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
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
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:
  - Characterized by physaliferous cells
    - Classical chordoma
      - 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: 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
Physaliferous cells contained PAS positive granules.
- Granules were diastase sensitive.
Negative for chromogranin A+B and GFAP
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

References


