RADIOLOGY

Interesting MSK Case

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History

***CC**: lower back pain*

***HPI**: 12 y.o. female presents to her PCP with persistent, worsening lower back / sacral pain 2 months after a fall. Pain is worse while sitting, at the end of the day, and when more active. She stands while eating dinner and frequently adjusts while sitting at school. No numbness, tingling, or difficulty with voiding or BMs.*

History

PMHx: *Acne vulgaris, eczema herpeticum, seasonal allergies, ADHD*

PSHx: *None*

Med: *Doxycycline, Valtrex, Cetirizine, Astaris*

All: *Egg, seafood, peanuts*

SHx: *Active. Great student. Adopted from Serbia at 5 y.o., Bosnian ancestry. Before 5, malnourished, dental issues, and heavy tobacco exposure*

Fam Hx: *Unknown*

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Physical Exam

- 85th percentile weight
- 92nd percentile height
- **MSK**: Firm mass in gluteal cleft, fixed and slightly tender
- **Neuro:** Unremarkable

→ XR Spine Sacrum Coccyx

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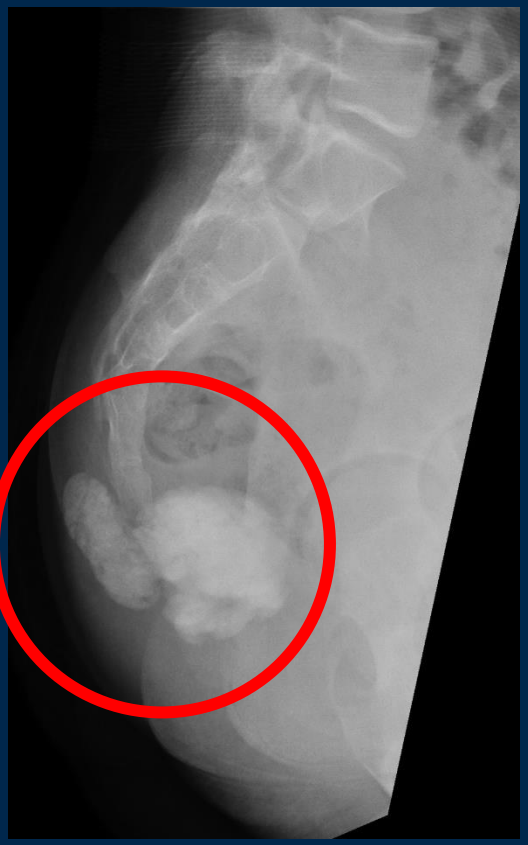
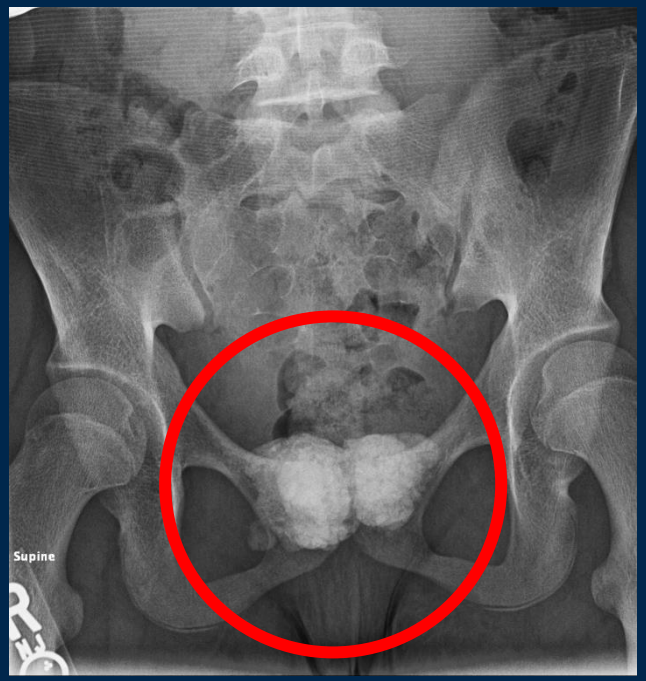
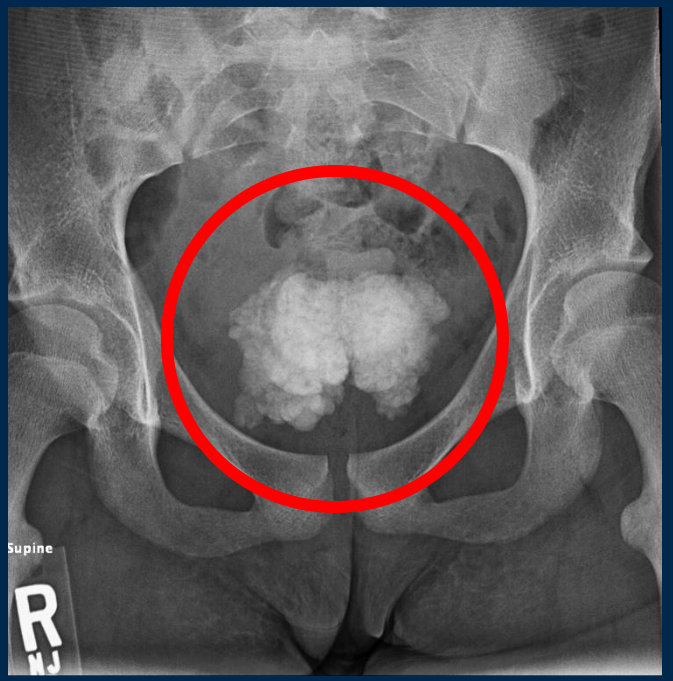
Radiography







→ CT Abdomen/Pelvis

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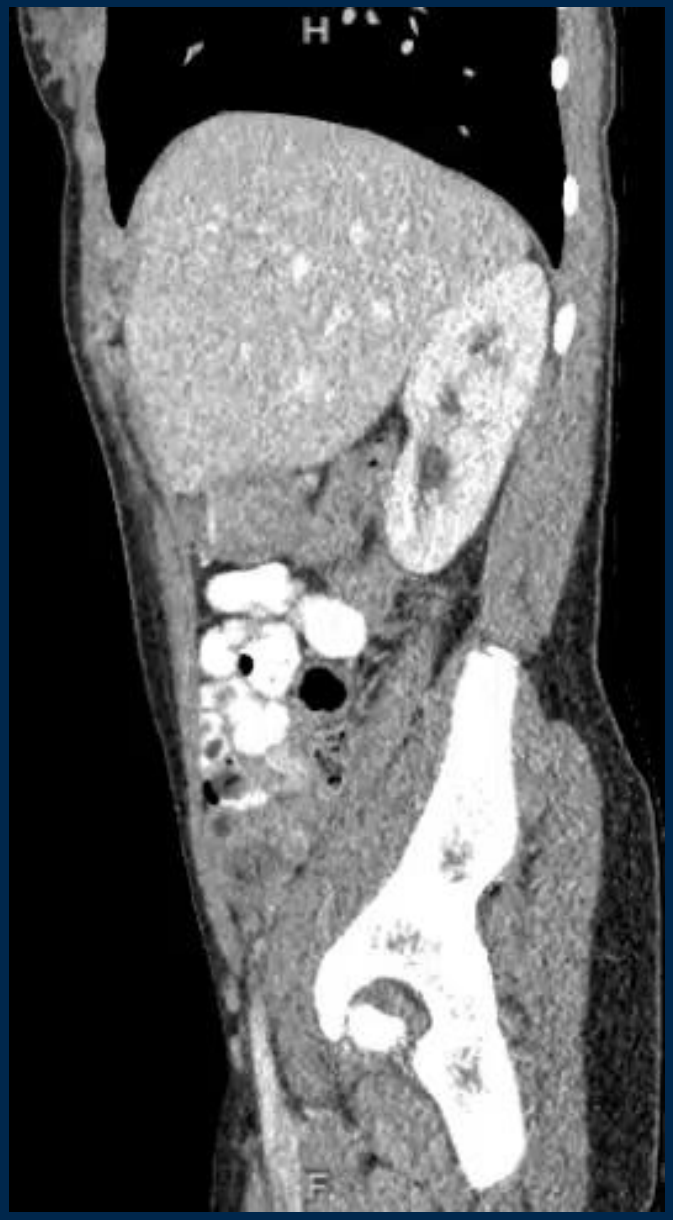
CT Abdomen/Pelvis with Contrast



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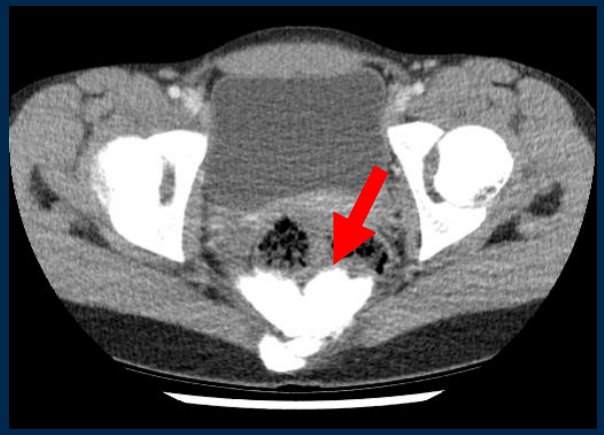
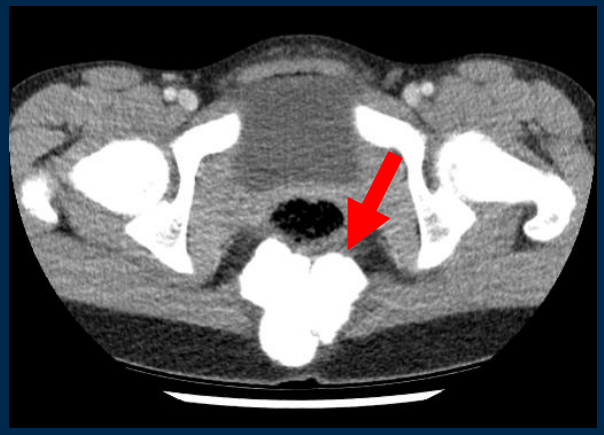
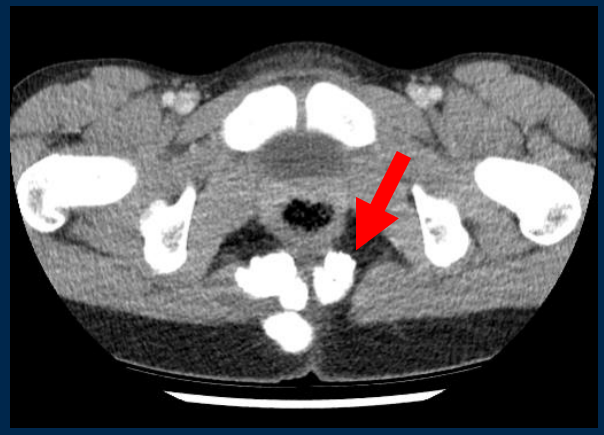
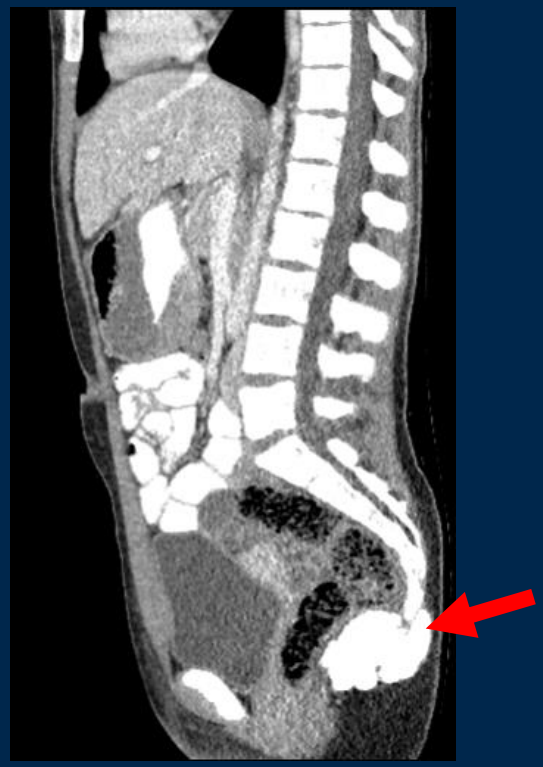
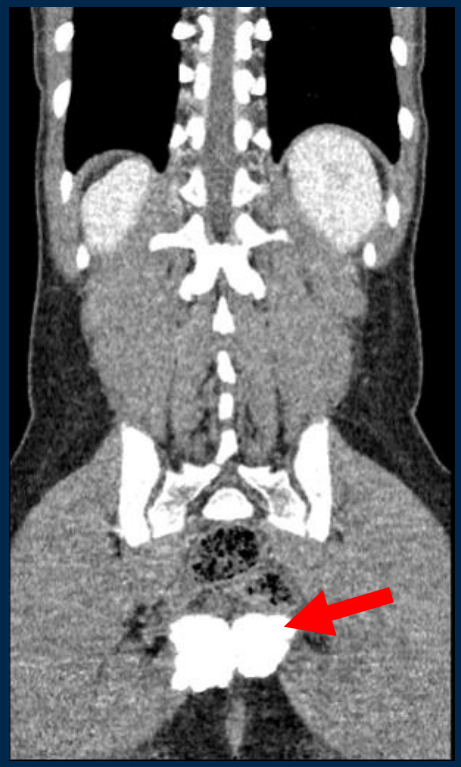
CT Abdomen/Pelvis with Contrast

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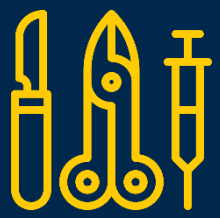
CT Findings

- Osseous, lobulated, calcified mass of the coccyx
- Mass size: 6.6 x 6.4 x 4.5 cm (L x AP x T)
- No evidence of fracture or trauma
- No definite fat or soft tissue component
- May reflect sequelae of sacrococcygeal teratoma, bone tumor with exuberant production of osseous matrix, or tumoral calcinosis

Multidisciplinary Work-Up & Diagnosis

- Pertinent labs
  - Phosphate mildly elevated
  - 1,25 $(OH)_2$ Vit D mildly elevated
  - Calcium, PTH, rheum labs, other tumor markers and labs unremarkable
- IR Biopsy → tumoral calcinosis. no malignancy



- PET: increased uptake sacrum-coccyx, lungs
  - Lungs → infectious
  - No other apparent calcinosis
- Added history: pain onset 1.5 years prior

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Tumoral Calcinosis (TC)

What is it?

- Rare, benign condition
- Calcium and phosphate deposition in soft tissues
- Often periarticular, or around joints or capsules

What causes it?

Genetics / Familial Inheritance

- Hyperphosphatemic
- Normophosphatemic

Secondary Causes

- Renal Failure
- 2^o or 3^o Hyperparathyroidism
- Rheumatologic

Idiopathic

Triggered by trauma?

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Tumoral Calcinosis

Presentation:

Most frequently with African or Middle Eastern descent

Most often in 1st/2nd decade of life

Reduced ROM, +/- pain

One lesion or many, progressively enlarge

+/- corneal calcification, dental abnormalities

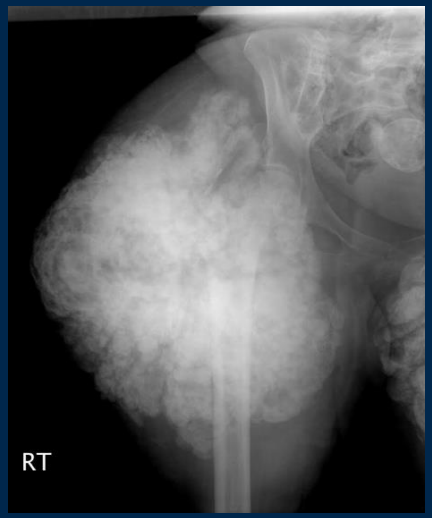
Locations:

Most common locations: hip, elbow, shoulder, foot, wrist

Case reports in TMJ, scalp, larynx, spine, sacrum, hand, knee

~7% cases involve spine → more likely to be painful

Coccyx is rare!



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Classic Imaging Findings:

Lobulated, heterogenous, well-demarcated calcification

Most often along extensor surfaces and periarticular

No erosion or osseous destruction

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Tumoral Calcinosis Diagnosis & Treatment

Role of imaging:

- Classic XR, CT, and MRI findings
- Help characterize concern for malignancy and need for surgery

Labs/Pathology:

- Phosphate, Vit D, Ca2+, PTH, GFR, rheum
- Characterize genetic concern
- Biopsy if uncertainty

Genetics Consultation:

- If concern for familial etiology

Treatment for Familial TC:

- Medical → low phos diet, phos binders
- +/- Surgical; recurrence possible

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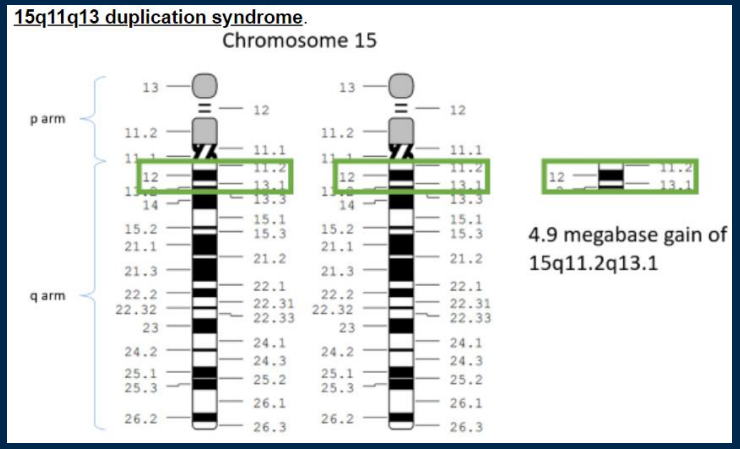
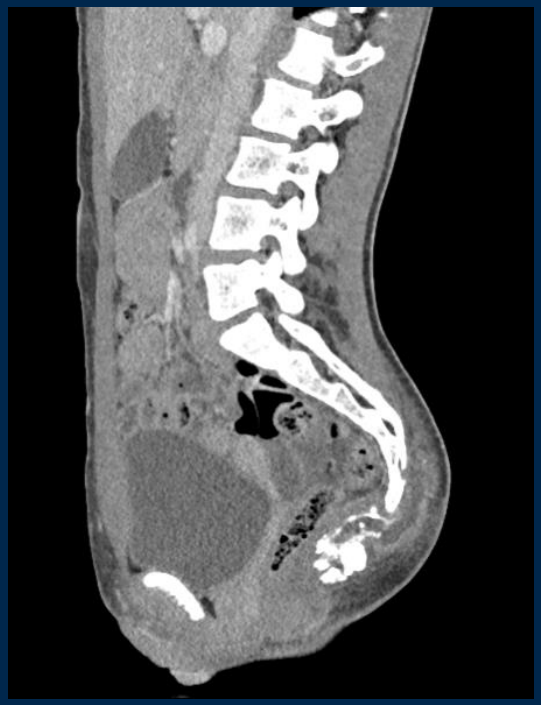
Back to the Case

- Genetics consult
  - Germline proximal chromosome 15 duplication (Prader-Willi and Angelman Syndromes region)
  - NOT thought to be cause of TC
  - Whole genome sequencing pending
- Medical management → lab abnormalities resolved
- Worsening pain, leg weakness, mass effect symptoms → surgical debulking





Post-op fluid collection and residual calcinosis

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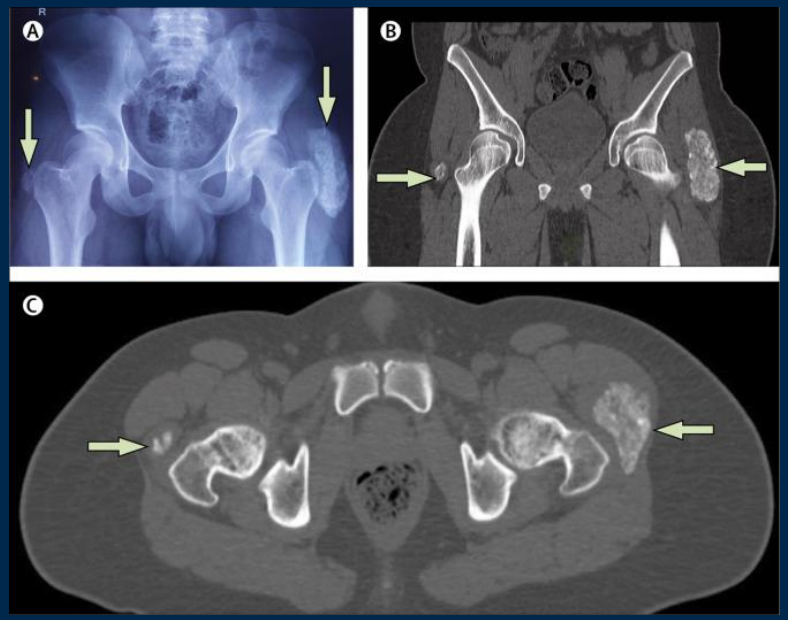
Tumoral Calcinosis Case Reports



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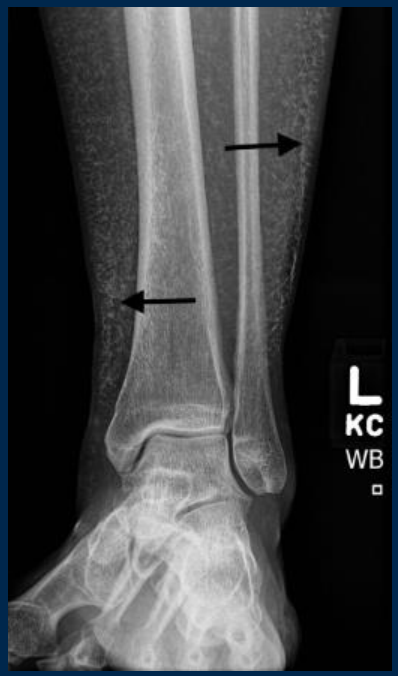

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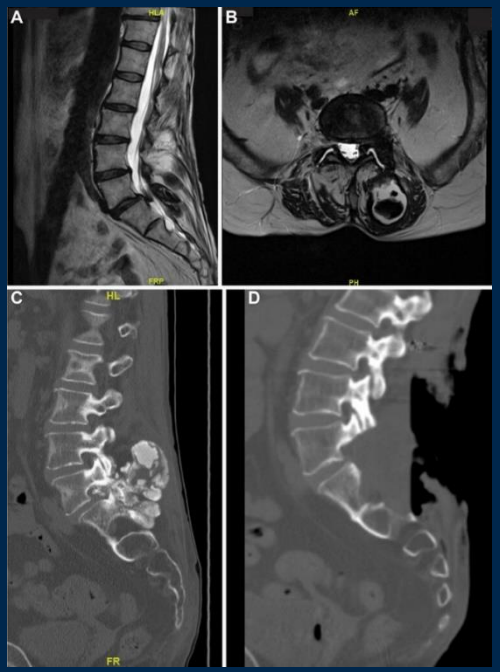

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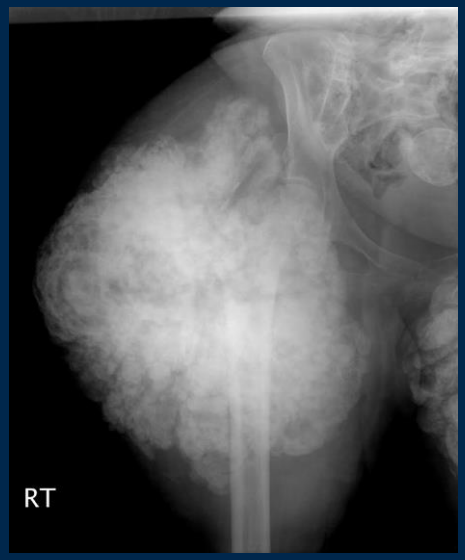

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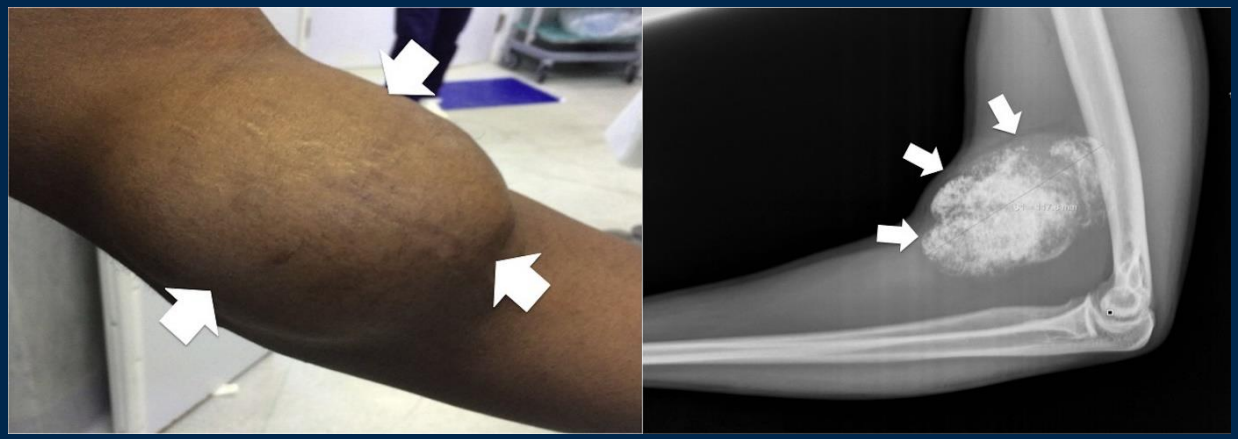

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Take Home Points

1. Radiologists should consider tumoral calcinosis in masses with lobulated, heterogenous calcifications despite abnormal location.
2. The diagnosis of hyperphosphatemic familial tumoral calcinosis is multimodal and includes clinical presentation, radiographic findings, lab work, pathology, and genetic findings.
3. Tumoral calcinosis is a rare diagnosis with typical management based on case-reports and case series. Incidence and prevalence is unknown.

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Thank you!

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