

Condroid Chordoma in a Ferret

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Signalment and History

- 6 year old male ferret
- 2-cm mass at the distal tip of the tail present for 1 week

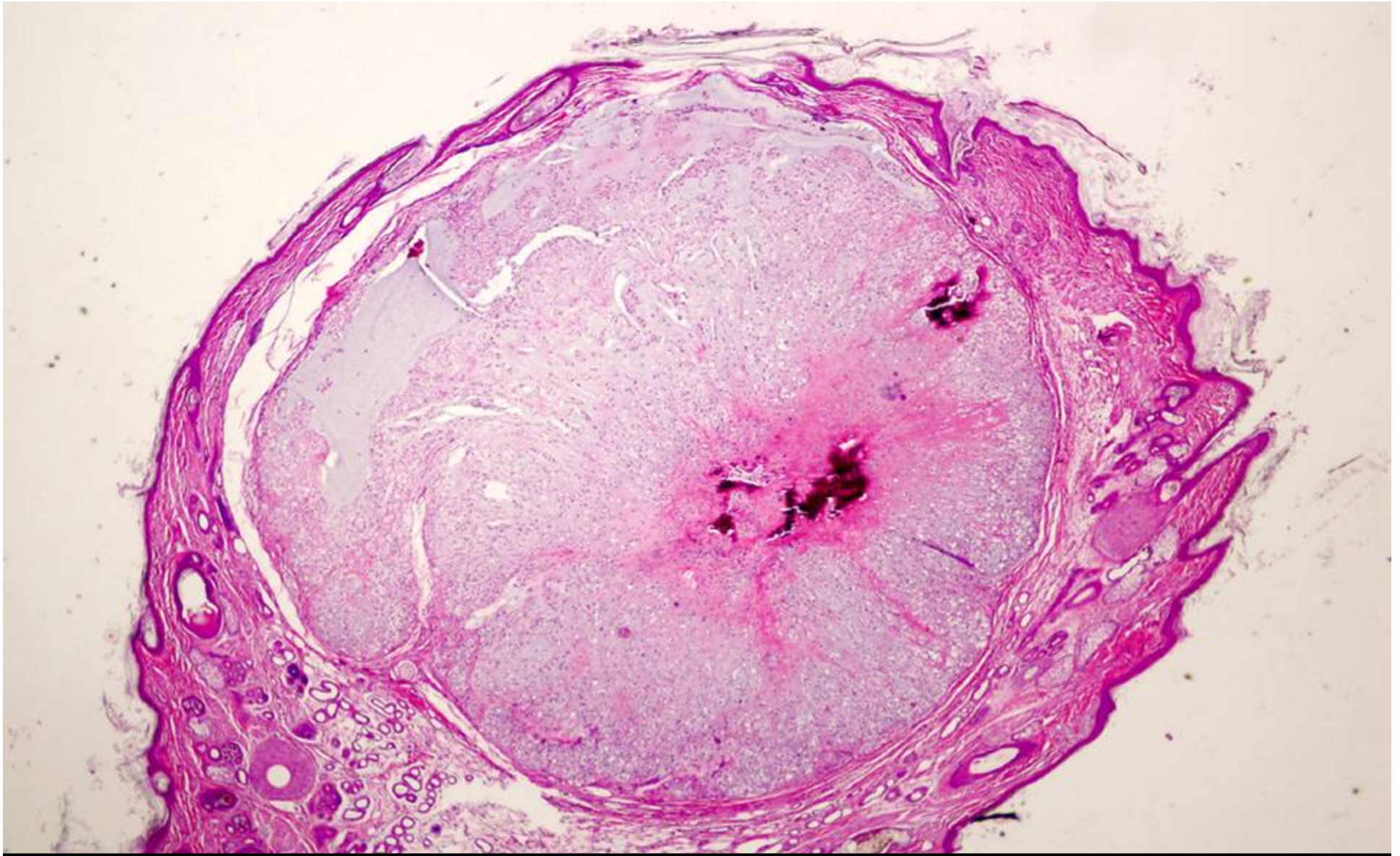


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Histopathology

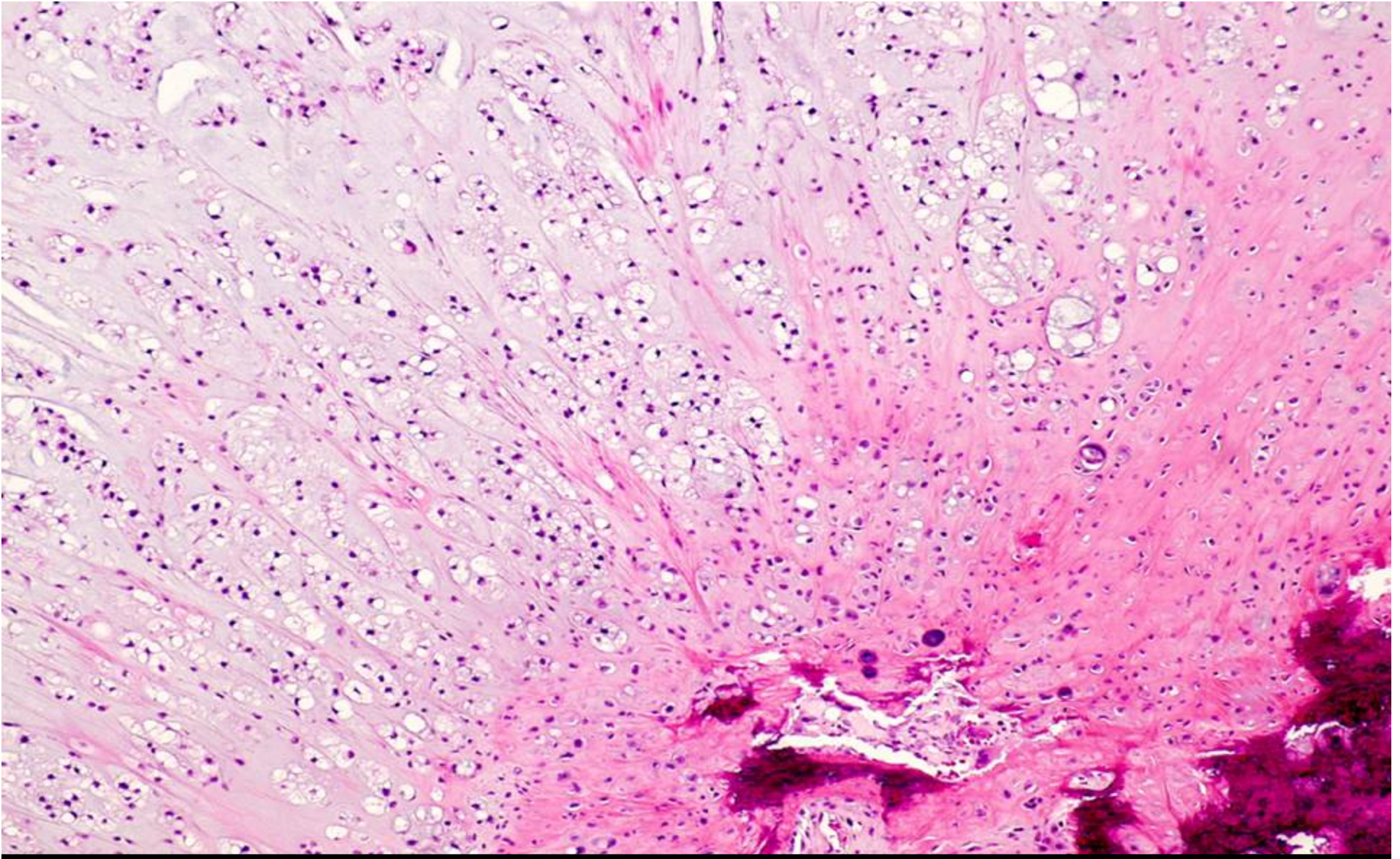
- **Lobulated mass with 3 concentric zones**
 - **Zone I: Discrete, vacuolated cells in a myxomatous matrix**
 - **Zone II: Small discrete cells in a hyaline matrix**
 - **Zone III: Central area of mineralization**



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Morphological Diagnosis

Ferret; distal tail, chondroid chordoma

Chordoma

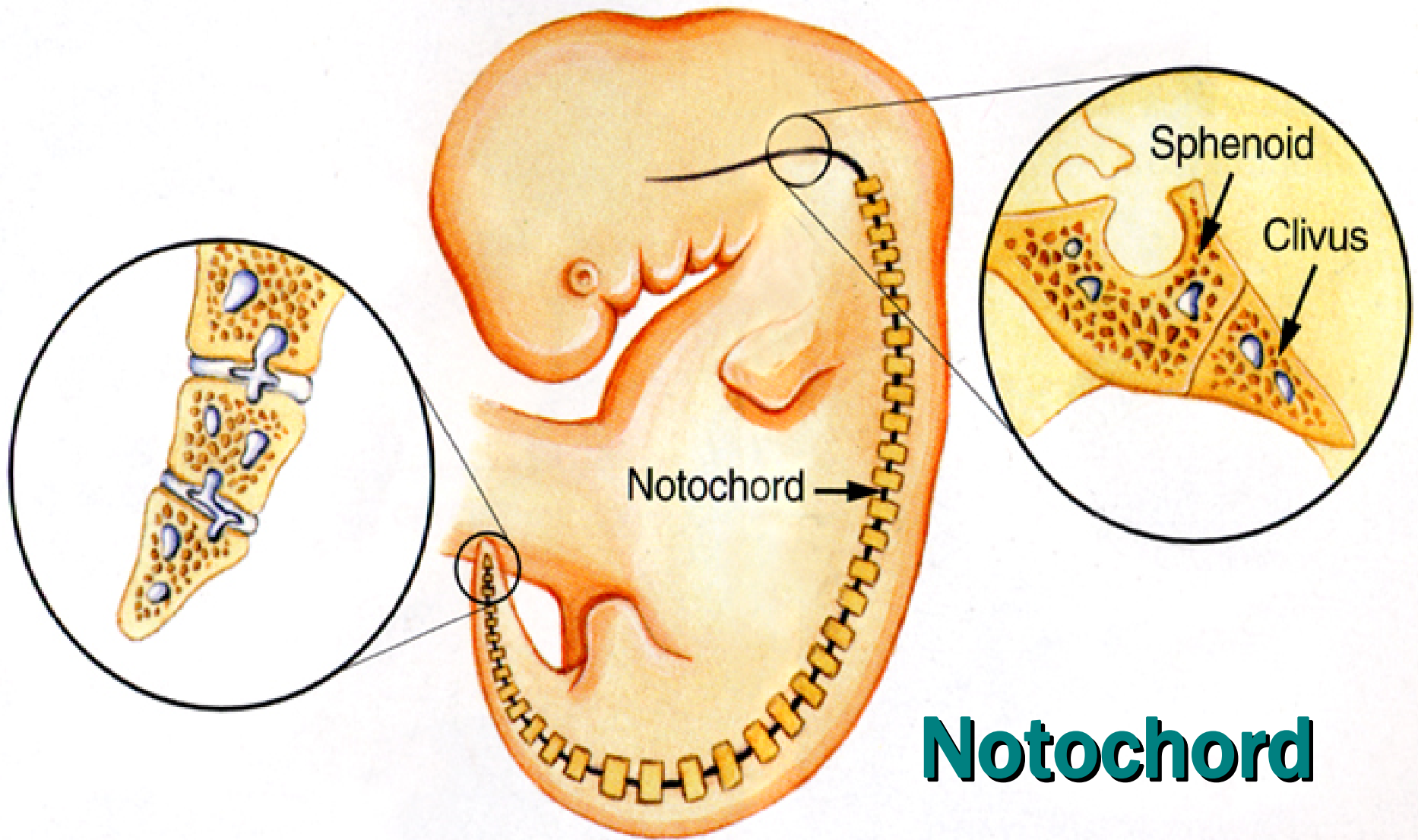
- **Neoplasm derived from remnants of notochord**
- **Most commonly found at the distal extremity of the tail in ferrets**
 - **Also reported in the cervical region**
- **Mean age: 3.4 years**
 - **All cases found in literature >3 years old.**
- **Slow growing**
- **1 reported case of metastasis**
 - **Skin near site of original mass**

Chordoma

- **3 variants of human chordoma:**
 - **Classical chordoma**
 - **Characterized by physaliferous cells**
 - **Slow growing, high rate of local recurrence**
 - **Dedifferentiated chordoma**
 - **Anaplastic, spindle cell component**
 - **Rapidly progressive disease**
 - **Chondroid chordoma**
 - **Area of cartilaginous differentiation**
 - **Sphenooccipital region**
 - **Improved prognosis**
 - **Equivalent to those found in ferrets**

Notochord

- **Rod shaped body of mesodermal cells**
- **Derived from the primitive streak**
 - **3rd week of development**
- **Functions:**
 - **The primordial axis**
 - **Support**
 - **Center of developmental organization**
- **Surrounded by developing vertebrae, regresses**
- **Persistent notochordal remnants**
 - **Nucleus pulposus**
 - **Ecchordoses physaliphora**



Notochord



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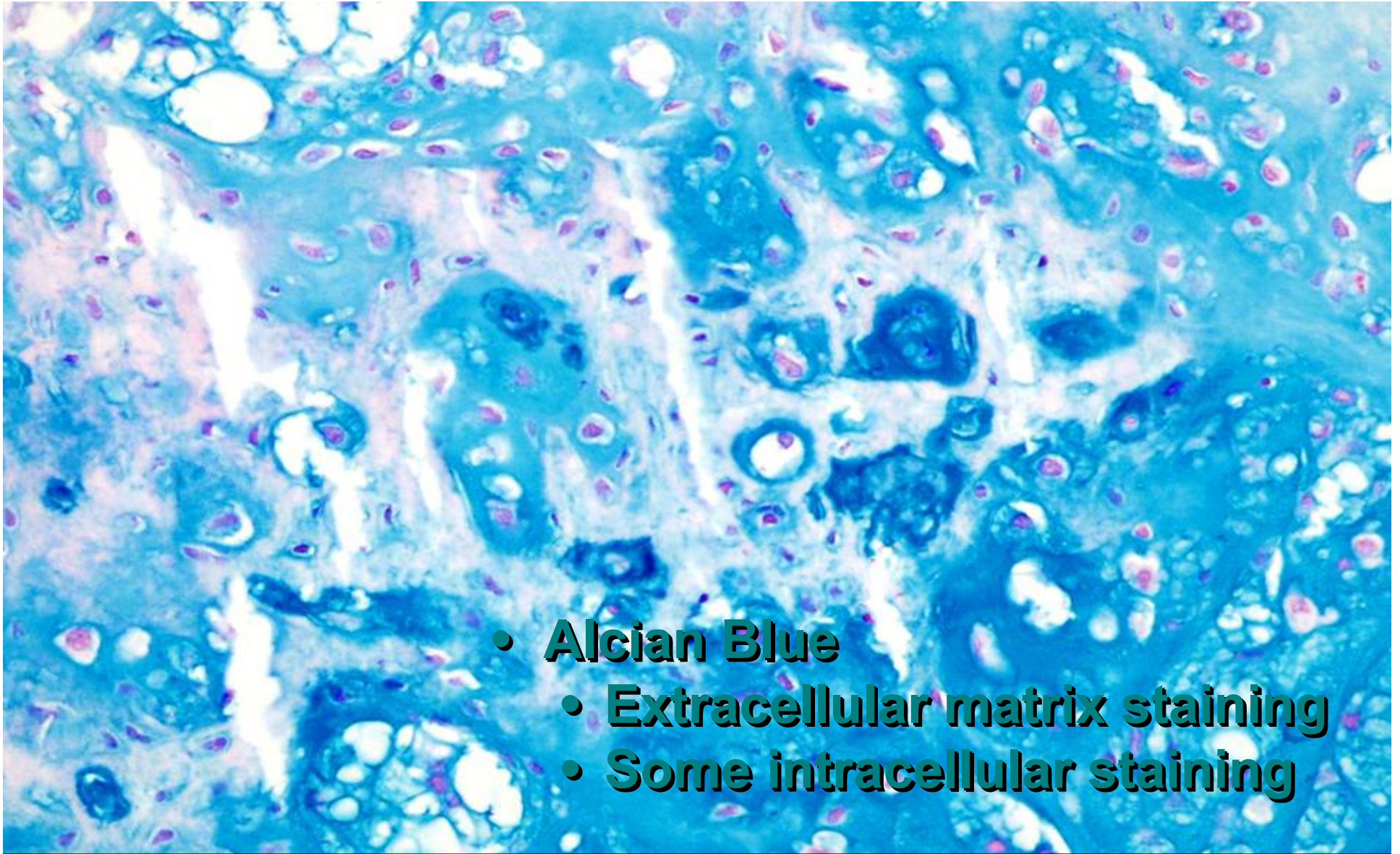


Notochord: Defining Characteristics

- **Early notochordal development**
 - **Epithelial morphologic characteristics**
 - **High levels of pancytokeratin expression**
 - **Low levels of vimentin expression**
- **Late notochordal development**
 - **Morphologic transformation into chorda reticulum (epithelial → mesenchymal)**
 - **Pancytokeratin expression**
 - **Increased vimentin expression**
- **Variable S-100 protein expression**

Characterization of Chordoma: Notochordal Origin of Chordoma

- **Histochemistry:**
 - **Alcian blue**
 - **PAS (w/diastase digestion)**
- **Immunohistochemistry:**
 - **Cytokeratin AE1/AE3**
 - **Vimentin**
 - **S-100 protein**
 - **Neuron specific enolase**
 - **Osteonectin**
 - **Osteocalcin**
 - **Chromogranin A and B**
 - **Glial fibrillar acidic protein**

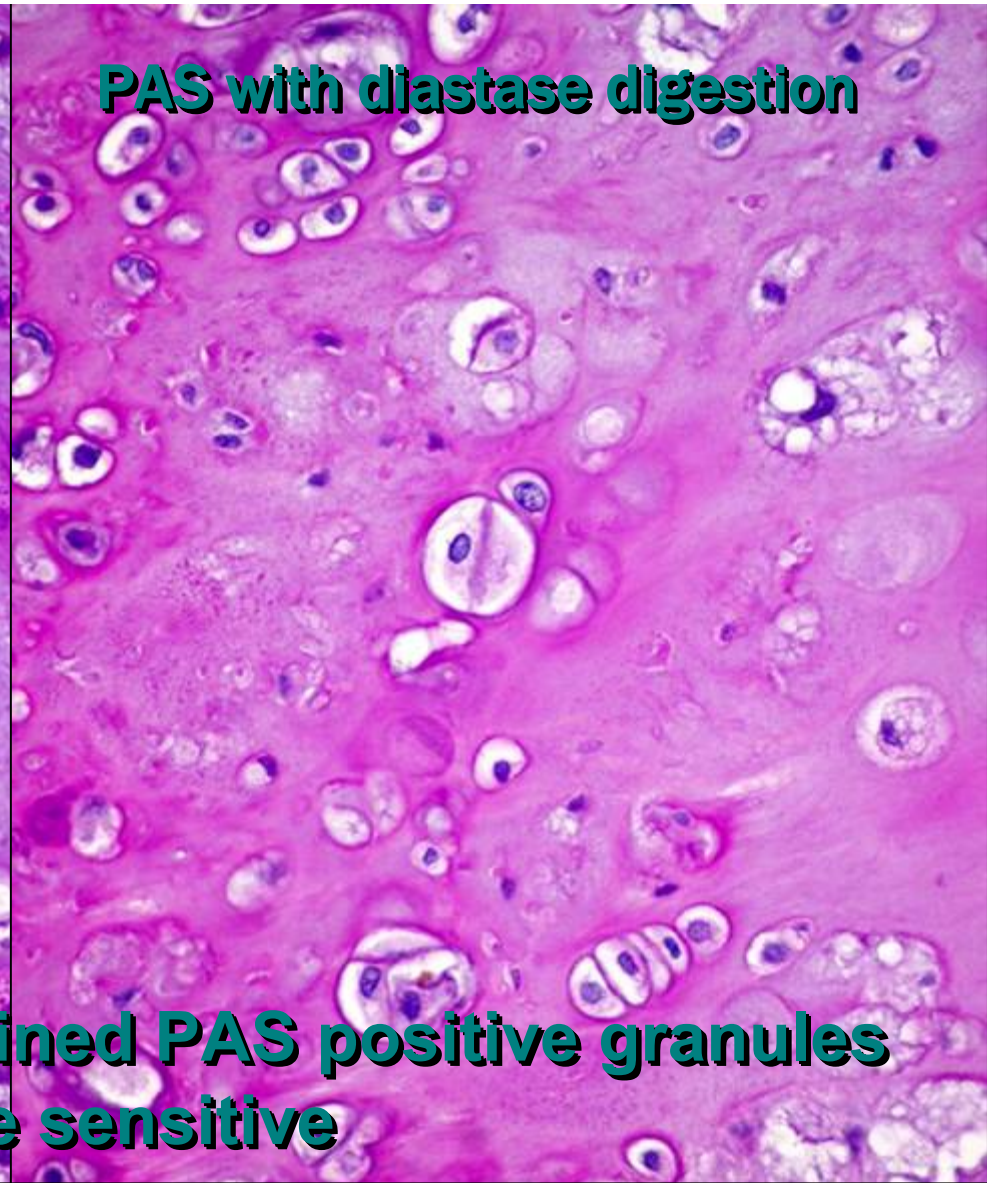
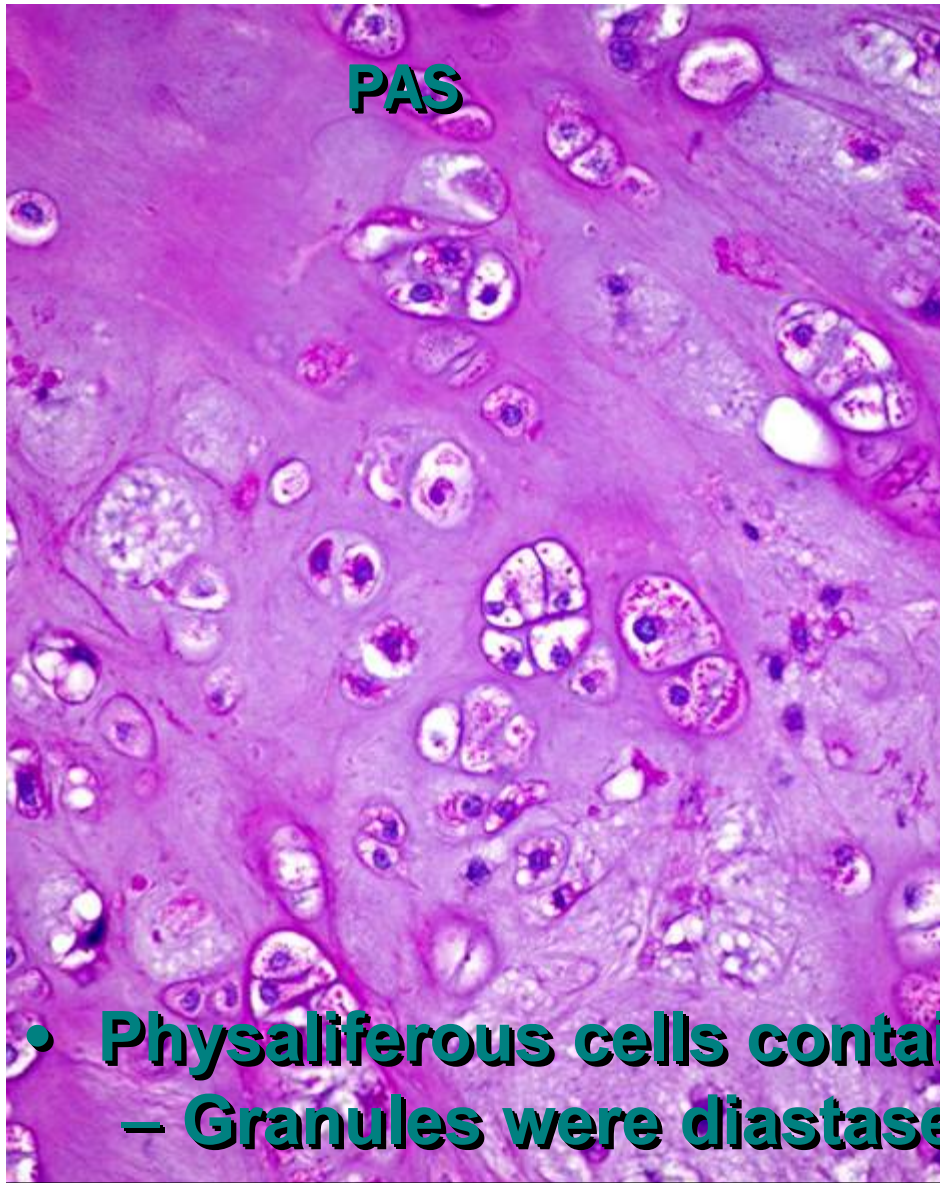


- **Alcian Blue**
 - **Extracellular matrix staining**
 - **Some intracellular staining**



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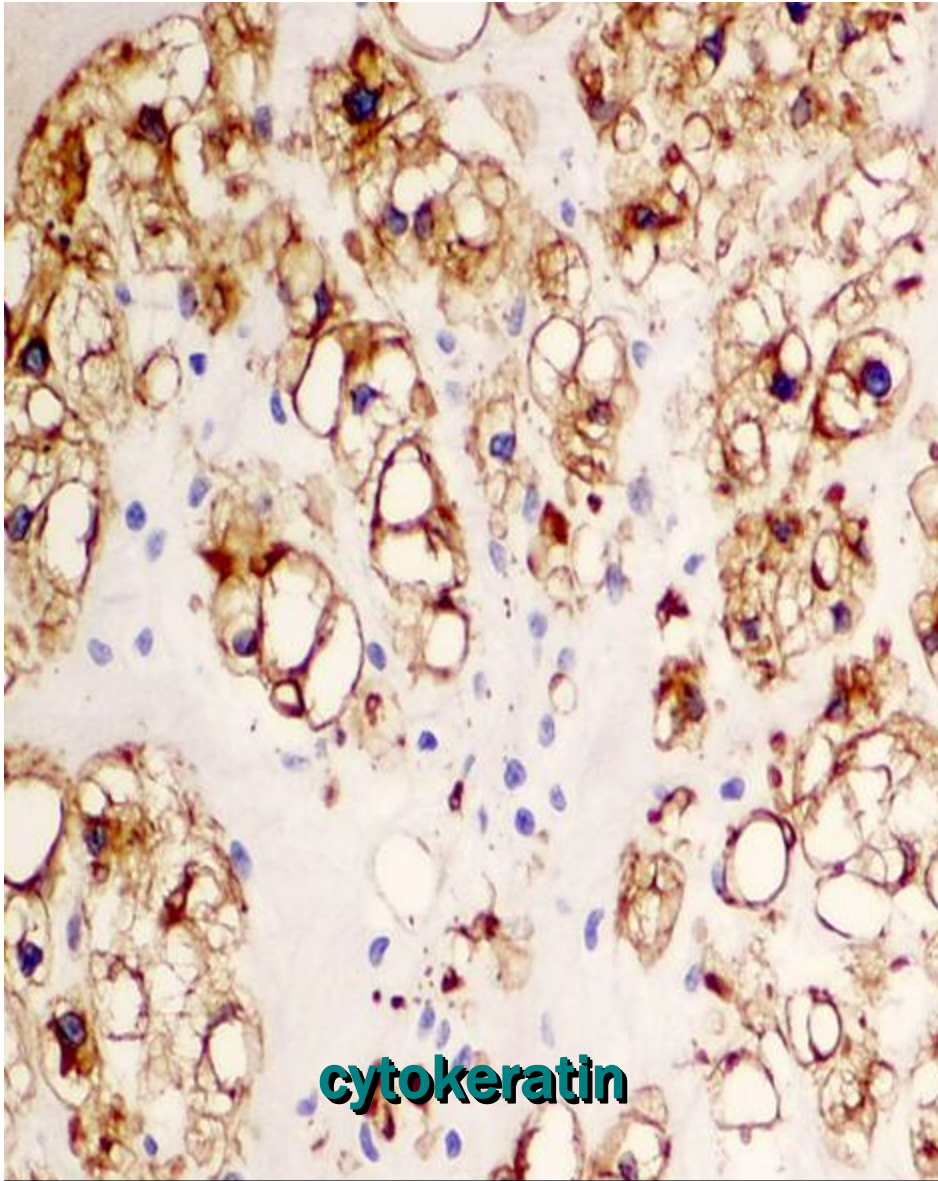


- **Physaliferous cells contained PAS positive granules**
 - **Granules were diastase sensitive**

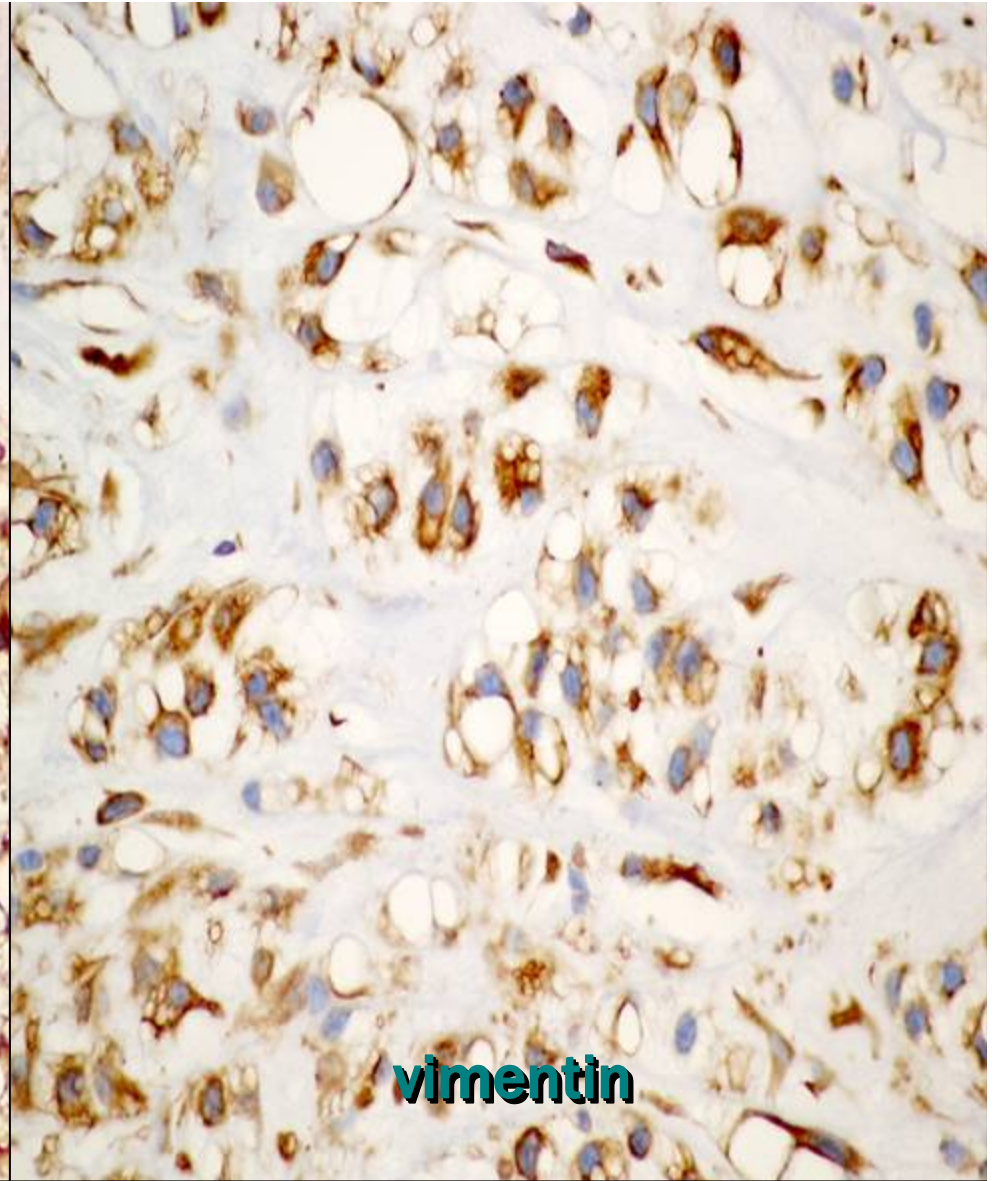


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cytokeratin

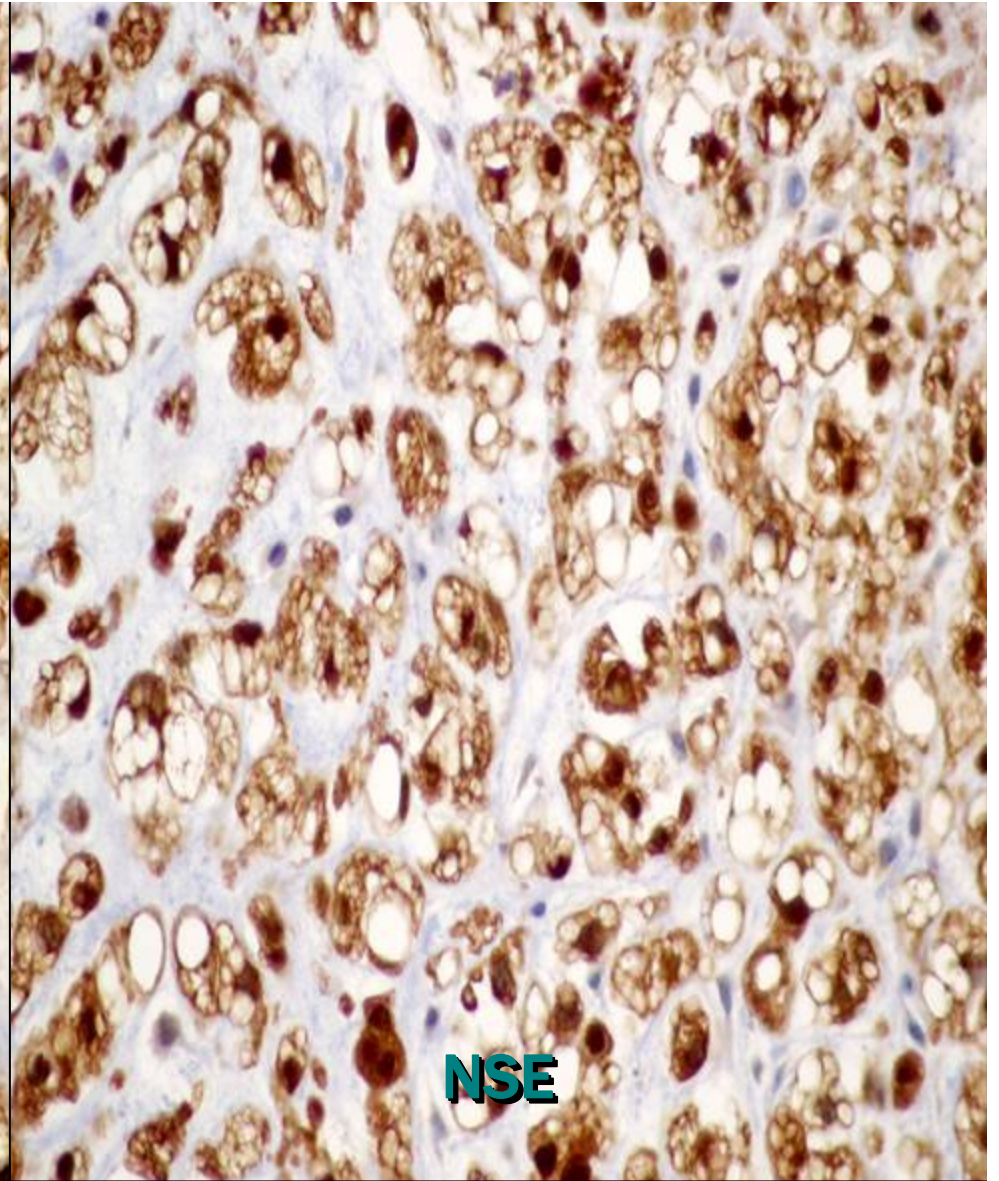
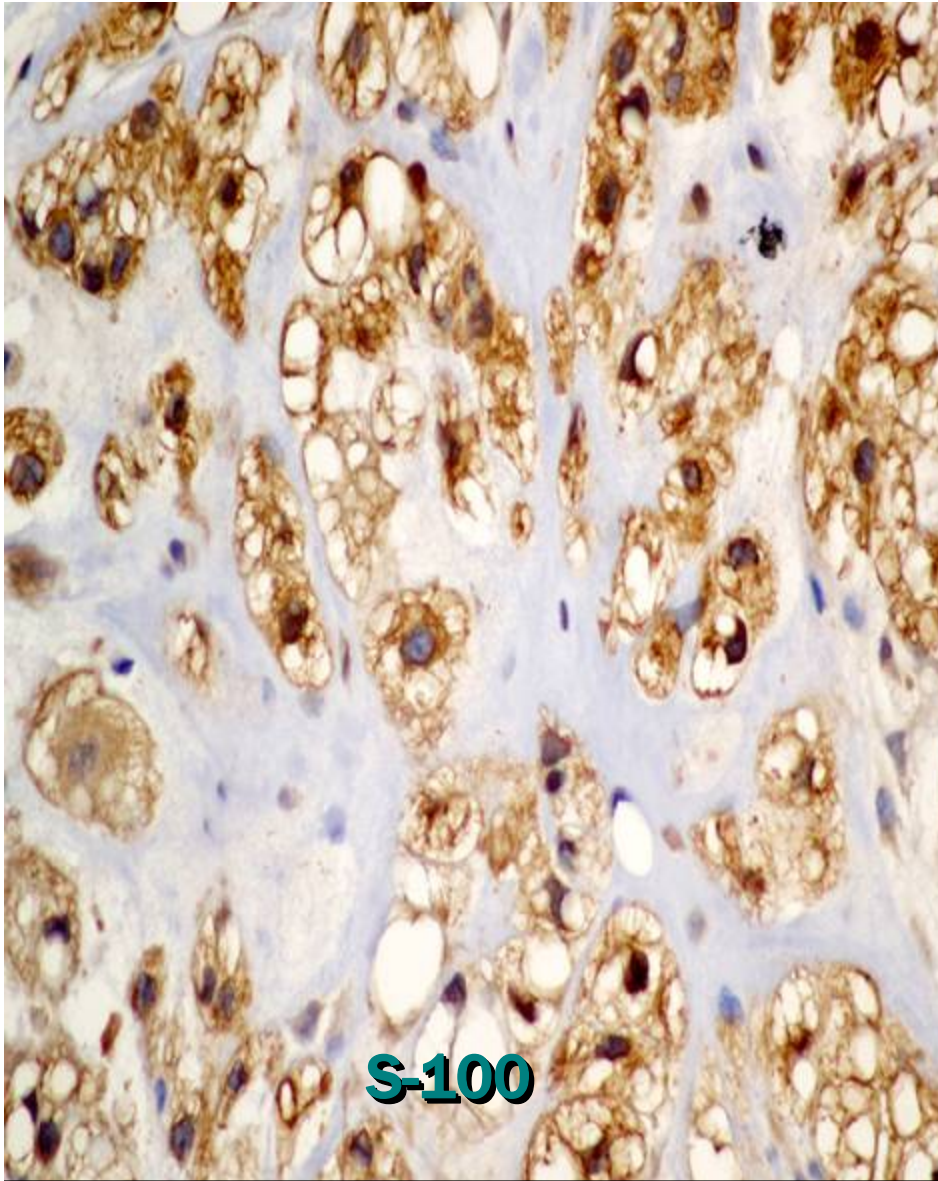


vimentin



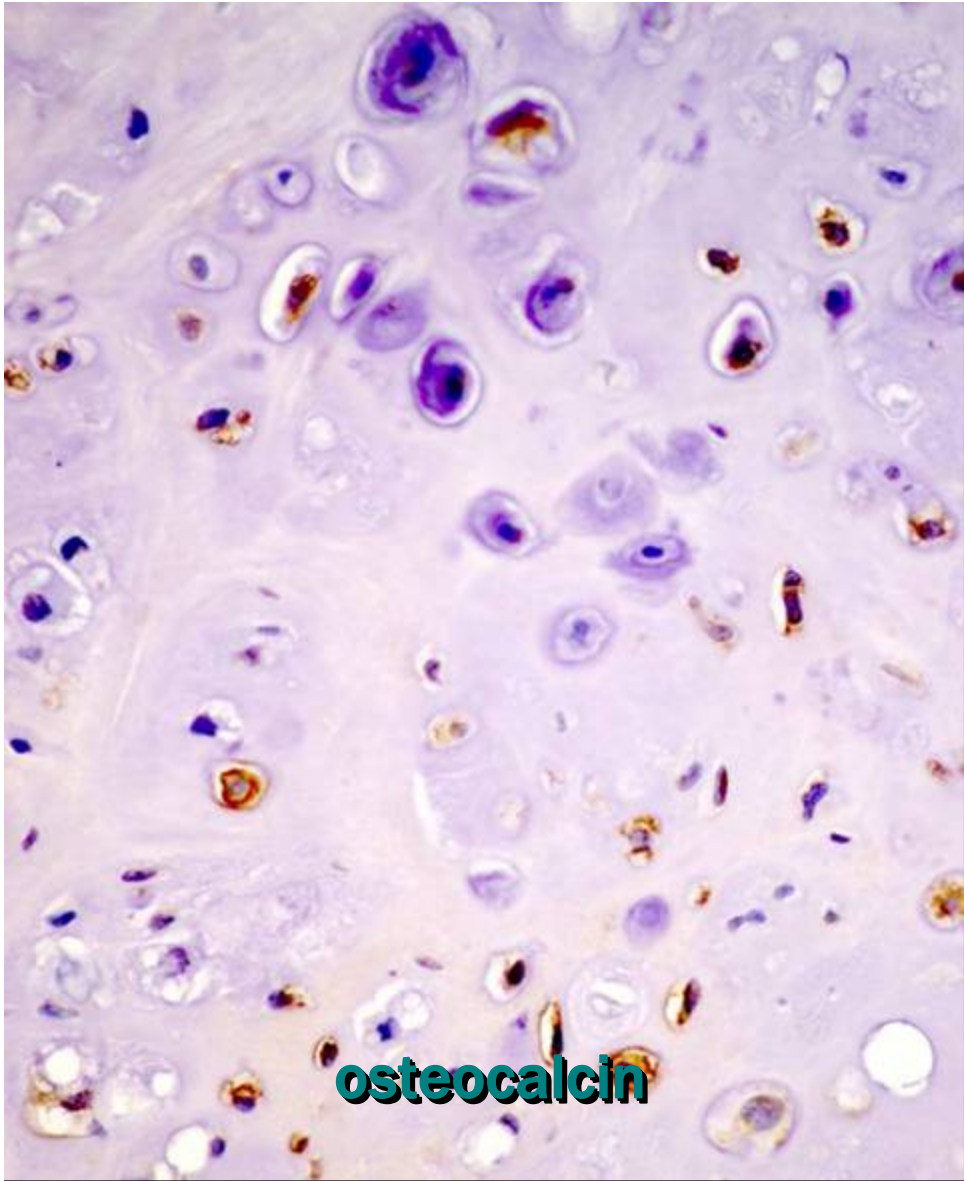
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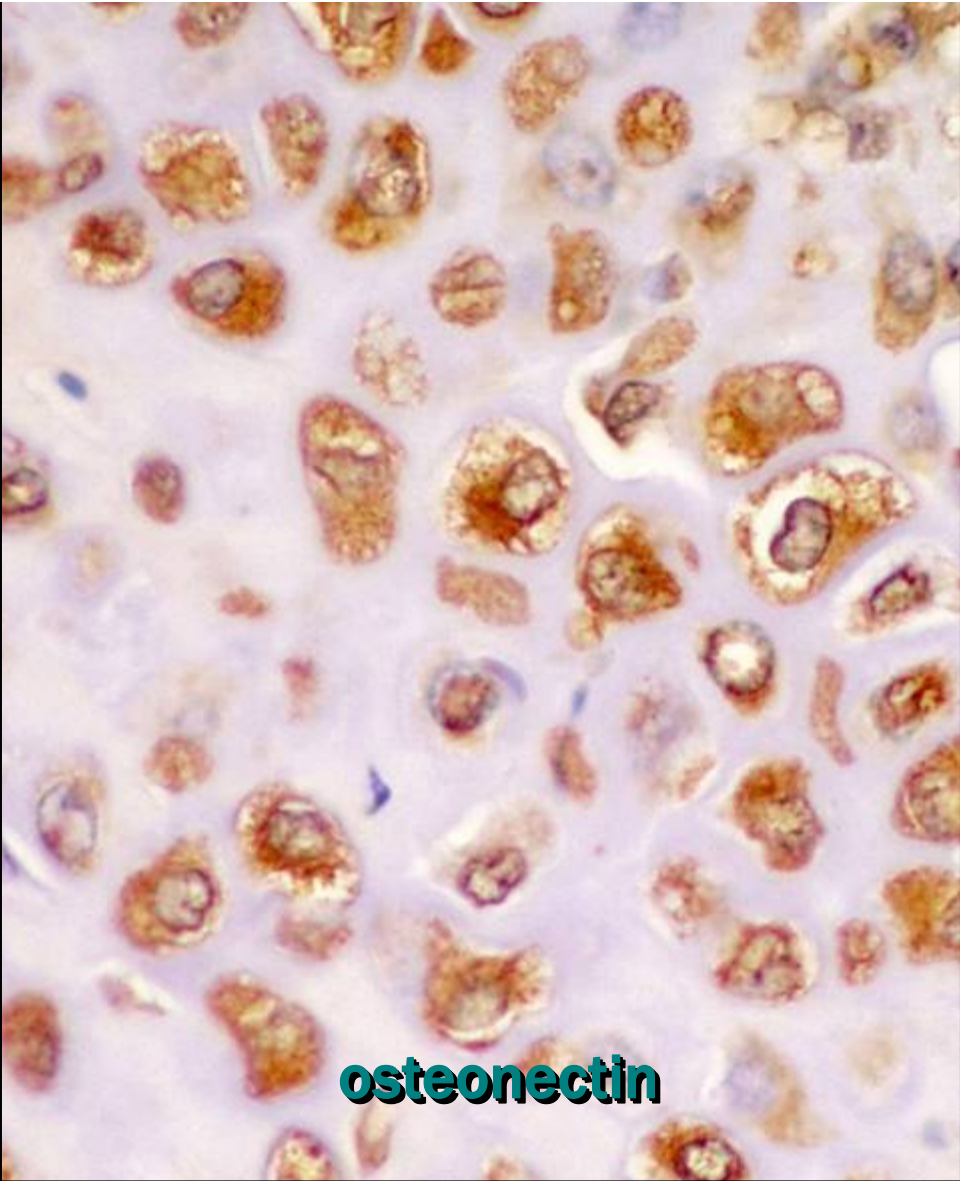


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osteocalcin

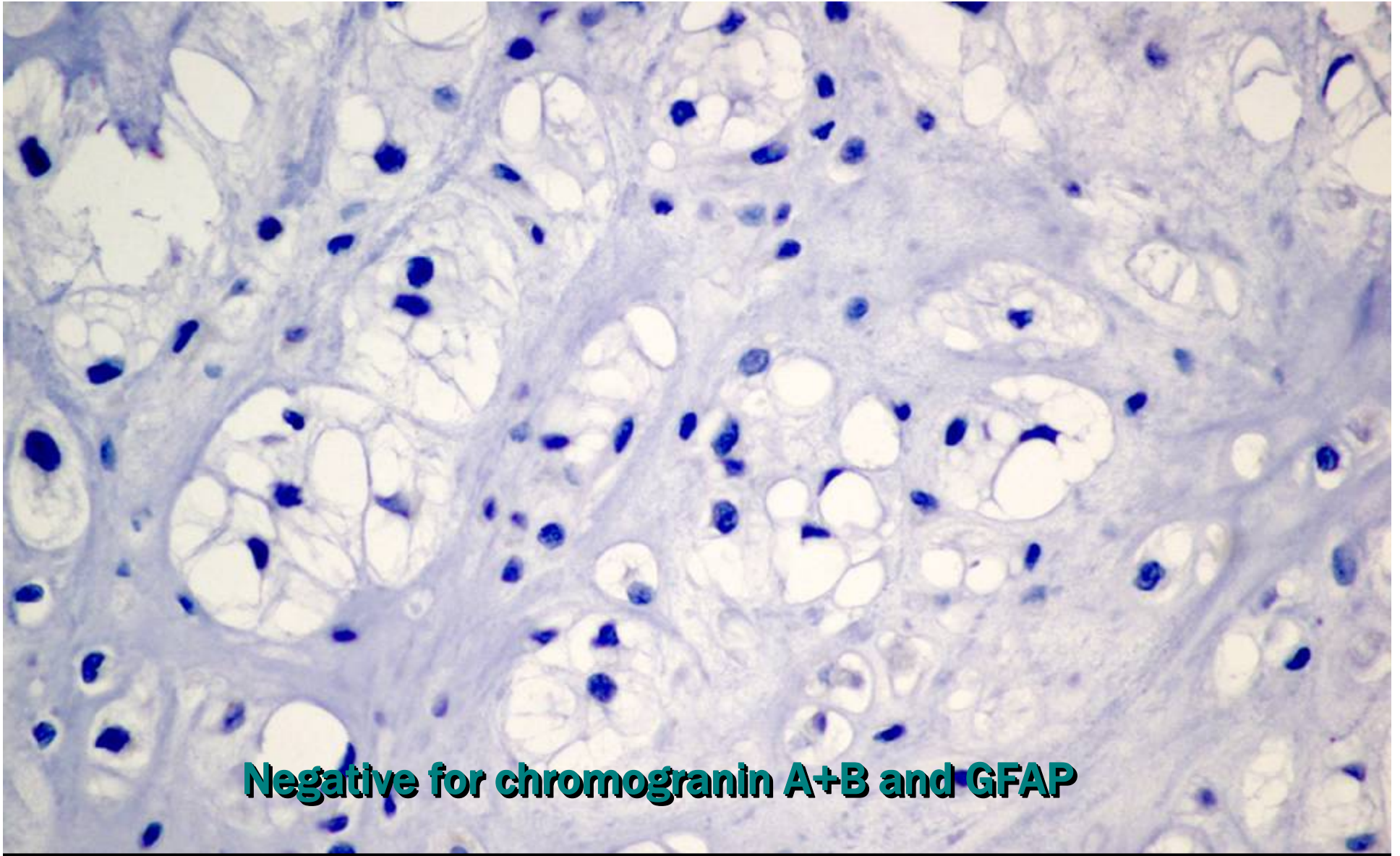


osteonectin



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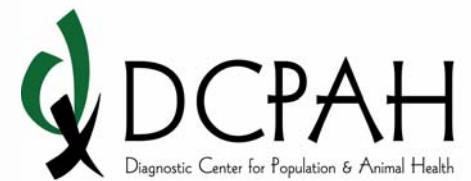
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Negative for chromogranin A+B and GFAP



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Conclusions

- **Supportive evidence of notochord origin of chordoma:**
 - **Axial location of tumors**
 - **Similar morphologic features**
 - **Cytokeratin and vimentin expression**
 - **Presence of epithelial and mesenchymal intermediate filaments**
 - **S-100 protein expression**

Conclusions

- **Additional characteristics of chordomas**
 - **Intra- and extracellular acidic mucopolysaccharides**
 - **Intracellular glycogen accumulation**
 - **Osteocalcin and osteonectin expression**
 - **Osteoid lineage**

References

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