

SL-02-1

Mimics of malignancy: An emphasis on microscopic features and best use of limited molecular tests

001 - Case - 1

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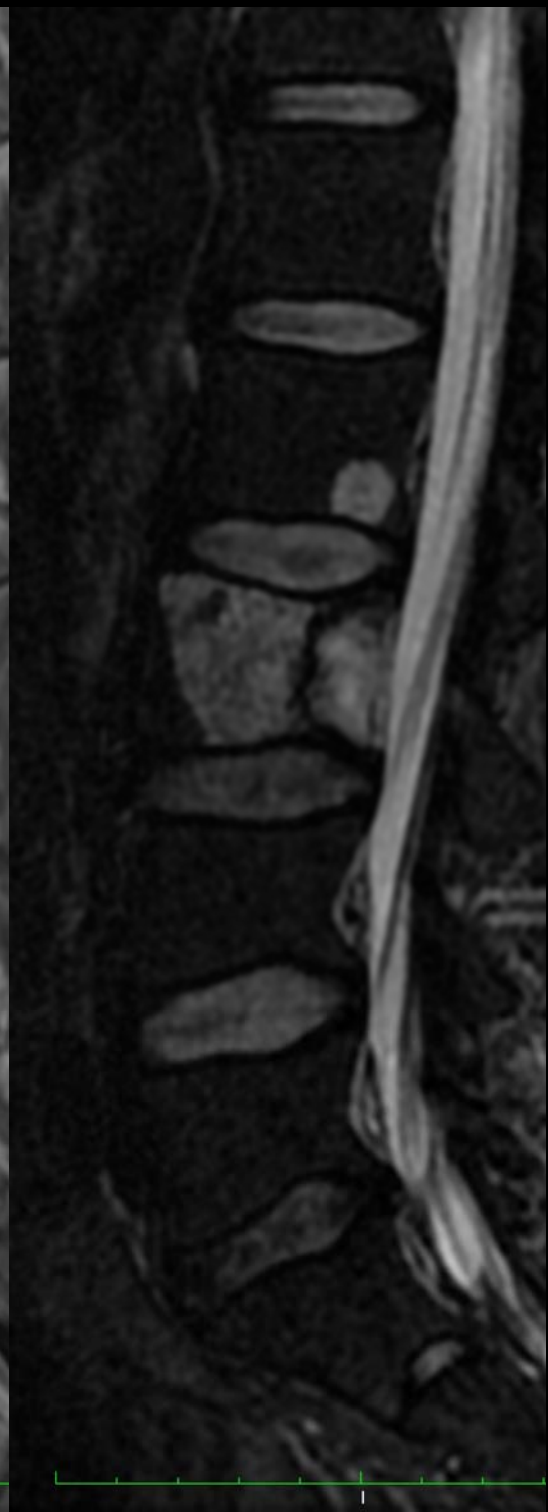
Clinical History

A 39-year-old man complained of low back pain following right knee pain. MRI demonstrated a bone tumor in the third lumbar vertebra. The tumor was biopsied and then excised.

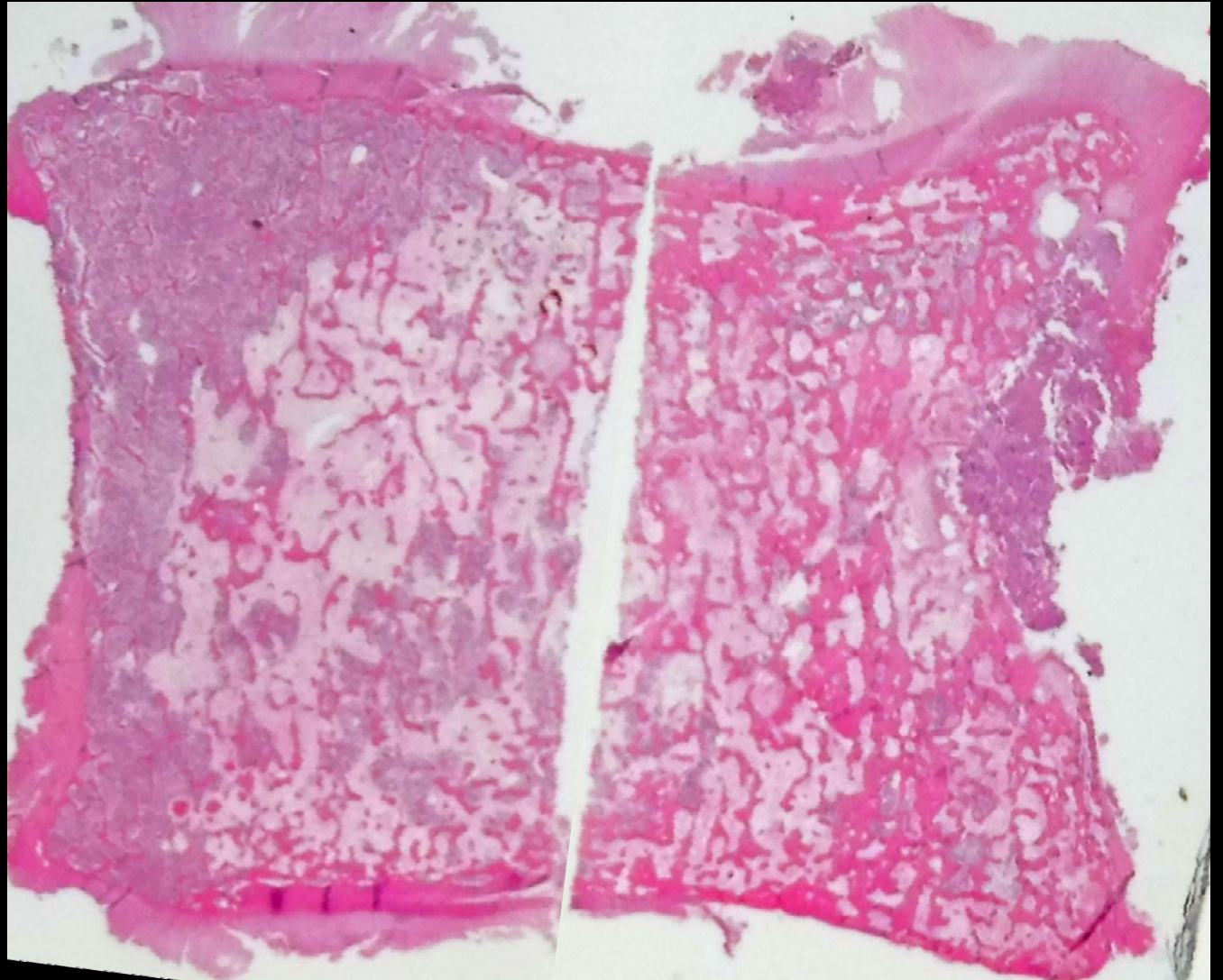
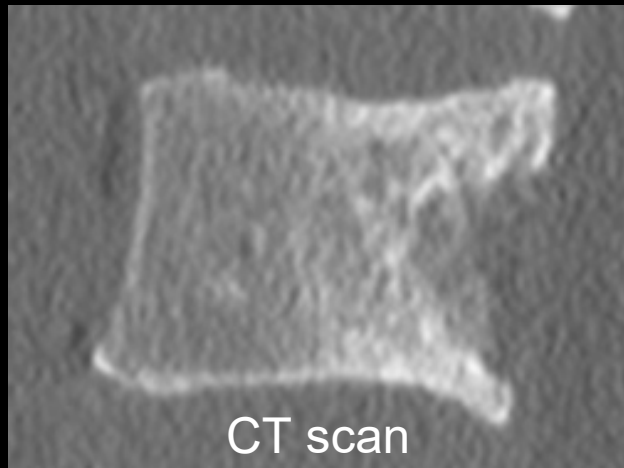
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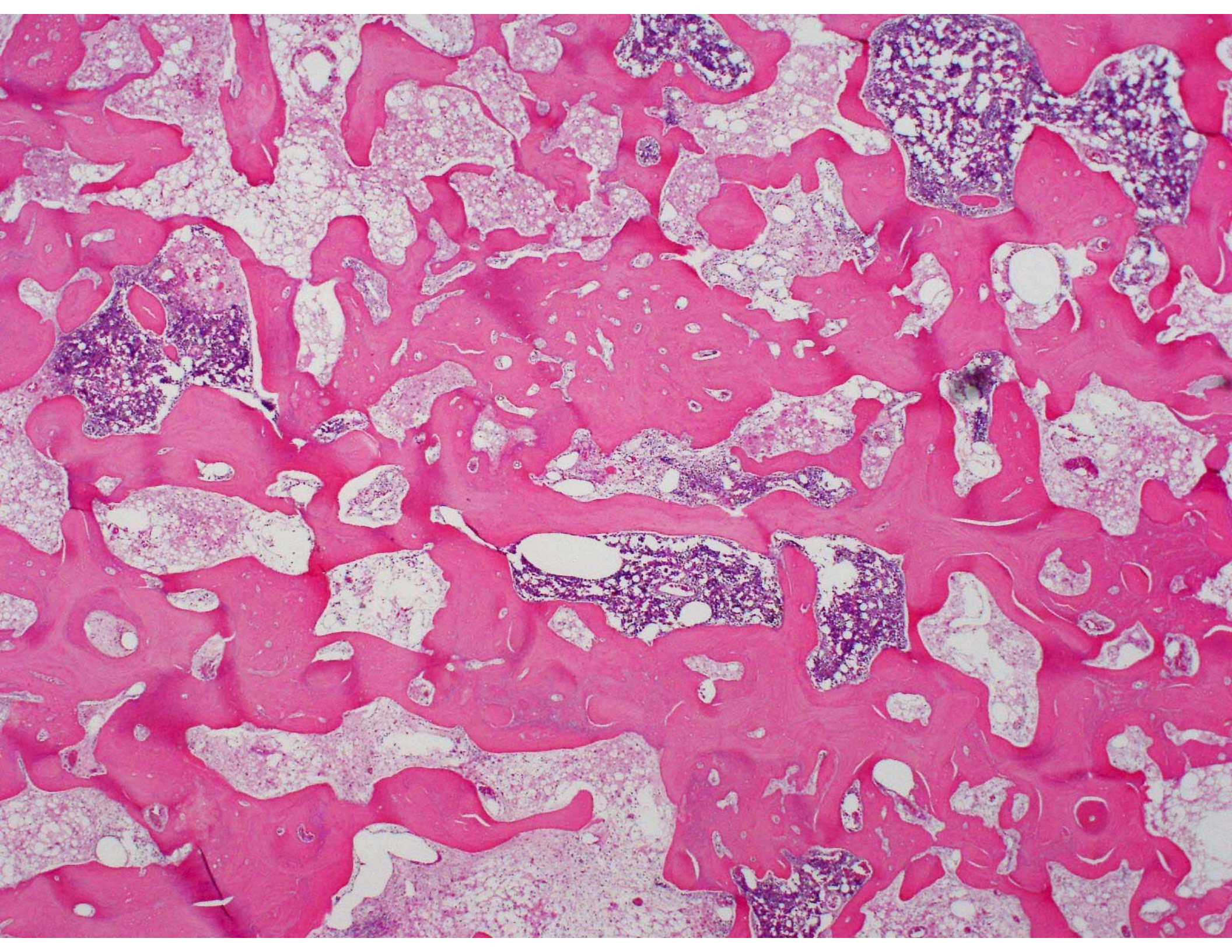


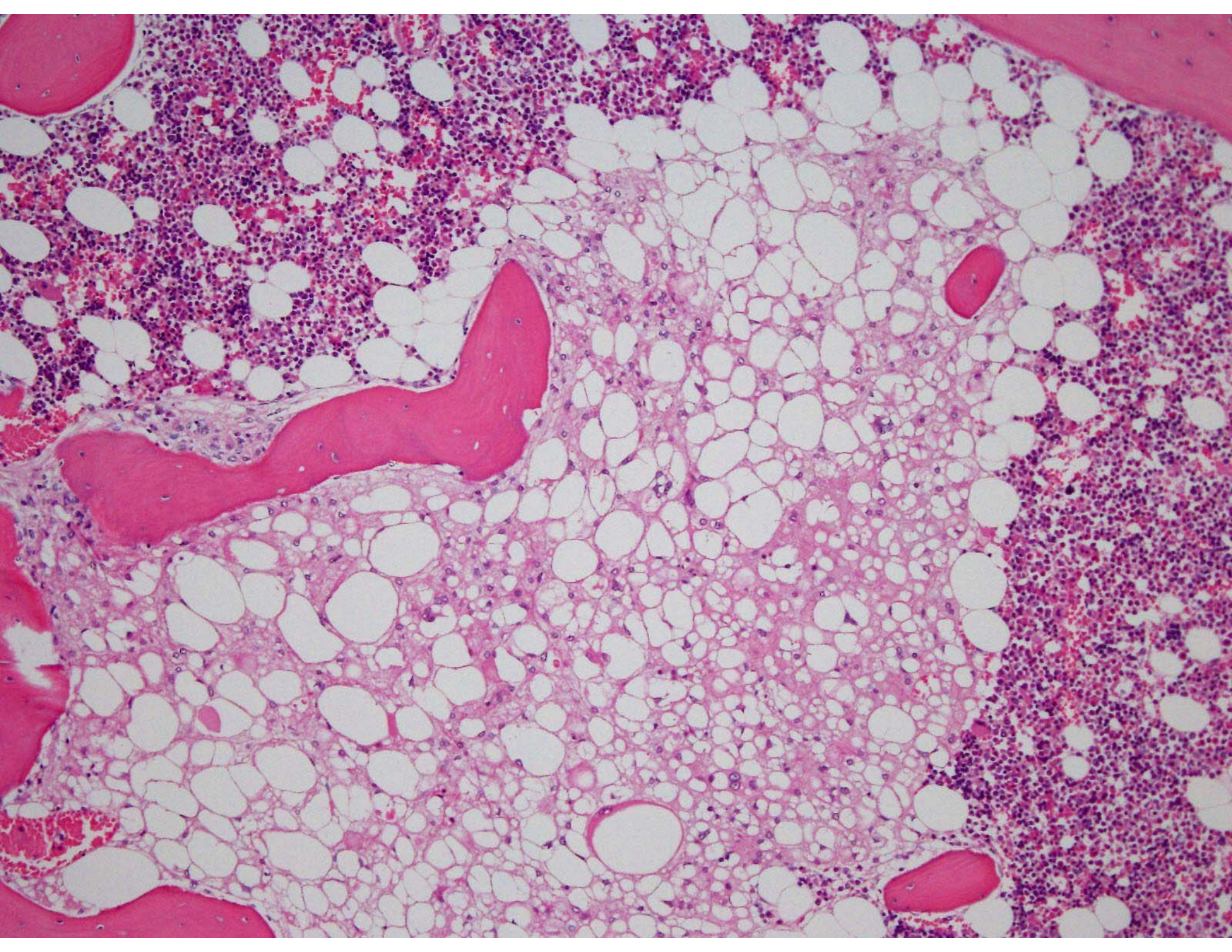
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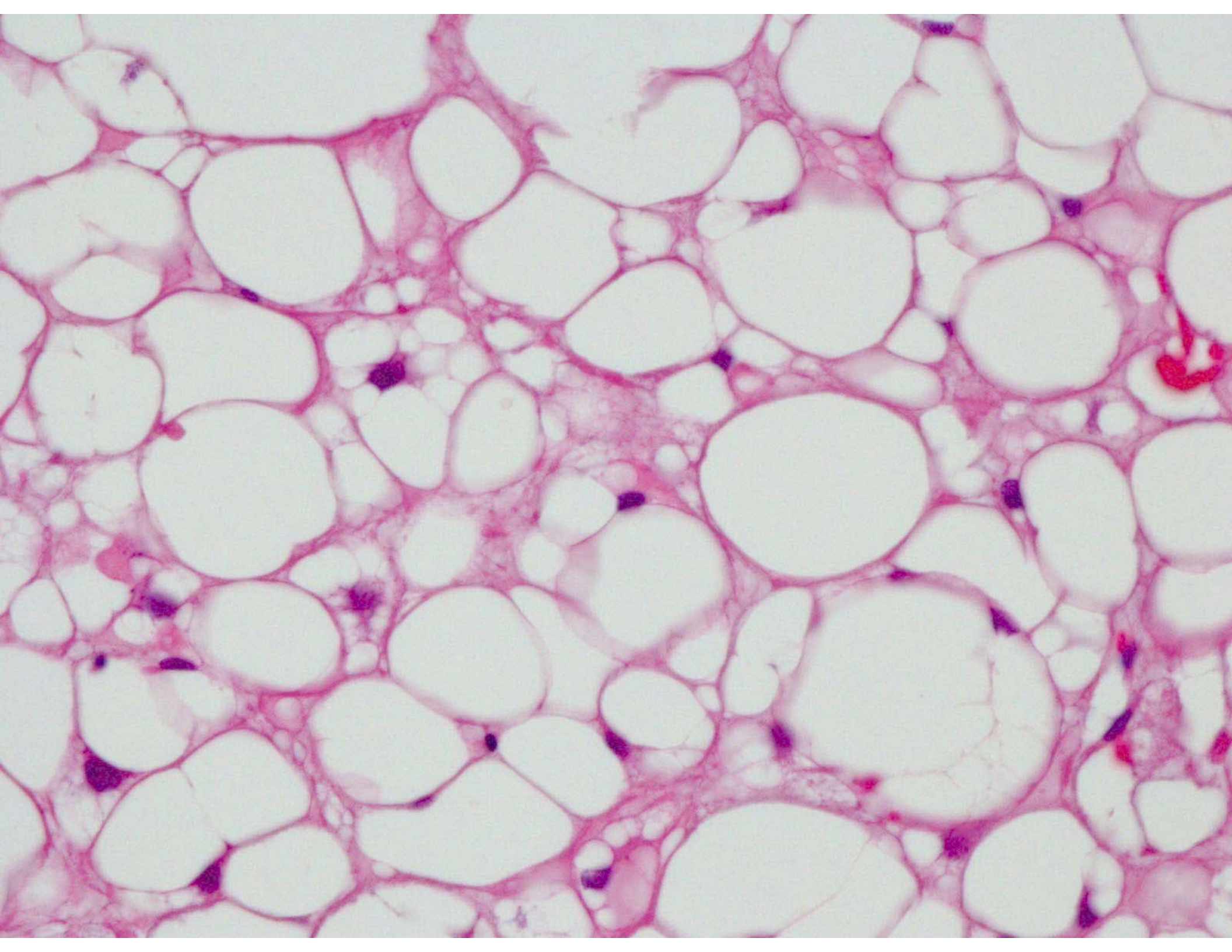


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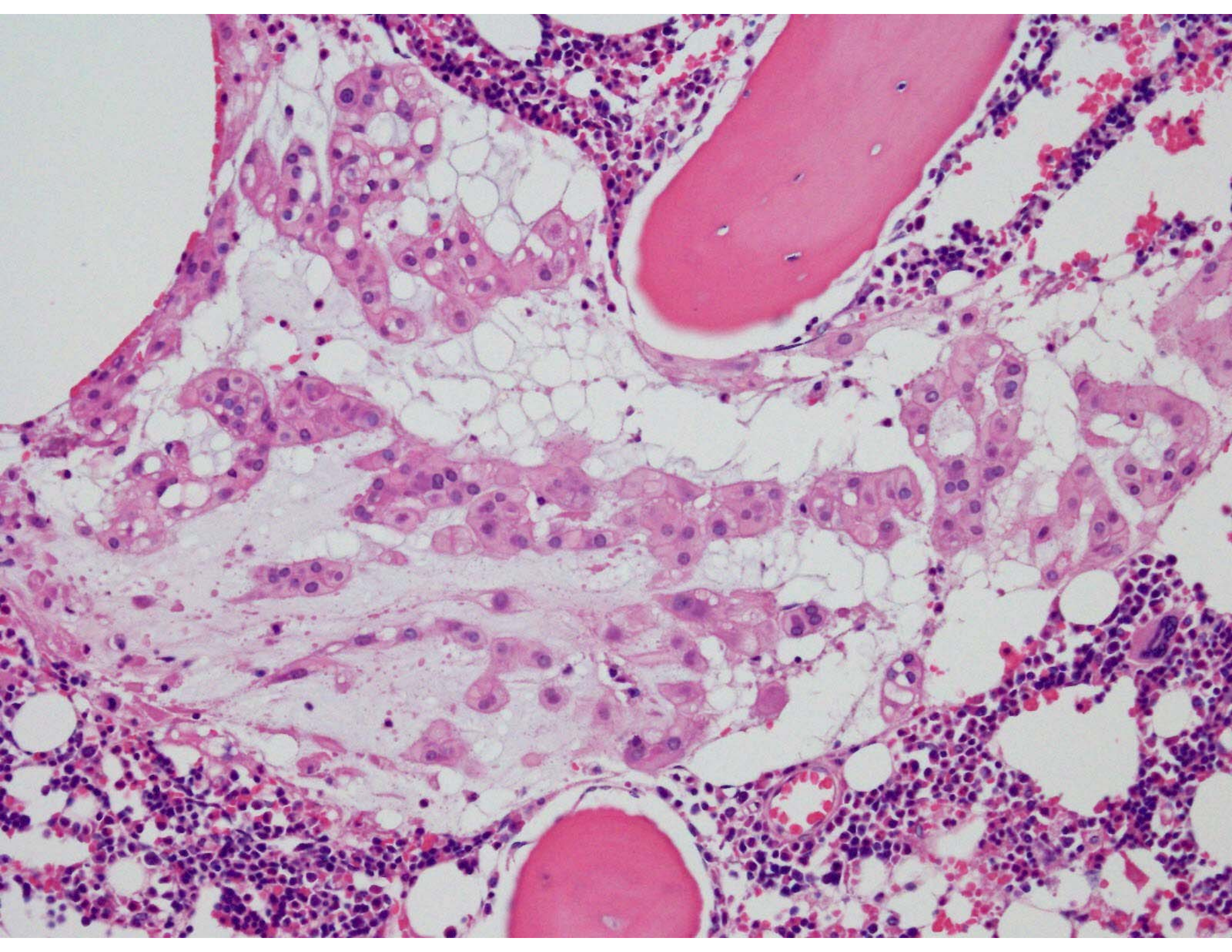


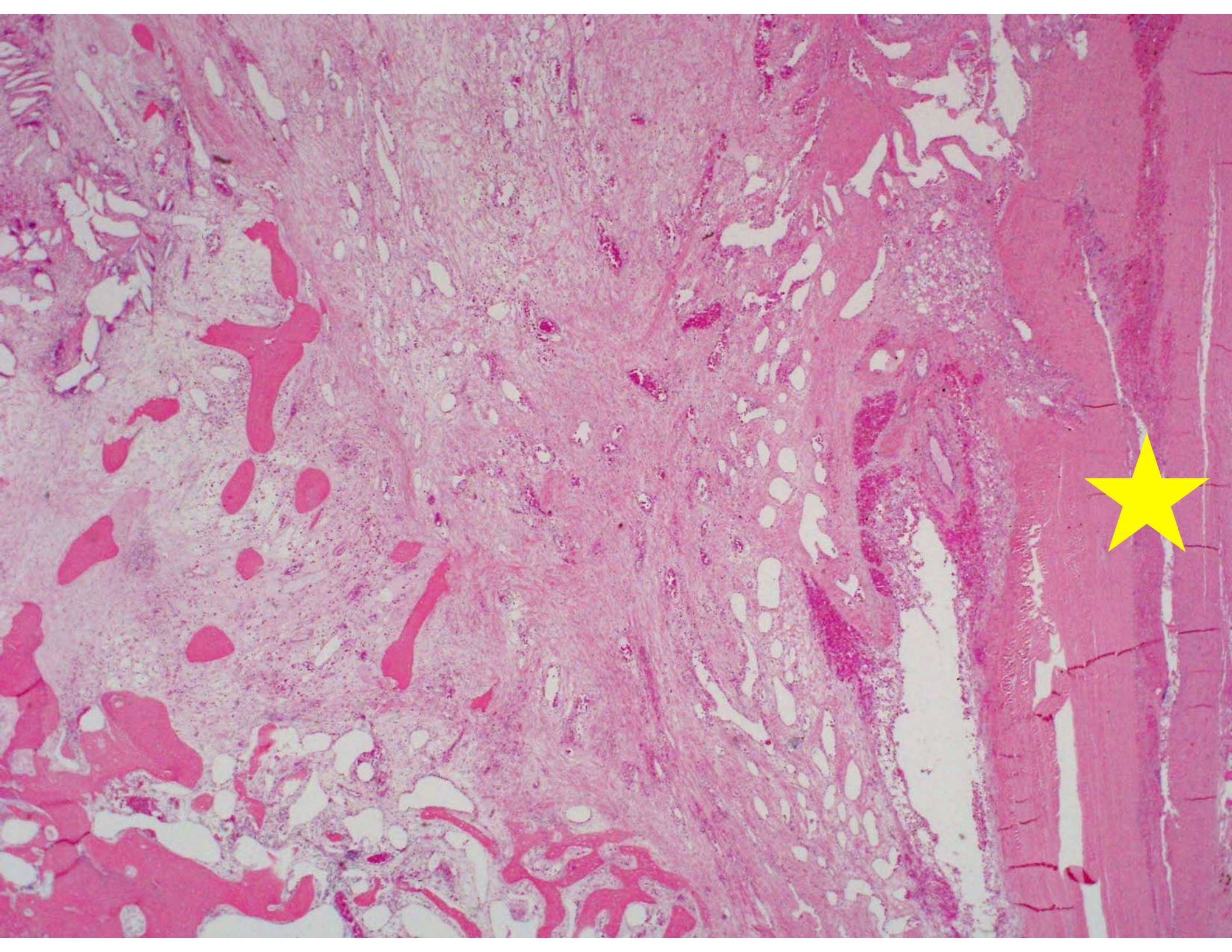


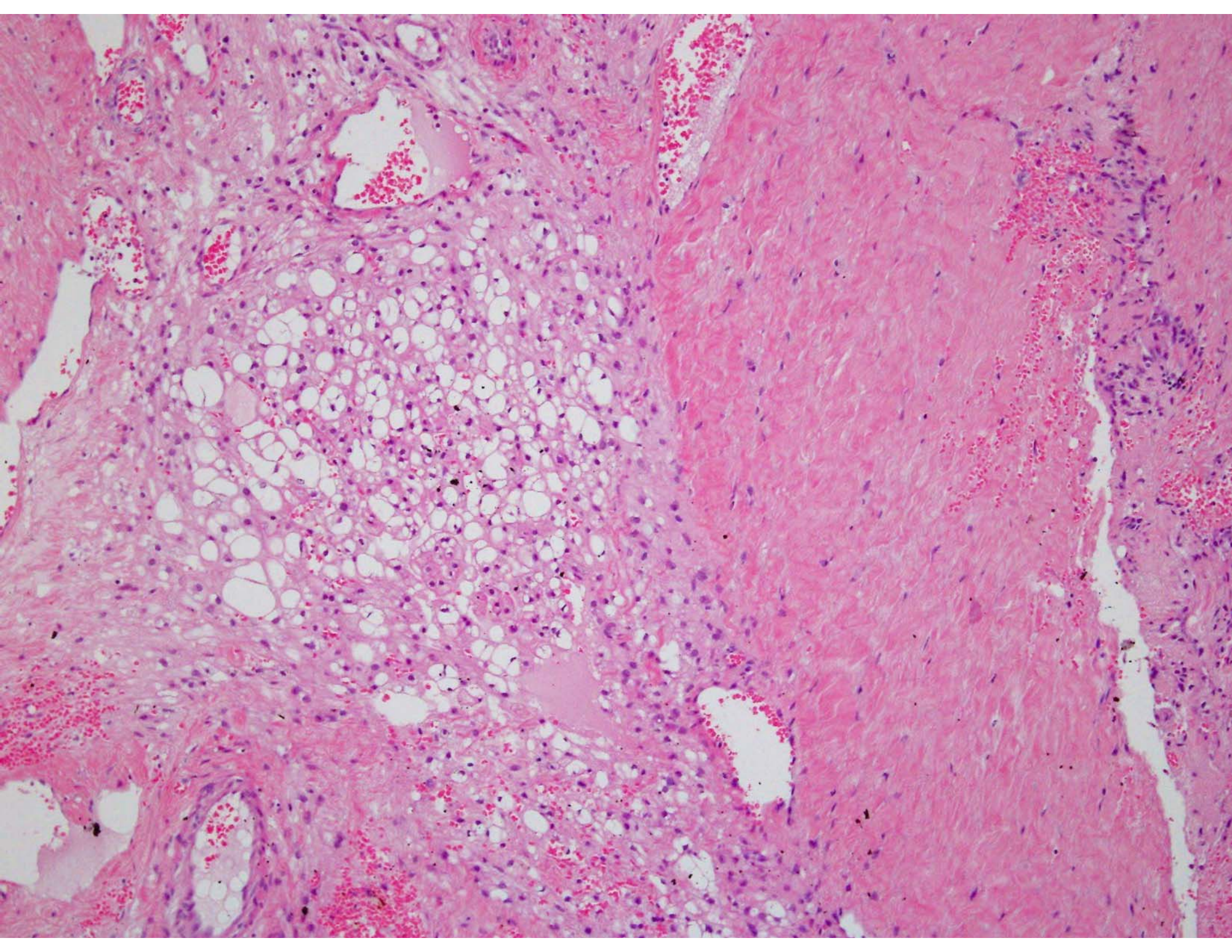


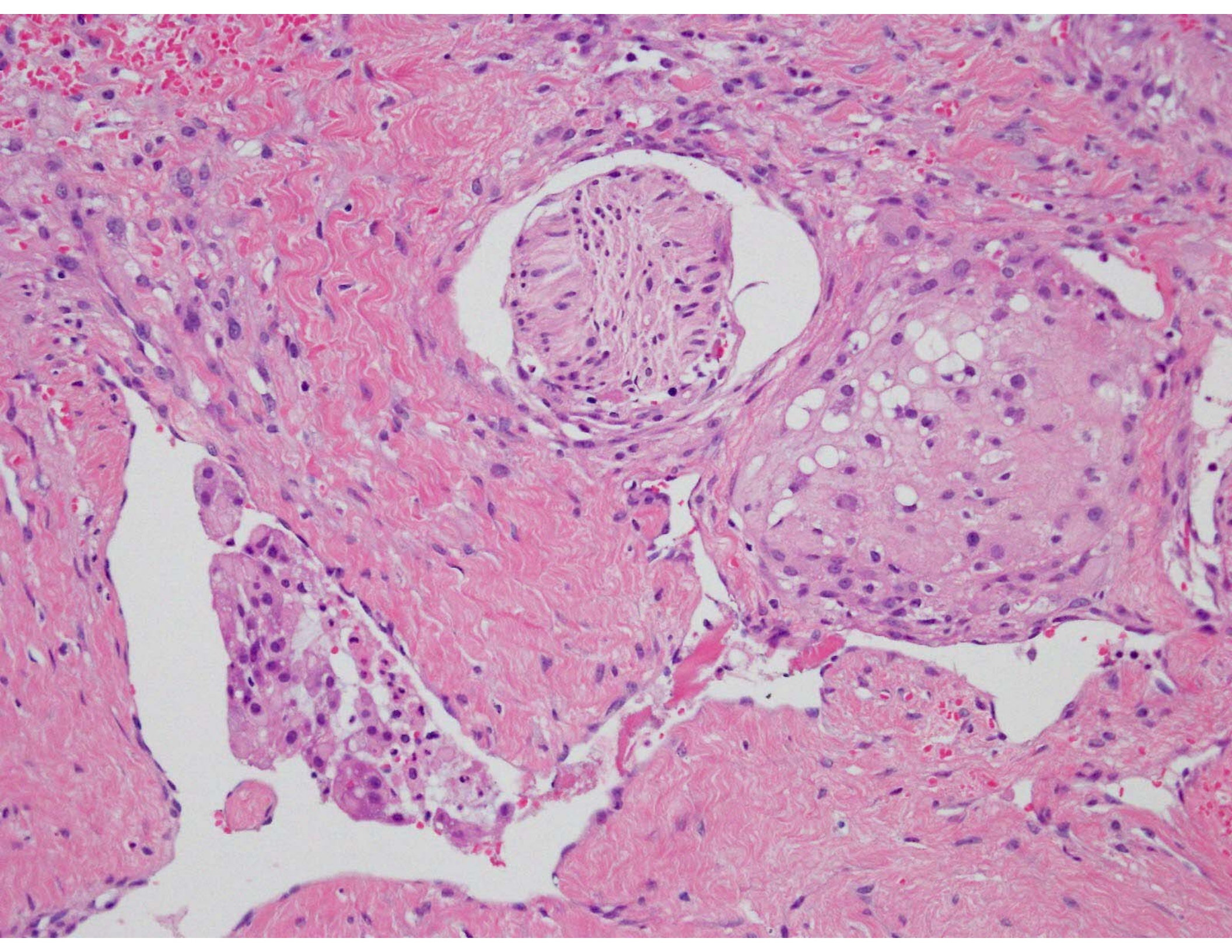


Benign Notochordal Cell Tumor

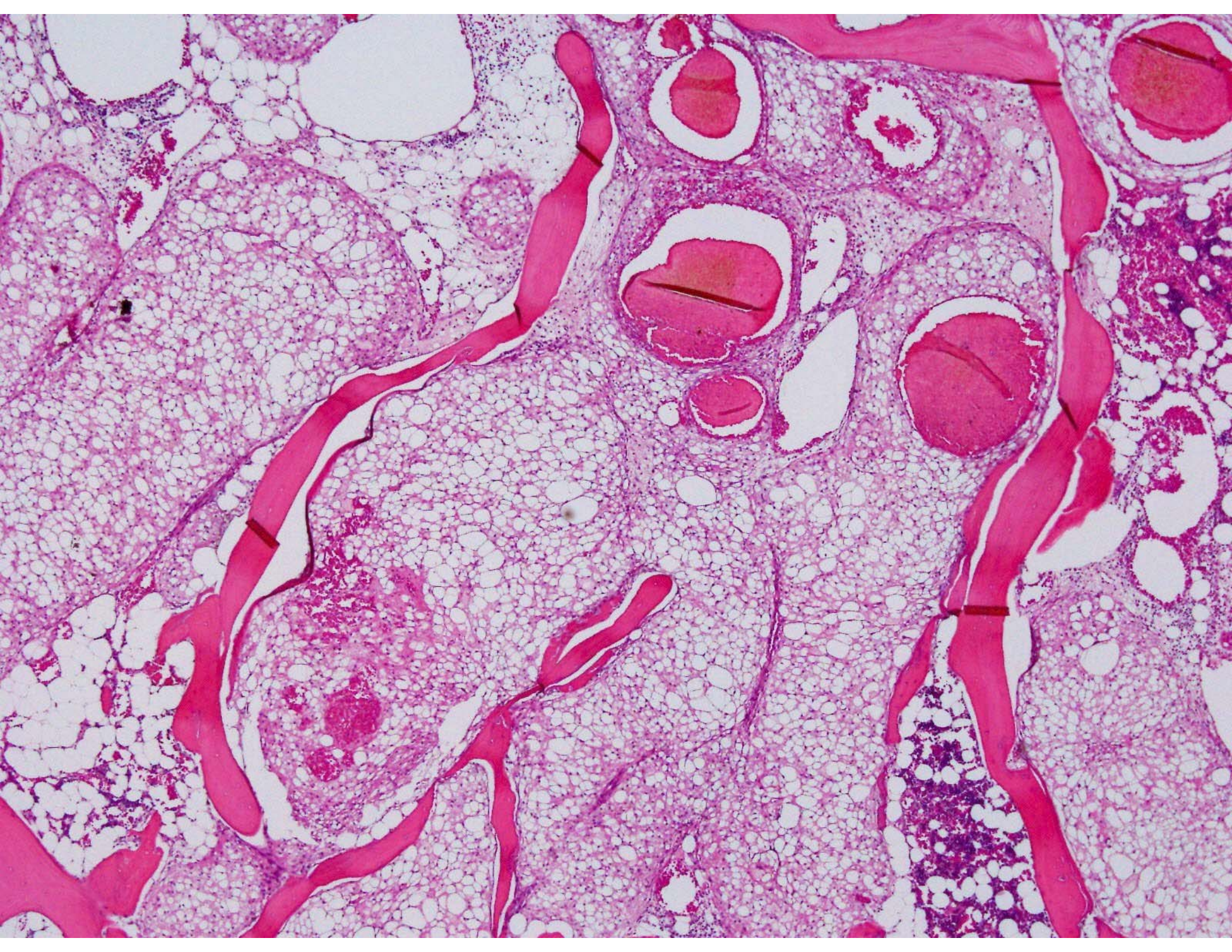


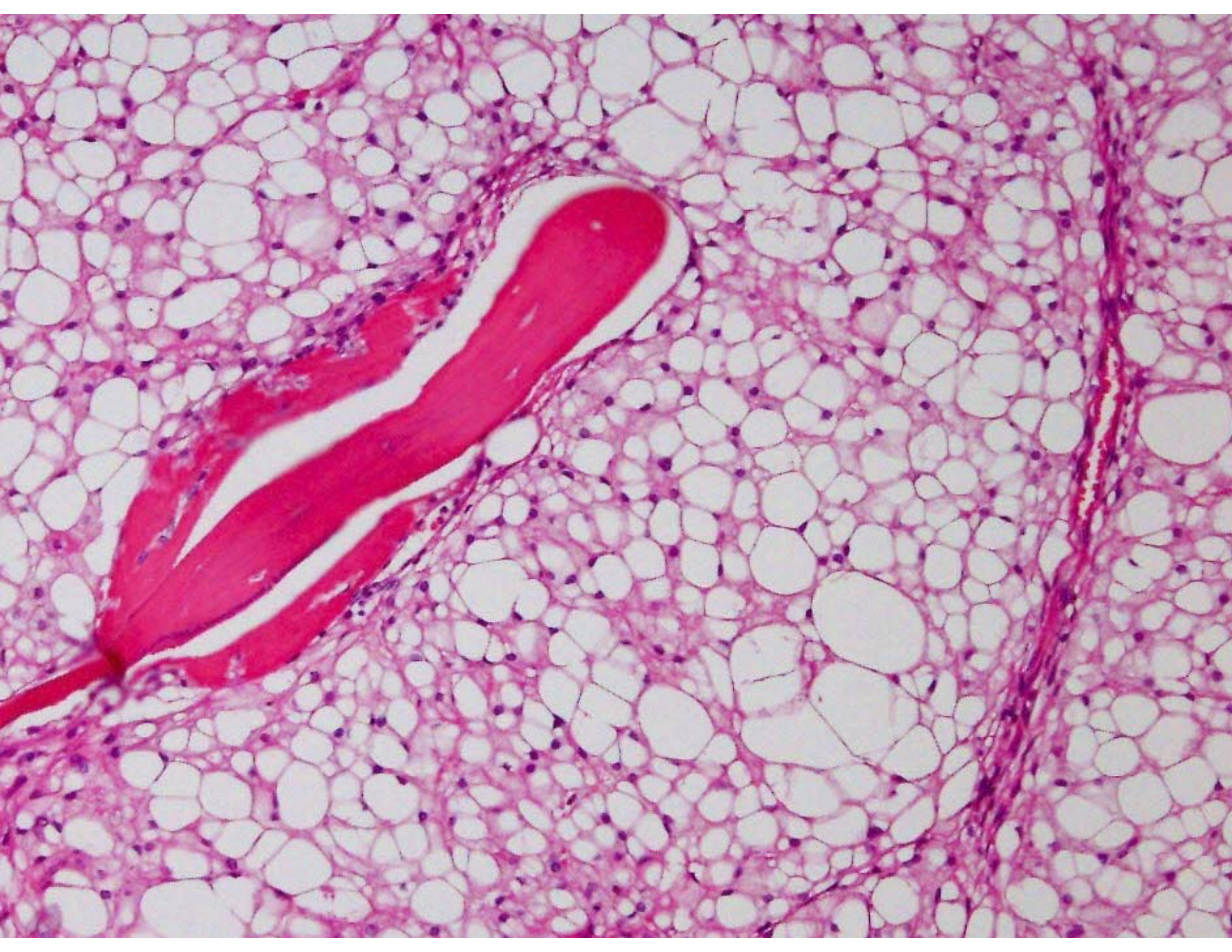




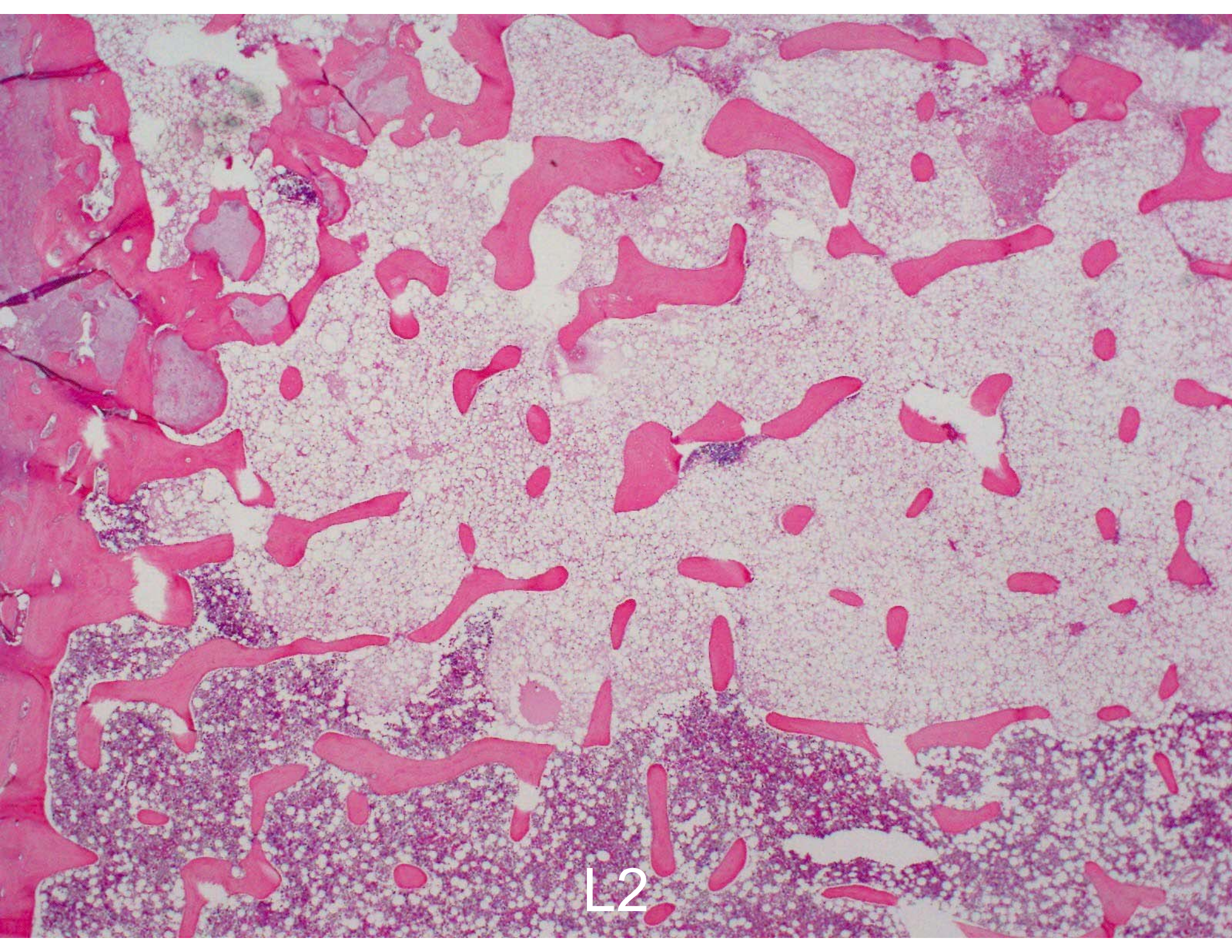


Chordoma

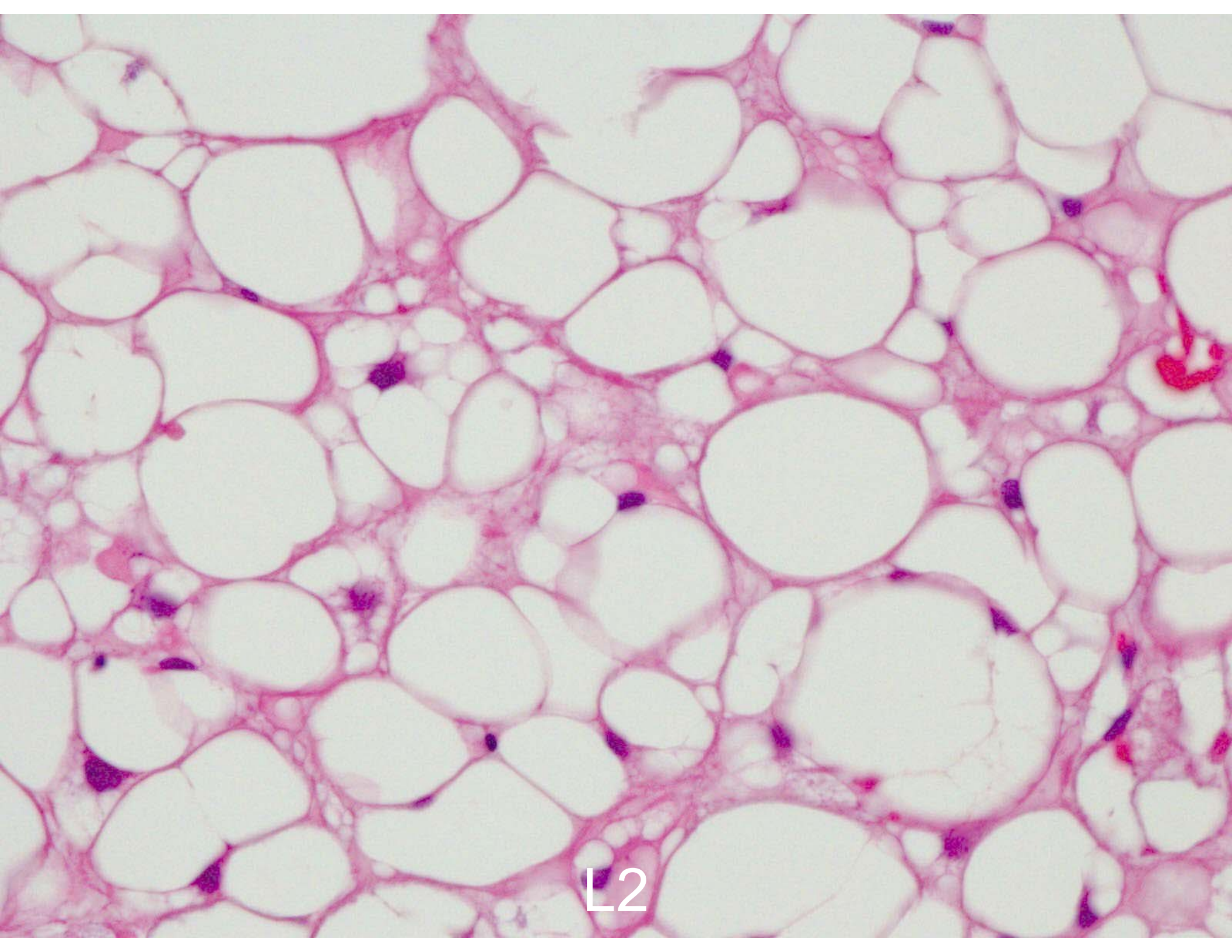




Chordoma, well
differentiated, associated
with BNCT components



L2



L2

Benign Notochordal Cell Tumor

Diagnosis

Chordoma, well differentiated,
associated with Benign
Notochordal Cell Tumors

Benign Notochordal Cell Tumor (BNCT)

- **Benign counterpart of chordoma**
- **Intraosseous benign tumor with notochordal differentiation**
- **may develop after birth**
- **different from notochordal rest**
- **Brachyury expression**

Benign Notochordal Cell Tumor (BNCT)

- **Location**
 - **Sacrococcyx and clivus (autopsy cases)**
 - **Cervical and lumbar vertebra (clinical cases)**
- **Imaging**
 - **X-ray: normal to sclerosis, occasionally presented as ivory vertebra**
 - **CT scan: normal to sclerosis, no bone destruction or extraosseous mass**
 - **MRI: homogeneous, low signal on T1-WI, high on T2-WI, no enhancement on enhanced T1-WI**
 - **Bone scan: no abnormal uptake**
- **Histology**
 - **Solid sheets of adipocyte-like vacuolated cells and less vacuolated cells with eosinophilic cytoplasm**
 - **No extracellular myxoid matrix, lobulation, or fibrous capsule**
 - **No significant nuclear atypia and no mitosis**
 - **Entrapped bone marrow**
 - **Sclerotic bone trabeculae affected**

Classification of Chordoma

- **Conventional**
- **Chondroid**
- **Dedifferentiated**

Classification of Chordoma

- **Conventional**
- **Chondroid**
- **Dedifferentiated**
- **Poorly differentiated**
INI-1 (SMARCB1) negative

Classification of Chordoma

- **Conventional**
 - ★ Well differentiated
 - ★ Moderately differentiated
 - ★ Poorly differentiated
- **Chondroid**
- **Dedifferentiated**

Differential Diagnosis Between BNCT and Well Differentiated Chordoma

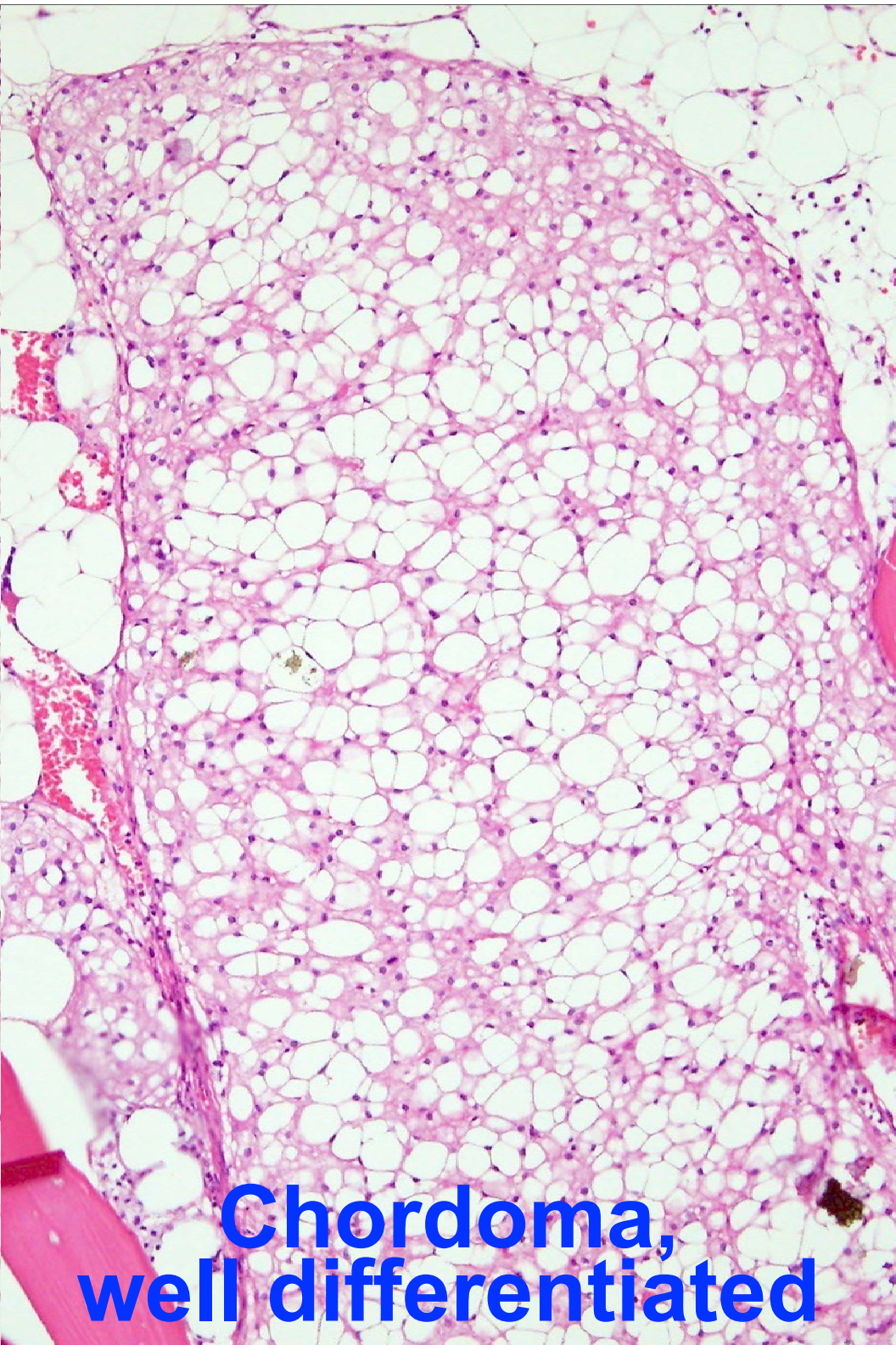
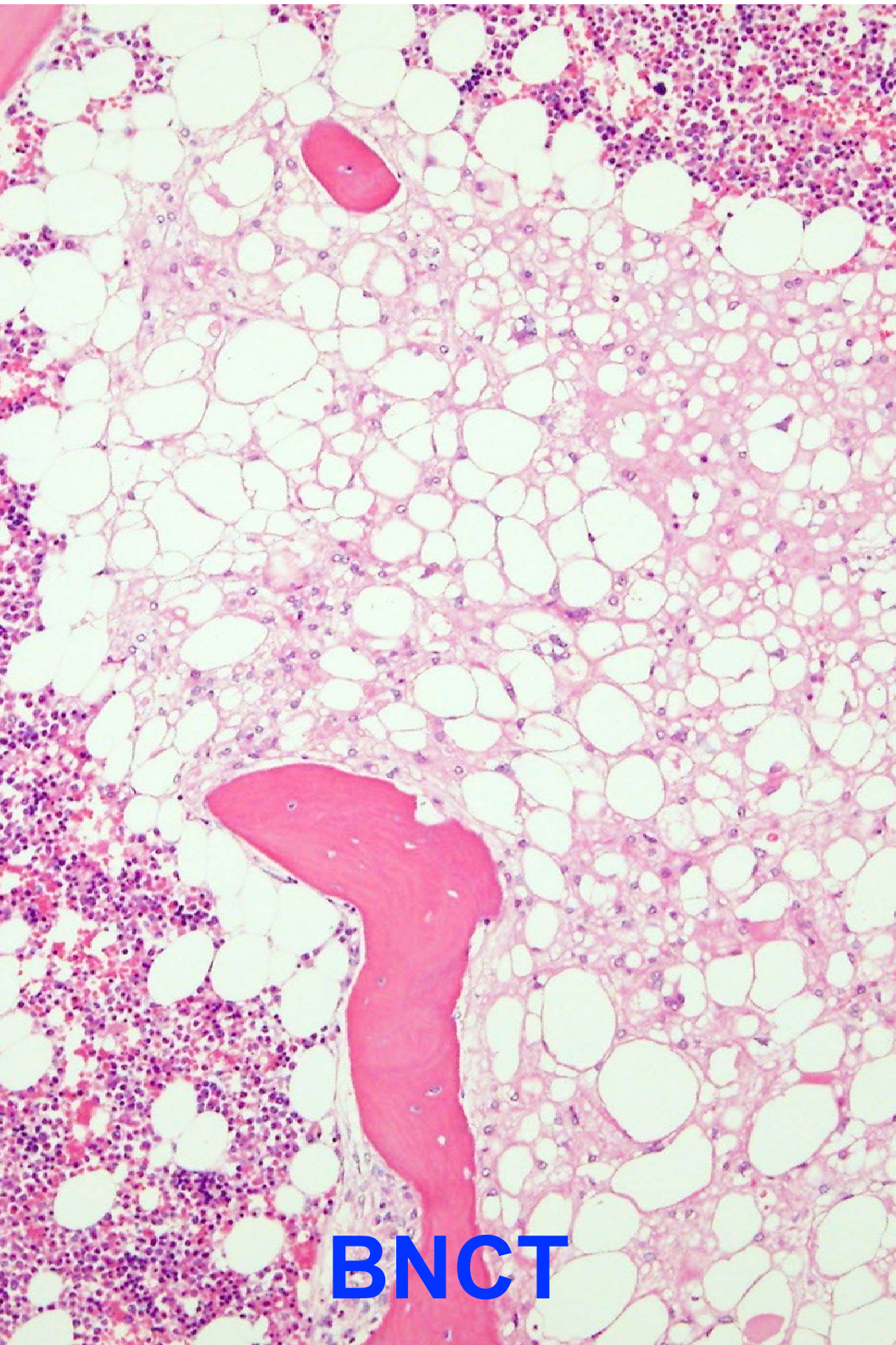
- Radiologically and morphologically overlap
- Can be distinguished



BNCT



Chordoma



Molecular Test

Unavailable, so far

Summary

- **A unique case of chordoma associated with BNCTs**
- **Chordoma may develop in BNCT**
- **Well differentiated chordoma mimics BNCT**
- **Careful imaging and microscopic interpretation is important for differentiating them**
- **No useful molecular test, so far**



Thank you!
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