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Medicyt, s.r.o.

Bioptické a cytologické laboratórium Trenčín

# Prípad SD-IAP č. 661



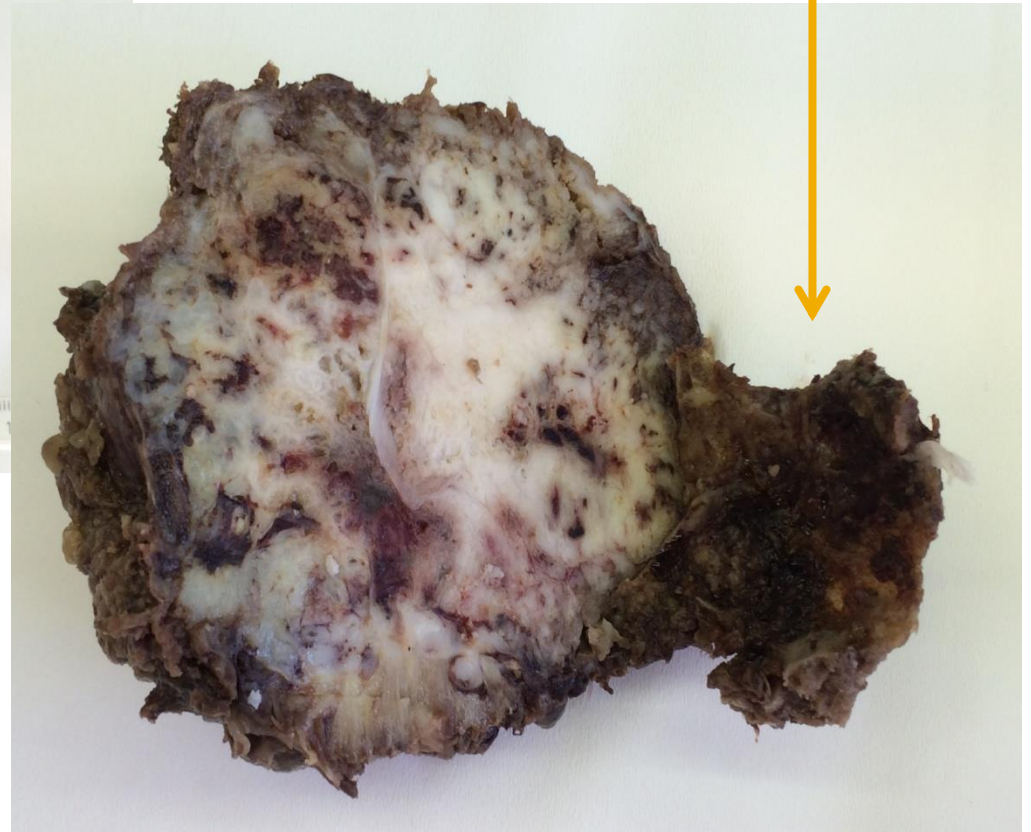
# Klinické údaje

- 71-ročný muž
- nádor v malej panve
- presakrálny, s resorpciou krížovej kosti
- pomalý expanzívny rast
- 12 cm extirpát nádoru s resekciou krížovej kosti

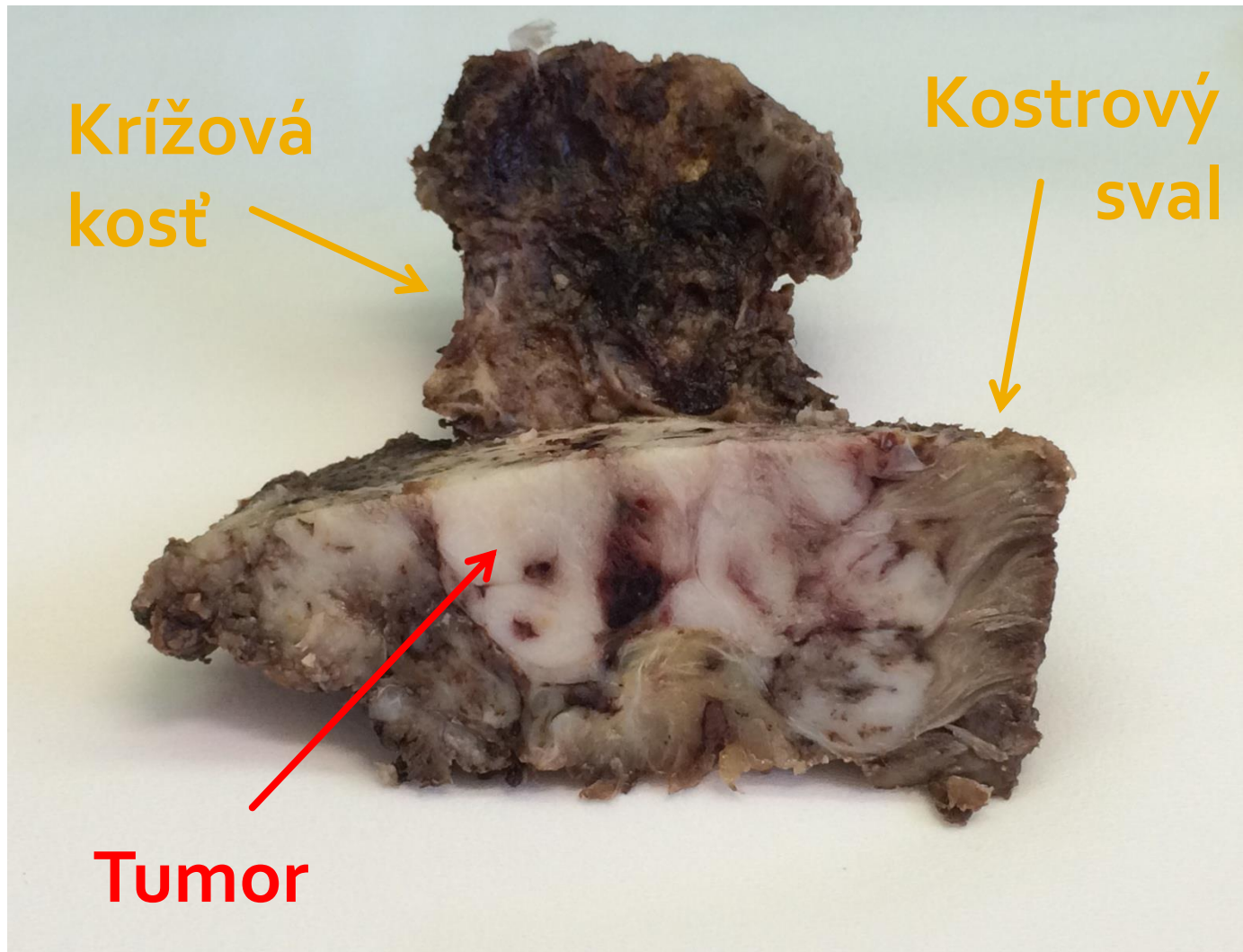
# Operačný materiál



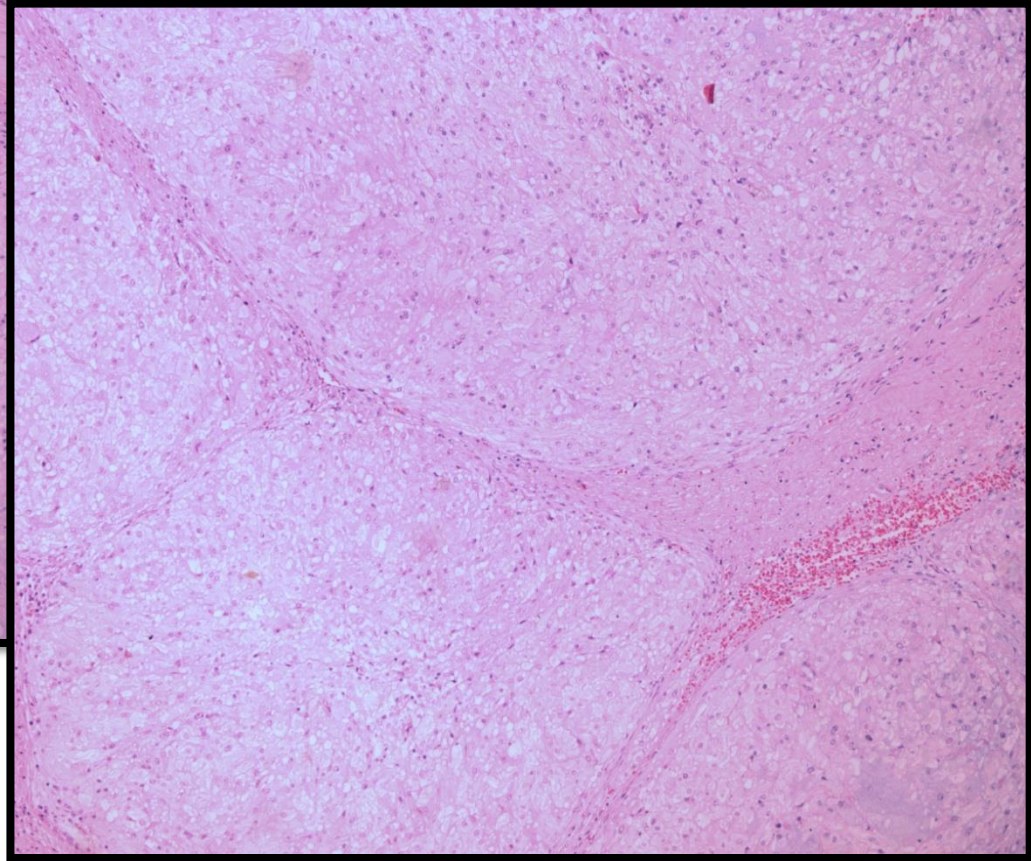
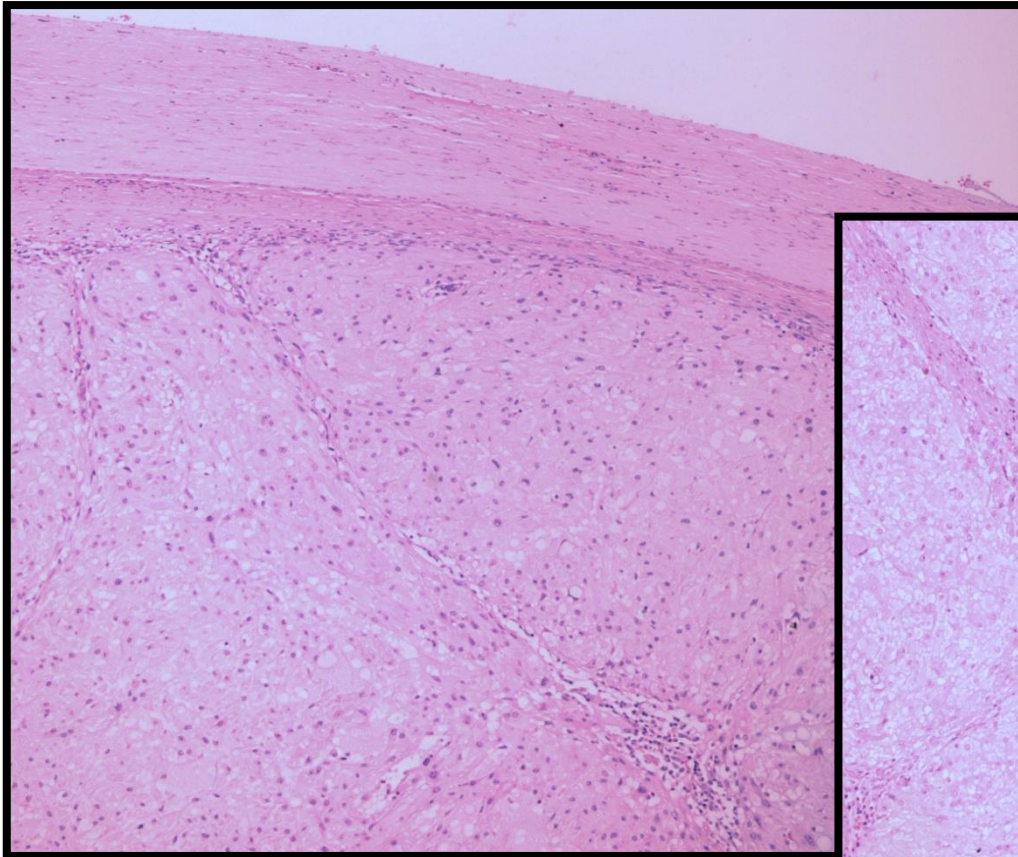
Krížová kosť



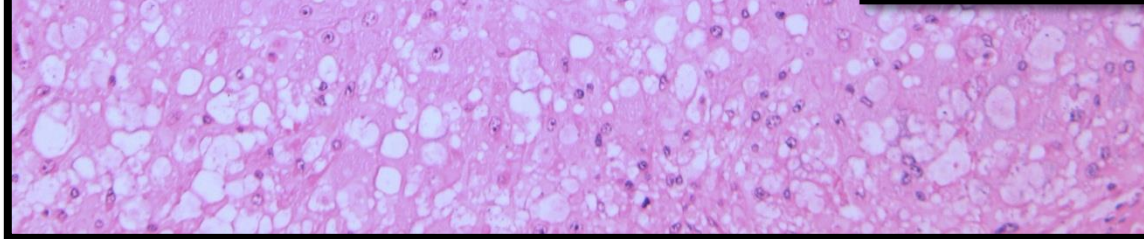
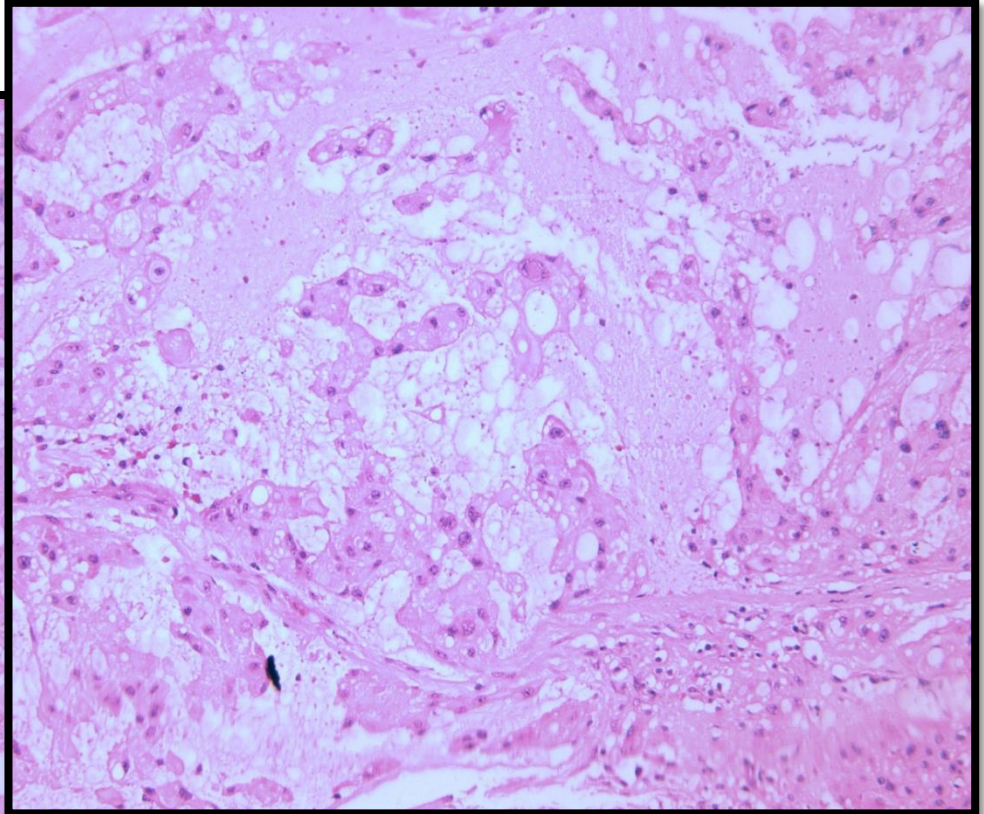
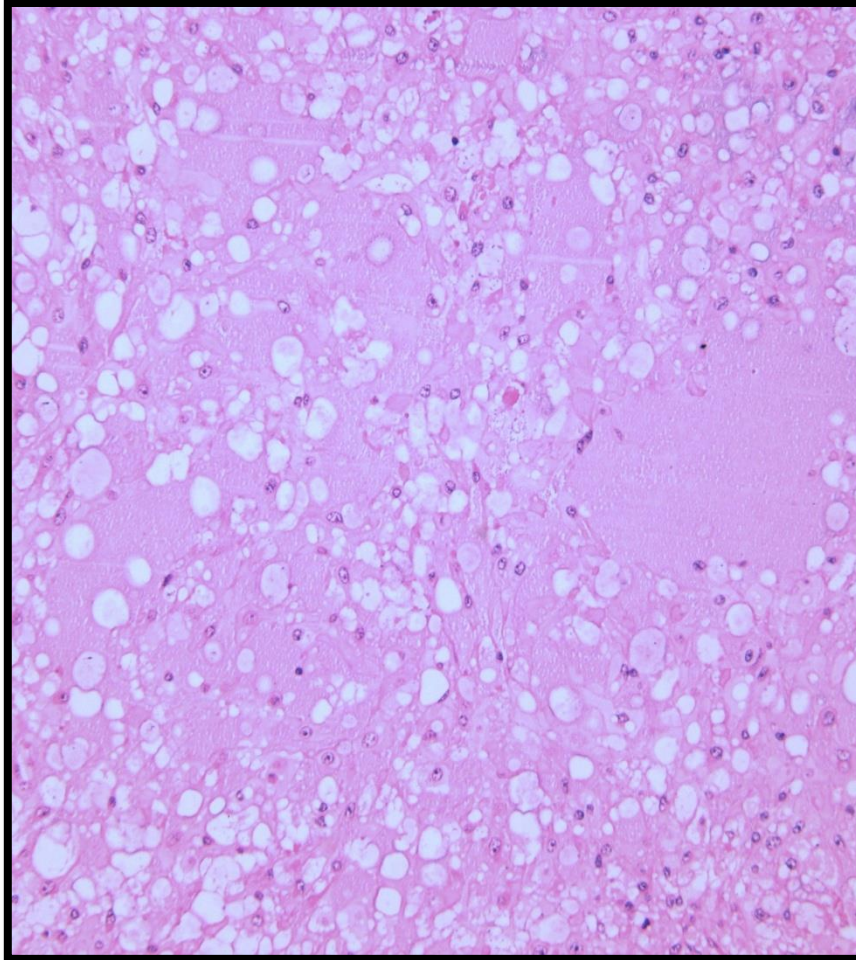
# Operačný materiál



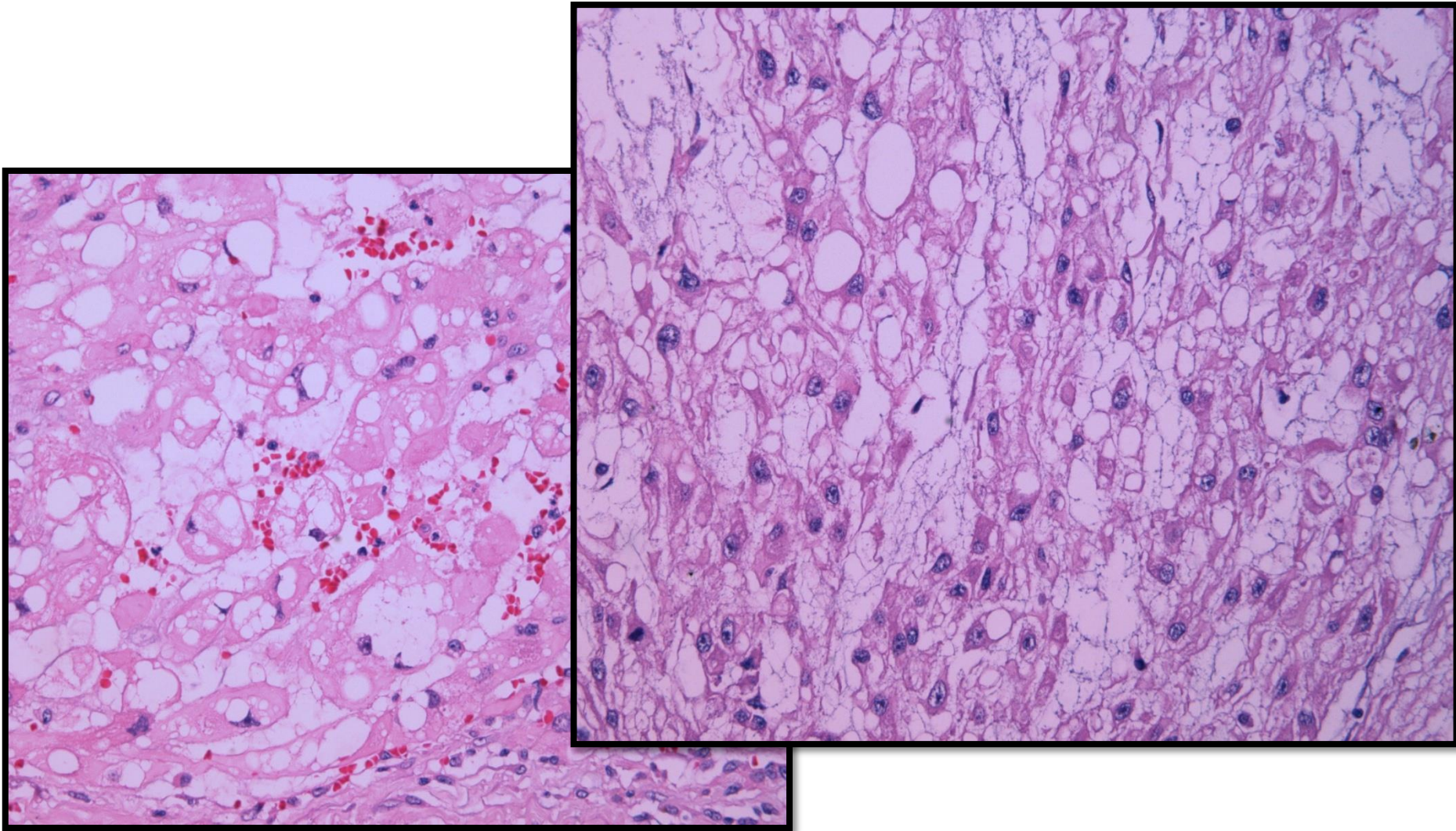
# Lobulárna architektónika



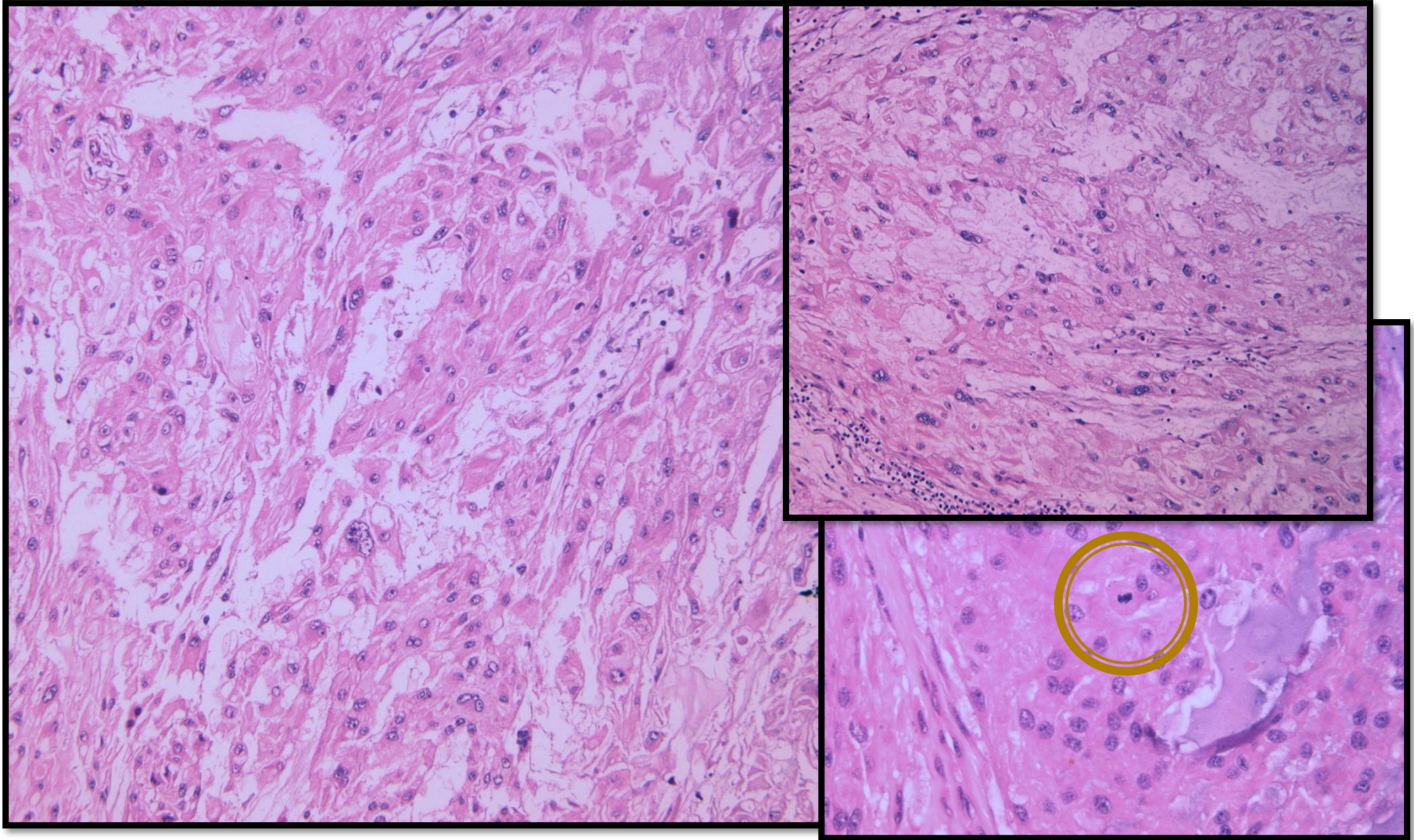
# Epiteloidné bb + mukoidná matrix



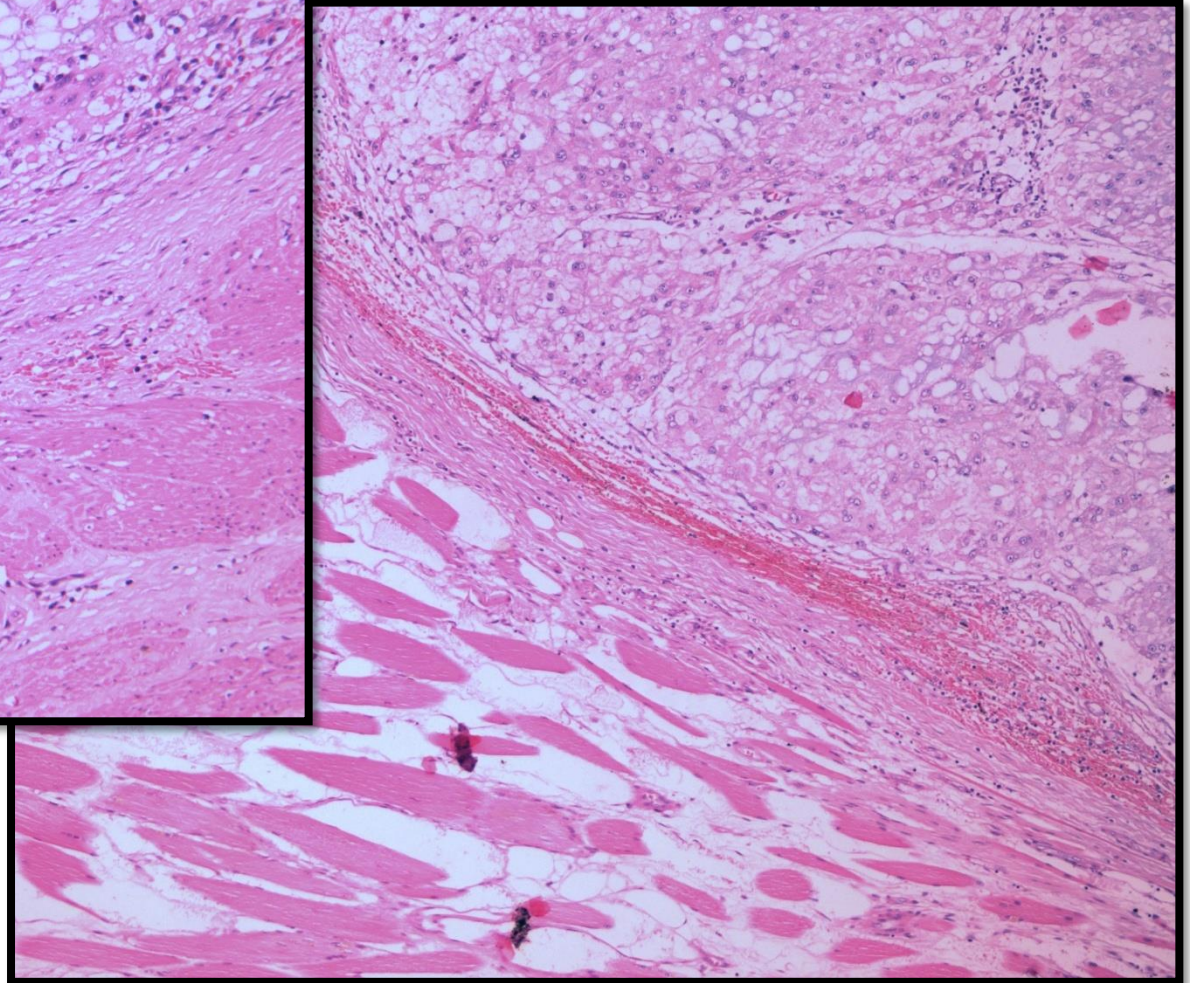
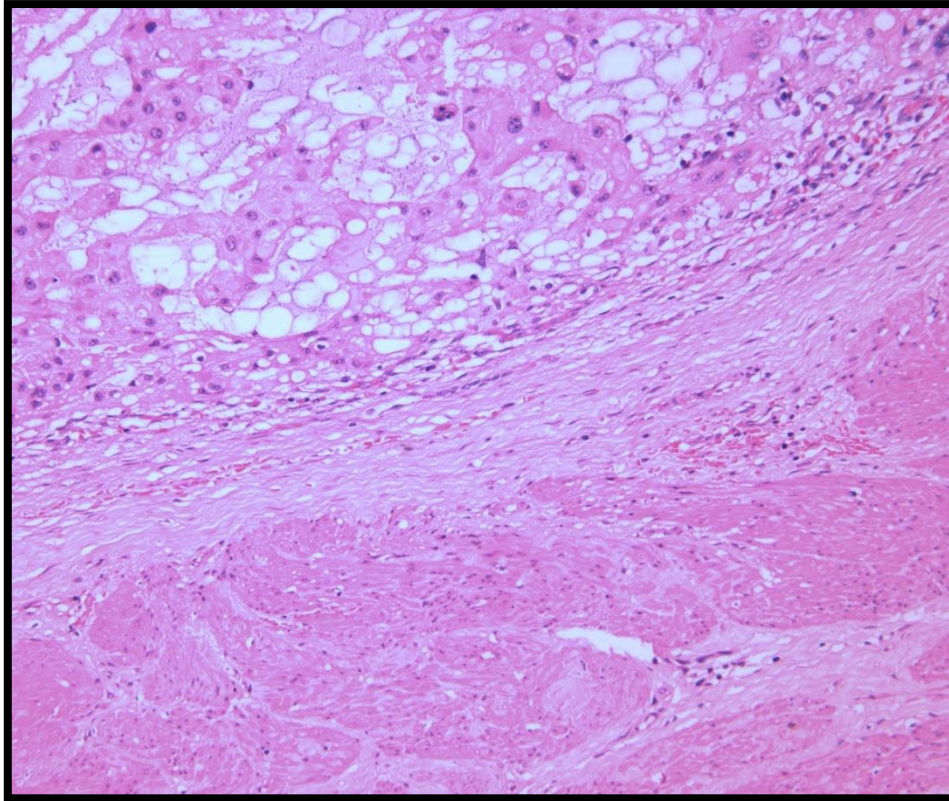
# „Physaliphorous cells“



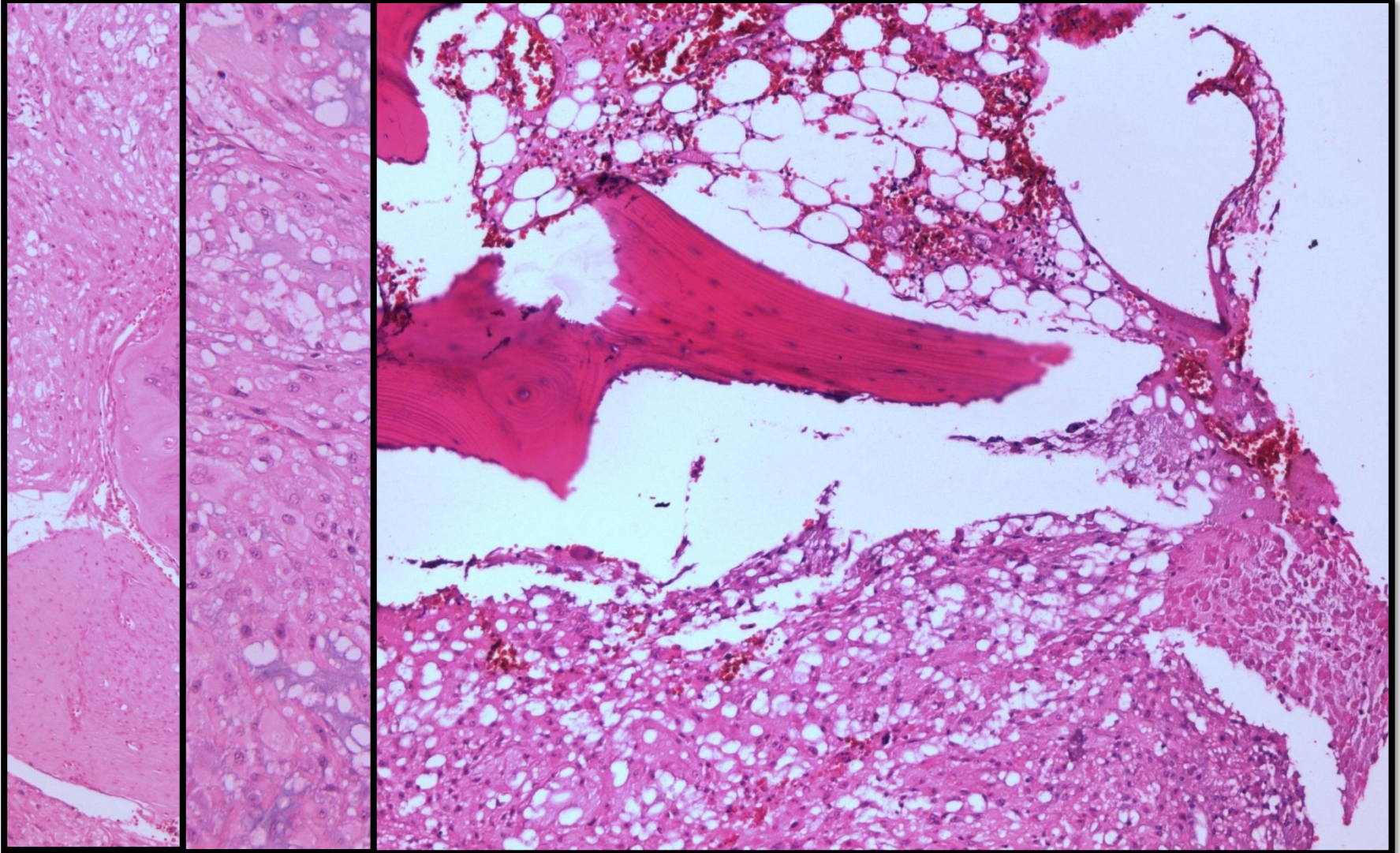
# atypie - mitózy



# Expanzívny rast



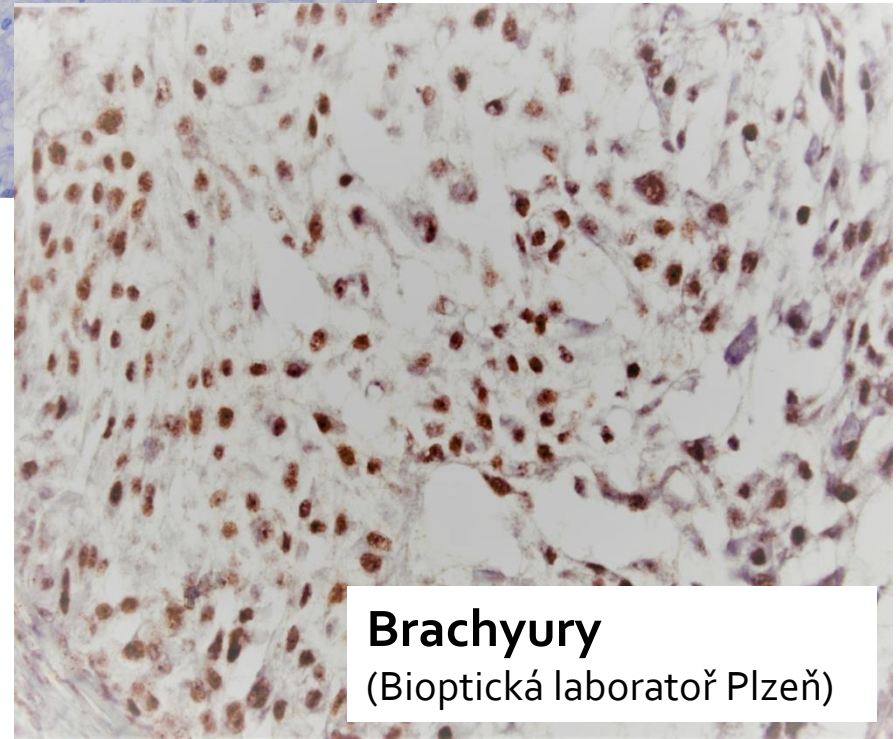
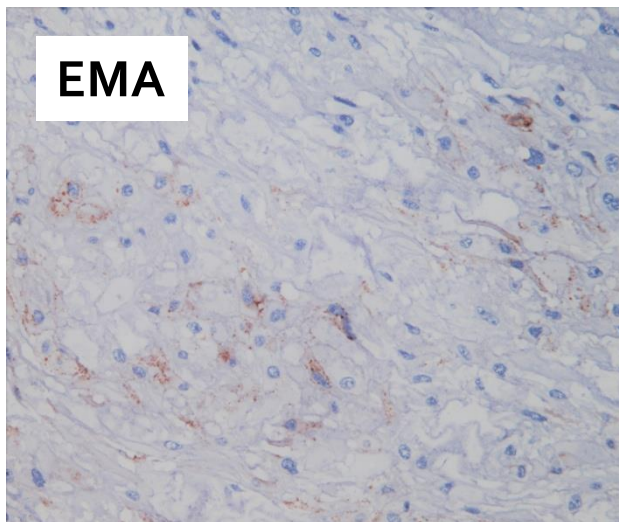
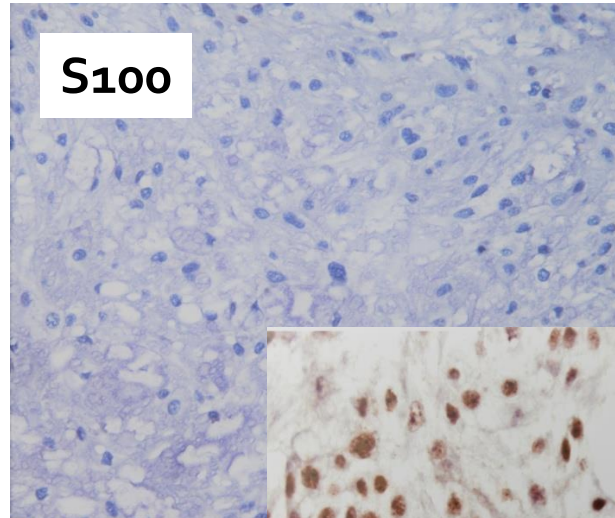
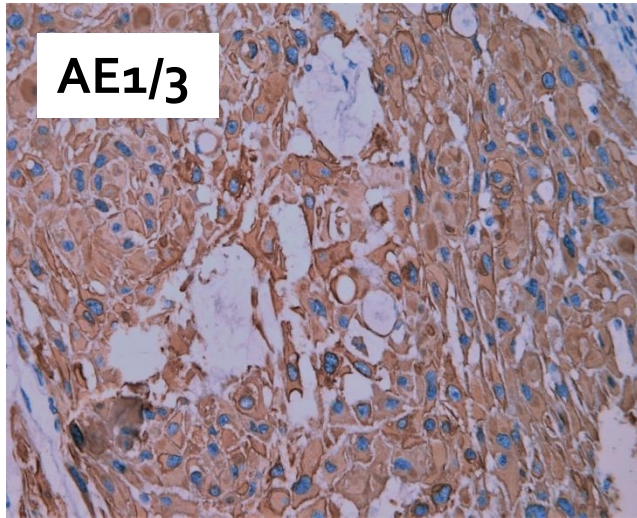
# Axiálny skelet



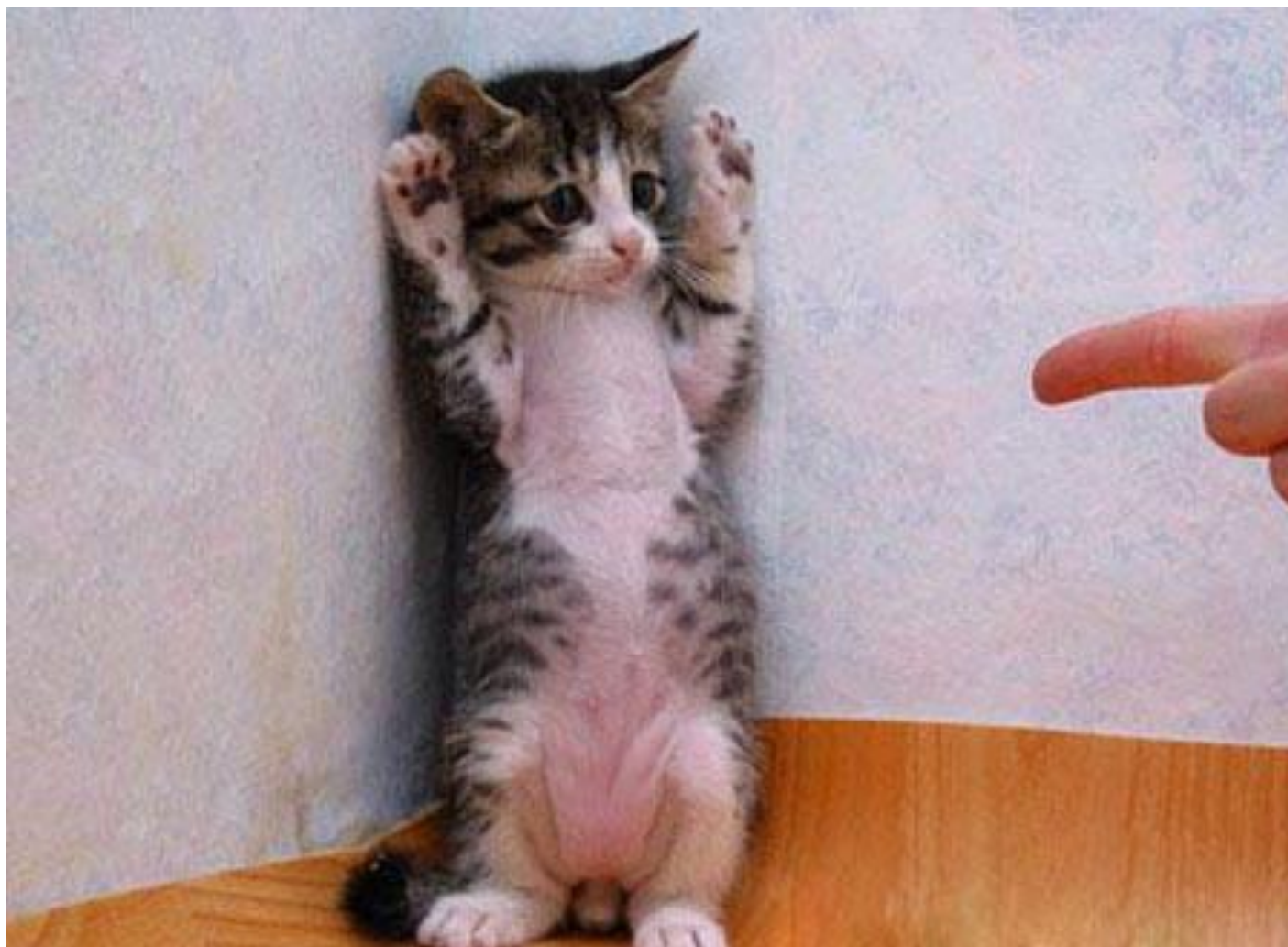
# Diagnostické návrhy



# Imunoprofil



# Diagnostický záver



# Chordoma

# Chordóm

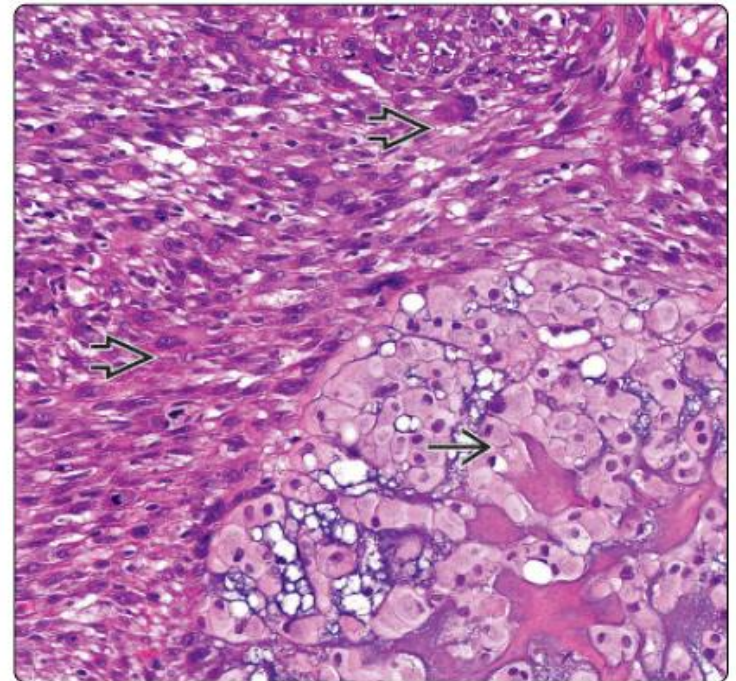
**Malígný** tumor vykazujúci notochordálnu diferenciáciu.

- starší ľudia (5.-7. dekáda)
- axiálny skelet (3/3, báza lebky, stavce, sakrokokcigeálna oblasť)
- bolesť (na podklade expanzívneho rastu)
- želatinózny, solídny, chondroidný, s nekrózami
- 7 – ročné prežívanie, 40% MTS (pľúca, kosti, LU, subkutis)

# Notochordálne tumory

- **Ecchordosis**
- **Benign Notochordal Cell Tumor**
- **Chordoma**
  - Not otherwise specified (NOS)
  - Chondroid chordoma
  - (Poorly differentiated)
  - **Dedifferentiated chordoma**  
(worse prognosis)

G.P.Nielsen, MD  
A.E.Rosenberg, MD  
Diagnostic pathology  
Bone, 2017



# IHC a genetické abnormality

## IMUNOFENOTYP

AE1/3 +  
EMA +  
S100 +

**Brachyury +**



po deklacifikácii



dediferencovaný

## MOLEKULÁRNE CHARAKTERISTIKY

**zriedka hereditárne** (autozom.dominantné)  
-/+ s duplikáciou brachyury génu (**T gene**)  
**sporadické (7% amplifikácia T génu)**  
diploidný / hypoploidný karyotyp  
viaceré numerické / štrukturálne preskupenia  
monozómia chromozómu 1, resp. nárast ch7  
70% strata CDKN2A a CDKN2B  
nárast kópií brachyury 7q33 lokusu a EGFR 7p12 lokus

Bez IDH1 a IDH2 mutácií

# Diferenciálna diagnostika

- AE1/3 +, brachyury +  Ecchordosis,  
Benign Notochordal Cell Tumor
- Conventional chordoma  Metastatic Adenocarcinoma  
„Parachordoma“  
„Chordoid sarcoma“
- Chondroid chordoma  Low-grade Conventional  
Chondrosarcoma
- „Poorly differentiated“  
Chordoma  Atypical Teratoid Rhabdoid  
Tumor

# Chordoma foundation

Understanding Chordoma × + -

chordomafoundation.org/understanding-chordoma

Ak tu chcete zobrazit' obľúbené položky, vyberte ikonu ☆ potom ikonu ☆ a presuňte položku do priečinka na paneli s obľúbenými položkami. Alebo ich importujte z iného prehliadača. [Importovať obľúbené](#)

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Chordoma is a rare type of cancer that occurs in the bones of the skull base and spine. It is part of a group of malignant bone and soft tissue tumors called sarcomas. Chordomas account for about 3 percent of all bone tumors and about 20 percent of primary spinal tumors. They are the most common tumor of the sacrum and cervical spine. A chordoma tumor usually grows slowly, often without symptoms at first, and then might cause symptoms for years before doctors find it.

Chordomas are complicated tumors to treat due to the involvement of critical structures such as the brainstem, spinal cord, and important nerves and arteries. They can also come back, or recur, after treatment — usually in the same place as the first tumor. This is called a local recurrence. In about 30 to 40 percent of patients, the tumor eventually spreads, or metastasizes, to other parts of the body.

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# Limitovaný odber



ĎAKUJEM ZA POZORNOST

