Case #7

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Presentation

• 31-year-old woman
  – Initially presented with worsening neck pain
  – She was seen in an urgent care clinic and treated for muscle spasm
  – The patient had persistent neck pain and headache and was transferred to Rush for further evaluation and diagnosis
Physical exam

- Resistant to any movement of neck
- Tenderness at midline from base of occiput to C5 area
- Labs unremarkable
  - Sodium 139
  - Potassium 3.5
  - Chloride 103
  - CO2 total 26
  - Urea nitrogen 8
  - Creatinine 0.50
  - Calcium 9.3
  - Spot urine phosphorus 37.4
She was found on imaging to have a large calcified clival mass which demonstrated mass effect on the right side of the medulla.
MRI

- 5.0 cm x 3.0-cm x 3.0-cm expansile, destructive mass in the right clivus and along the right anterolateral margin of the foramen magnum
Surgery

• A biopsy was performed
  • Showed a spindle cell lesion with extensive calcification
• Proceeded with resection
Diagnosis?
Differential Diagnosis of calcified lesions of the neural axis

- Astrocytoma
- Cavernous malformation
- Chondrosarcoma
- Chordoma
- Ganglioglioma
- Infection/granuloma
- Meningioma
- Oligodendroglioma
- Osteosarcoma
- Phosphaturic mesenchymal tumor
- Tumoral calcinosis
- Vestibular schwannoma
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- No cartilage or osteoid material
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- No cavernous blood filled spaces
Differential diagnosis

- Neoplastic
  - Chordoma
  - Meningioma

- Reactive / other
  - Infectious granulomatous disease
  - Phosphaturic mesenchymal tumor
  - Tumoral calcinosis
Chordoma

• **Similarities**
  – Epithelioid cells
  – Abundant pale pink to clear cytoplasm, central nuclei

• **Differences**
  – Nests and cords of cells
  – Myxoid matrix
  – S100 and cytokeratin positive

Our case
Meningioma

- **Similarities**
  - Whorled or lobulated architecture
  - Indistinct cell borders

- **Differences**
  - Round to oval nuclei
  - Psammoma bodies
  - S100 positive

*Our case*
Granulomatous Reaction

- **Similarities**
  - Multinucleated histiocytes
- **Differences**
  - Rim of lymphocytes
  - Necrosis
  - Giant cells are localized

Our case
Phosphaturic Mesenchymal Tumor

• Similarities
  – Spindle cell lesion
  – Calcification
  – Giant cells

• Differences
  – Osteomalacia, phosphaturia and hypophosphatemia
Tumoral calcinosis

- **Similarities**
  - Large calcified areas
  - Multinucleated giant cells
- **Differences**
  - Usually a skin lesion
  - Usually familial and associated with hyperphosphatemia

AFIP atlas of nonneoplastic Disorders of Bones and Joints

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Immunohistochemistry

• Negative
  – S100
  – Desmin
  – Pan-keratin
  – EMA
  – GFAP

• Positive
  – SMA in reactive stromal myofibroblasts
  – CD163 highlights histiocytes
Calcifying pseudoneoplasm of the neural axis (CAPNON)
Calcifying pseudoneoplasm

Epidemiology

• Very rare
  – About 32 cases reported worldwide
• First described by Rhodes and Davis in 1978
• Found along the neural axis
  – Anywhere from L2 to frontal lobe
• Age range is highly variable
  – 6-83 years
  – Mean age 46.2
• Clinical symptoms
  – Most commonly pain
  – Other compressive symptoms possible
Radiology

- Calcification
- Hypointense appearance on T1 and T2 weighted images without gadolinium enhancement
- Minimal linear enhancement or partial rim enhancement
- Often showing destruction or erosion of adjacent bone
- Radiologic differential is broad and nonspecific

Histology

- “Classic” histology
  - Calcification
  - Palisading spindle to epithelioid cells
  - Fibrous stroma
  - Foreign body giant cells
- Can also show
  - Chondromyxoid matrix in a nodular pattern
  - Osseous metaplasia or psammoma bodies
- These patterns are variable and not always present

• Hydroxyapatite
  – $\text{Ca}_5(\text{PO}_4)_3(\text{OH})$

• Calcium Pyrophosphate
  – $(\text{Ca}_2\text{O}_7\text{P}_2)$
Etiology

- Originally though to be an unusual expression of tumoral calcinosis
- Suggested to be a healing response
  - Trauma
  - Infection
  - Inflammation
- Possible tissue of origin includes arachnoid or fibroblasts in the choroid plexus stroma
- Calcifying pseudoneoplasm of the neural axis
  - Vs Calcifying pseudotumor of the neural axis
Treatment

• Surgical resection
  – No chemotherapy or radiation

• Benign course is typical
  – One case has shown recurrence
  – 2 fatalities reported due to CAPNON, due to complications related to anatomic location
Follow up

• Patient is 6 months post op with no recurrence
References


- Special thanks to Dr. Unni