Hashemite University
H · U · M
Medical School Committee

Scientific Team
الفريق العلمي

PATHOLOGY

DONE BY :shaden Fadda

Cartilage-Forming Tumors

*OSTEOCHONDROMA (EXOSTOSES) :

- **Definition** :
  a relatively common slow-growing benign tumor , cartilage-capped outgrowths attached by a bony stalk to the underlying skeleton

- **It has 2 types** :

1) Solitary osteochondromas >> - diagnosed in late adolescence & early adulthood
   - M/F ratio = 3/1
   # rarely progress to chondrosarcoma or other sarcoma

2) Multiple exostoses >> * become apparent during childhood
   * occur as multiple **hereditary** exostoses, an autosomal dominant disorder
   * etiology : Inactivation of both copies of the EXT tumor suppressor gene in chondrocytes in sporadic & hereditary osteochondromas
   # pts. are at ↑ risk of malignant transformation.

- **Tumor site** :

- Exostoses develop in endochondral bones (any bone that develops in and replaces cartilage) at the metaphysis from the epiphyseal growth plate of long tubular bones, especially **about the knee**
  ~.~ they tend to **stop growing** once the normal growth of the skeleton is completed

- OCCASIONALLY they develop as a sessile tumors from bones of the **s**capula & rib**s** & pelvi**s**
  RARELY they involve the short tubular bones of hands & feet

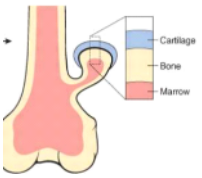
Important note : bone outgrowth may have a stalk or stem that sticks out from the normal bone.
>> If the tumor has a stalk : the structure is called **pedunculated**.
>> If the tumor outgrowth is attached to the bone with a broader base, it is called **sessile**
اذا sessile : اسسسسستغنى عن ال ssssssstalk

- **Gross appearance** :

- 1 to 20 cm in Ø

- The development of an osteochondroma begins with outgrowth from the epiphyseal growth plate that forms mushroom-shaped bony outgrowth which consists of :

1) The **cap** is : benign hyaline cartilage, resembling disorganized growth plate undergoing endochondral ossification.
2) the newly formed bone forms the **inner portion** of the head & stalk
3) the stalk cortex merge with the cortex of the host bone



- **Clinically :**

- Osteochondromas are slow-growing masses so they are asymptomatic & detected incidentally
- they becomes painful when :- 1)the stalk is fractured OR 2)they impinge on a nerve

- REMEMBER :) Solitary Osteochondroma **rarely progress to chondrosarcoma** or other sarcoma
  BUT patients with the hereditary exostoses syndrome are at ↑ **risk of malignant transformation**

---

*CHONDROMA:

❖ **Definition** :
Benign T of hyaline cartilage, developing probably from slowly proliferating rests of growth plate cartilage

❖ **Types** :
1) enchondromas >> within the bone medulla
2) juxtacortical (ecchondroma) >> on the bone surface
3) Ollier disease >> multiple chondromas that involve one side of the body (Unilateral)
4) Maffucci syndrome >> multiple chondromas that is associated with benign soft tissue angiomas

❖ **Grossly** :

● Enchondromas :
1- It affect 20 to 50y age group
2- typically solitary
3- located in the metaphyseal region of tubular bones, specially of the short tubular bones of hands & feet.
4- gray-blue, translucent nodules usually smaller than 3 cm.
5- **HISTOLOGY**-there is :-
*well-circumscribed hyaline matrix
*cytologically >> benign chondrocytes
*at the periphery >> endochondral ossification
*at the center >> calcifies & dies

● Hereditary multiple chondromatoses :
1- the islands of cartilage exhibit greater cellularity & atypia
2- difficult to distinguish them from chondrosarcoma

❖ **Clinically :**

● Enchondromas : *Most of them are asymptomatic & detected incidentally
*Occasionally: they are painful & cause pathologic fractures

● On X-ray :
1) the unmineralized nodules of cartilage appear as (O-ring sign) which is well-circumscribed oval lucencies surrounded by thin rims of radiodense bone
الجزء الداخلي البيضوي بكون radioluscent
اما الجزء يلي بحوط الدويرة بكون radiodense
2) Calcified matrix exhibits irregular opacities

● The growth potential of chondromas is limited, & most remain stable, although they can recur if incompletely excised

● Solitary chondromas >> rarely undergo malignant transformation
Enchondromatoses >> ↑ risk of malignant transformation

Maffucci syndrome >> ↑risk of developing other types of malignancies, like ovarian ca & brain gliomas

---

*CHONDROSARCOMA :

- **Definition** : malignant T that produce neoplastic cartilage
  - they occur roughly half as frequently as OS — عدد المرات ال عدد المرات التي يحدث فيها ال condrosarcoma يساوي نصف عدد المرات التي يحدث فيها ال sarcoma
  - most patients are age 40 or older
  - M/F ratio = 2/1.

- **Morphology :**

1) **Conventional ChS** :
   *arise within the medullary cavity to form an expansile glistening mass that often erodes the cortex.
   *they exhibit malignant hyaline & myxoid cartilage

2) **myxoid ChS :**
   *viscous and gelatinous & the matrix oozes from the cut surface
   *spotty calcifications are typically present
   *central necrosis can create cystic spaces اكياس فاضية
   *the adjacent cortex is thickened or eroded متآكلة
   *the tumor **grows with broad pushing fronts** into marrow spaces & the surrounding soft tissue.

3) **other histological variants :** clear-cell & mesenchymal ChSs

- **Tumor site :** pelvis, shoulder, & ribs
  in contrast to enchondromas, ChSs rarely involve the distal extremities

- **Metastesis** : hematogenously, to the lungs & skeleton

- **T grade :**
  *it is determined by : cellularity & cytologic atypia & mitotic activity

  *RULES : 1) the more radiolucent the T is, the greater the likelihood that it is high grade
  2) Low-grade T : resemble normal cartilage.
  3) Higher grade T : contain pleomorphic chondrocytes with frequent mitotic figures.
  multinucleate cells are present with lacunae containing two or more chondrocytes

  *10% of conventional low-grade ChS have areas of, second high-grade poorly differentiated components (dedifferentiated ChS) that include foci of fibro- or osteosarcomas (This is called, **tumor heterogeneity**).

  *The a direct correlation between grade & biologic behavior of the tumor :
  1) most conventional ChSs are lowgrade, & indolent with a 5-year survival rate of 80% to 90%
  BUT grade 3 tumors have 43% survival rate
  2) grade 1 tumors rarely metastasize, whereas 70% of the grade 3 tumors disseminate.

  *The size is another prognostic feature :
  tumors larger than 10 cm being significantly more aggressive than smaller tumors

- **TTT** :

1) surgical excision

2) chemotherapy >> it is added for the mesenchymal & dedifferentiated variants
(because of their aggressive clinical course)