Les chondrosarcomes de l'enfant

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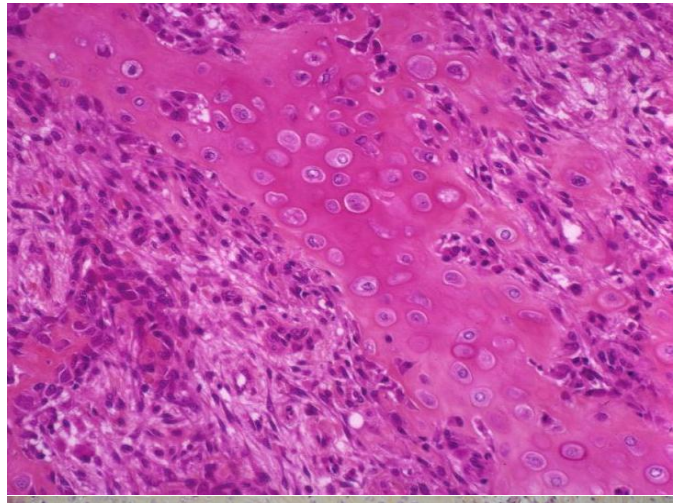
Journée « Tumeurs rares en pédiatrie »

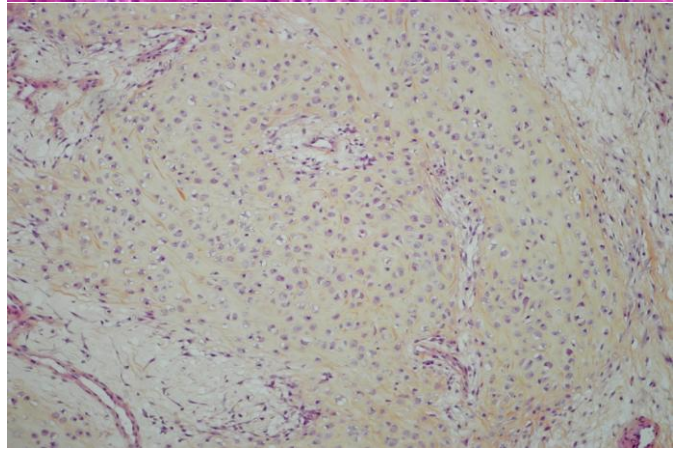
15 avril 2011, Paris

Diagnostic différentiel d'une lésion cartilagineuse chez l'enfant

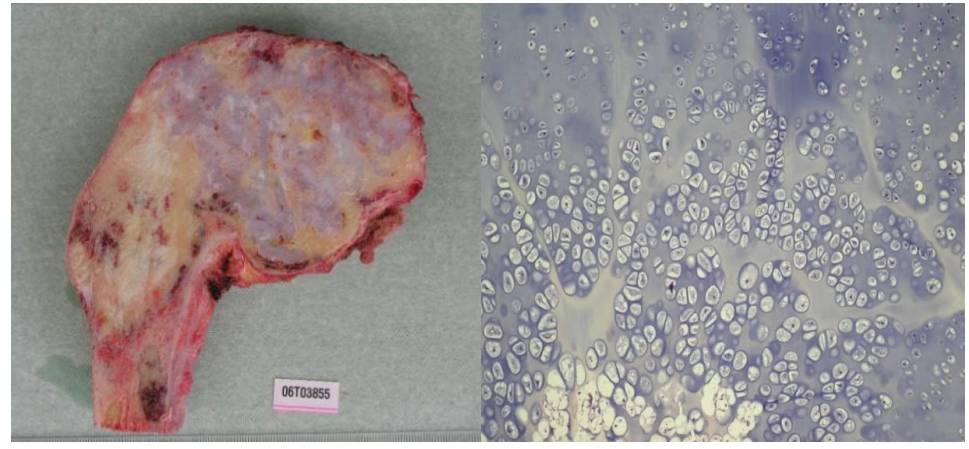
1- Eliminer une lésion pseudotumorale

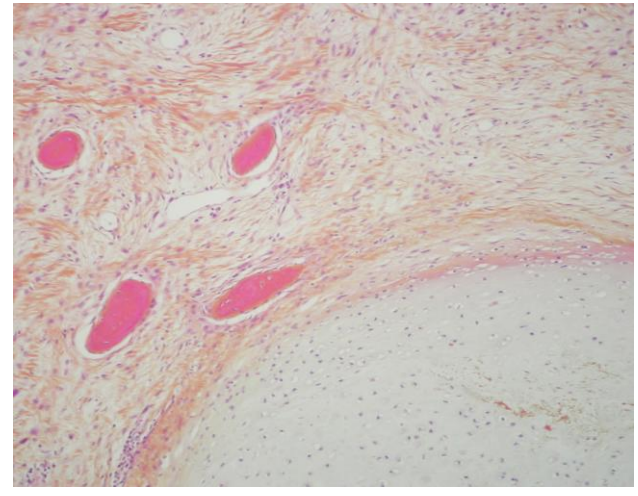
Cal avec composante cartilagineuse





Dysplasie fibro-cartilagineuse





Diagnostic différentiel d'une lésion cartilagineuse chez l'enfant

2- Si tumeur cartilagineuse

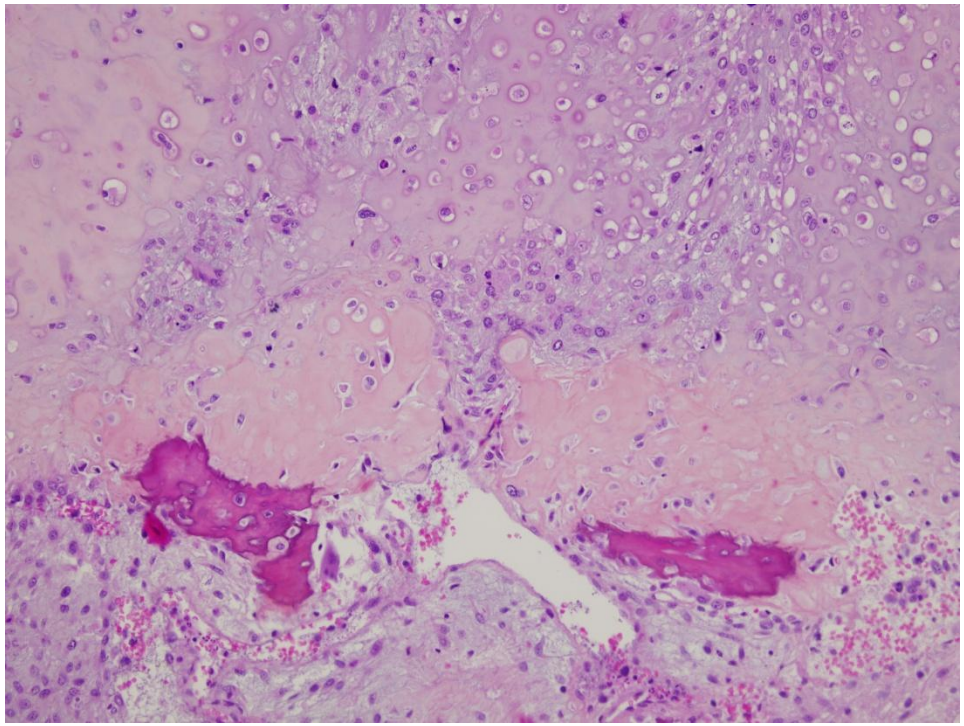
- Tumeurs bénignes
  - Ostéochondrome (découverte 10-15 ans +++)
  - Enchondrome/Chondrome périosté (découverte après la 2ème décade)
  - Chondroblastome (90% 2ème 3ème décades)
  - Fibrome chondromyxoïde (2ème 3ème décades/aile iliaque)
- Si critères en faveur de la malignité:

  Diagnostic à évoquer en premier lieu:

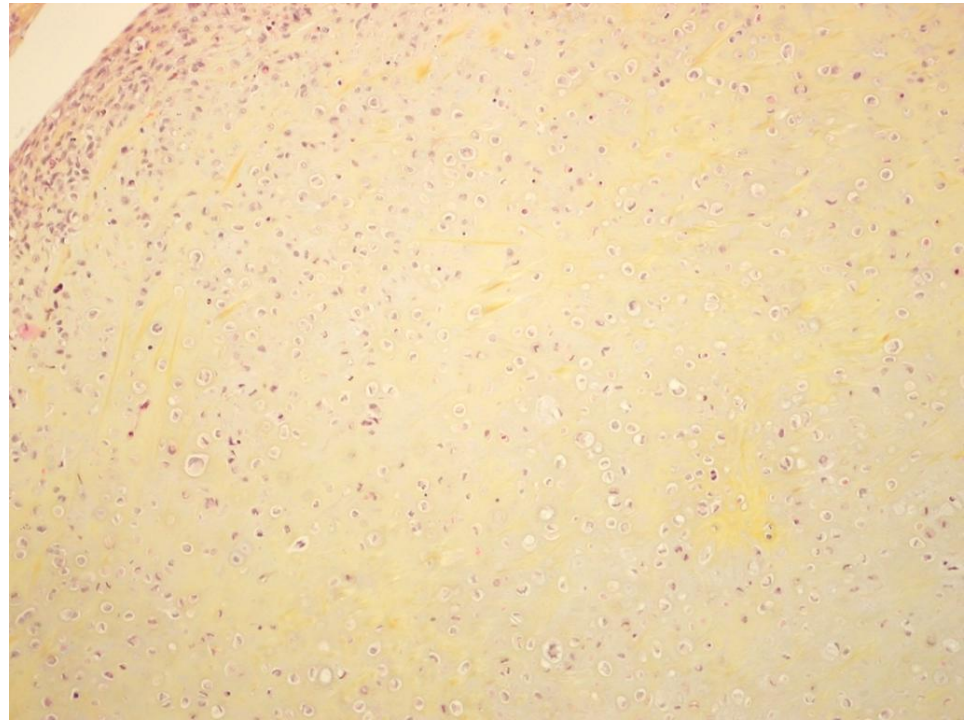
  ostéosarcome chondroblastique

Critères morphologiques suggérant un ostéosarcome chondroblastique / chondrosarcome

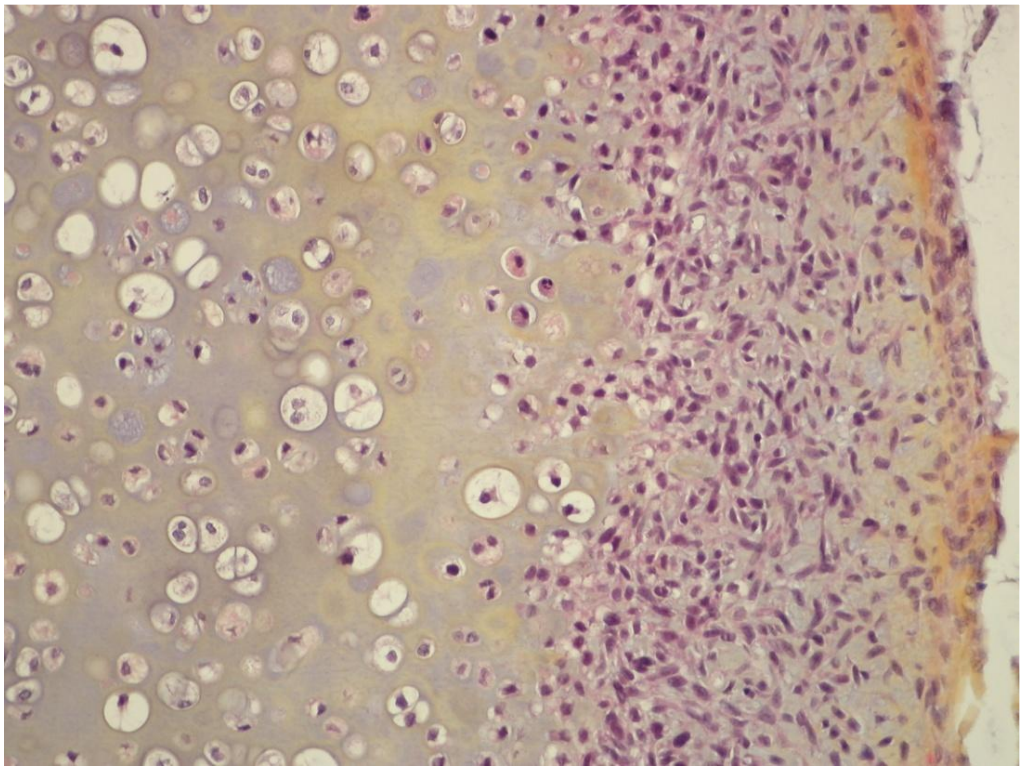
1- Secteurs d'ostéogenèse tumorale (peuvent être minimes)



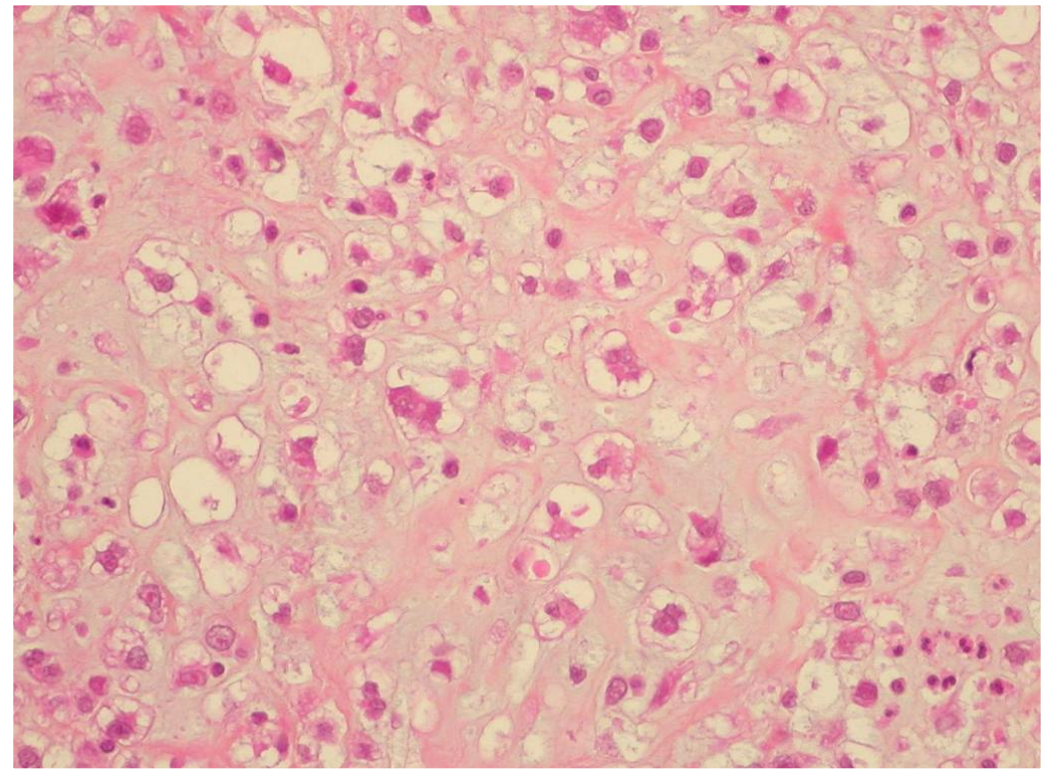
2- Cartilage tumoral organisé en larges plages de cartilage fibro-hyalin



3- Zones d’ hypercellularité à la périphérie des plages de cartilage tumoral : cellules fusiformes atypiques, mitoses.



4- Cartilage tumoral de haut grade, de grade 3



5- Marqueurs immunohistochimiques

EZRINE
Produit du gène *VIL2* (6q25-26)

GALECTINE-1
Lectines de type S Ca ind

Impliquées dans:
le maintien de la forme et de la polarité cellulaires (ezrine)
l'adhérence cellulaire
l'apoptose
La croissance cellulaire
la transduction du signal

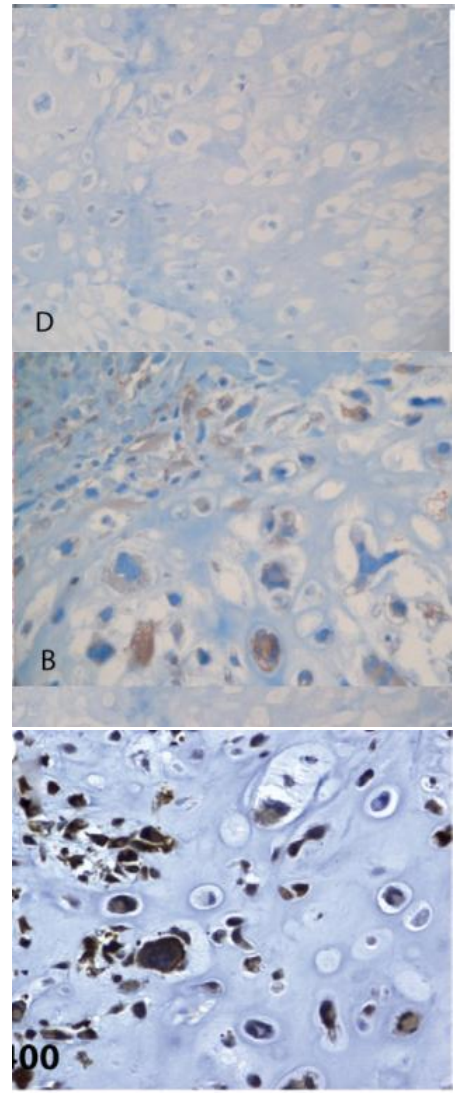
Fortement exprimées par les ostéoblastes

Diagnostic différentiel
ostéosarcome chondroblastique/chondrosarcome

Chondrosarcomes conventionnels centraux (n=101)
Grades 1-2-3
Ezrine non exprimée

Ostéosarcomes chondroblastiques (n= 16)
Ezrine exprimée dans 10/16 cas soit **62,5%**

Salas et al, Virchows Arch 2009

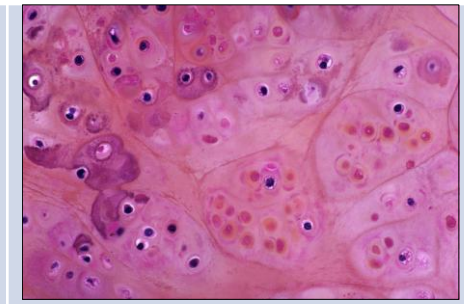
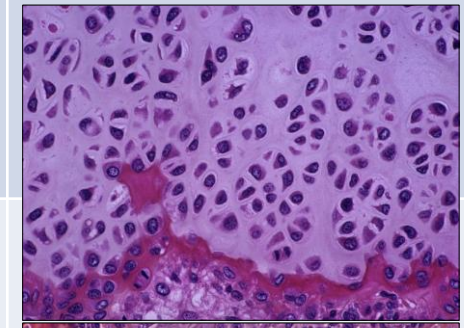
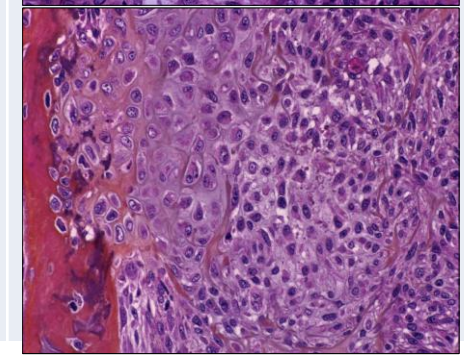


Chondrosarcomes conventionnels centraux (n=66)
Galectine-1 exprimée dans 12/66 soit 18%

Ostéosarcomes chondroblastiques (n= 25)
Galectine-1 exprimée dans 22/ 25 cas soit **88 %**

Gomez-Brouchet et al, Hum Pathol 2010

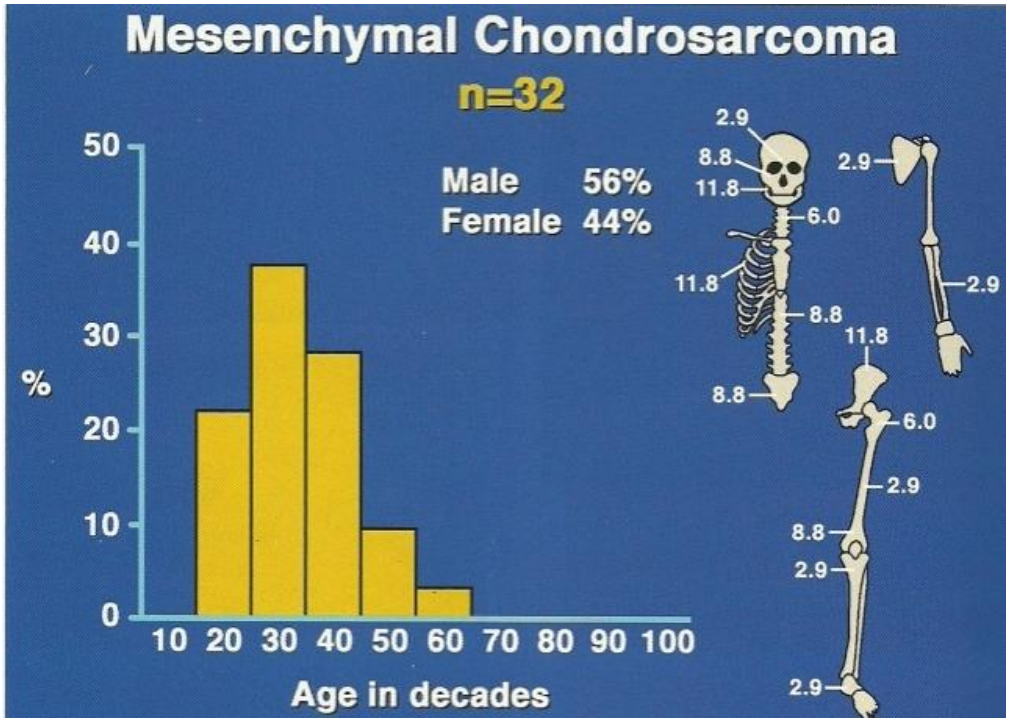
Classification des chondrosarcomes

Type	Incidence (/ensemble des CS)	Grade (O'Neal et Ackerman)	
Chondrosarcome conventionnel	85 %		
Central	**85%**	**Grade 1 (26 à 50%)** Grade 2 (30-60%) Grade 3 (8 à 25%)	
Périphérique	15%		
Chondrosarcomes (variantes rares)	15%		
Dédifférencié	6-10%	Assimilé grade 3	
Mésenchymateux	2%	Assimilé grade 3	
Périosté	2%	Grade 1-2	
À cellules claires	1%	Assimilé grade 1	

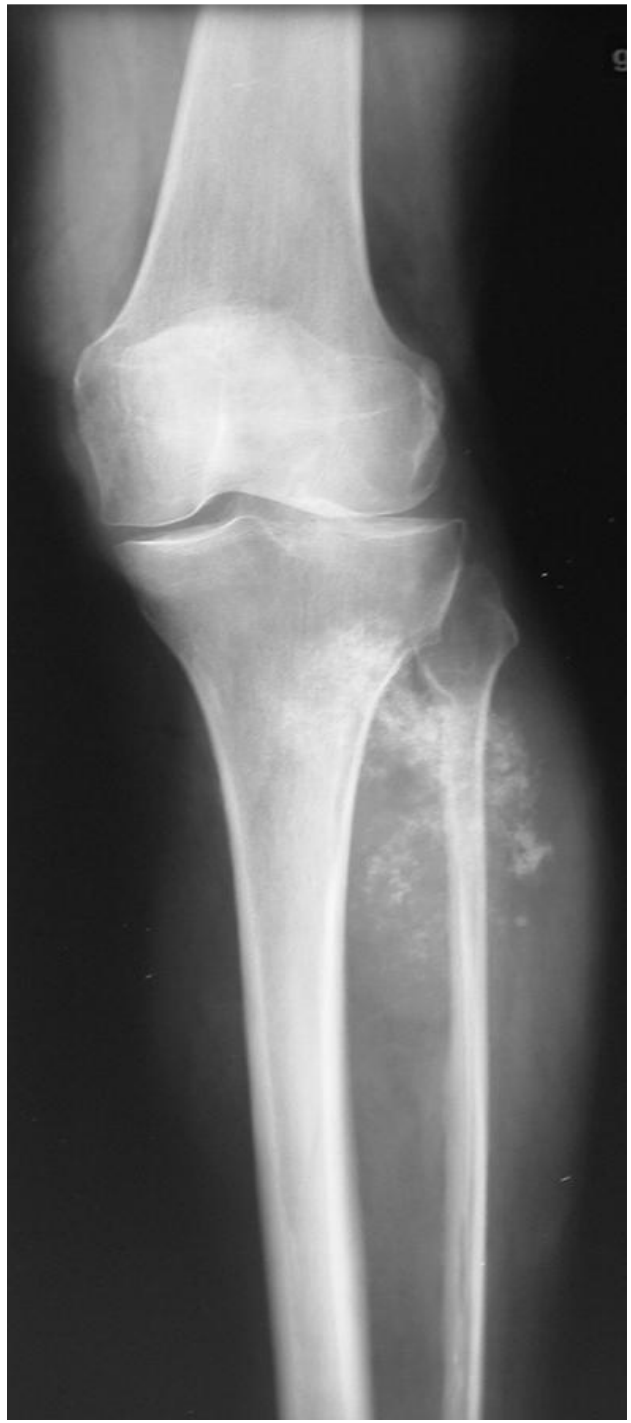
2005-2010 / 50 chondrosarcomes < 30 ans

AGE	LOCALISATION	TYPE CHONDROSARCOME	GRADE
H11 ans	Aile iliaque	Conventionnel central	1
F16 ans	Côte	Chondrosarcome périphérique sur ostéochondrome	1
F12 ans	Tibia	Conventionnel central	1
F16 ans	Talon (calcanéum, astragale, métat)	Conventionnel	2
H14 ans	Condyle fémoral	Conventionnel central	2
H15 ans	Clavicule	Périosté	1
H17 ans		Chondrosarcome périphérique sur ostéochondrome	1
F17 ans	Fémur proximal	Conventionnel central	1
F18 ans	Condyle fémoral	Conventionnel central	1

CHONDROSARCOME MESENCHYMATEUX



< 3% des CS
10-30 ans

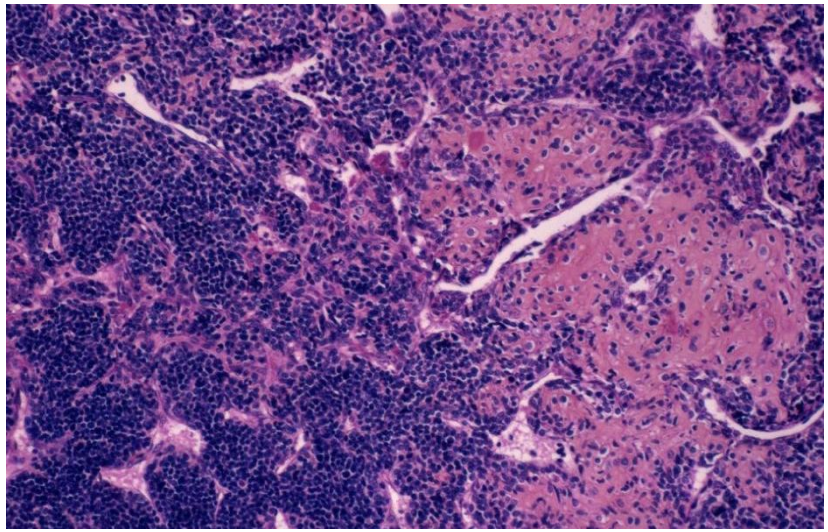


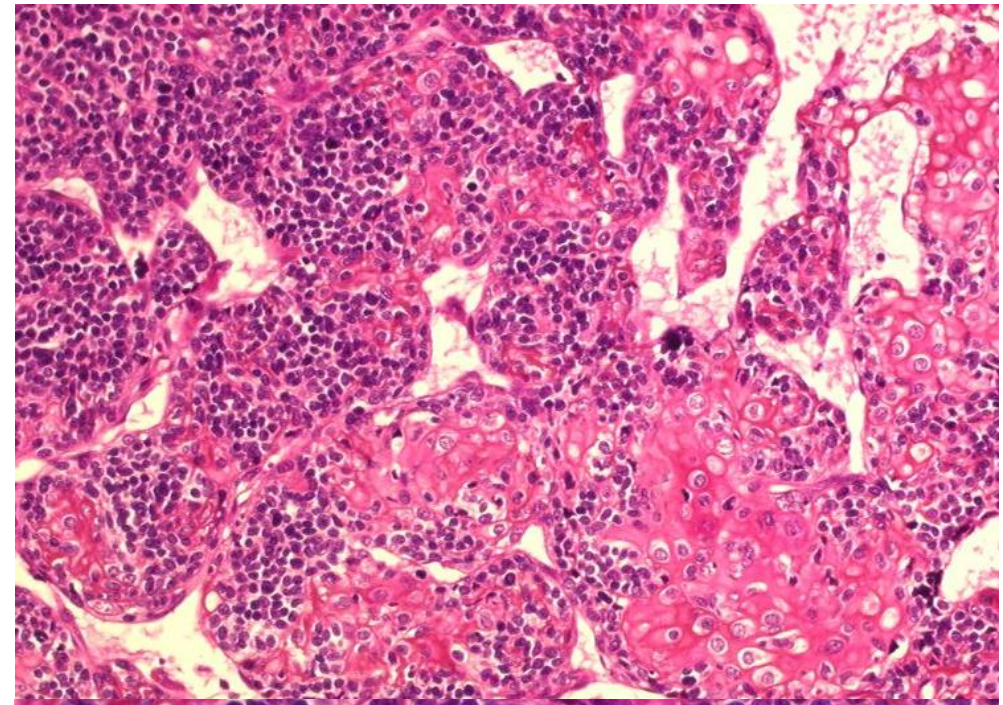
Squelette axial
(cranio-facial, côtes, rachis, iliaque)
Atteinte osseuse multifocale possible
Atteinte extra-squelettique (20 à 30% des cas)
Méninges ++

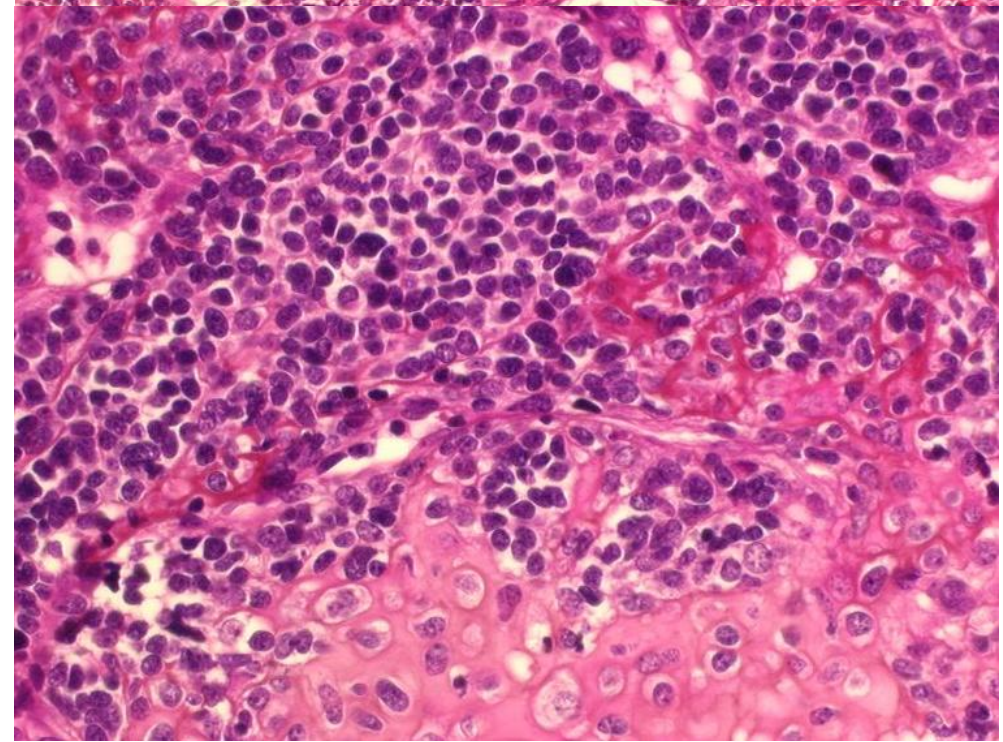
Petites cellules rondes

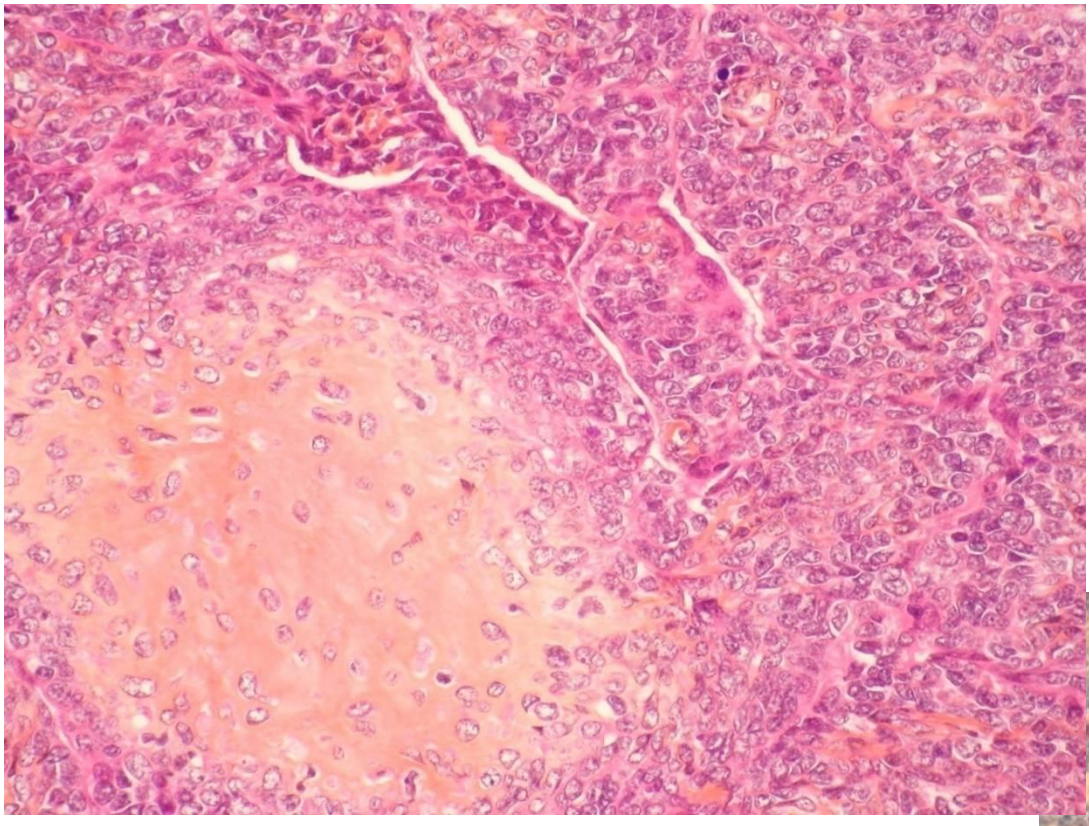
Ilots de chondrosarcome de bas grade

Vascularisation de type hémangopéricytaire

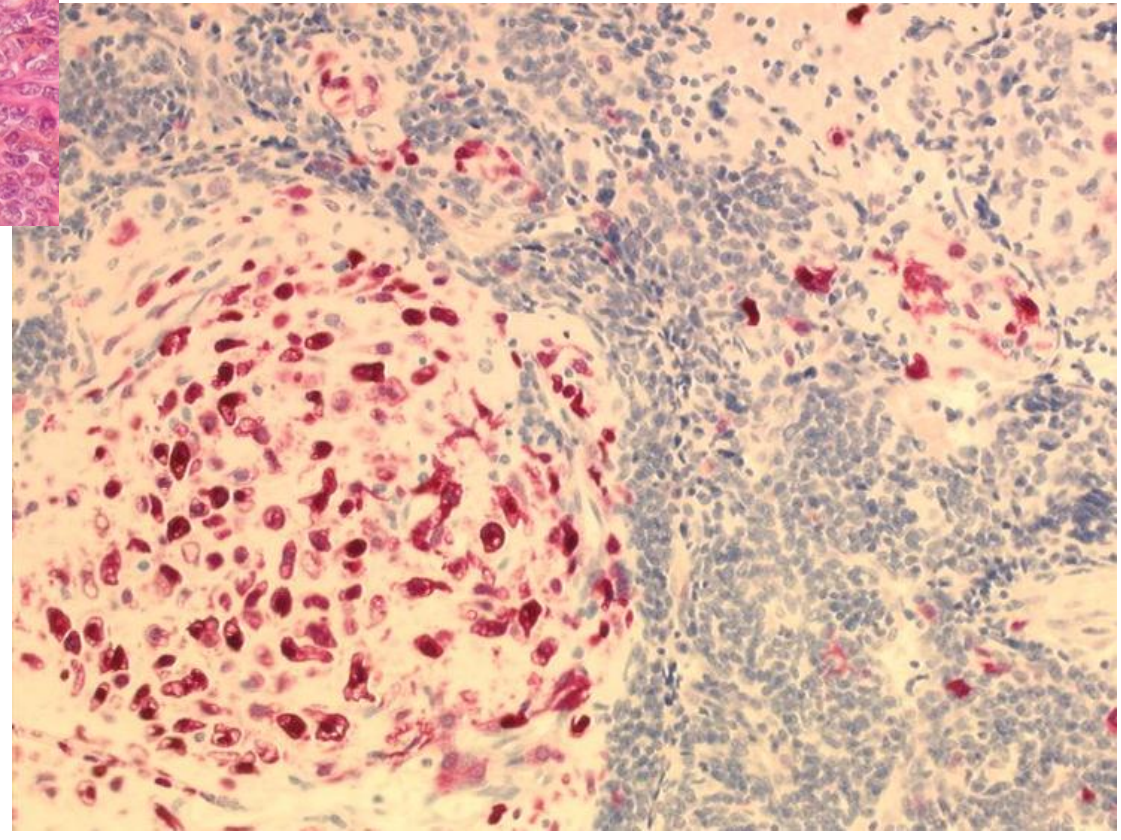






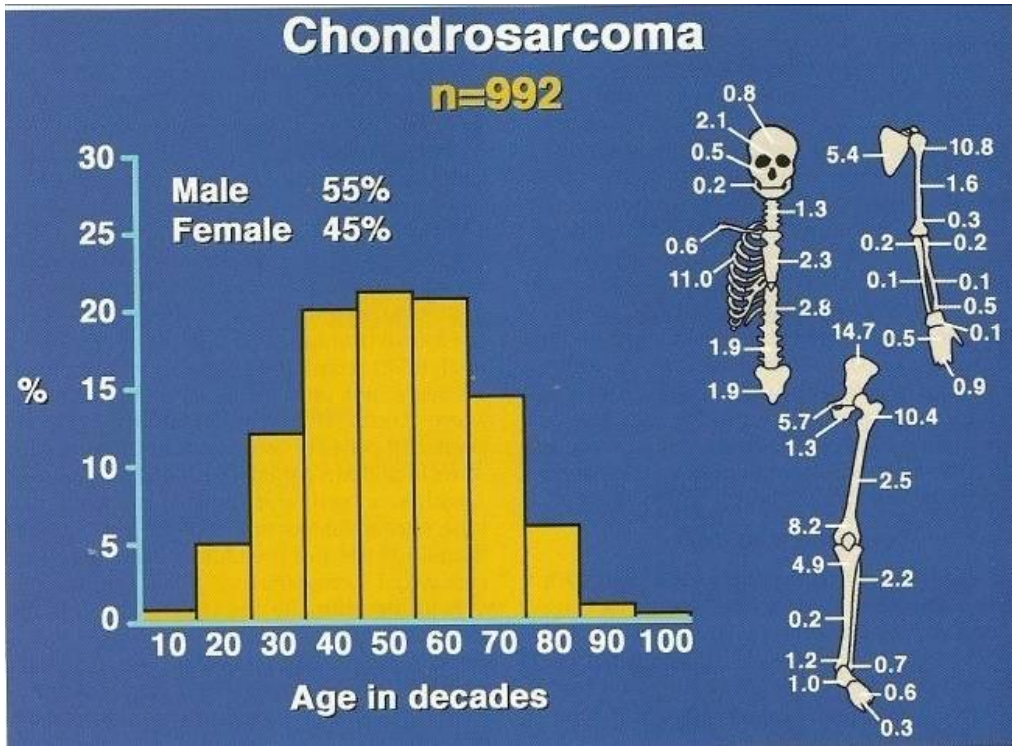


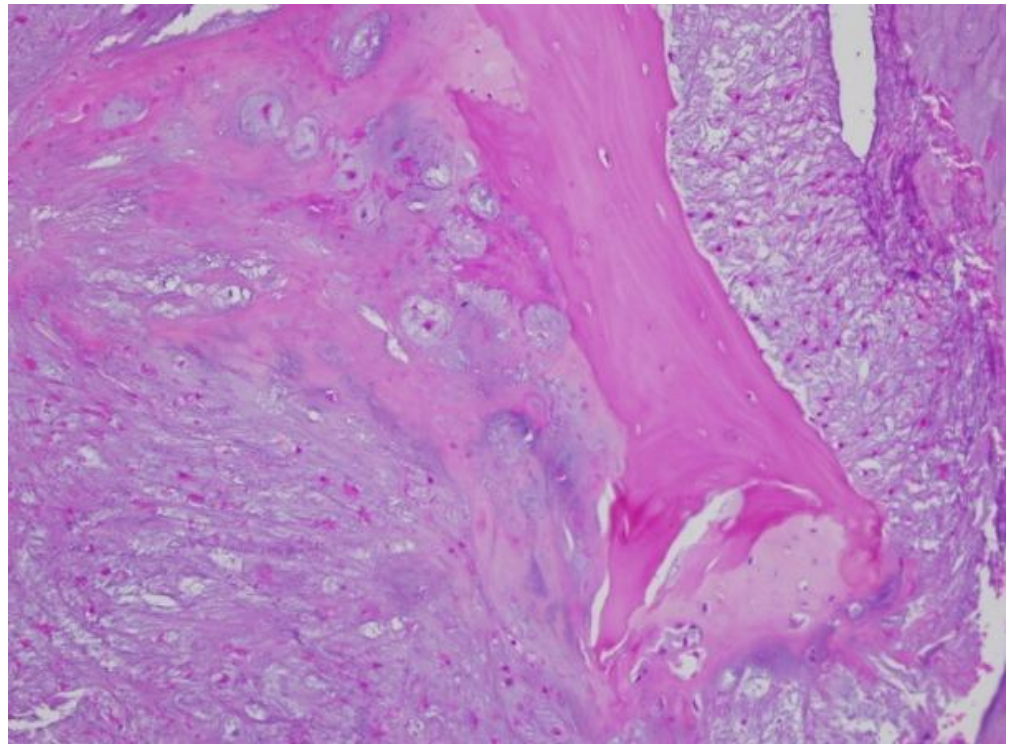
PS100 exprimée par la composante chondrosarcomateuse
Pas de marqueur diagnostique fiable Concernant la composante à cellules Rondes (positivité possible de Sox 9)



Pas de translocation ou anomalie moléculaire spécifique connue.

Chondrosarcomes conventionnels centraux (5 cas)



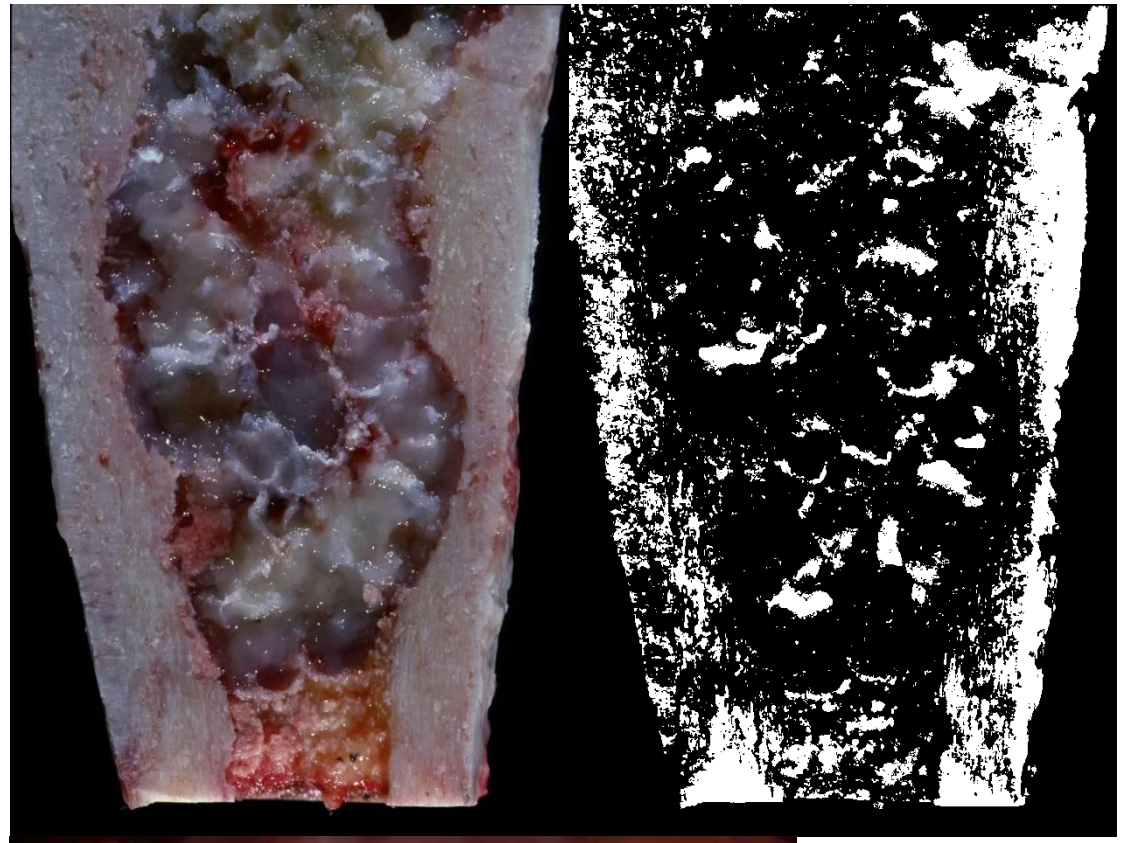


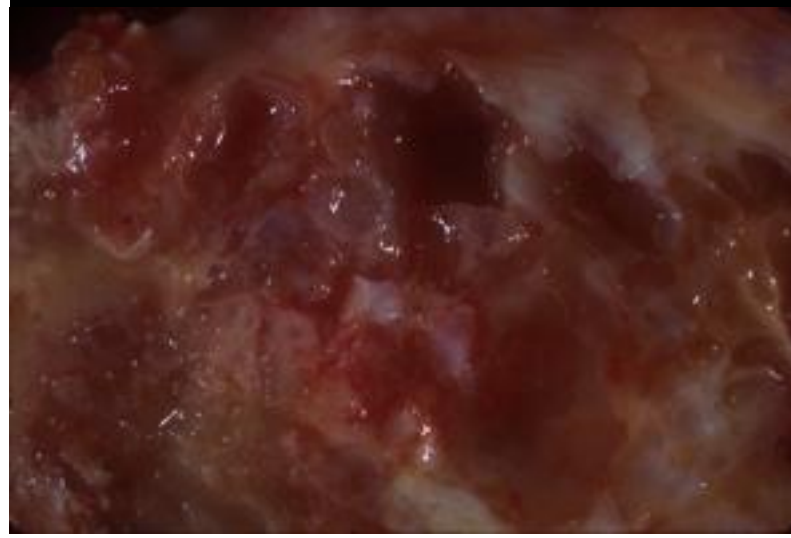
Cytologie

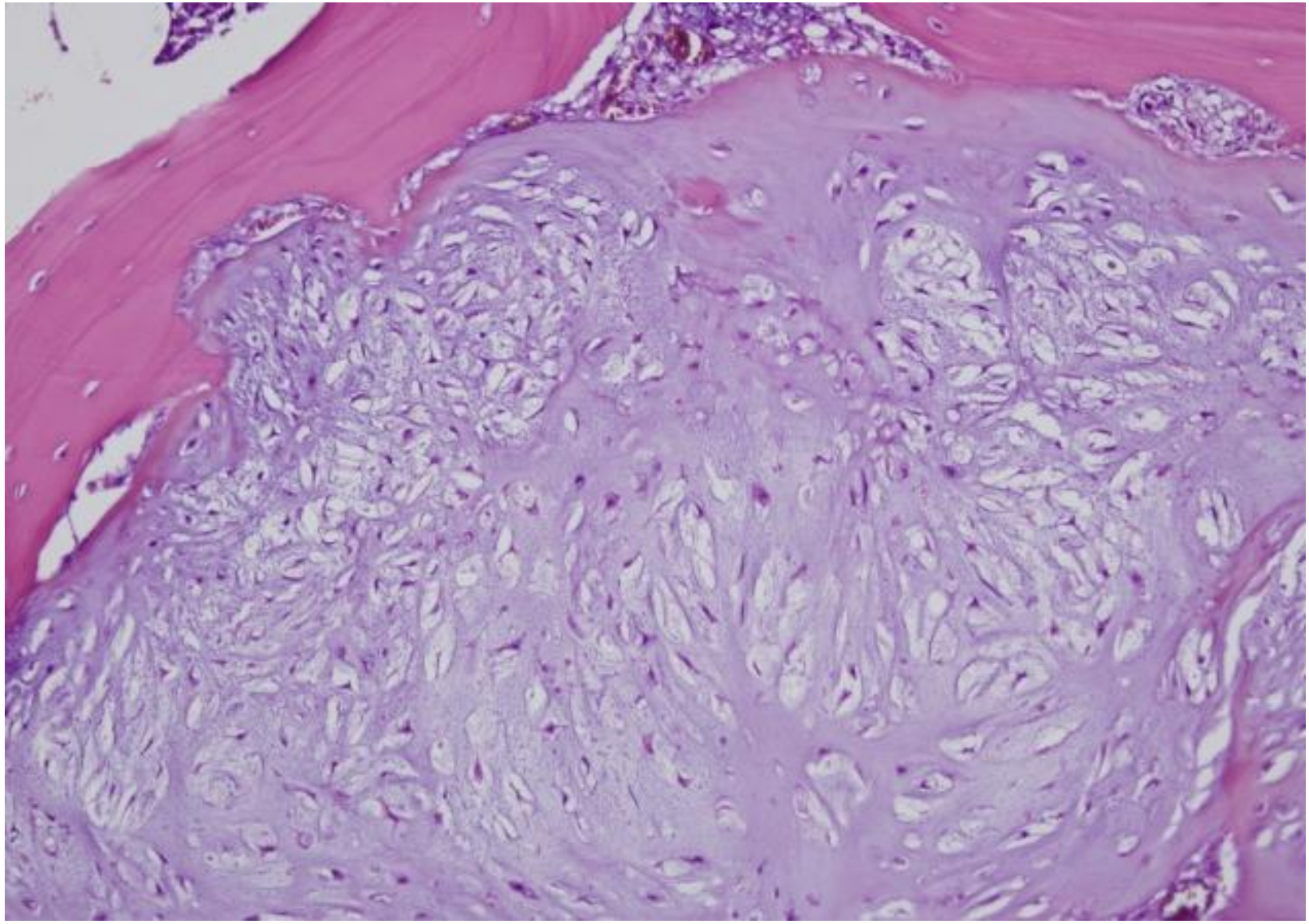
Atypies cyto-nucléaires (grades 2 et 3)

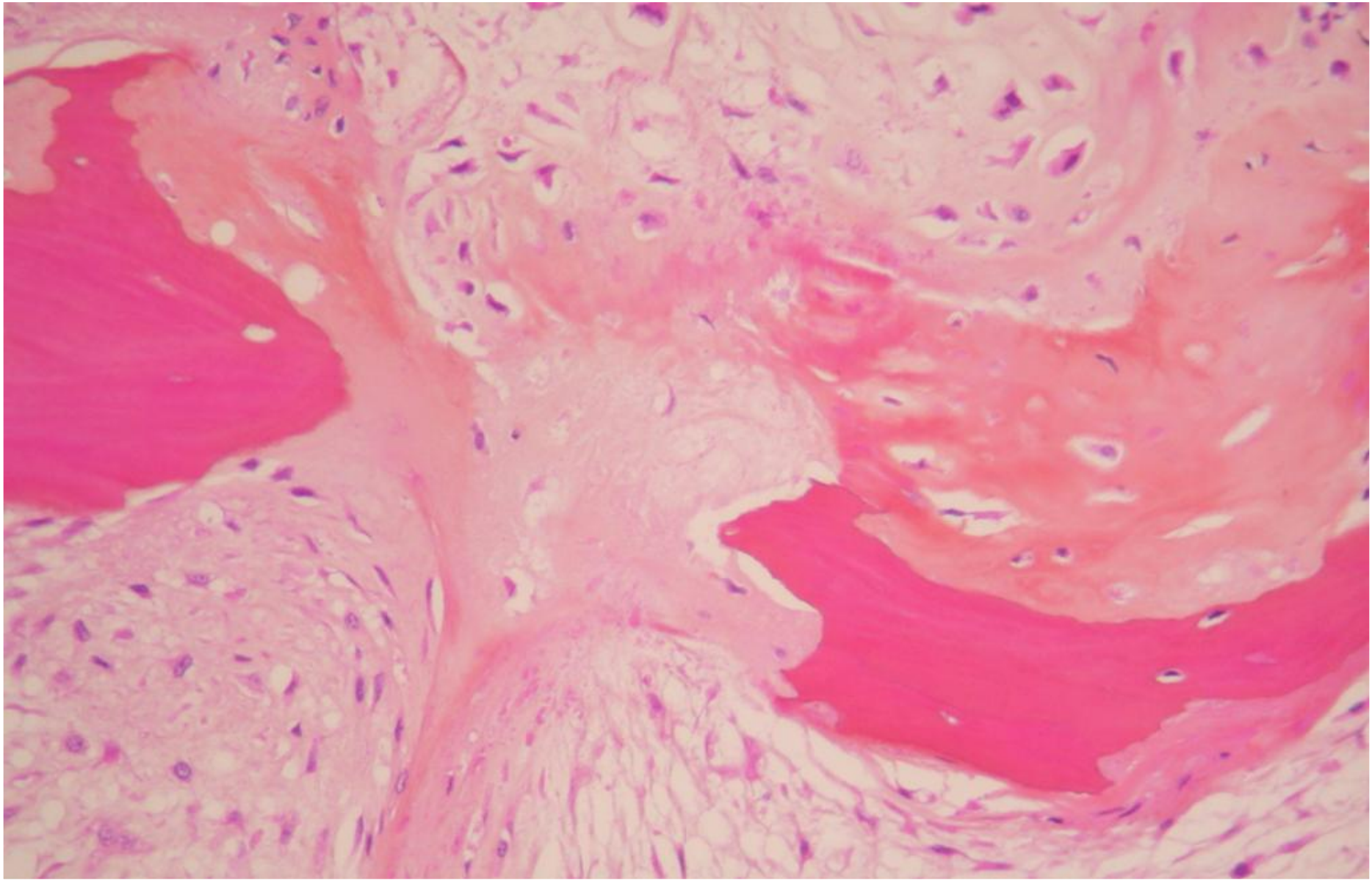
Résorption par la tumeur de l'os pré-existant (trabéculaire du spongieux, compact de la cortical)

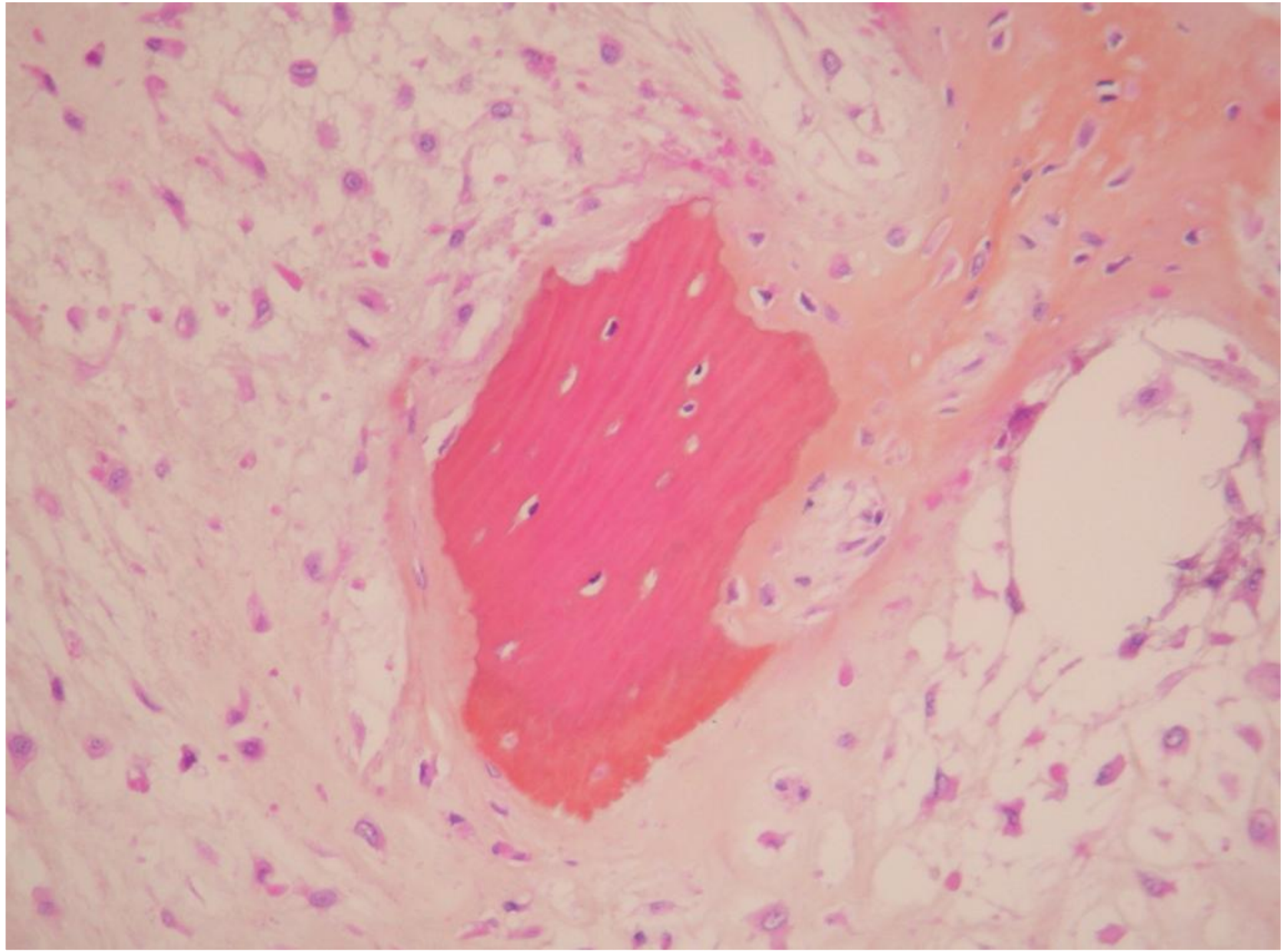
Remaniements myxoïdes étendus (>20% de l'échantillon tumoral examiné)

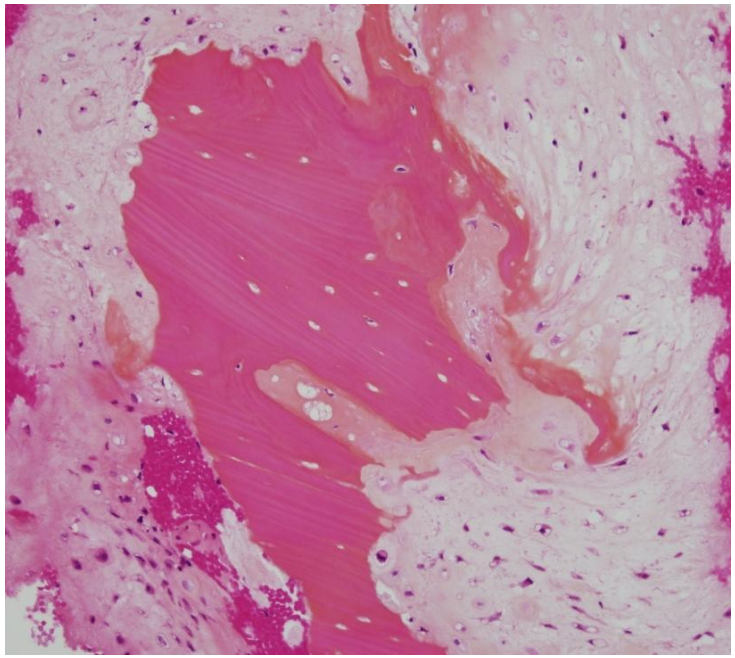
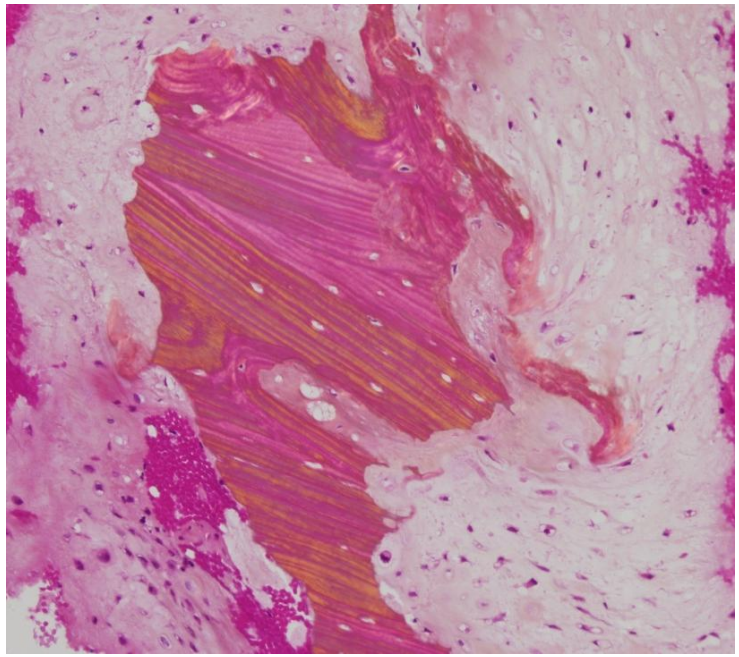
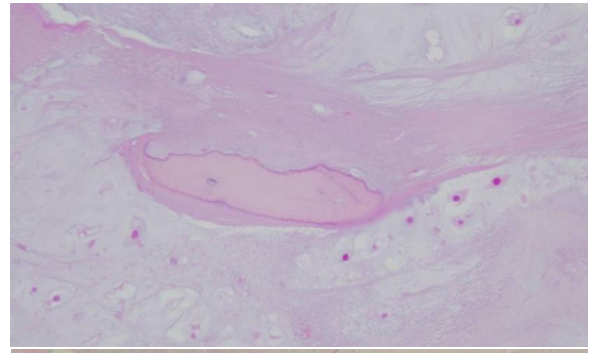
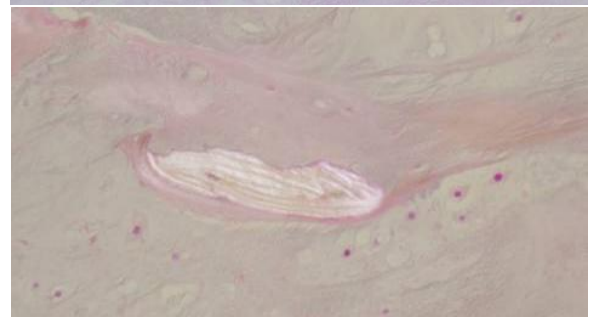
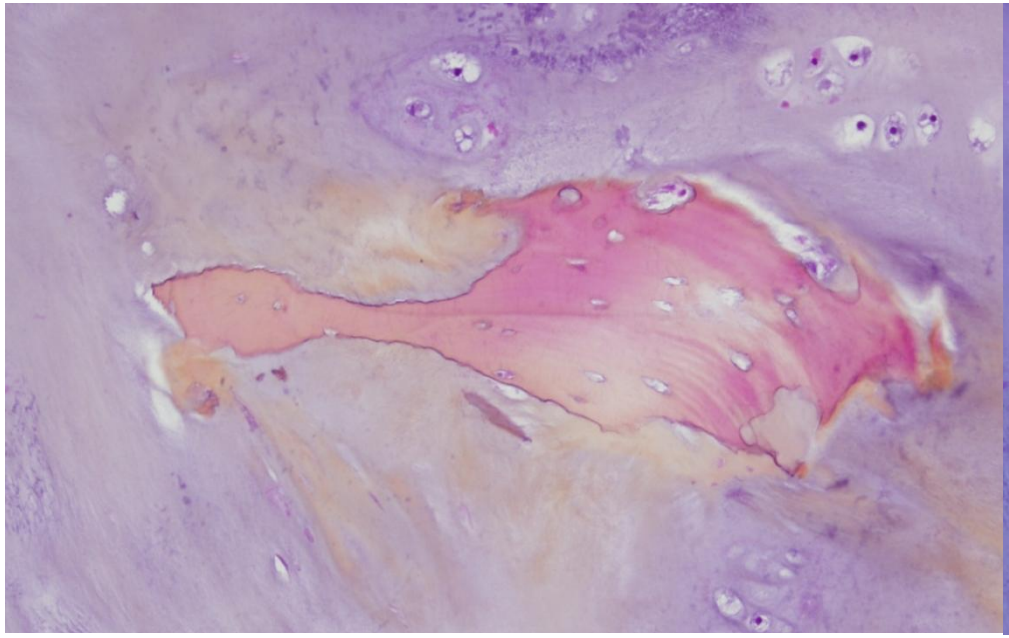
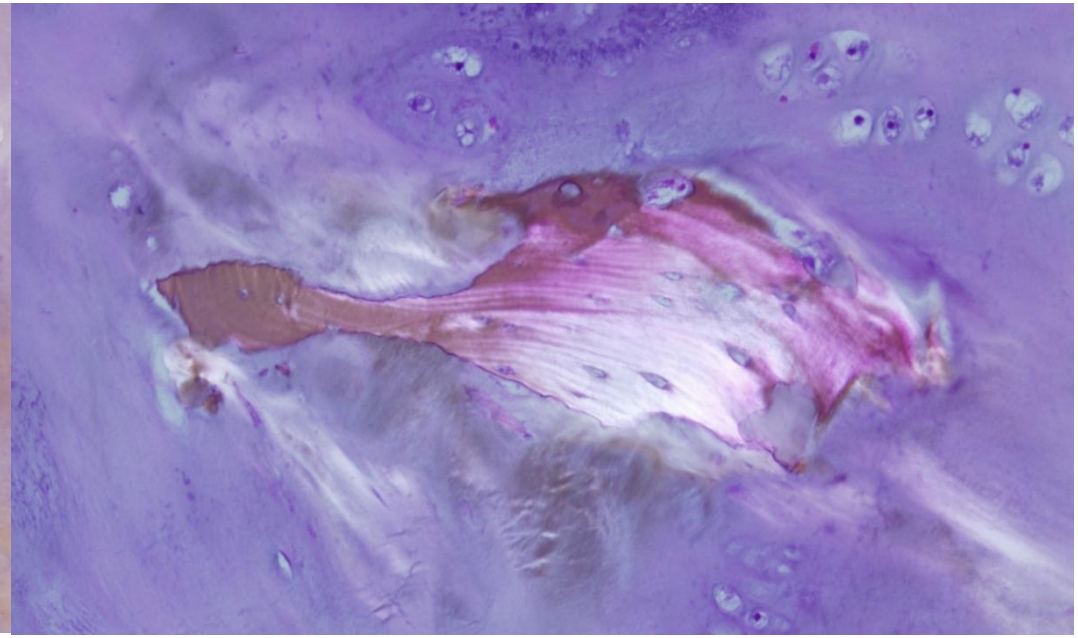




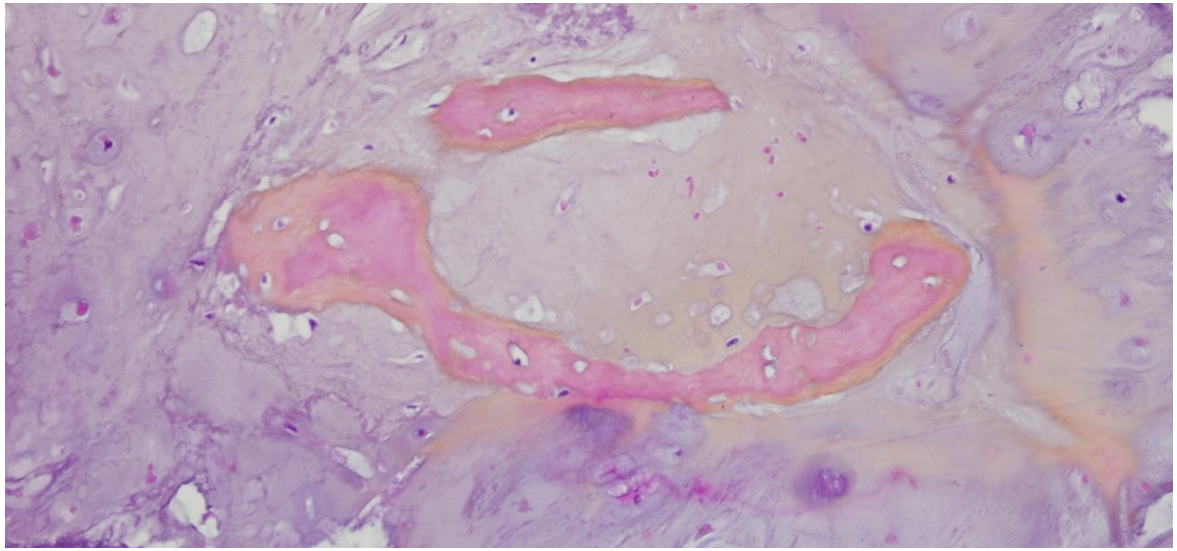


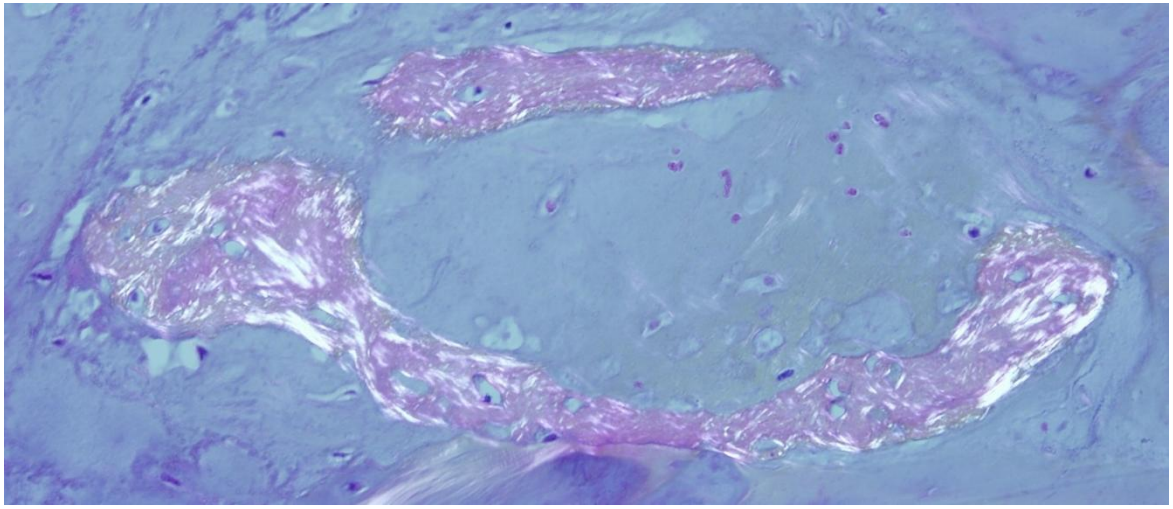


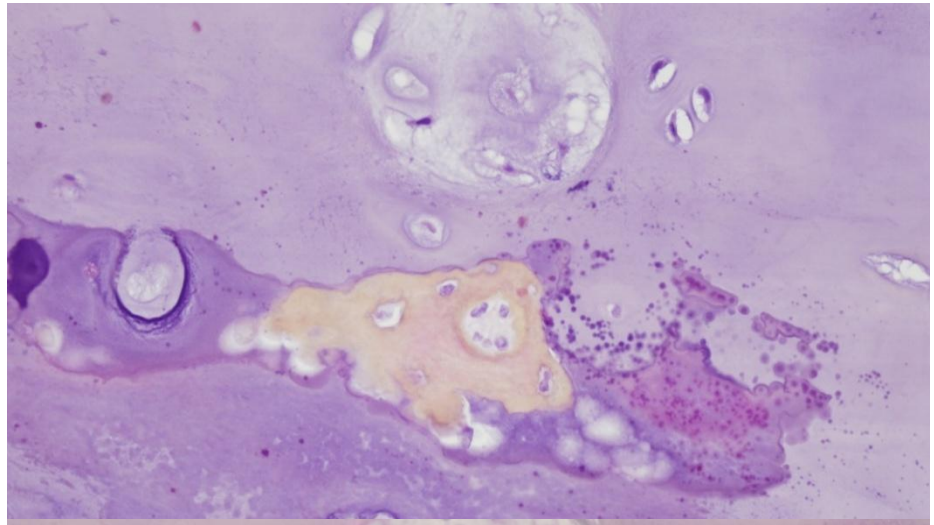


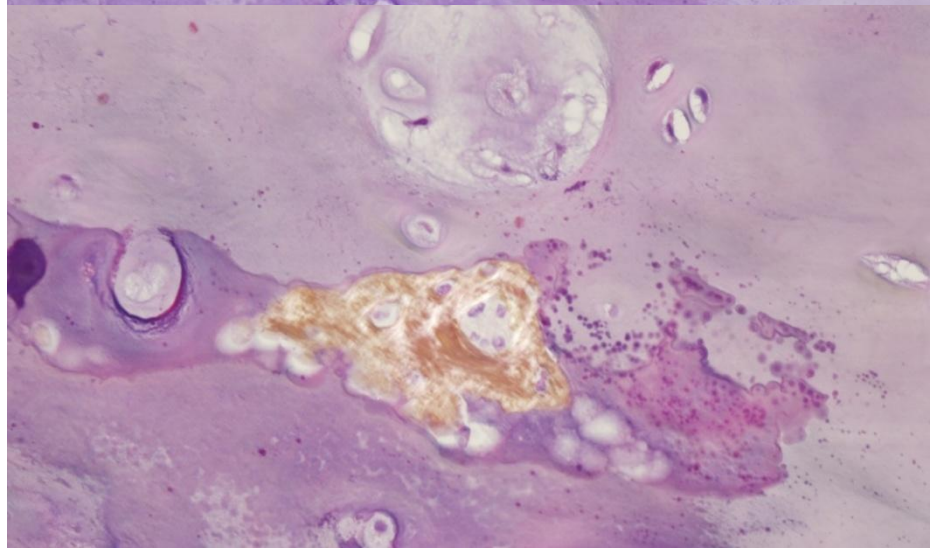







Diagnostic différentiel: l'ossification enchondrale architecture fibrillaire de l'os néoformé

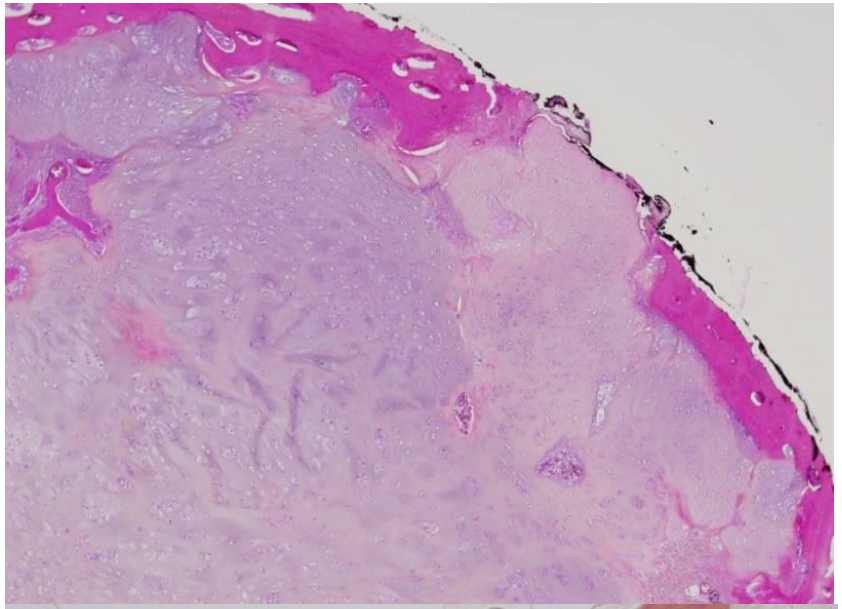


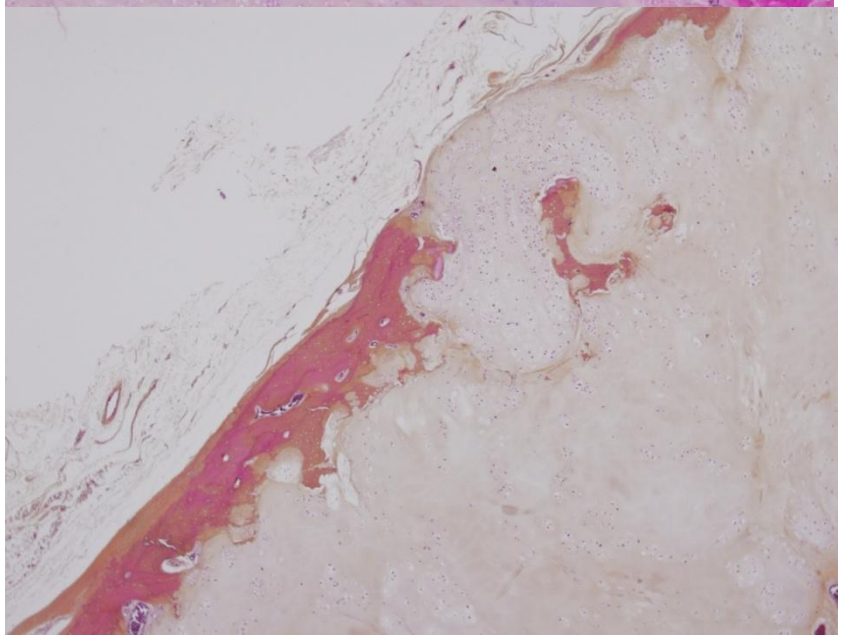


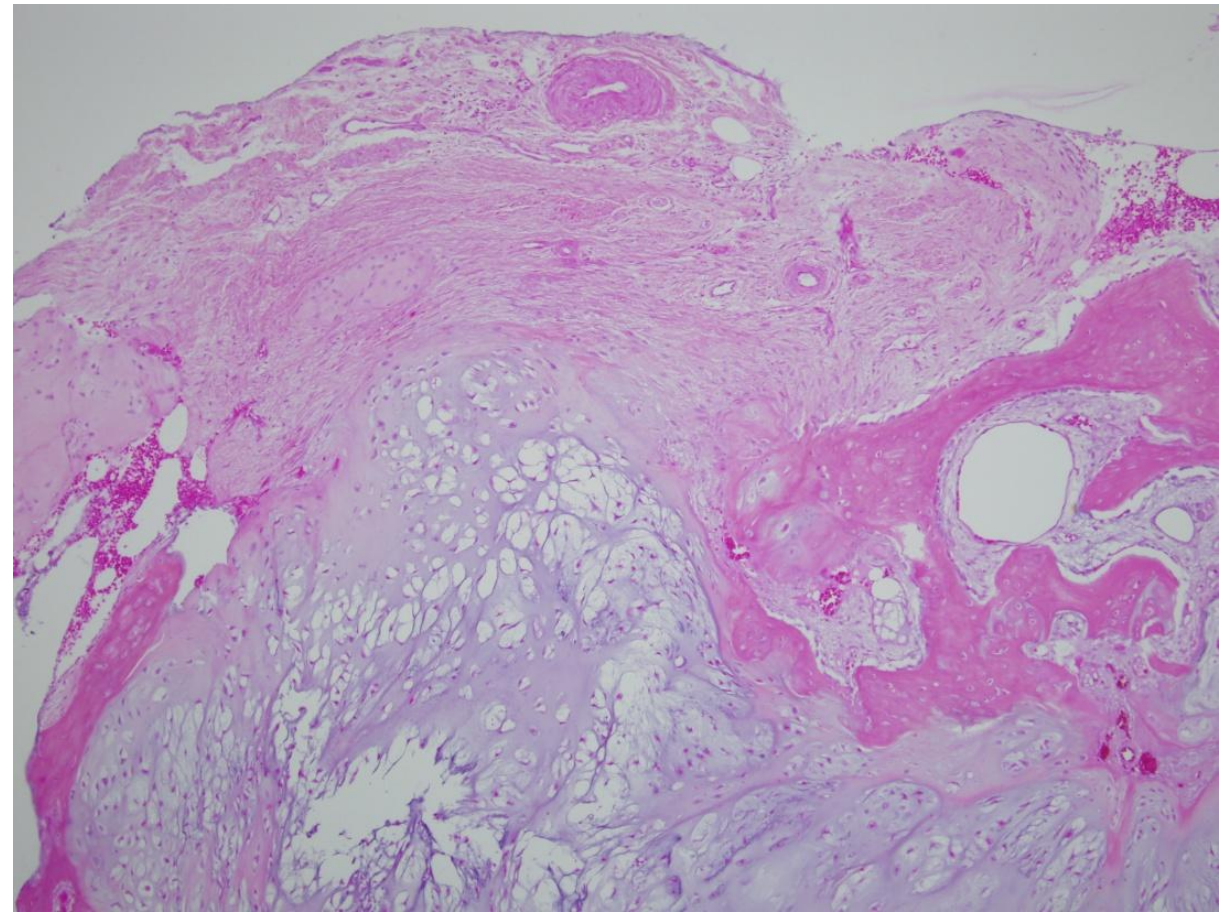


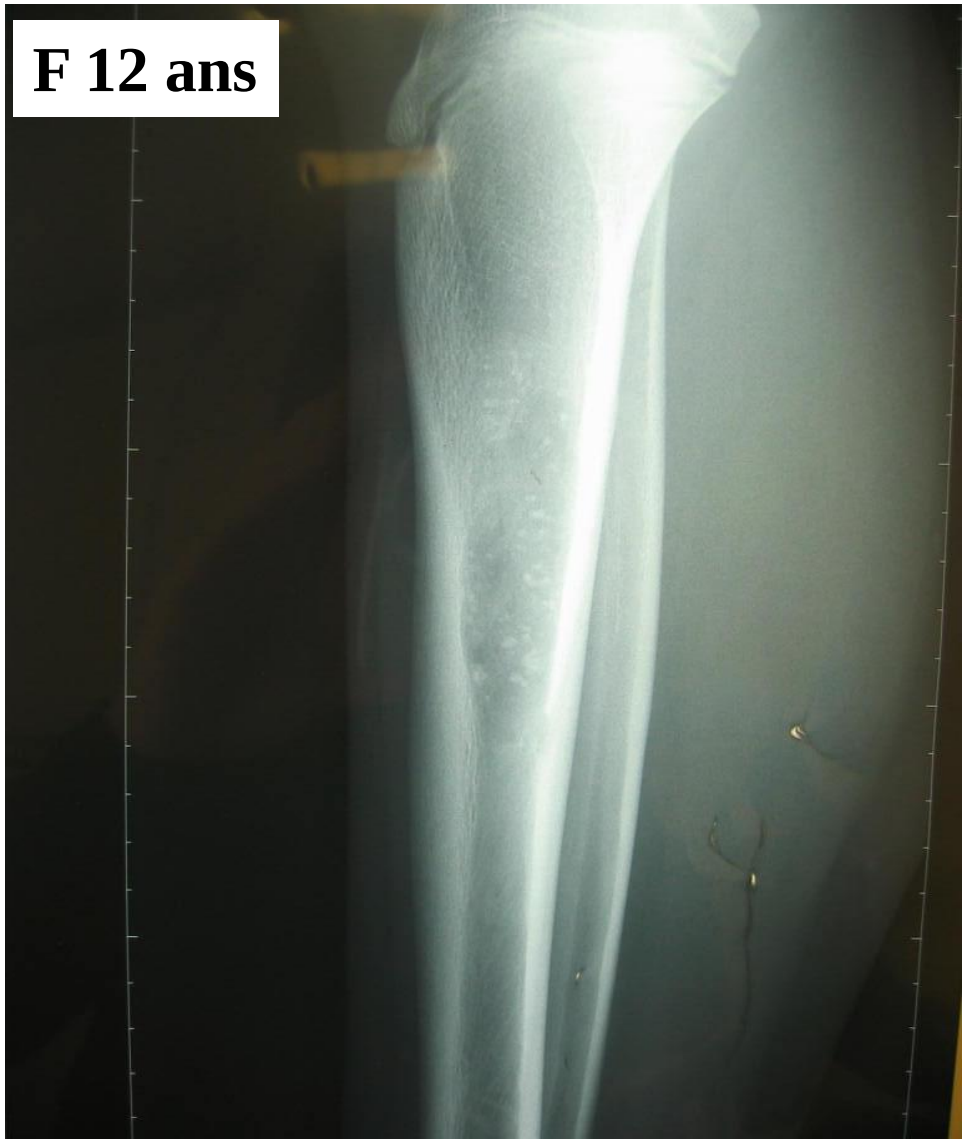


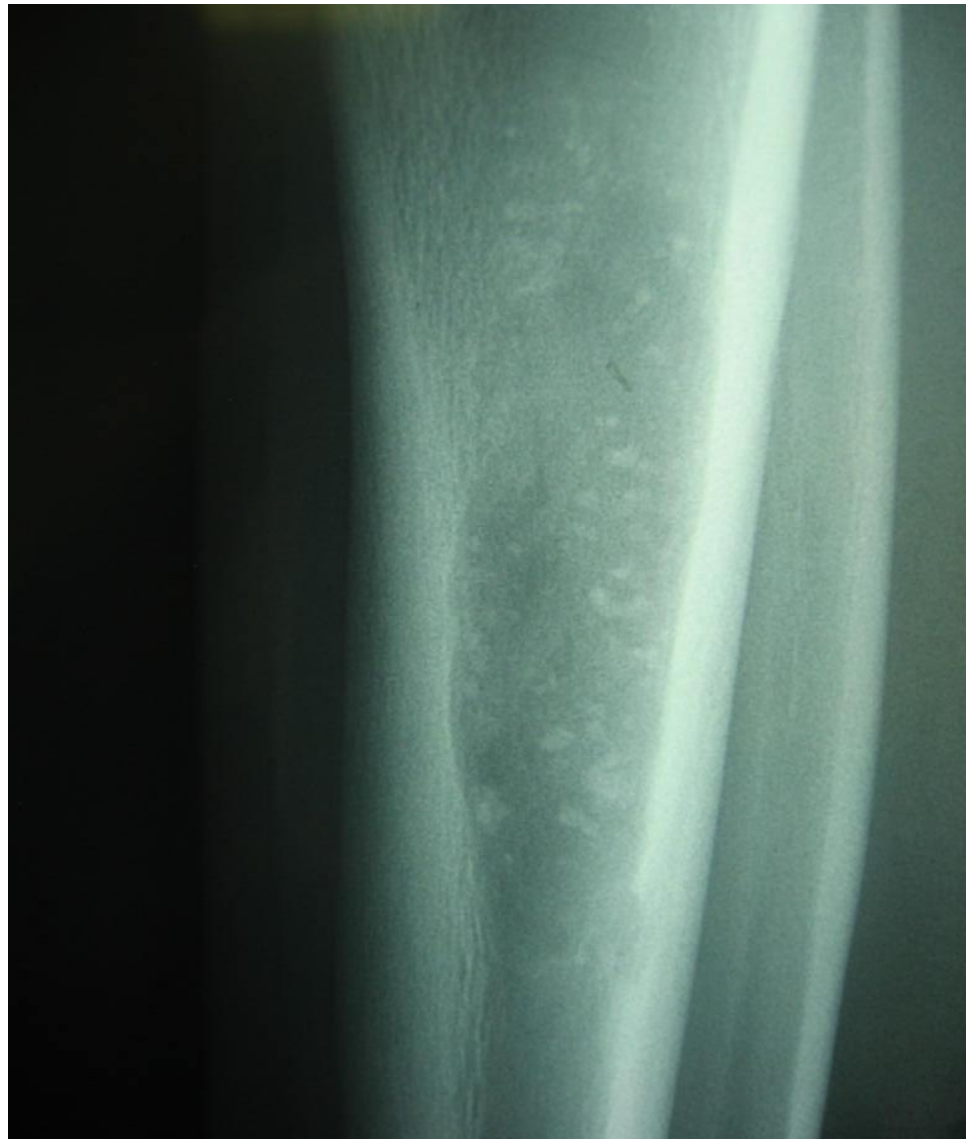
Erosion et rupture de la corticale

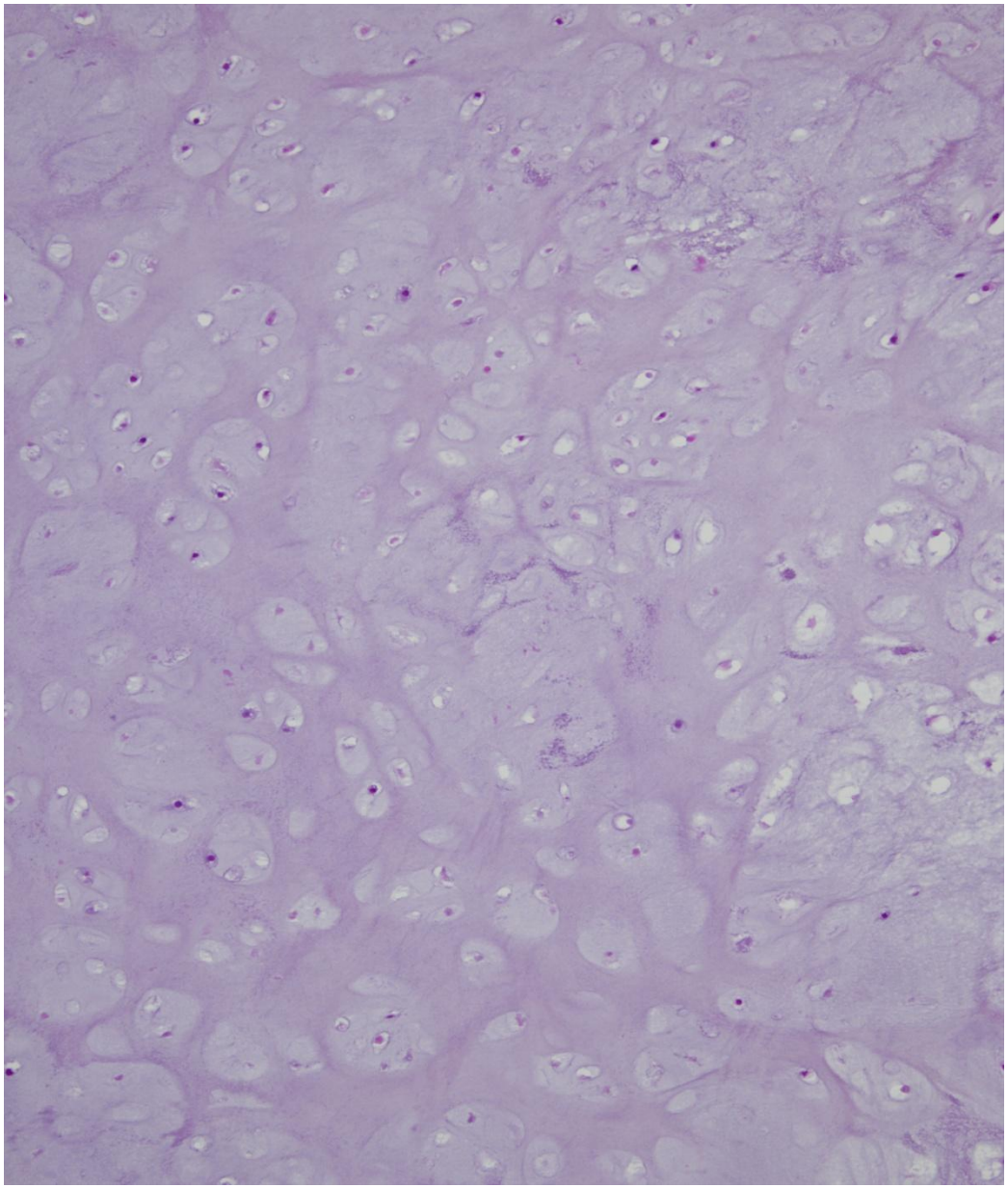


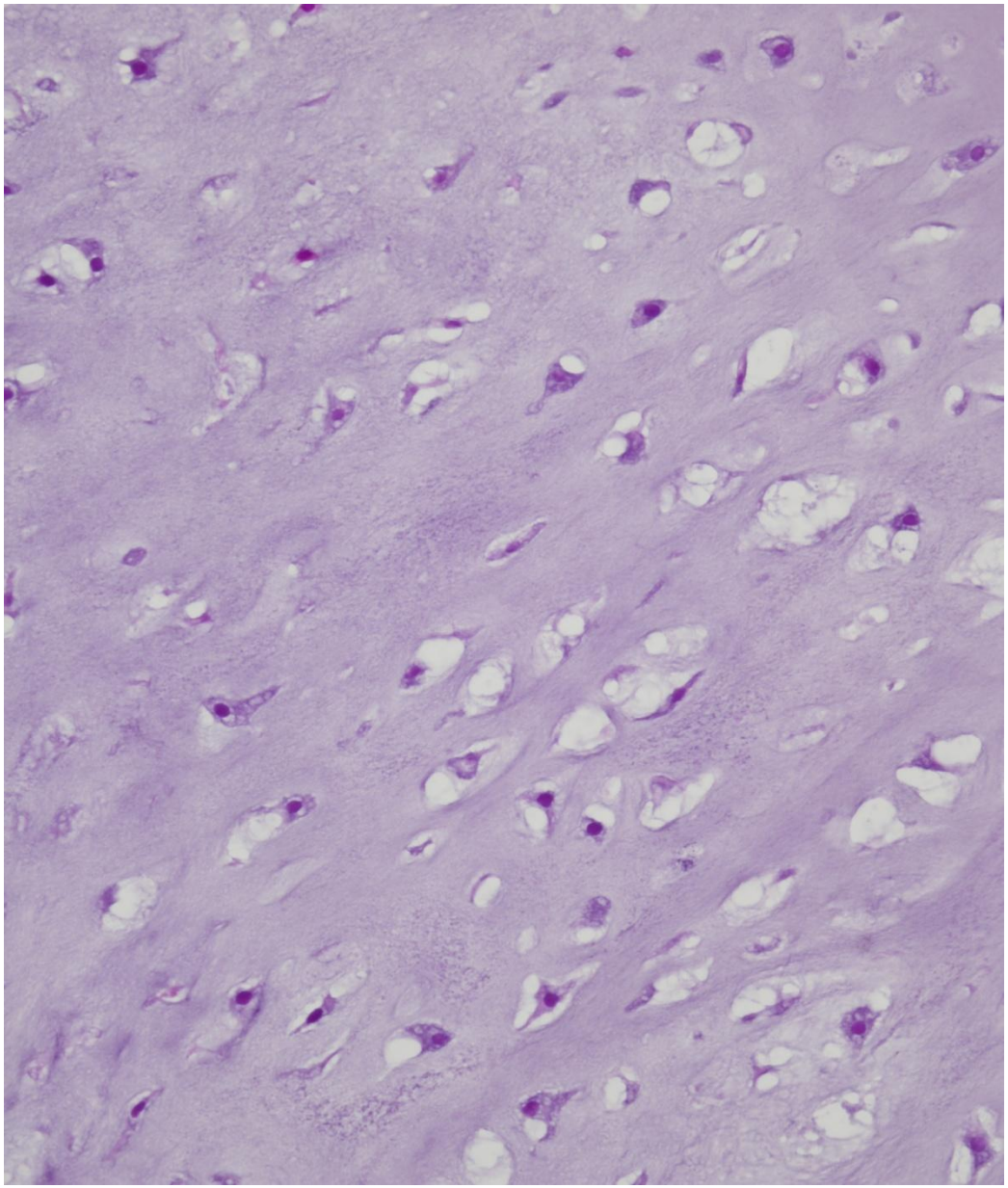


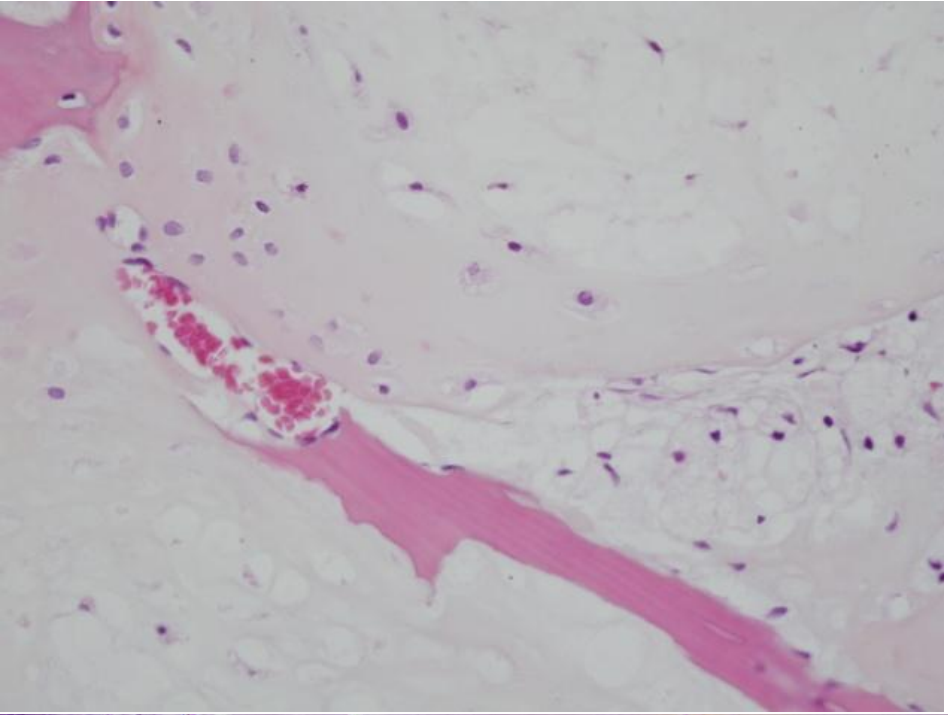


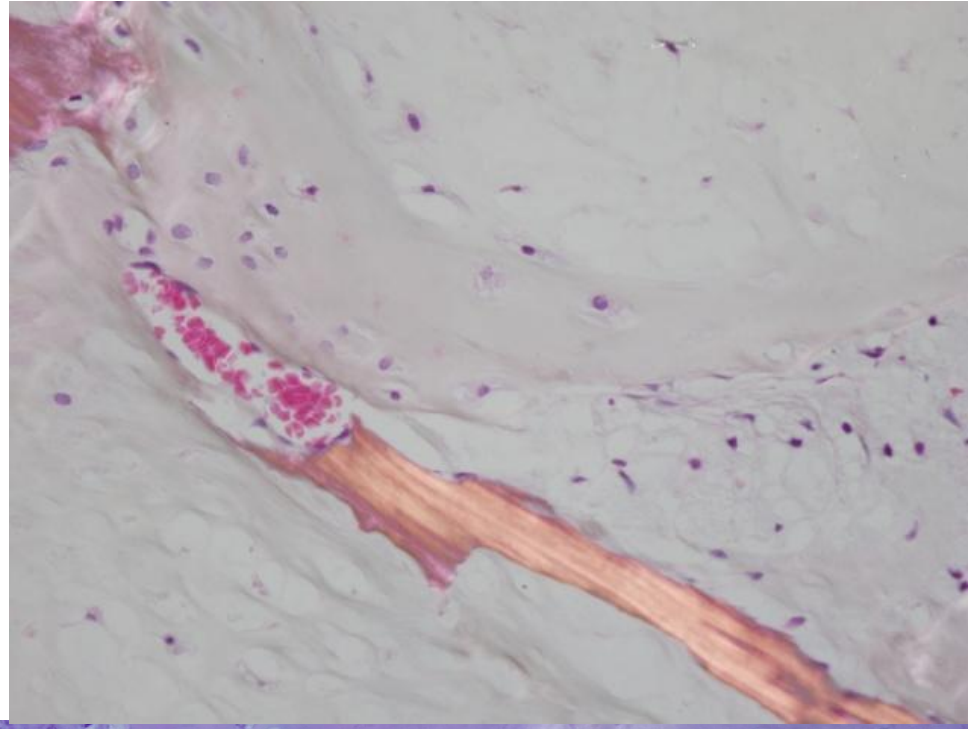

F 12 ans

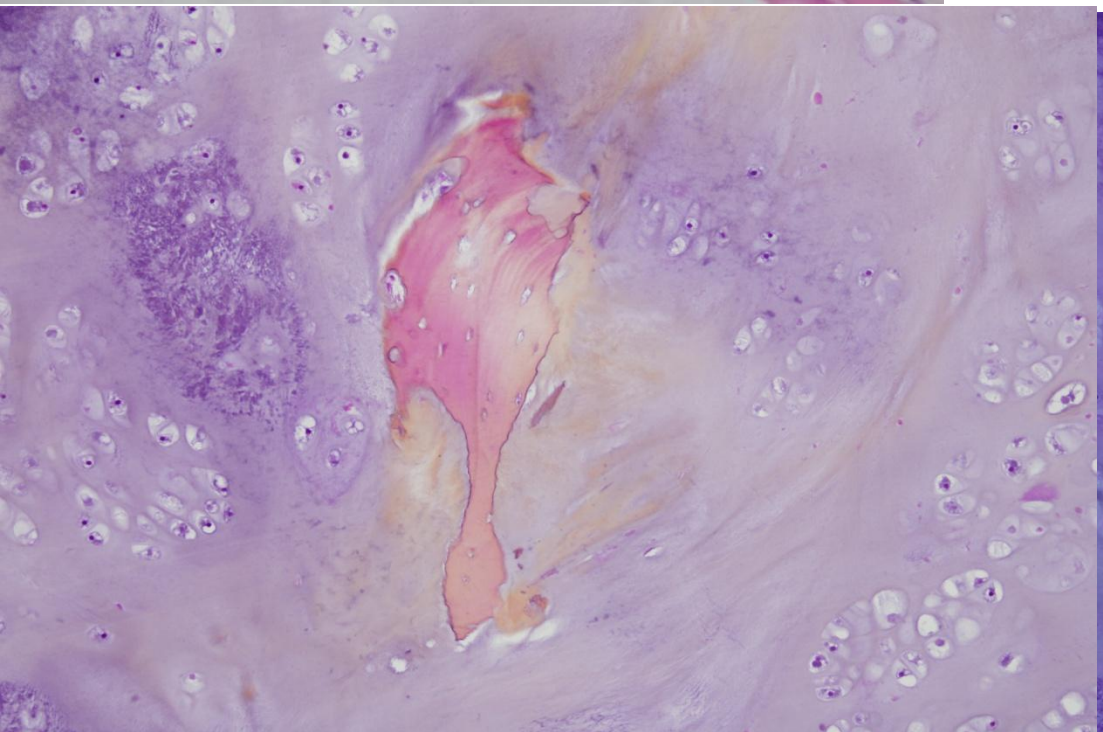


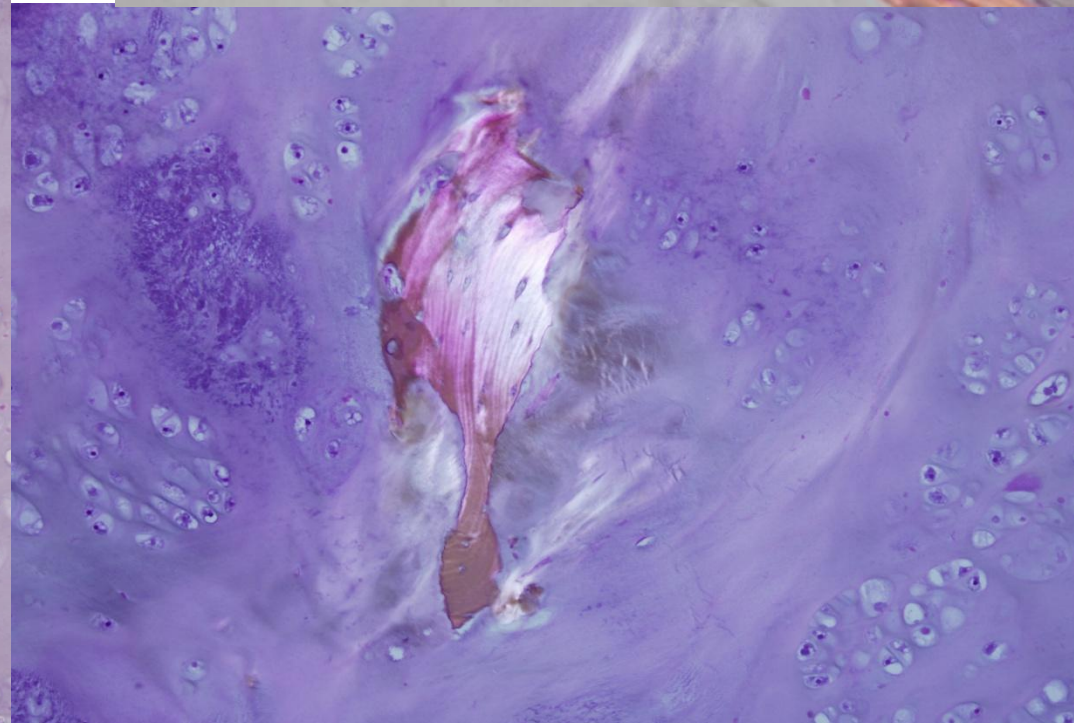




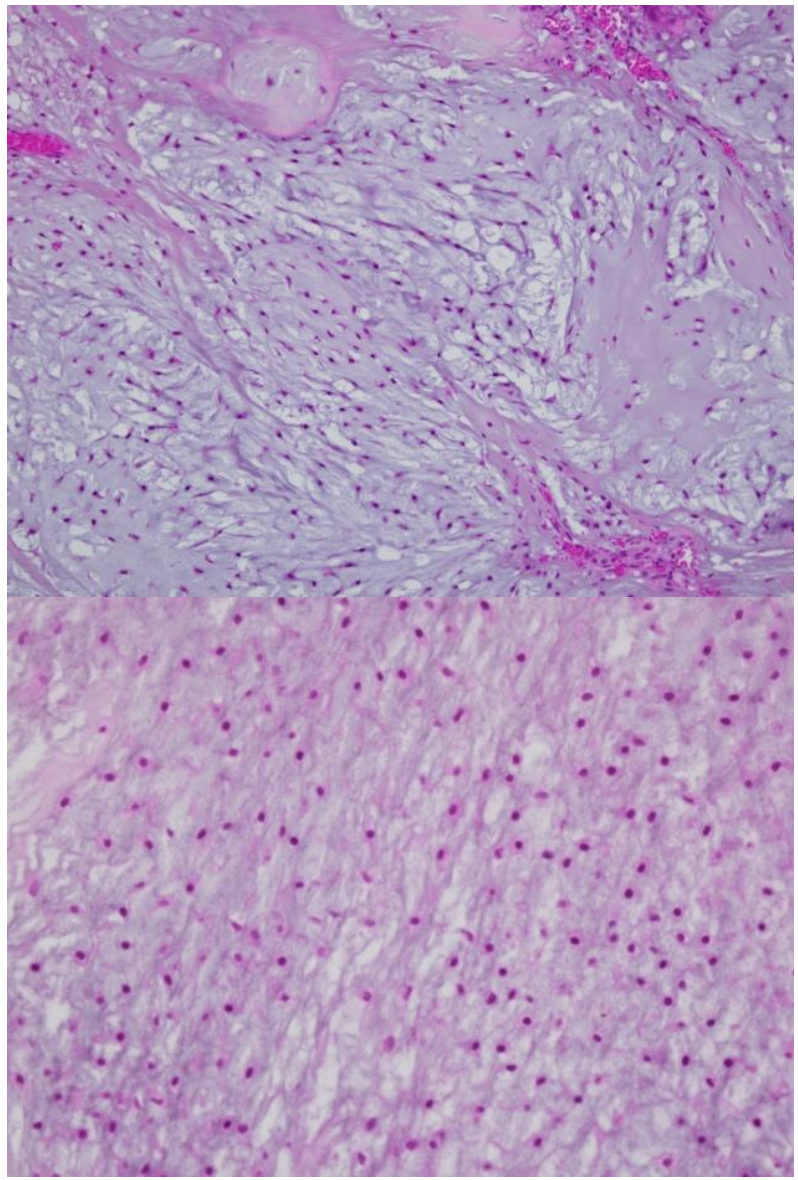


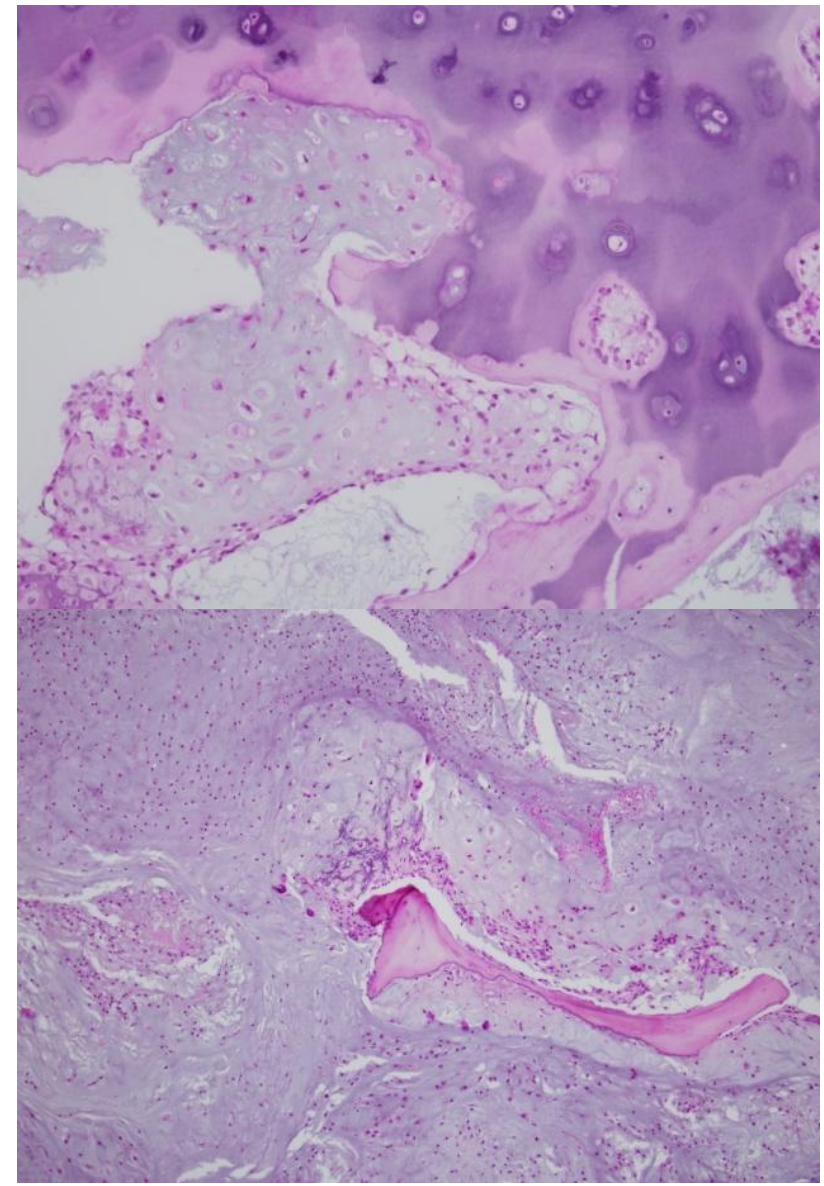




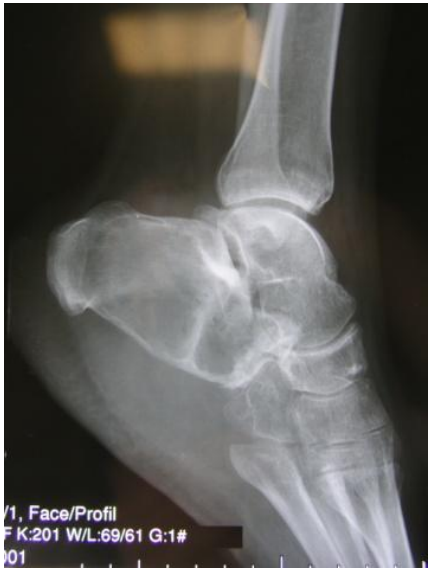


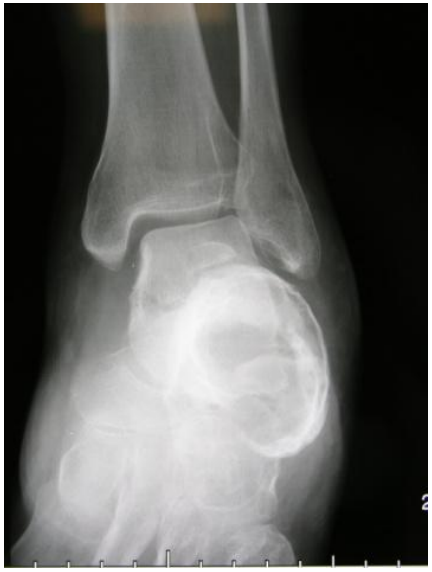
H14 ans Condyle fémoral

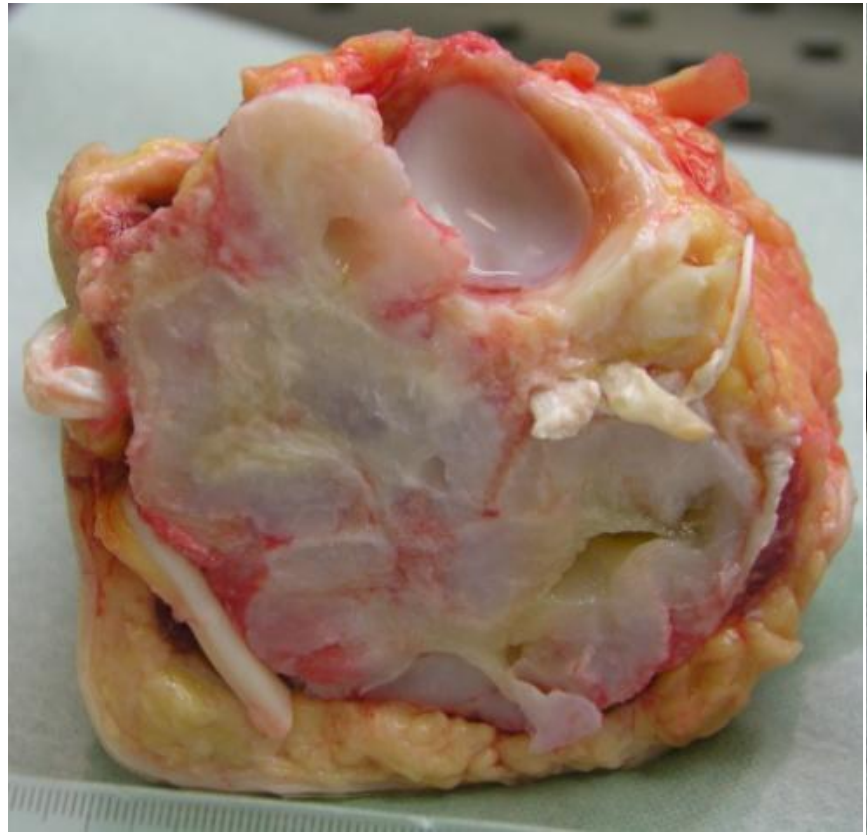


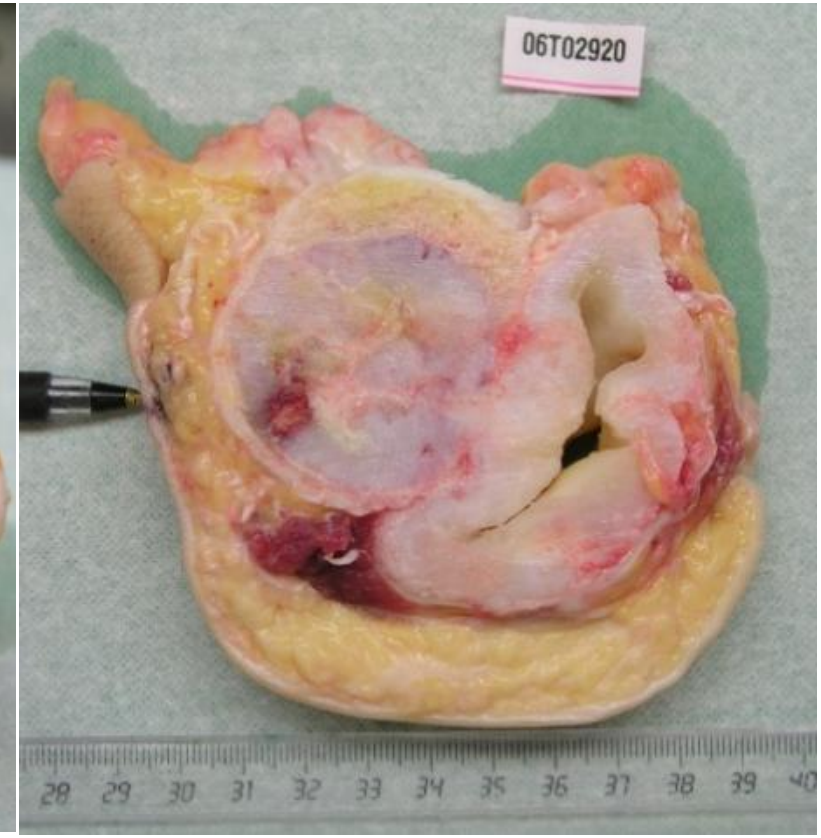


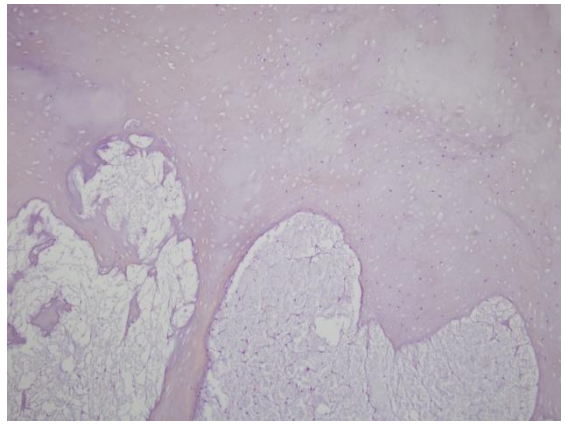
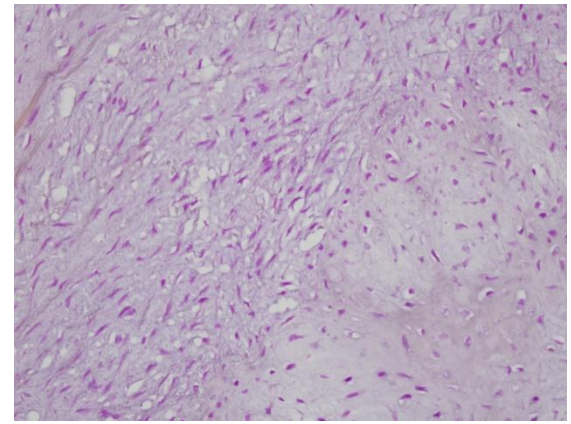
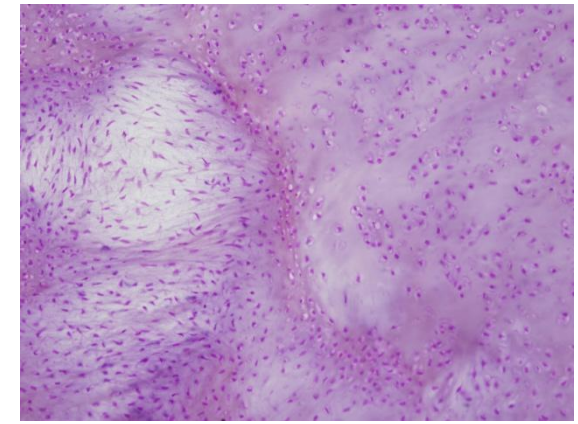
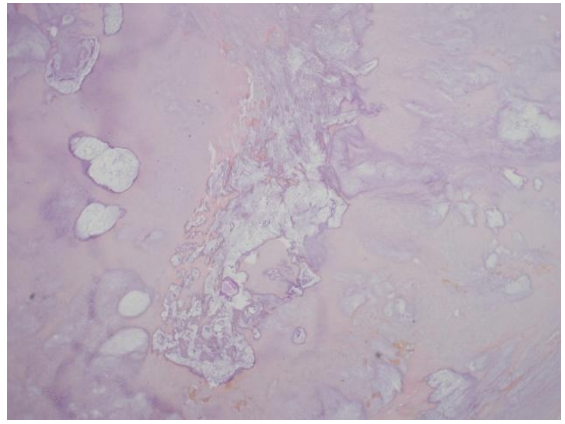
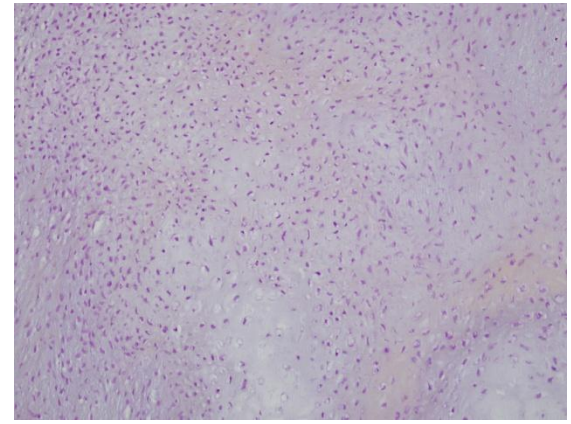
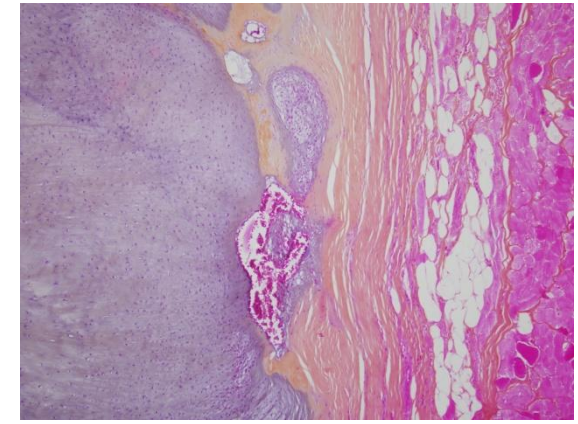
F16 ans Volumineuse tumeur du « talon »



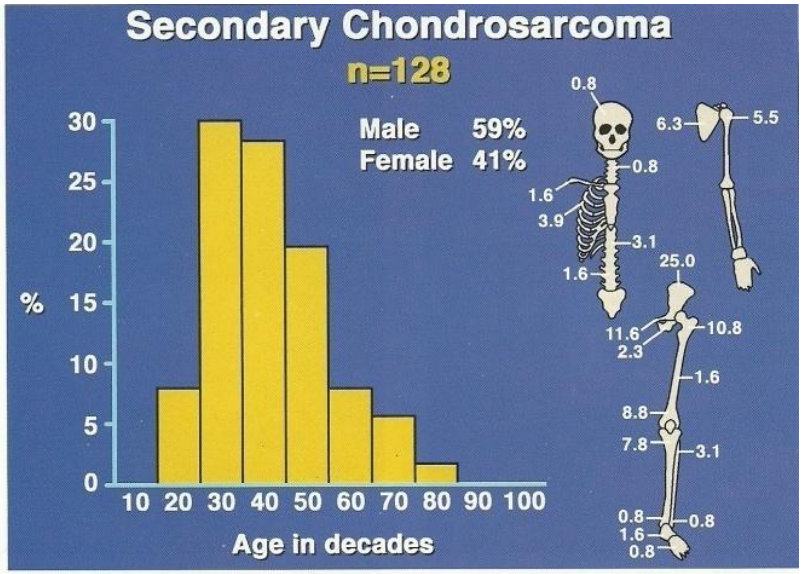






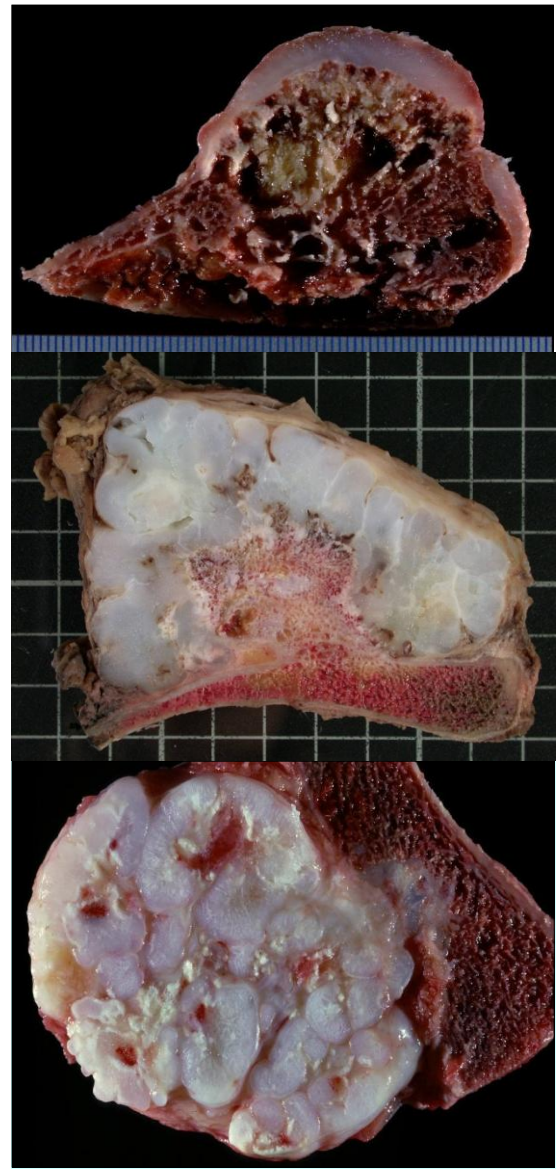
Chondrosarcome conventionnels périphériques (secondaires, développés sur ostéochondrome)



Ostéochondrome

Chondrosarcome
périphérique
de bas grade (1)
(sur ostéochondrome)

Chondrosarcome
périphérique de
haut grade (2-3)

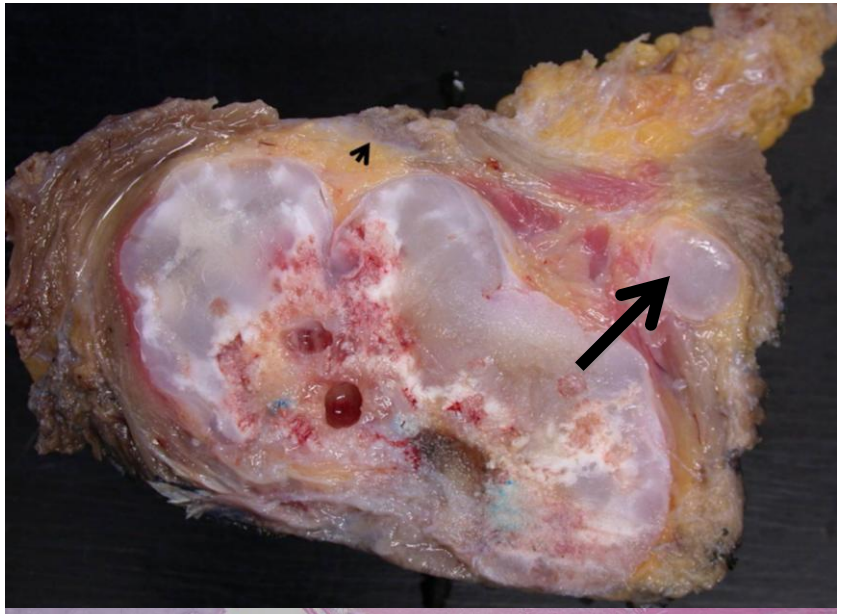


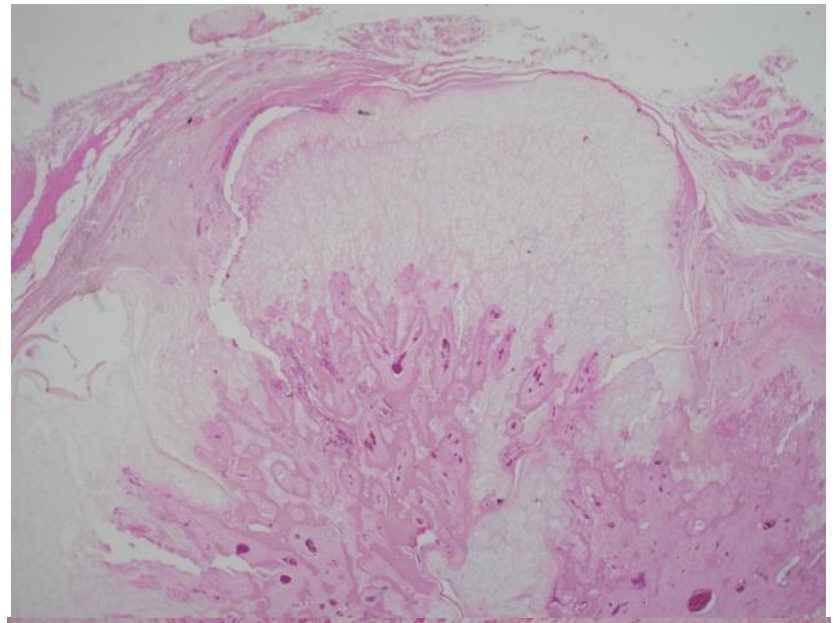
Diagnostic de transformation chondrosarcomateuse d'un ostéochondrome

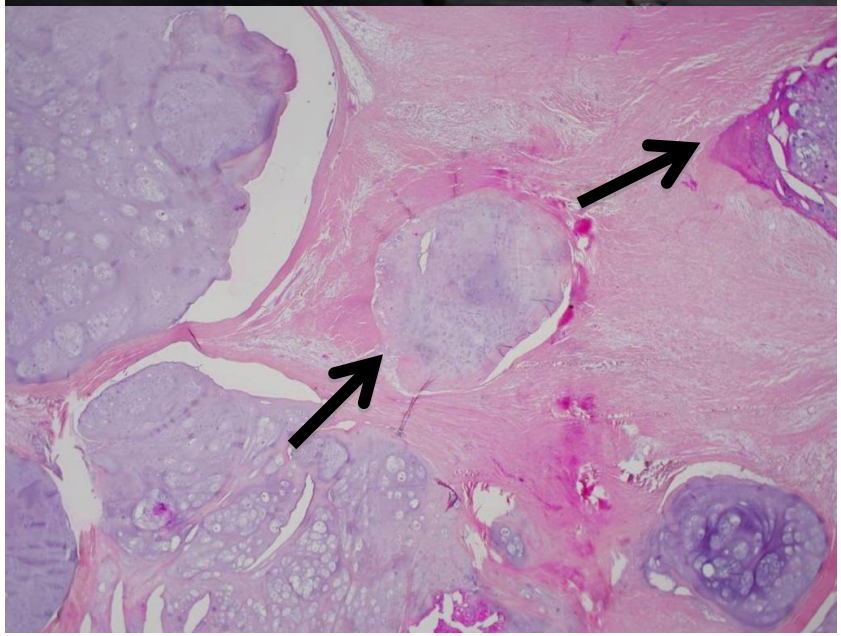
Biopsie inutile (chondrosarcome très bien différencié)
Diagnostic avant tout macroscopique: nécessite l'étude de la pièce d'exérèse.

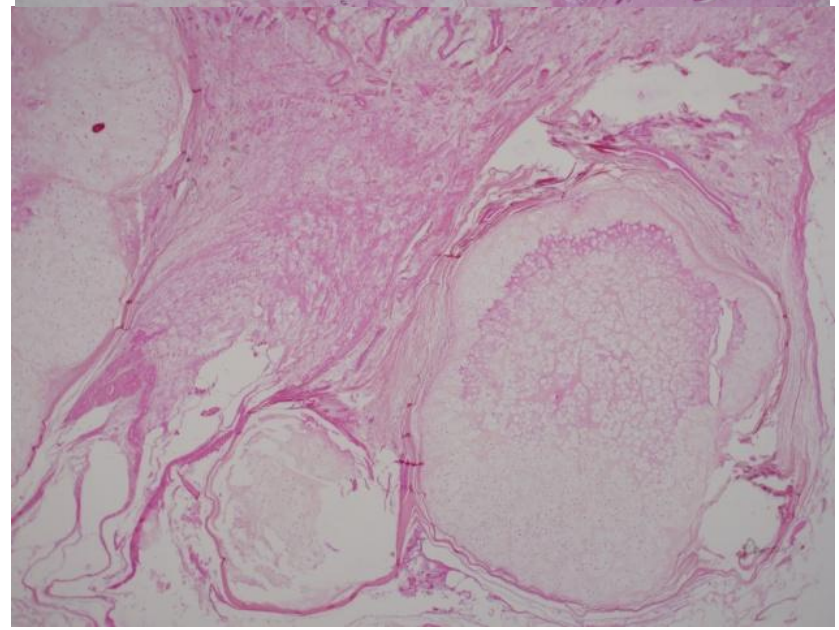
Les deux critères architecturaux permettant ce diagnostic:
- Epaisseur de la coiffe (> 2cm)
- Lobulation de la coiffe +++

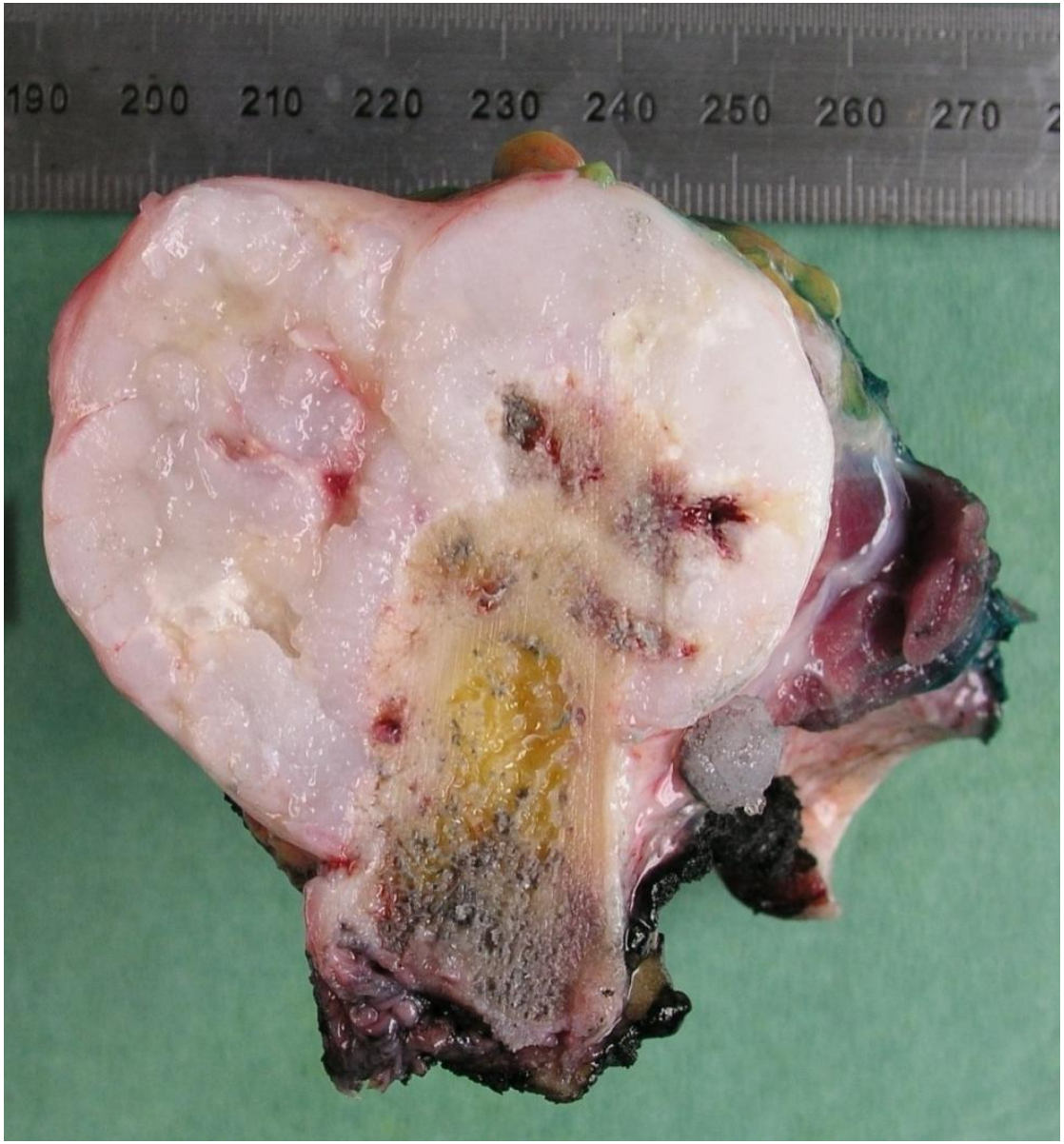
F 16 ans Tumeur costale







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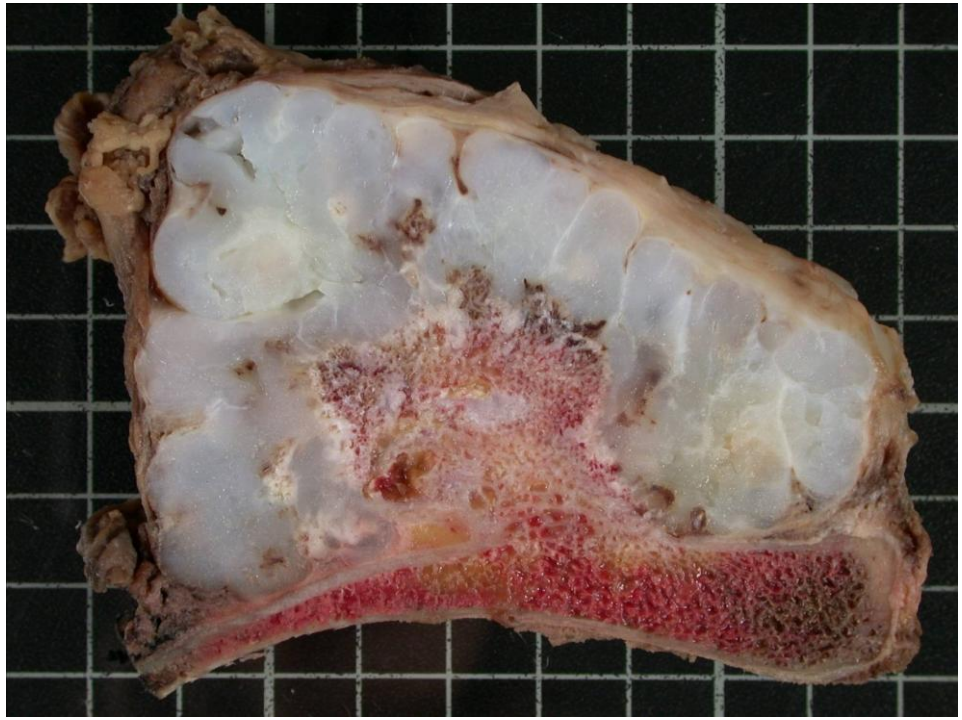
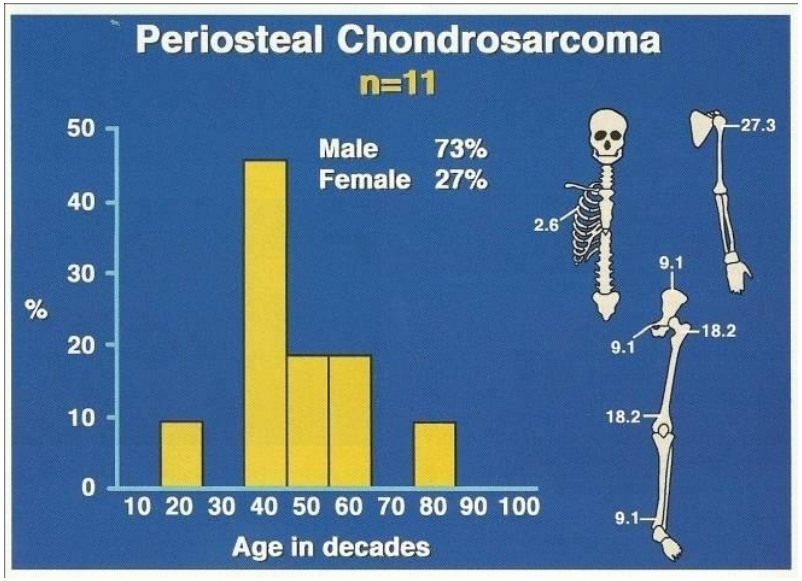
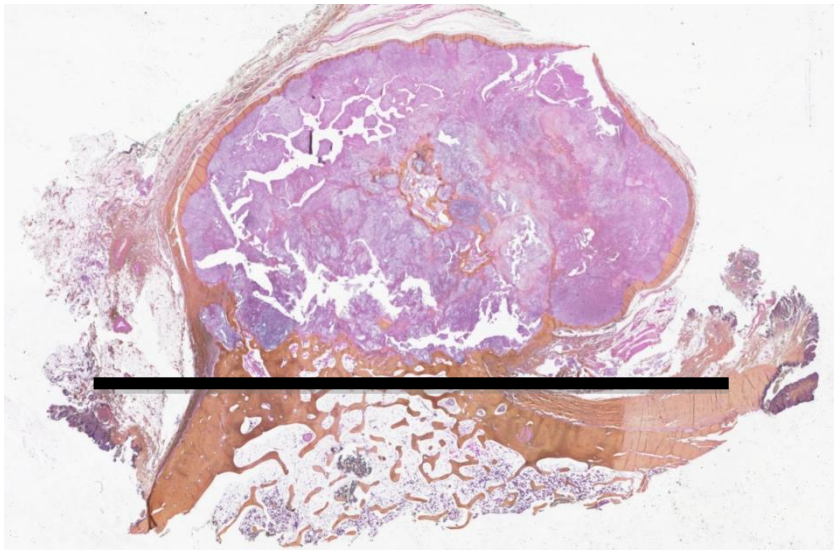
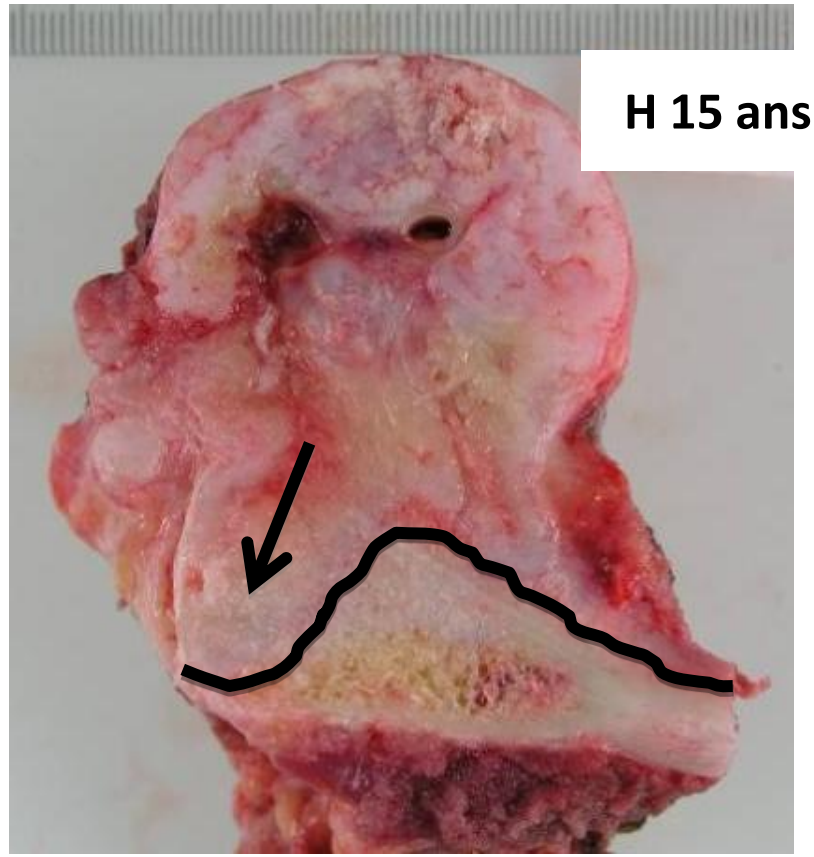


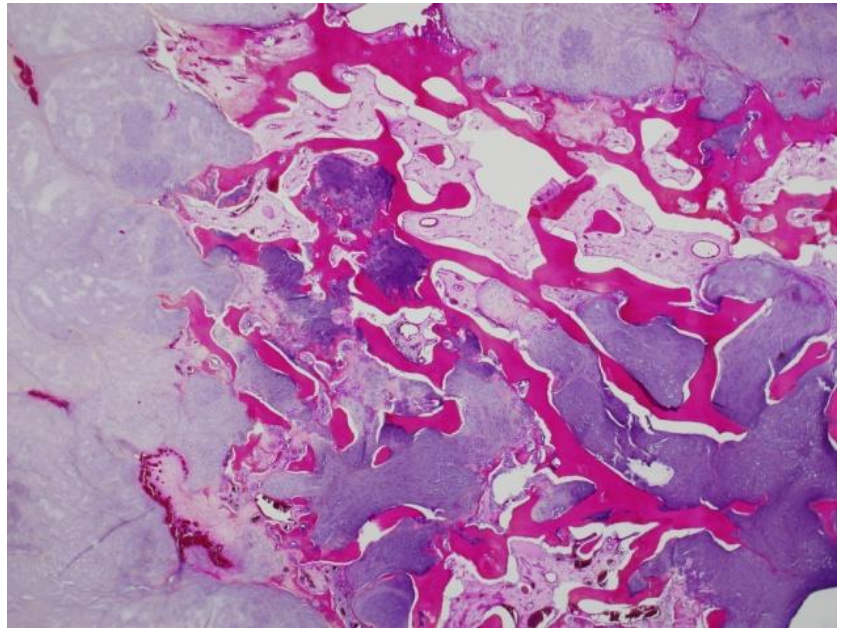


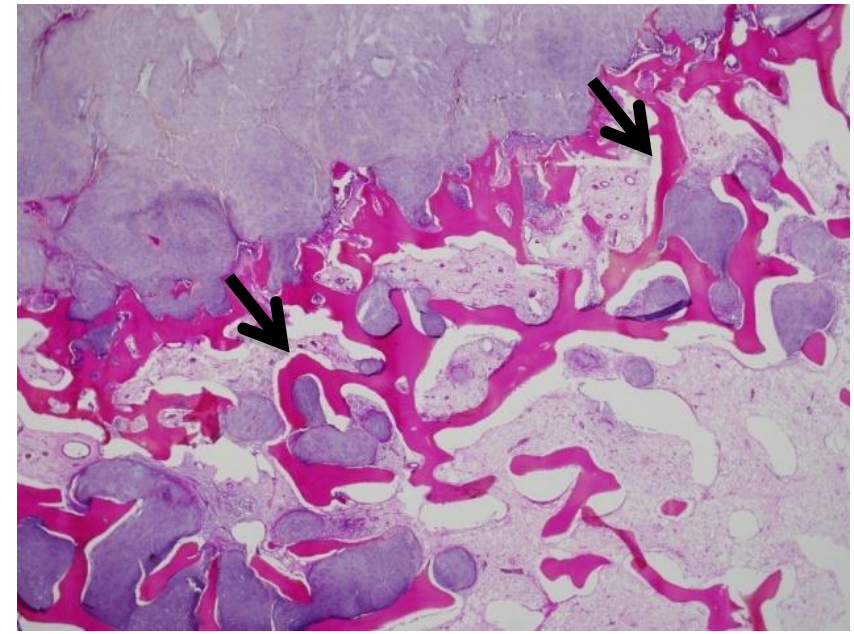
Chondrome périosté
< 5 cm
Pas d'extension endomédullaire

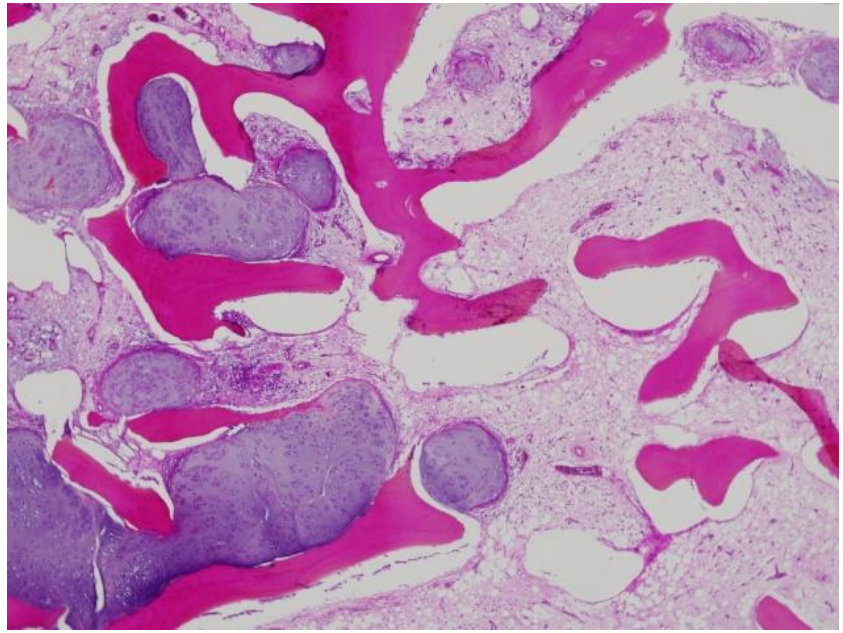


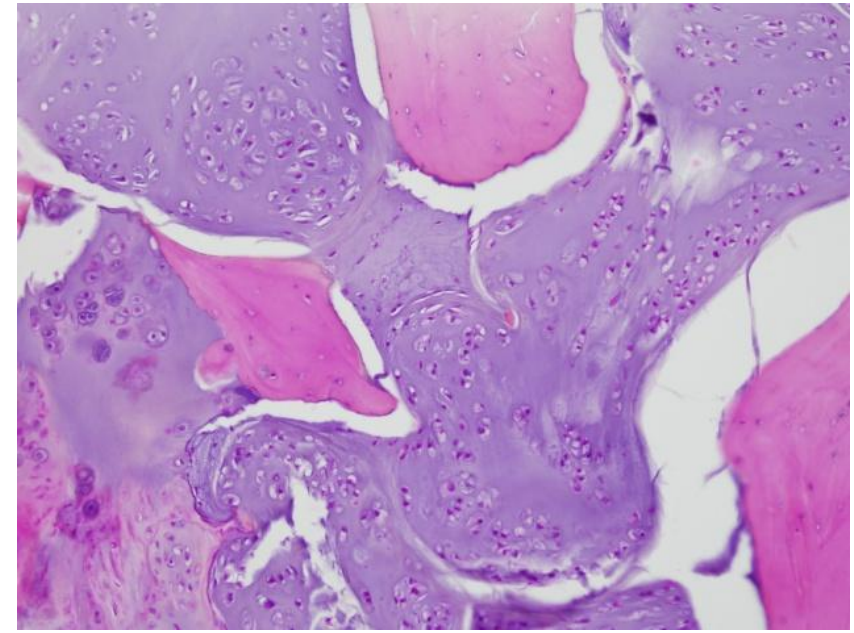
Chondrosarcome périosté
>5 cm
Extension endomédullaire

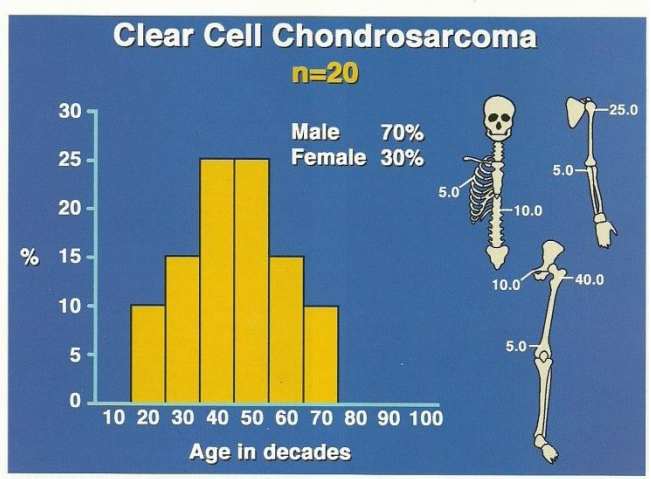












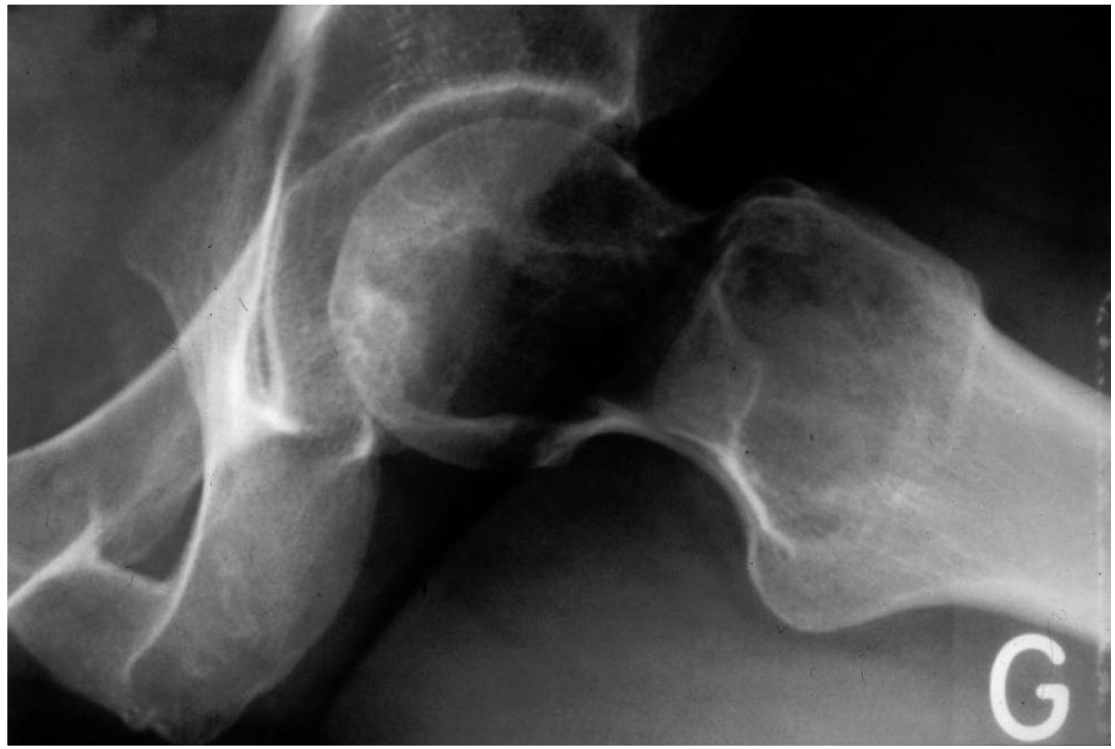
CHONDROSARCOME A CELLULES CLAIRES

2% des CS

25 – 50 ans

Epiphyses

Tête humérale 2/3
Tête fémorale



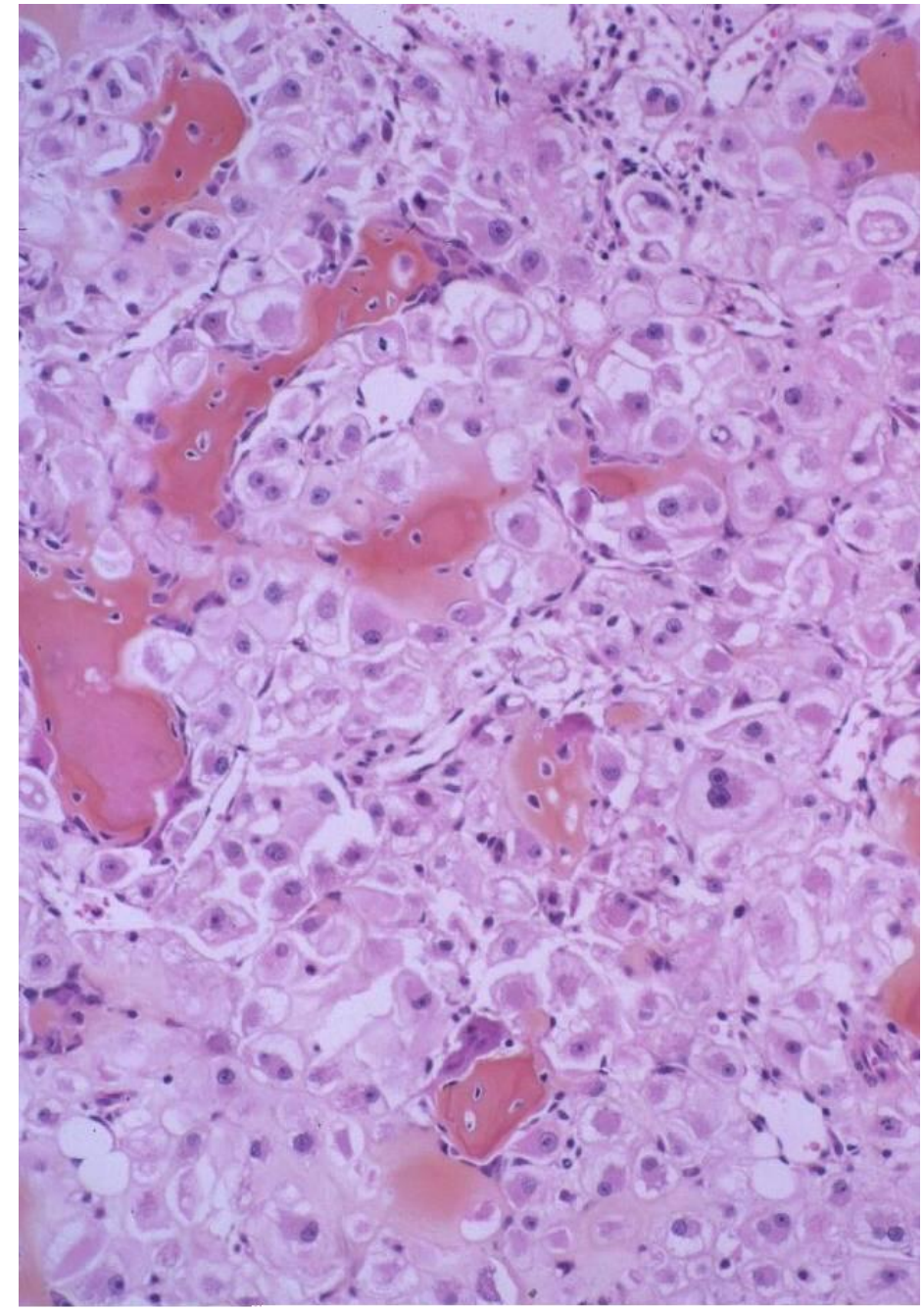
Cellules claires

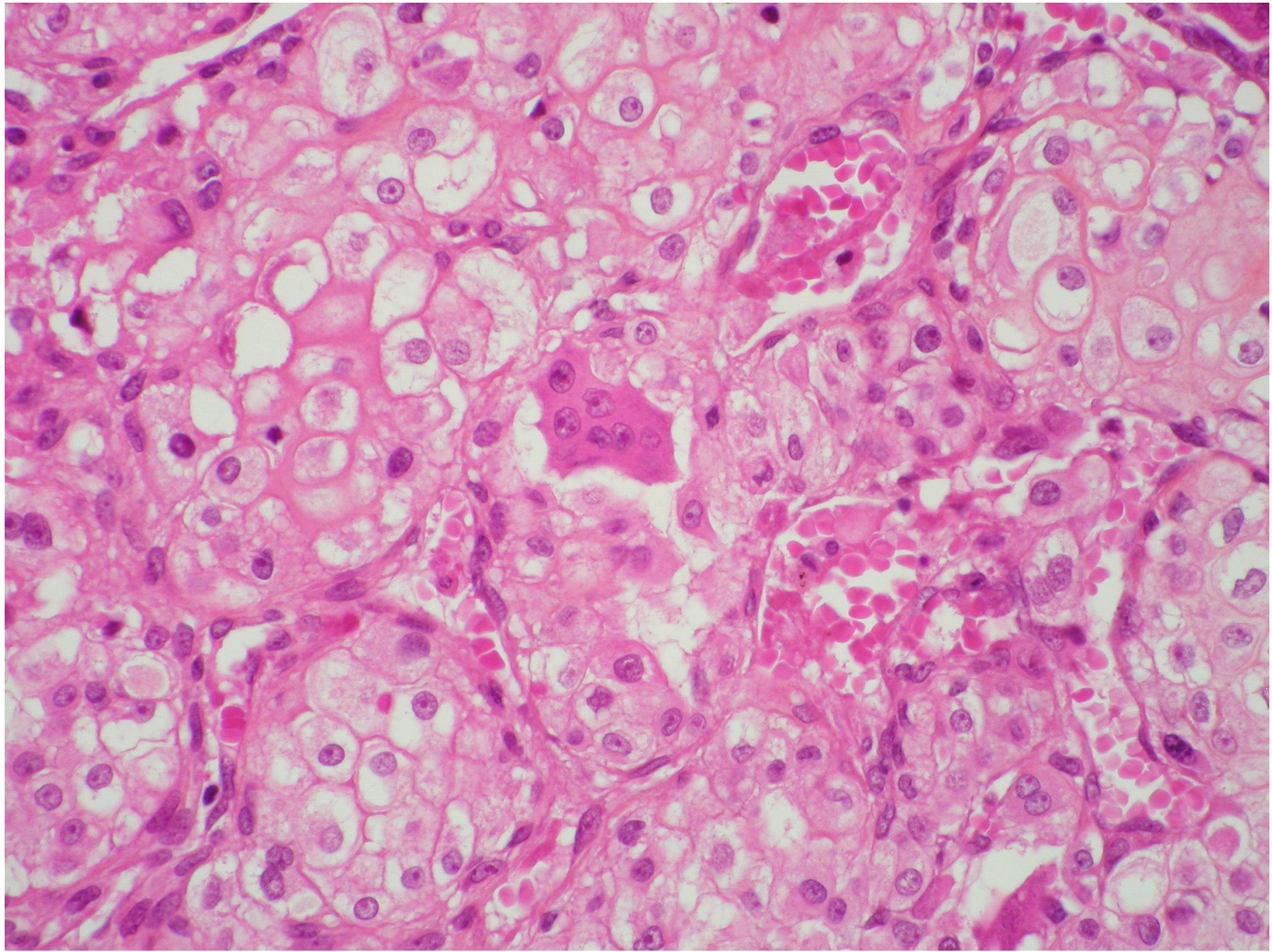
Ostéogenèse d'allure réactionnelle +++

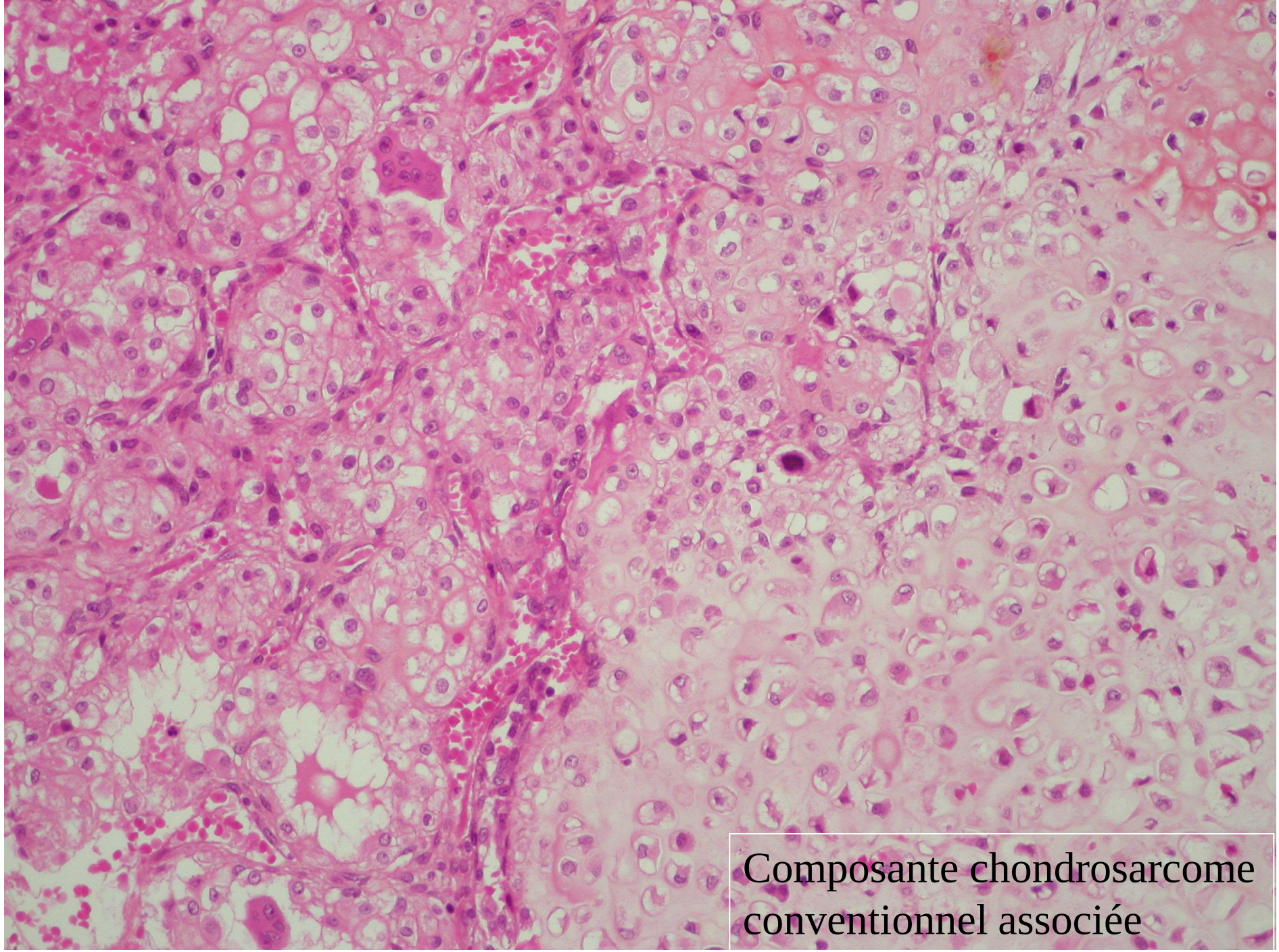
Matrice cartilagineuse très réduite

Stroma vasculaire

Cellules géantes de type ostéoclastique







Composante chondrosarcome conventionnel associée

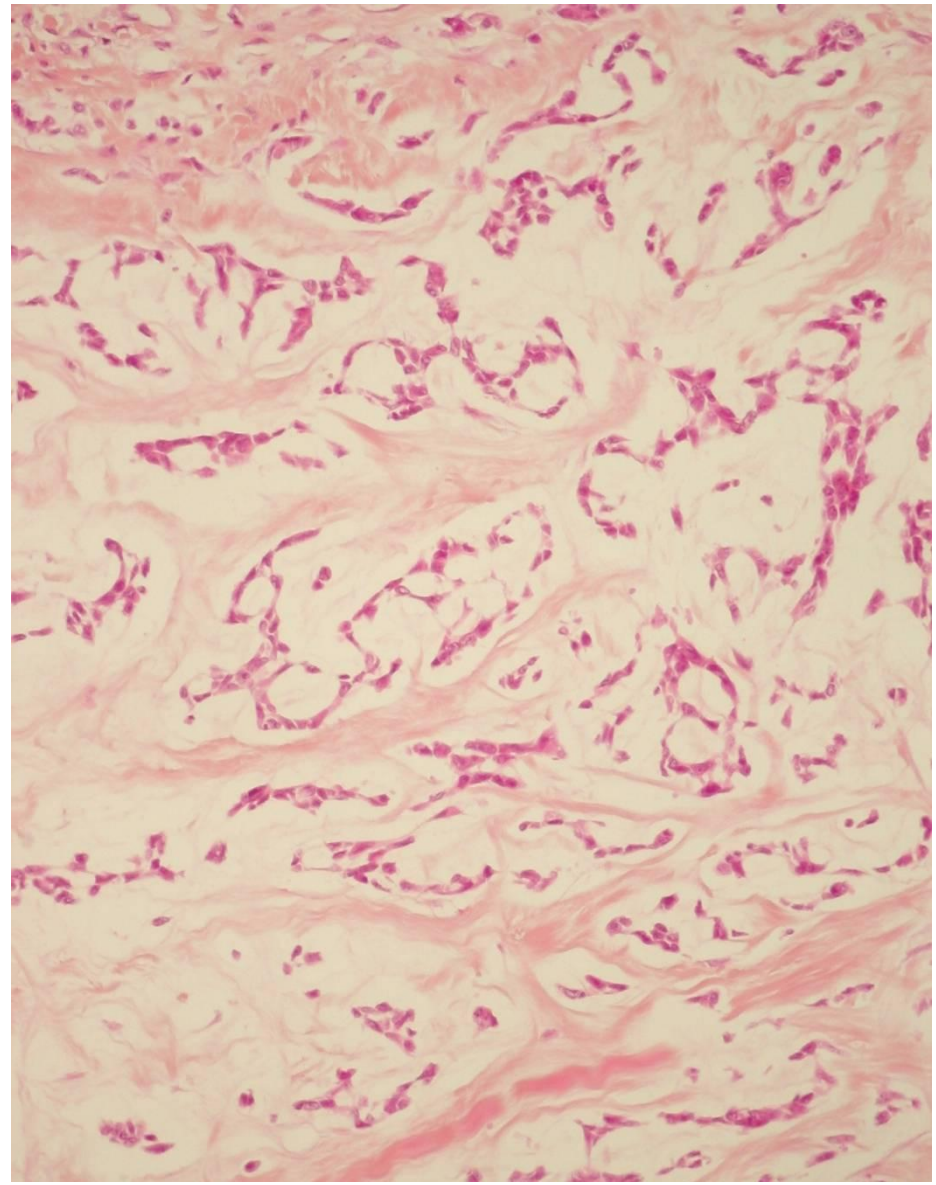
Chondrosarcome myxoïde Extra-squelettique

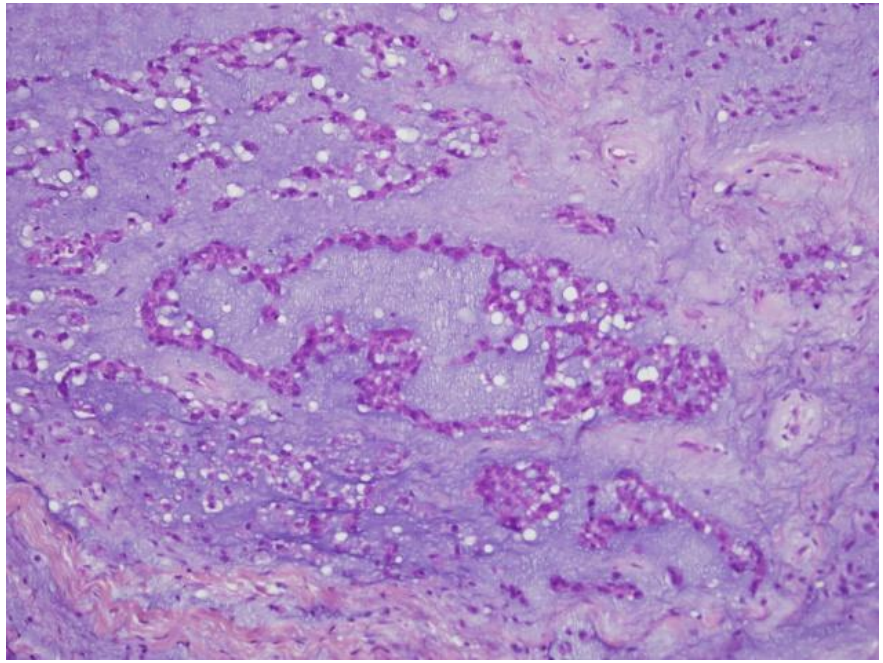
Tumeur de l'adulte
Rares cas décrits chez l'enfant et l'adolescent

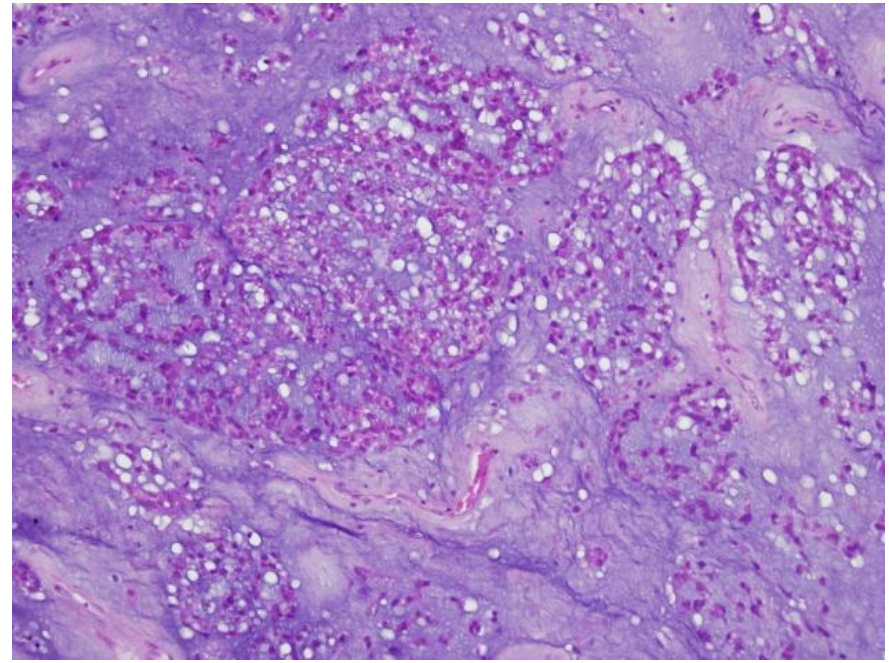
Tissus mous profonds
(cuisse +++)

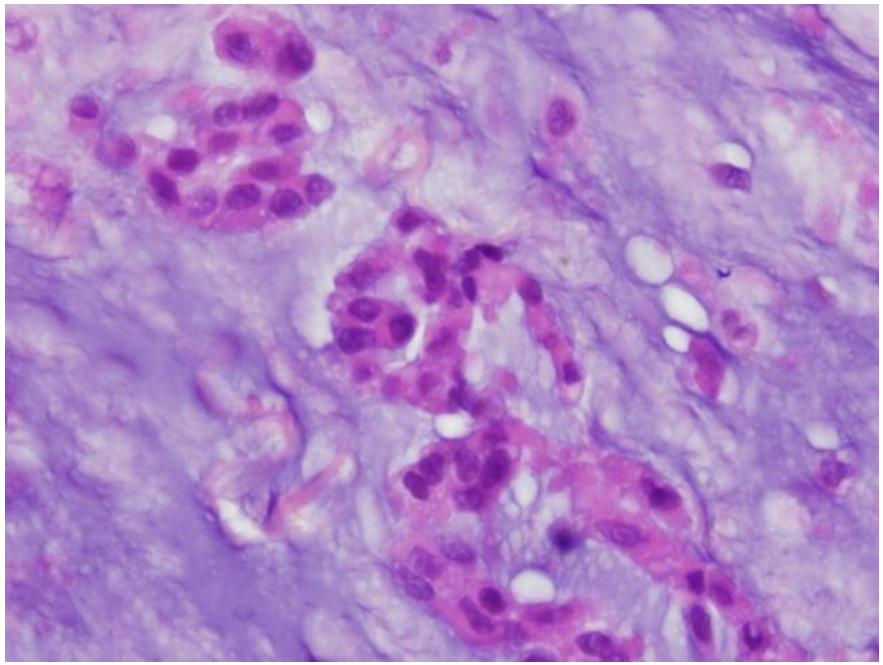
Translocation spécifiques
t(9;22) (q22;q12/EWS) 50% des cas
T(9:17)(q22;q11)

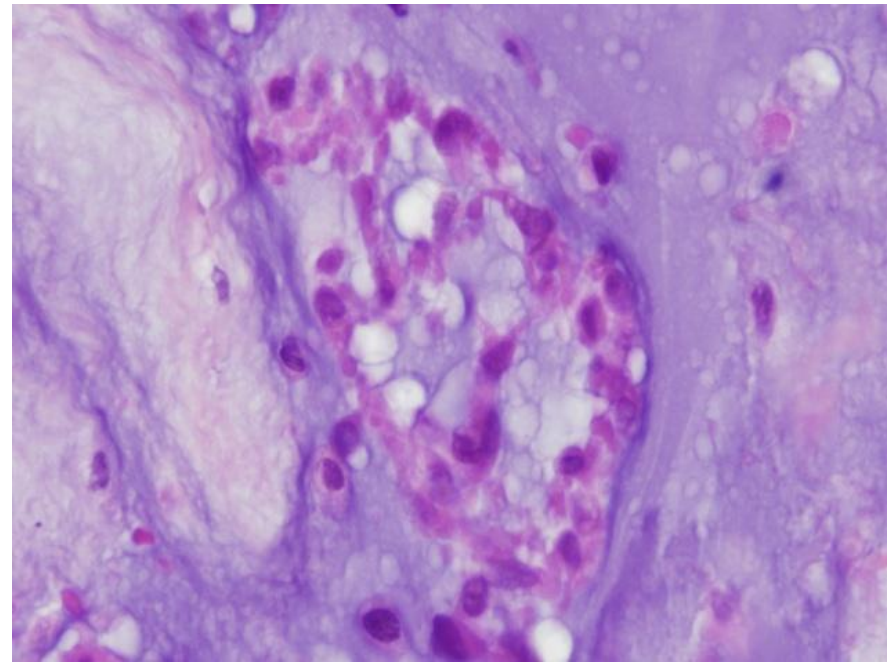
Origine tumorale non déterminée



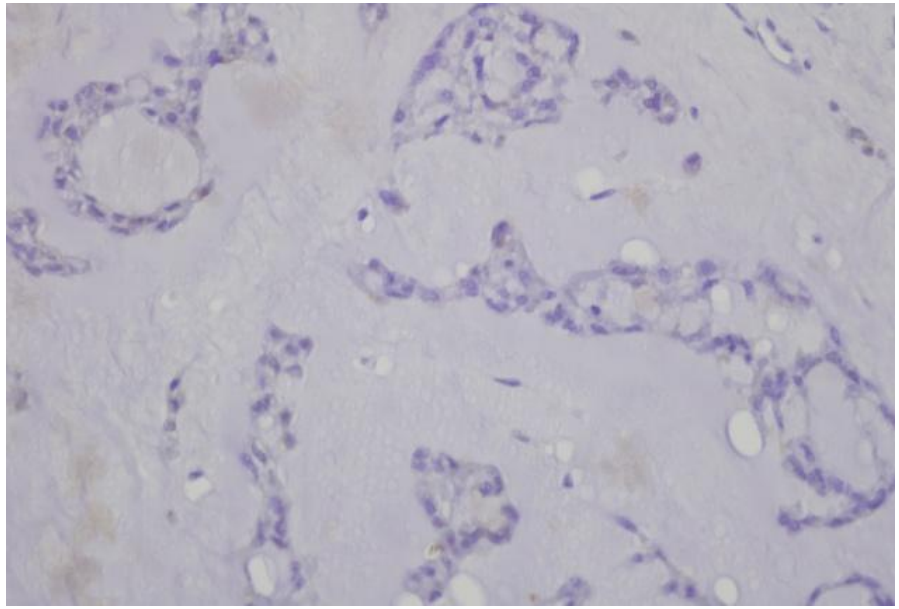


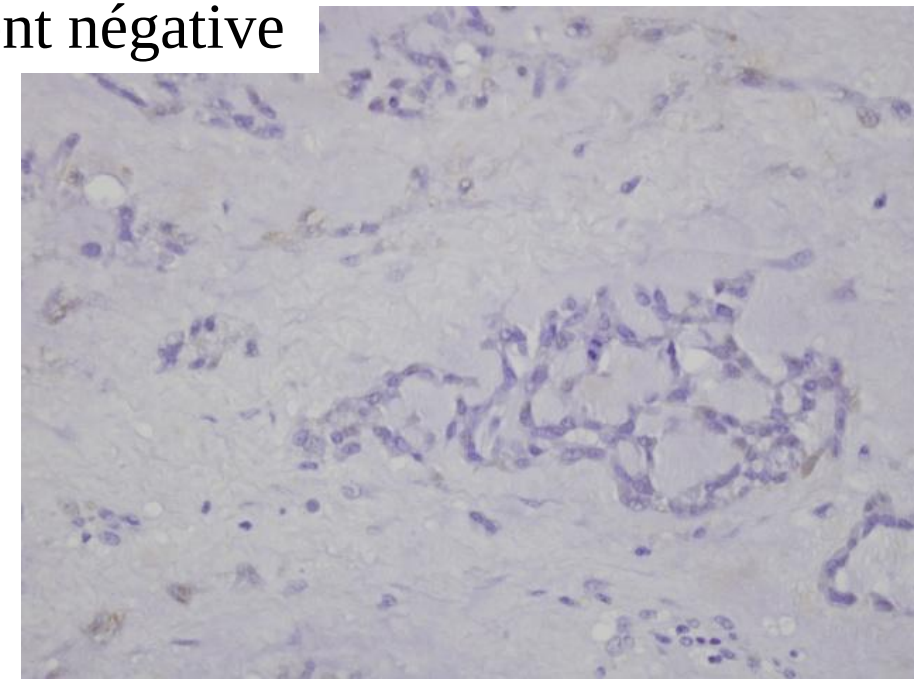


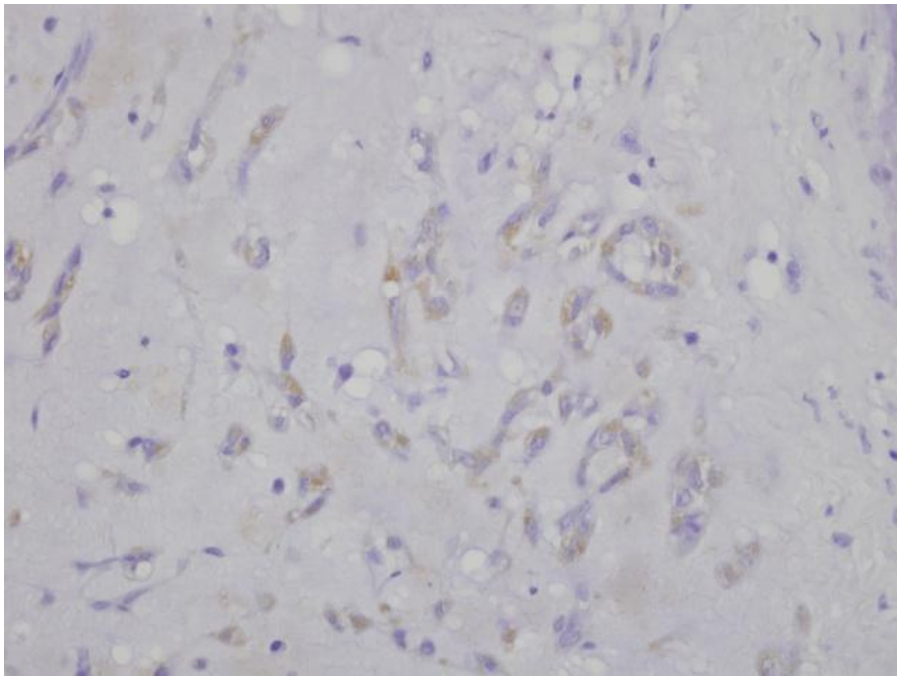


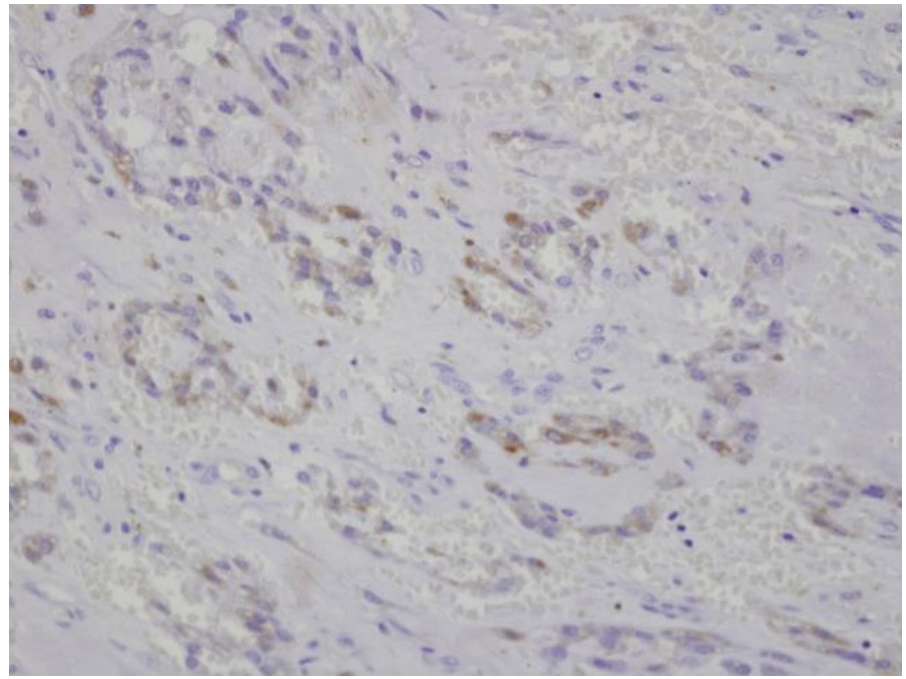


Immunohistochimie: PS100 le plus souvent négative

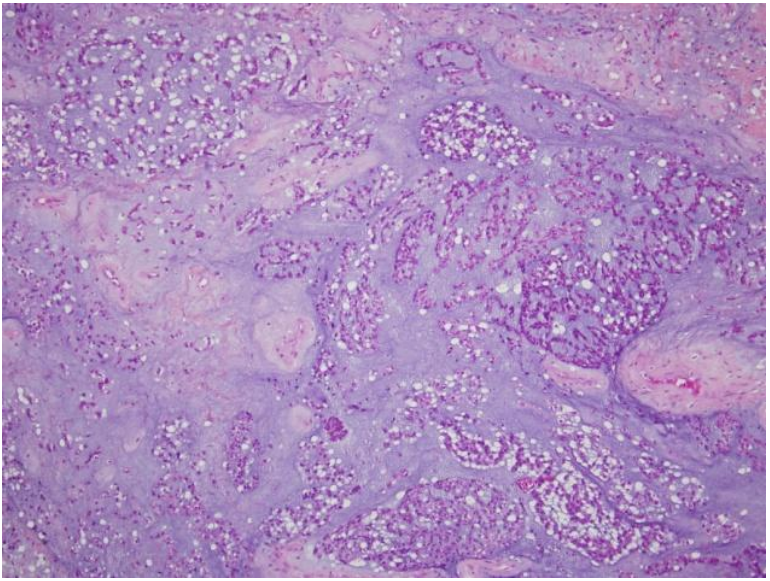


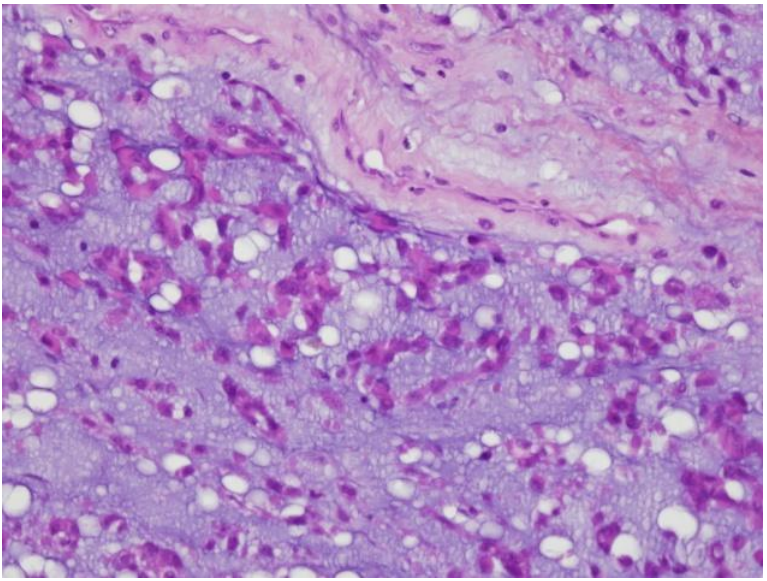






Chondrosarcome myxoïde extra-squelettique





Chondrosarcome conventionnel osseux avec remaniements myxoïdes