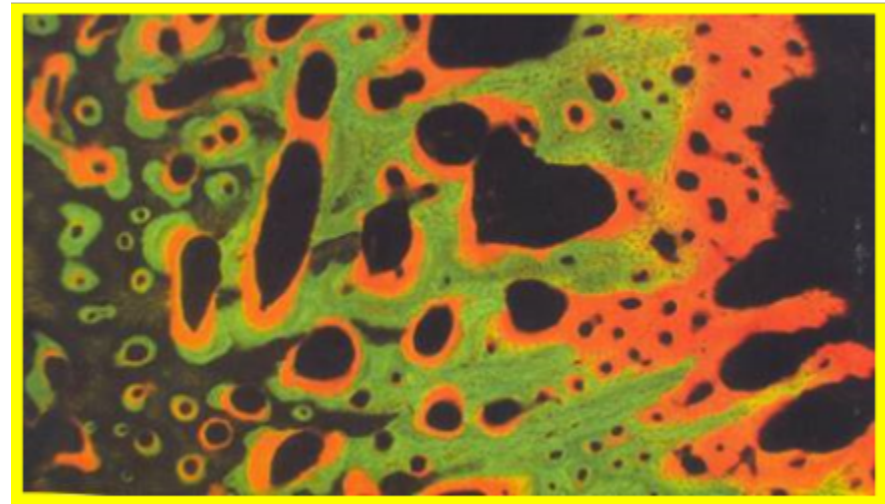
First phase of woven bone formation



Remodeling of bone trabecula



Later phase in woven bone formation



Accretion and remodeling of bone

## 6-Osteoarthritis

A degenerating disease. It will become painful because there is no joints that decrease the friction.

An obese 56-year-old woman presented with bilateral localized pain to her knees, hands and difficulty in walking.

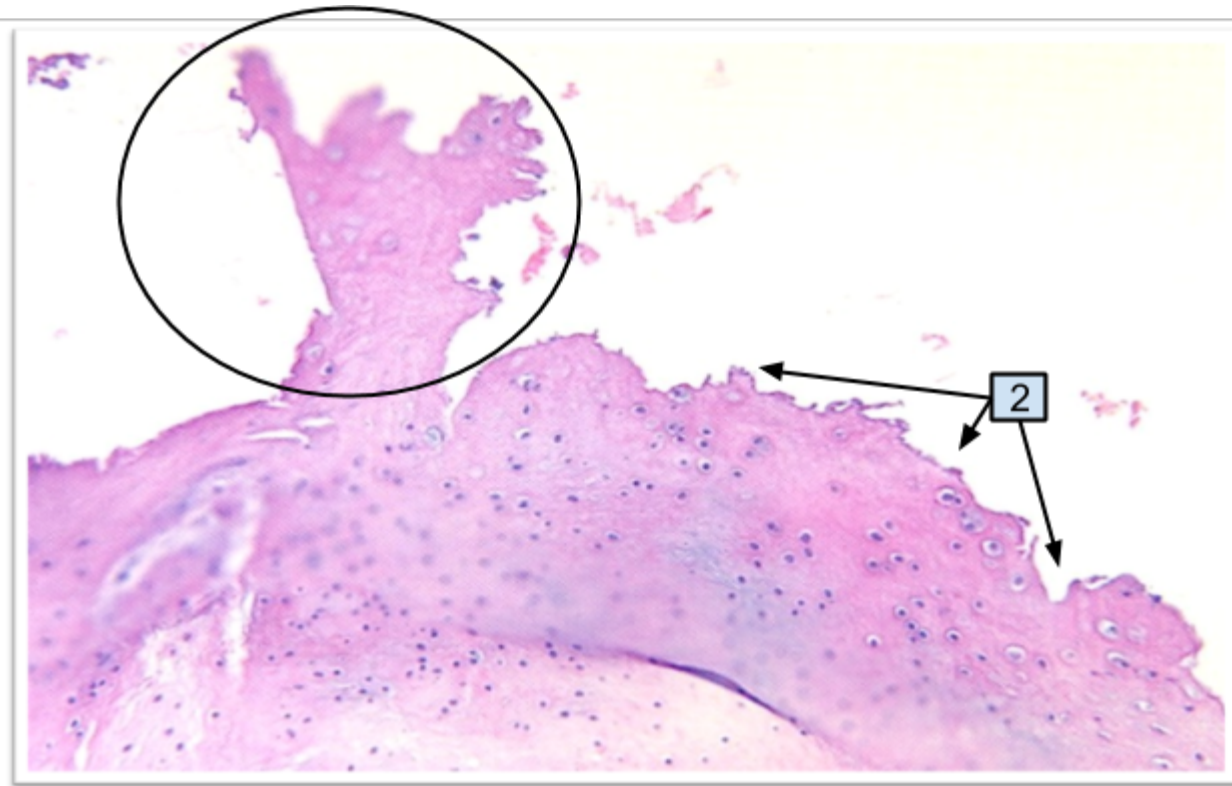
**Q: Is Osteoarthritis an inflammatory, and where does it occur?**

**A: Osteoarthritis is not a real inflammation, and it occurs in large bearing joints.**

1- Subchondral cyst with fluid.

2- Eburnated /Erosions in the articular surface.





1- Inflammatory cells are **absent**.

2- Irregular articular surface .

3- Irregular outgrowth of osteophytes. [black circle]



## 7-Rheumatoid Arthritis



### Describe:

1-Deformity in small joints.

2-Swelling.

- Same symptoms as TB arthritis.. But tb arthritis affects mainly the spine.
- Morning pain
- deformity in small joints.

Rheumatoid arthritis affecting the head of the femur.

The synovium becomes edematous, thickened and hyperplastic and transforming its smooth contour to one covered by delicate and bulbous fronds .

**Q: Define Rheumatoid Arthritis.**

**A: It is an autoimmune disease that attacks normal joints tissue and as a response, it causes inflammation.**

**Q: What is the main keys from the symptoms to detect Rheumatoid Arthritis?**

**A: 1)Morning Pain**

**2)Swelling and pain in the small joints**



Case:

A 45 -year- old woman complains of low grade fever , malaise and stiffness in her joints each morning .

1-Edematous.

2- thickened.

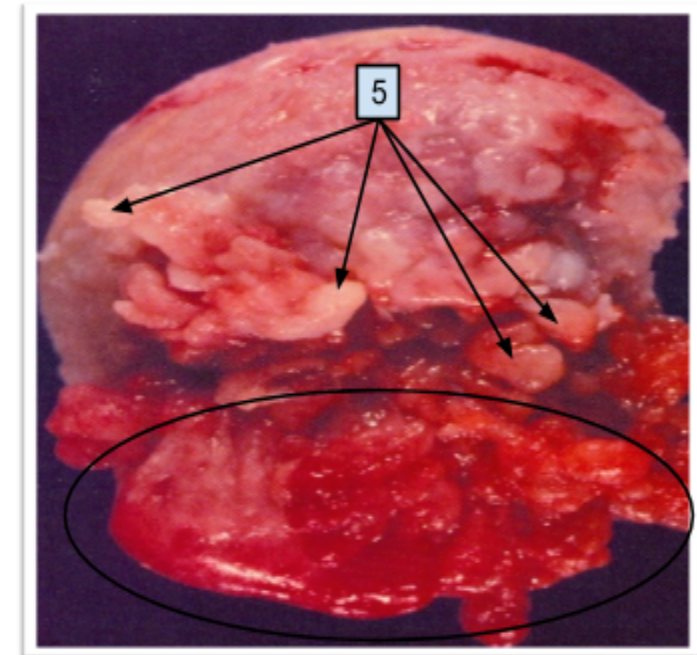
3- Hyperplastic.

4-Bulbous.

5- There is finger of synovial hyperplasia.

6- Hemorrhage.

7-Eroded bone and cartilage.



Rheumatoid arthritis affecting the head of femur.

The synovium becomes

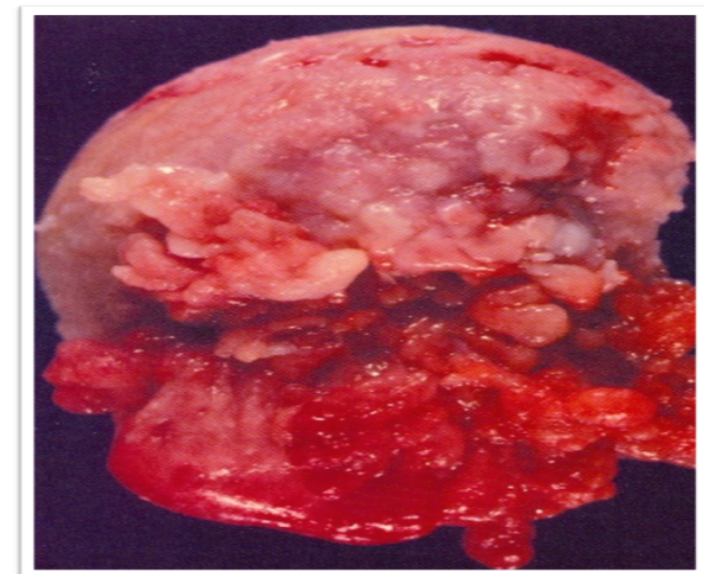
edematous,

thickened and

hyperplastic

note the presence of

inflammatory protrusions on the synovial surface.

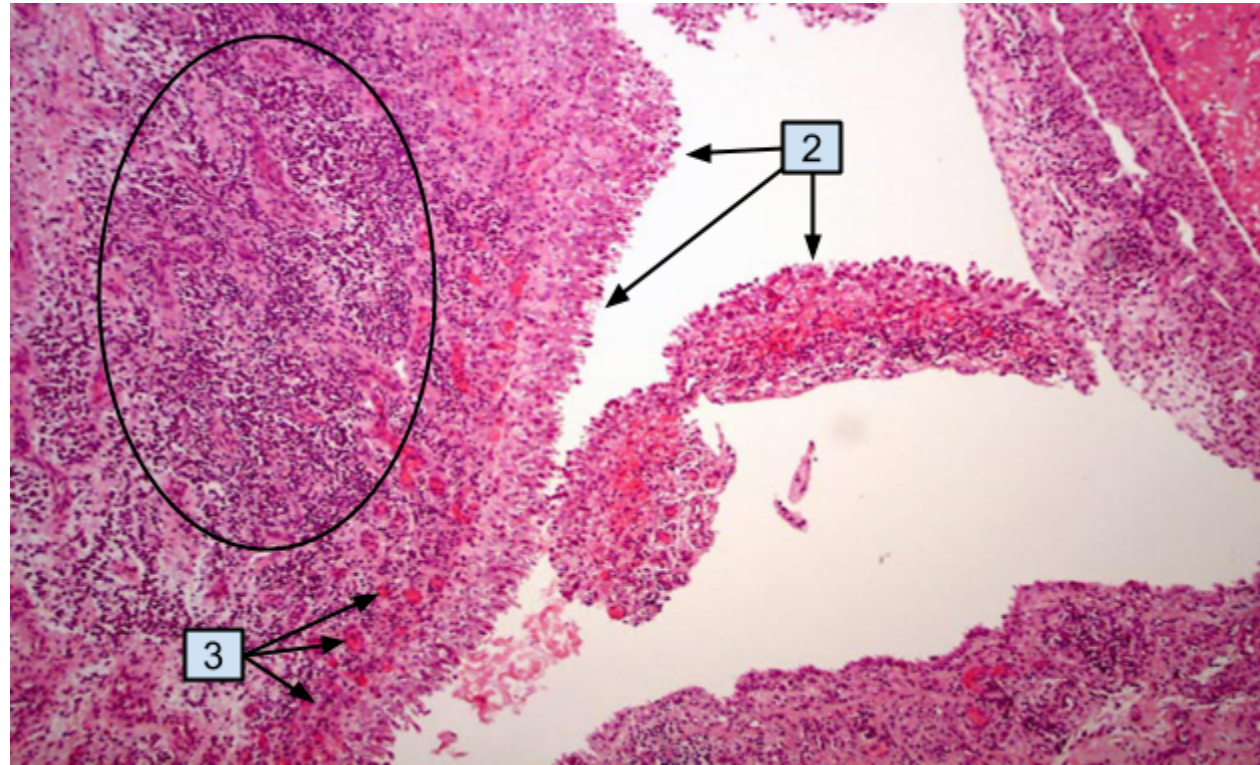


1- Extensive Inflammatory Cells (Lymphocyte).

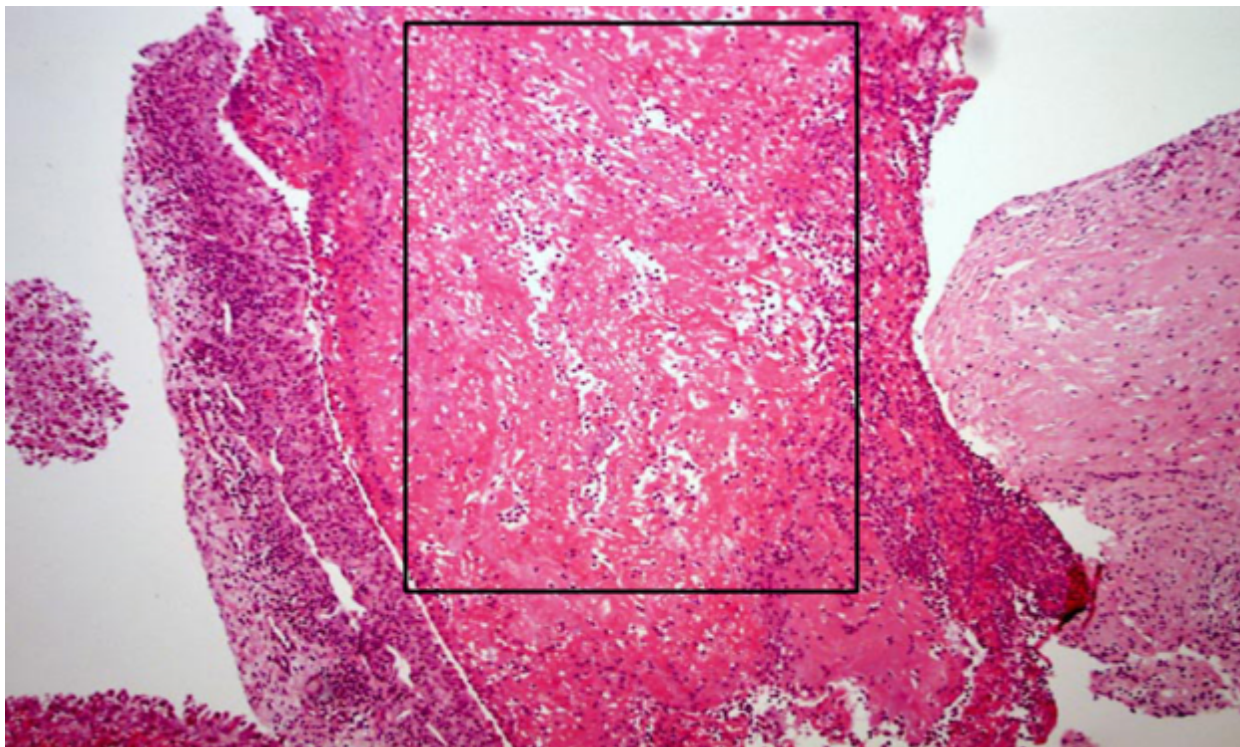
2-Synovial layer hyperplasia.

3- vascular congestion. [small red dots]

Hyperplastic synovial lining with villous like projections: note the presence of dense lymphoplasmacytic infiltration and extreme vascular congestion. This biopsy was taken from a classical case of active rheumatoid arthritis.







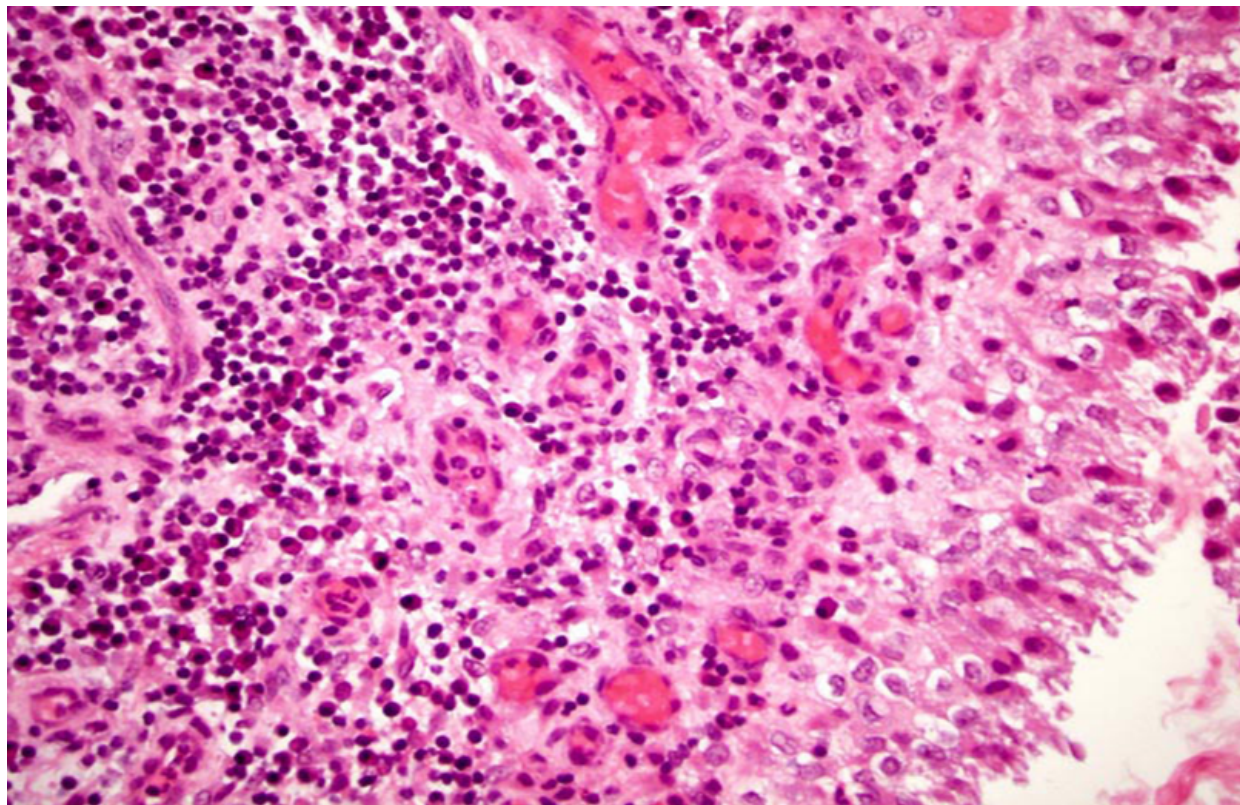
### 1-Pannus Formation.

- pannus formation: inflammatory fibrinous exudate.
- permanent ankylosis adhesions (requires knee replacement).

Pannus consisting of **fibrinous inflammatory exudates** and later on, the pannus may fill the joint space and undergo fibrosis , calcification and causes permanent ankylosis (adhesions).

The serological tests that can be done to diagnose this disease are:

- Rheumatoid factor.
- Cyclic citrullinated peptides



- 1- Plasma cells.
- 2-Inflammatory Cells (Lymphocytes).
- 3- Congested Blood Vessels.

Hyperplastic synovial lining associated with plasma cells and lymphocytic infiltration with vascular congestion.

## 8-Gout

Gout is a syndrome caused by the inflammatory response to tissue deposition of monosodium urate crystals (MSU).

**Q: What is the cause of Gout?**

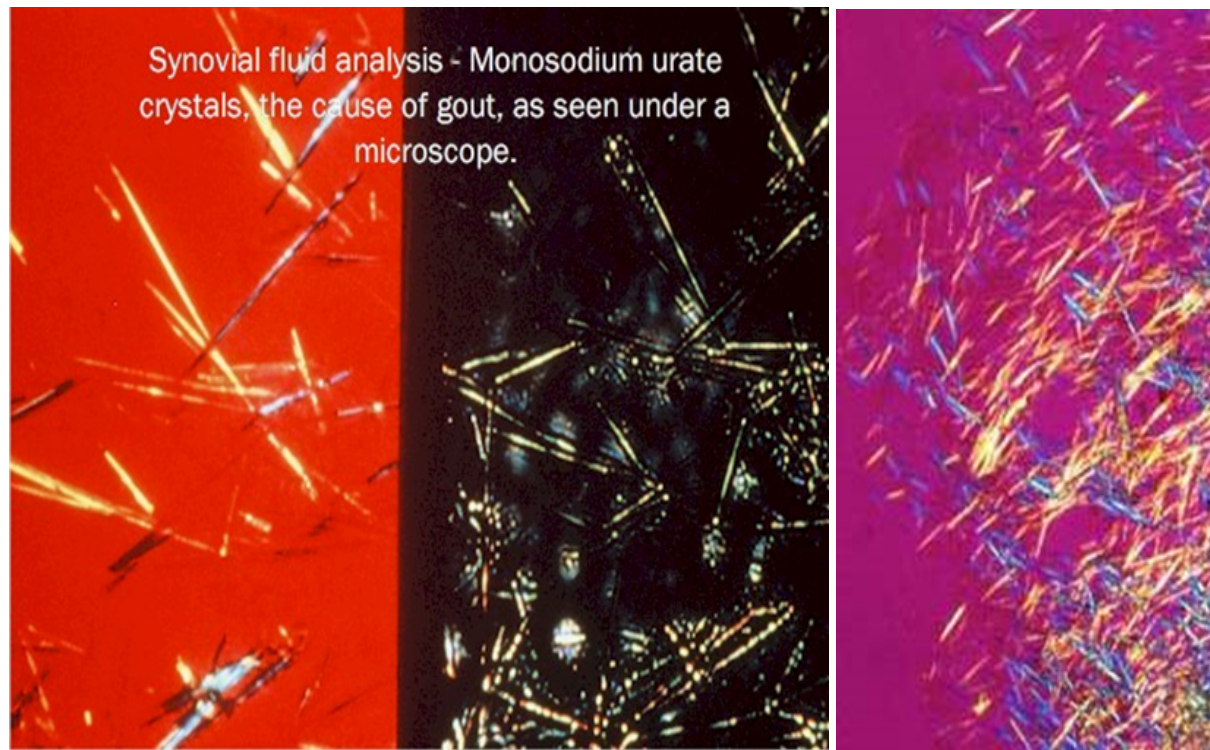
**A: Accumulation of uric acid in the joints, especially the small joints.**

### **Acute Gout Features:**

- 1-Many areas filled with swellings.
- 2-Deformity in small joints [SPECIFY].







- 1-Needle-shaped crystals.
- 2-In left polarized [black] and in right unpolarized.

## 9- Osteomyelitis

A 22 years old male presented with localized pain above his right knee joint with recurrent fever. Later on, he developed discharging sinuses from the skin overlying the right knee.

What is the most likely diagnosis?

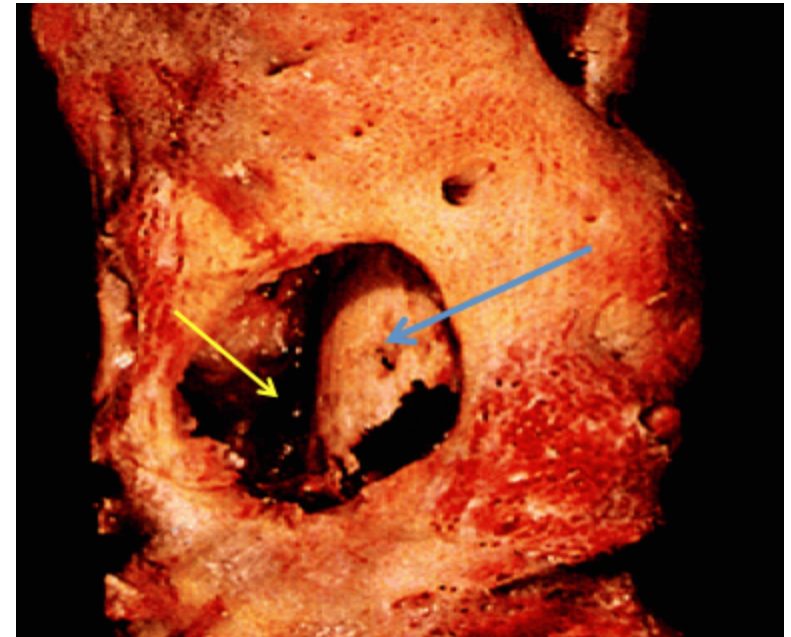
### Describe:

- 1-Involucrum (New bone).
- 2-Sequestrum (Dead bone).

### Direct infection of bone.

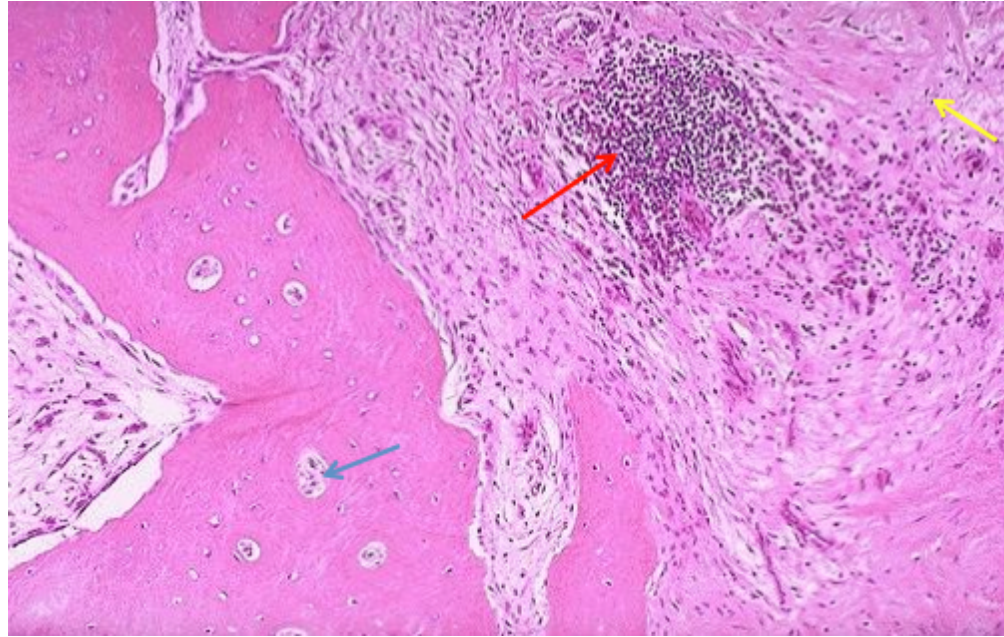
Bacterial most often:

- Staphylococcus
- Salmonella
- Sickle Cell Disease
- Tuberculosis
  - Spine first
  - Syphilis
  - Periosteum



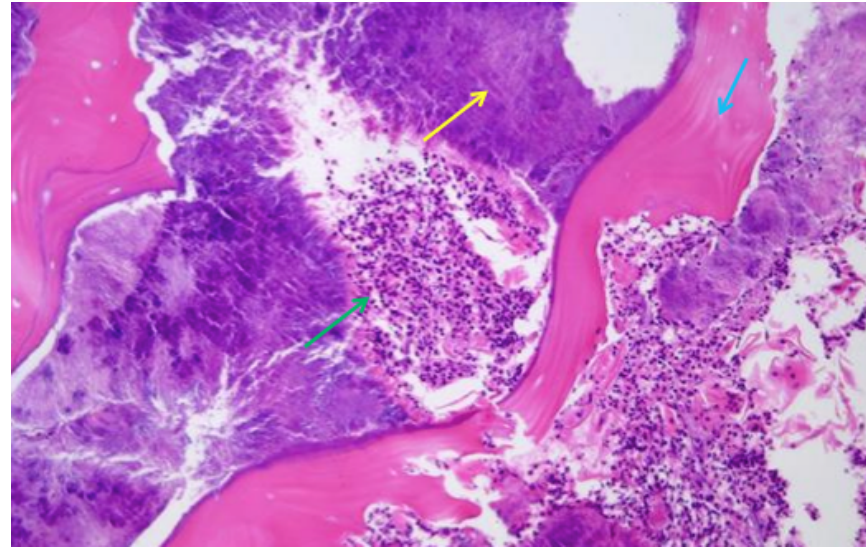
Advanced chronic osteomyelitis of the femur bone.

The blue arrow points to an area of reactive new bone formation (involucrum). The yellow arrow points towards an area of necrotic bone (sequestrum) surrounded by hemorrhage. The patient also had discharging sinuses on the surface of the skin which were draining pus from the diseased bone.



- 1- inflammatory cells.
- 2- bacterial colonies.
- 3- fibrosis.
- 4- new bone formation.

Chronic osteomyelitis: note the fibrosis of the marrow space (yellow arrow) accompanied by chronic inflammatory cells (red arrow). The blue arrow points towards an involucrum consisting of active new bone formation. Chronic osteomyelitis can be complicated by amyloidosis.



1-Inflammatory Cells.

2-Fibrosis.

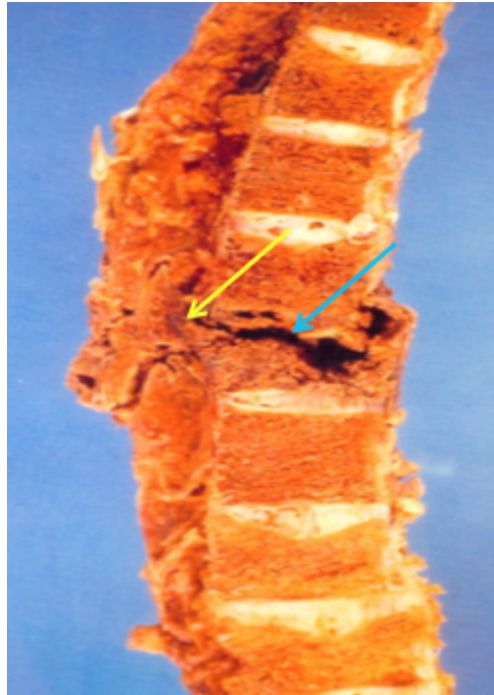
Acute pyogenic osteomyelitis:

The yellow arrow shows colonies of Gram positive bacteria. The blue arrow points towards a trabeculae of dead bone (note the presence of empty lacunae). This type of diseased bone is called sequestrum. The green arrow points towards clusters of neutrophils admixed with fibrin (acute fibrinous inflammatory exudate).



## 10- Spinal TB - Potts Disease (Tuberculous osteomyelitis)

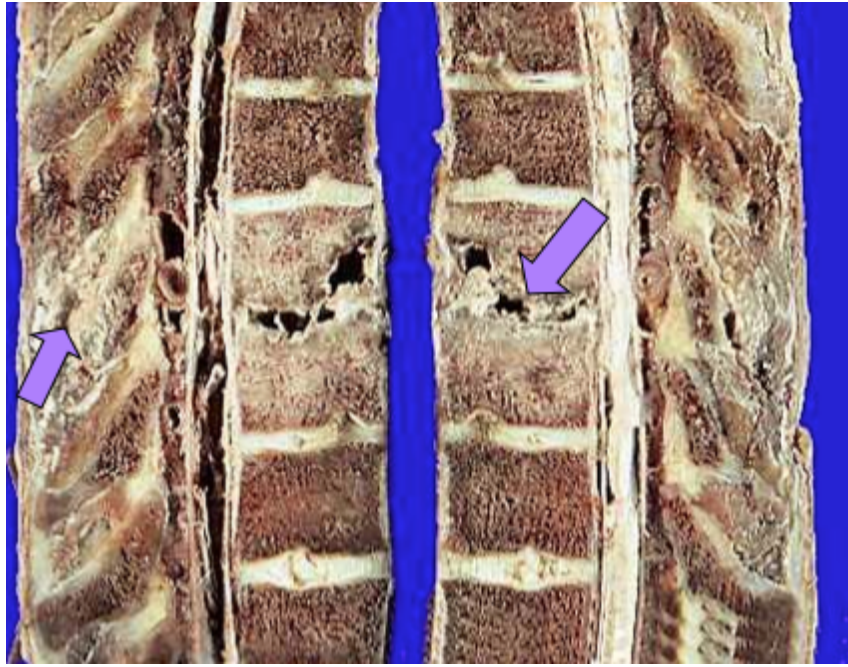
A 35 years old debilitated man presented to the orthopedic clinic with back pain, low grade fever, marked elevation of sedimentation rate and recent kyphosis with mild scoliosis . The patient has a history of coughing up blood, mild fever, chills and night sweats. He told his doctor that he also has a tendency to get tired very easily.



- 1- fracture vertebrae
- 2-hemorrhage.
- 3-necrosis.

A fractured vertebrae associated with an abscess with fibrosis in the surrounding soft and muscular tissue. A biopsy taken from this lesion showed evidence of tuberculous osteomyelitis of the spine (also called Pott's disease).

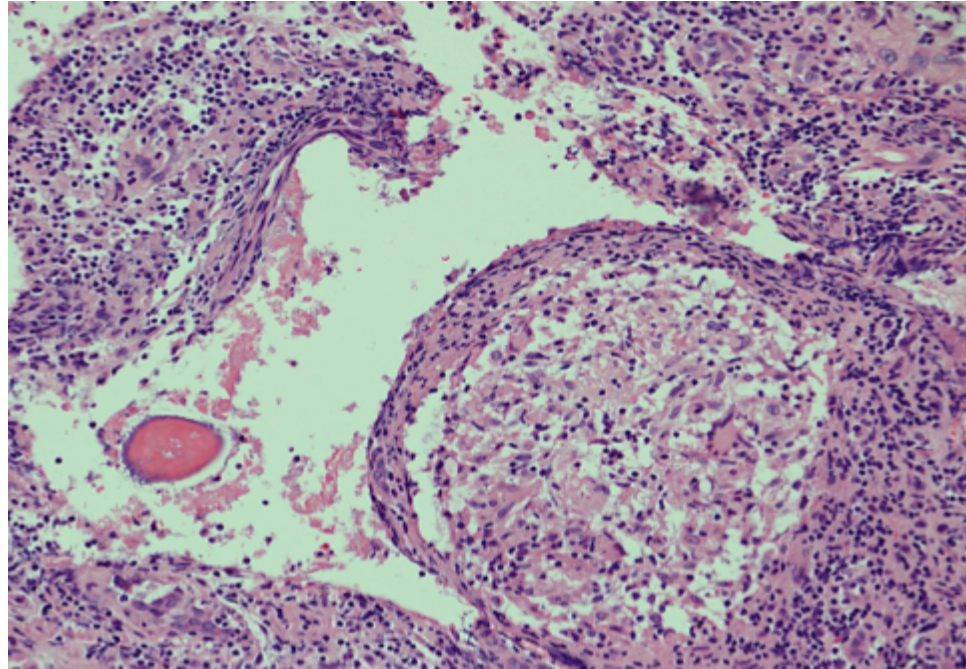




1- necrosis.

2- fracture.

The vertebrae shows a lytic lesion with cavitation and areas of necrosis.

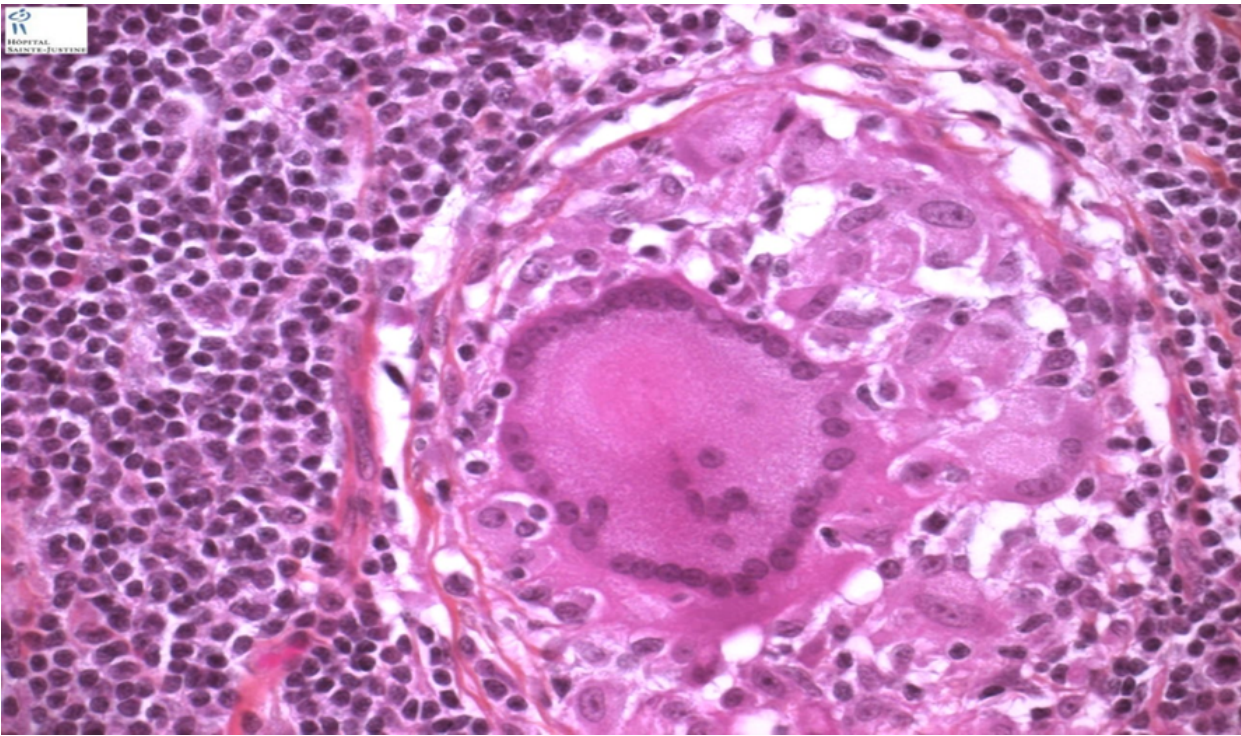


**Describe:**

**Granuloma which consisting of :**

- 1- lymphocytes.
- 2-Langhans cells (Giant cell).
- 3-Epithelioid macrophages.

Section of bone shows granuloma formation with epithelioid like cells , langhans-type giant cells and a rim of lymphocytes. The features are consistent with granulomatous inflammation secondary to tuberculosis.

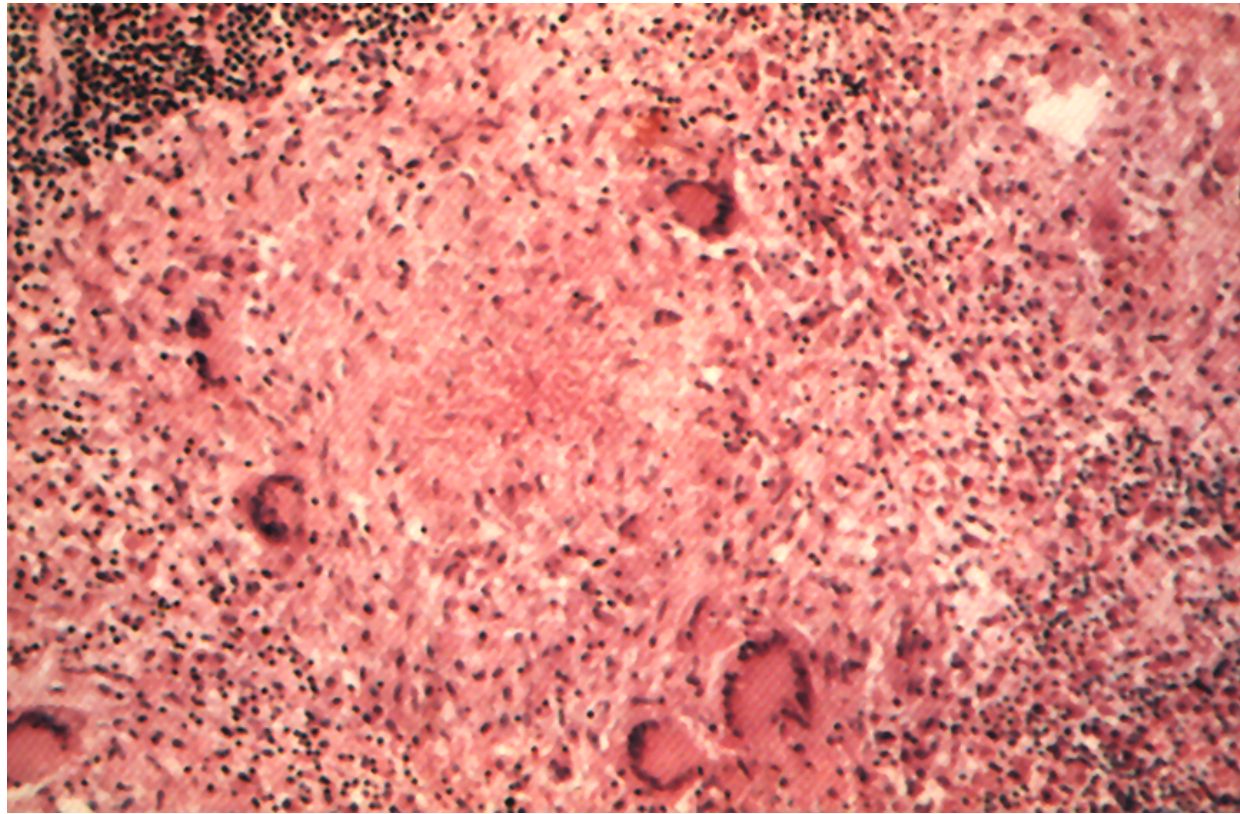


**Describe:**

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**Describe:**

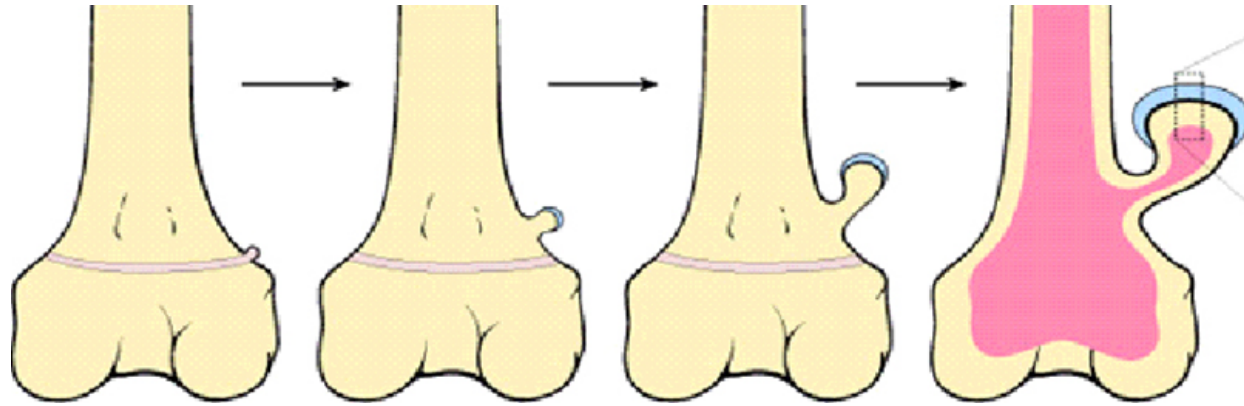
**Granuloma which consisting of :**

- 1- lymphocytes.
- 2- Langhans cells (Giant cell).
- 3- Epithelioid macrophages.



## 11-Osteochondroma (osteochondroma exostosis)

A 16 -year-old male was found to have a small swelling protruding from upper part of his leg with local pain .



**1-Bony prominence.**



1-MRI picture showing bony swelling arising from upper of fibula.



1-X-ray picture showing bony swelling.



**1-X-ray picture of Tibia showing bony protrusion.  
-Benign Lesion.**

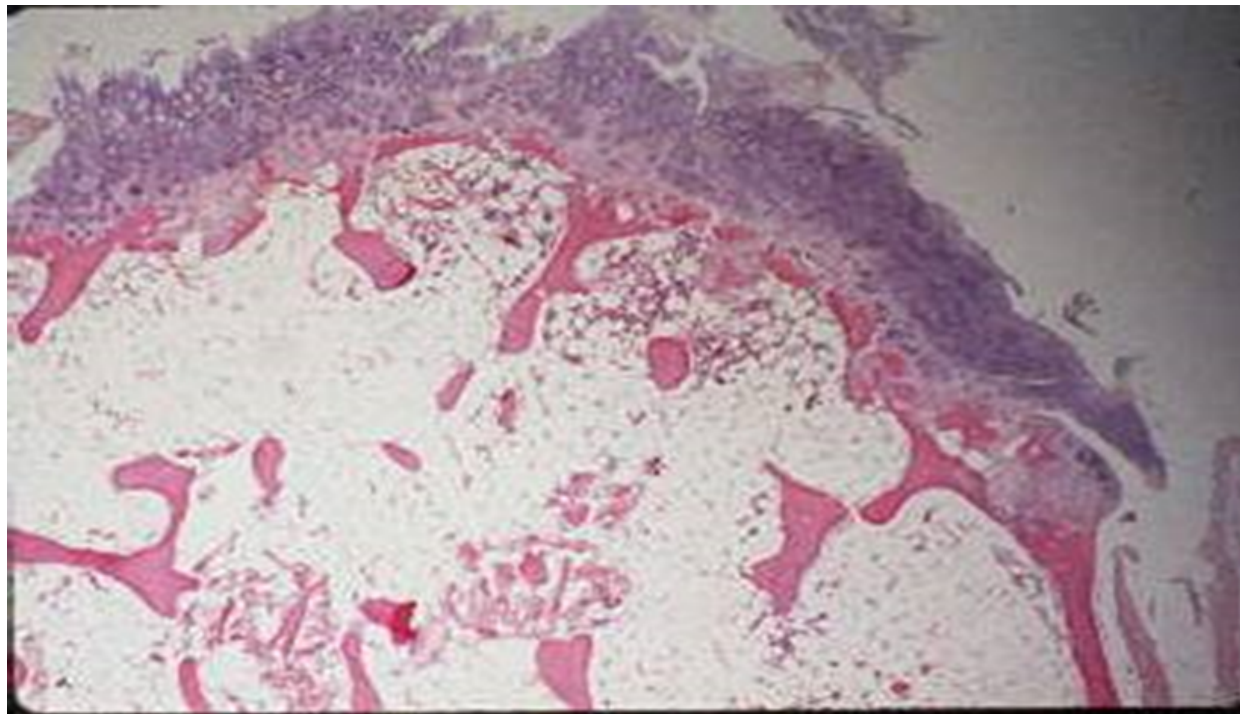




**1-Multiple Cartilaginous Exostoses.**



- 1- benign (Prognosis is good).
- 2- Bone stalk covered by cap of cartilage.



- 1- bone.
- 2- cartilage.
- 3- fibrous tissue cap.

-It is Benign tumor , the risk from transferring to malignant.

-

## 12-Osteosarcoma

- “Weight bearing
- “Long bones
- “Young people
- “Osteoblast is malignant cell
- “Genetics of tumor being unraveled

An 18-year-old female presented to the rheumatology clinic with 2 months history of pain and swelling in her upper thigh with weight loss.





•**Malignant tumor.**

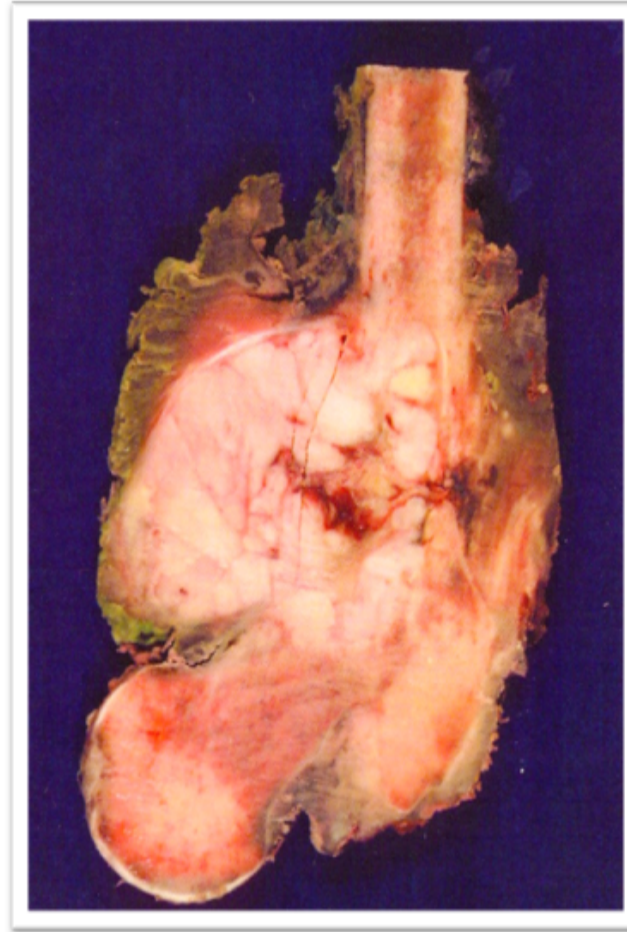
1- hemorrhage.

2- necrosis.

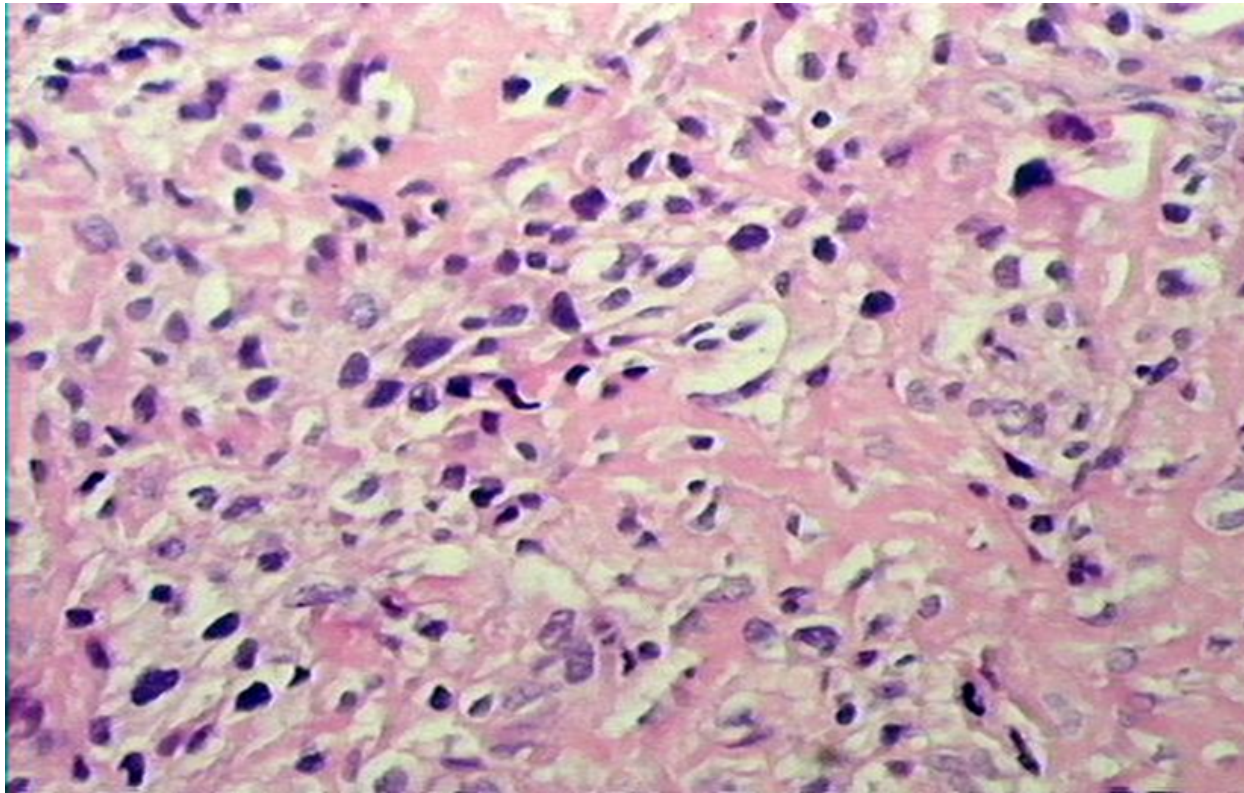
3- intramedullary irregular bone tumor.

4- Inflated cortex.

5- metaphysis and epiphysis of Tibia.

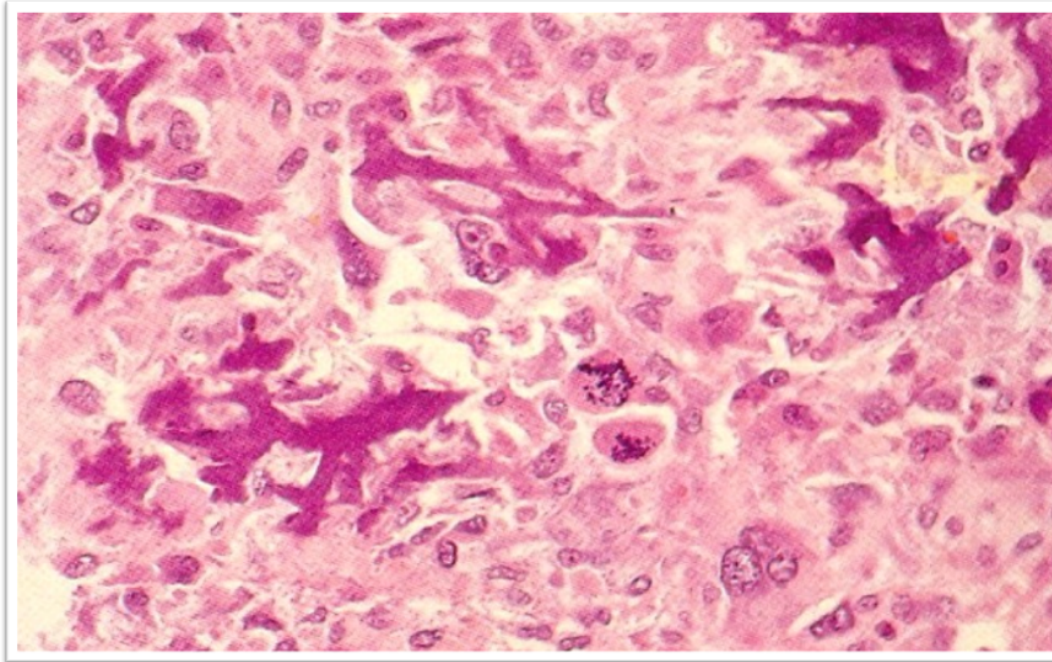


- 1-Necrosis.
- 2-Hemorrhage.
- 3-cyst area.



**1-Spindle shaped cells producing osteoid.**





1- mitosis.

2-prominent nuclei.

3-Spindle shaped cells .



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Good Luck!

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