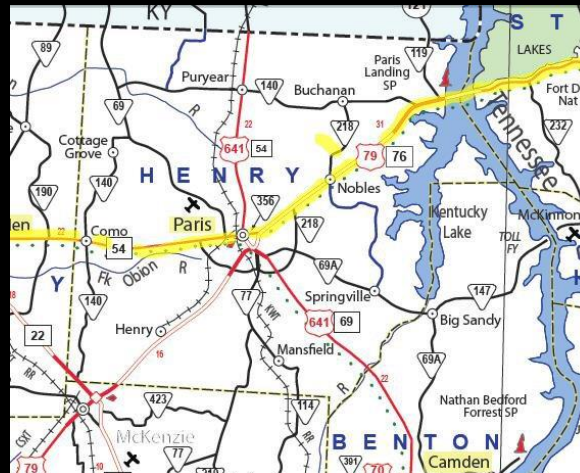




Middle aged female with leg pain.



Chris Bruno
October 28, 2016



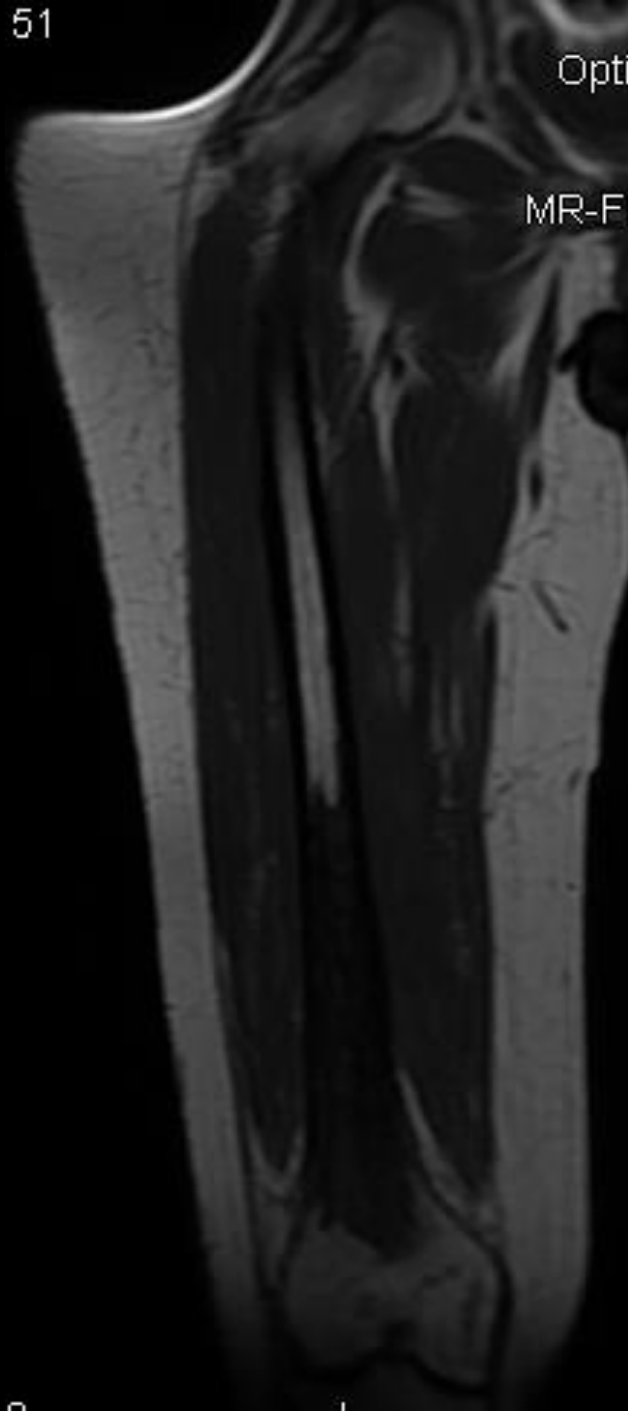
51

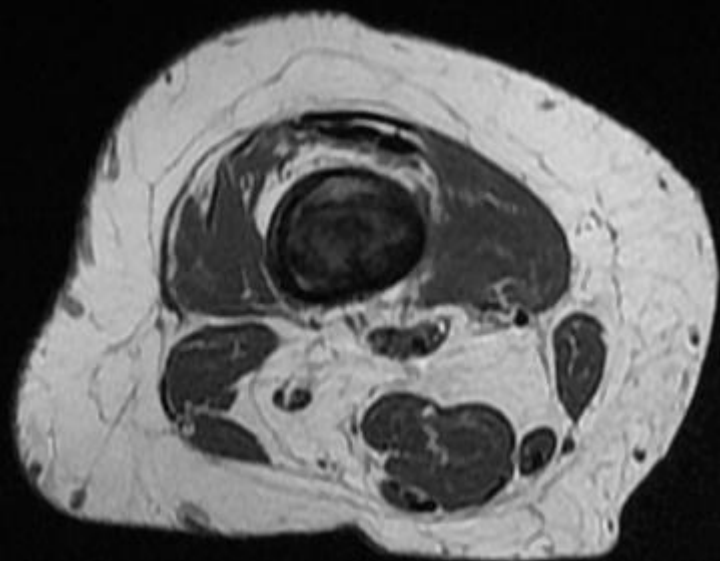
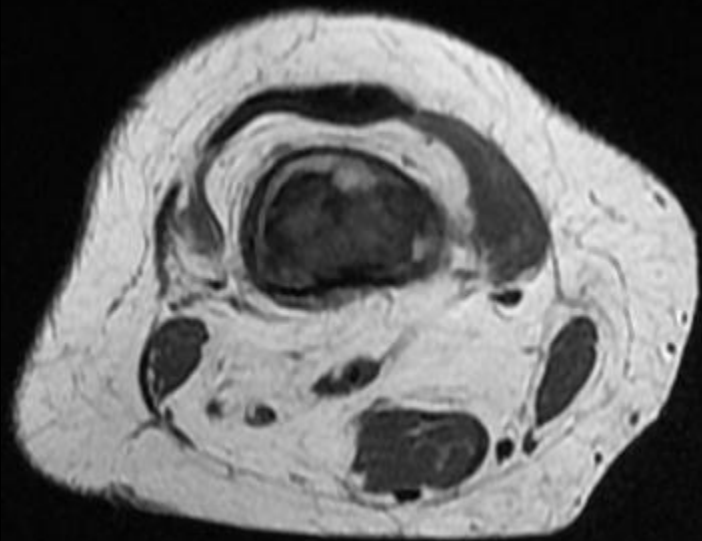
Opti

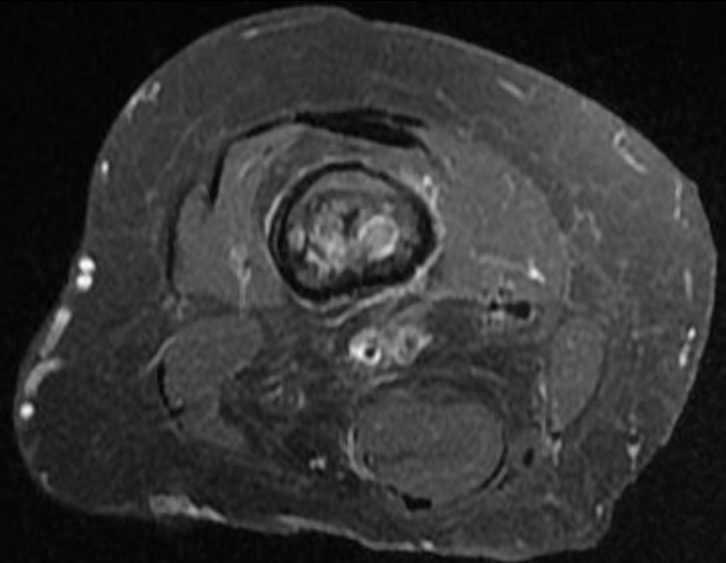
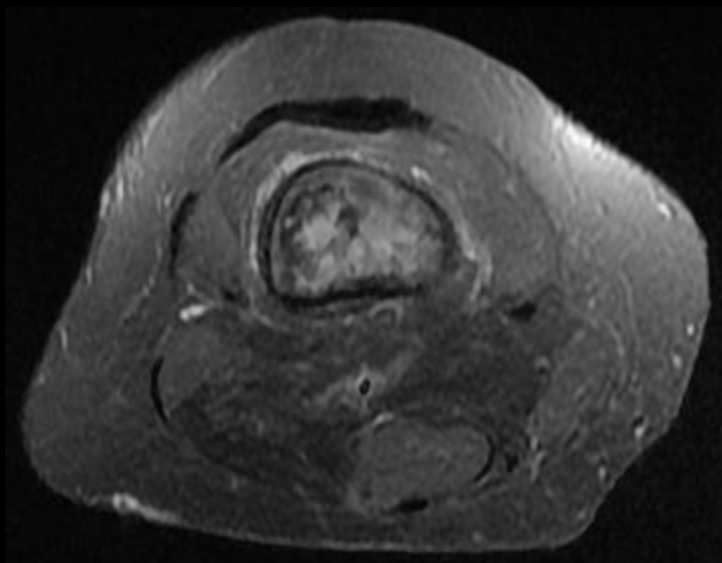
MR-FI

8

1





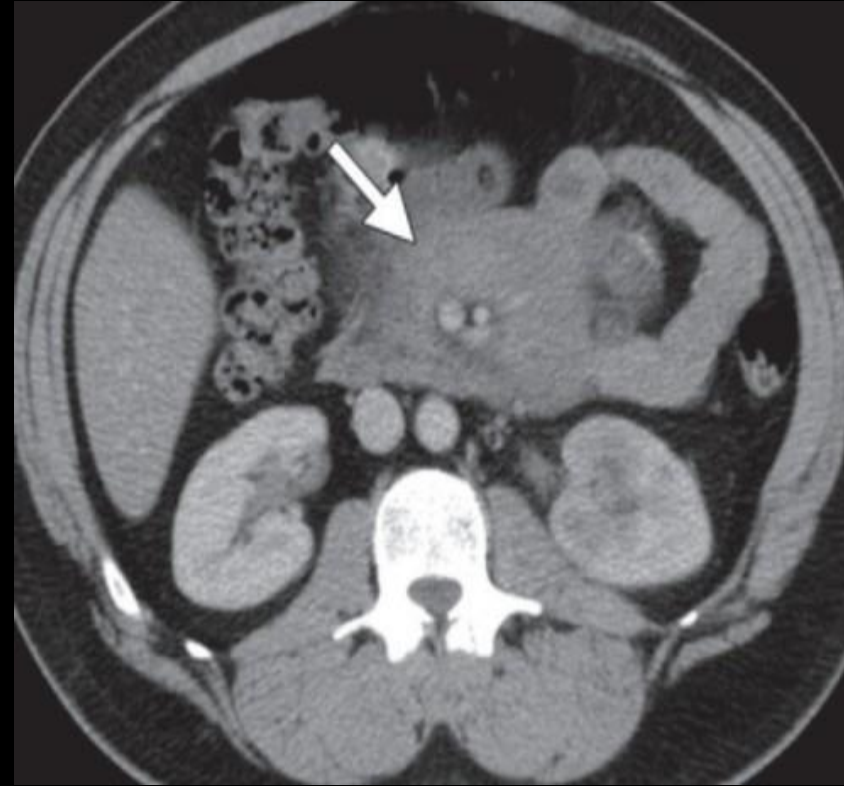


Erdheim-Chester Disease (ECD)

- rare form of non Langerhans dendritic cell histiocytosis
- unknown etiology
- primarily affects adults between 5-7th decade of life; slight male predominance
- most common symptom is bone pain; other general symptoms include: fever, weakness, weight loss, night sweats, fatigue
- classic triad: bone pain, exophthalmos, diabetes

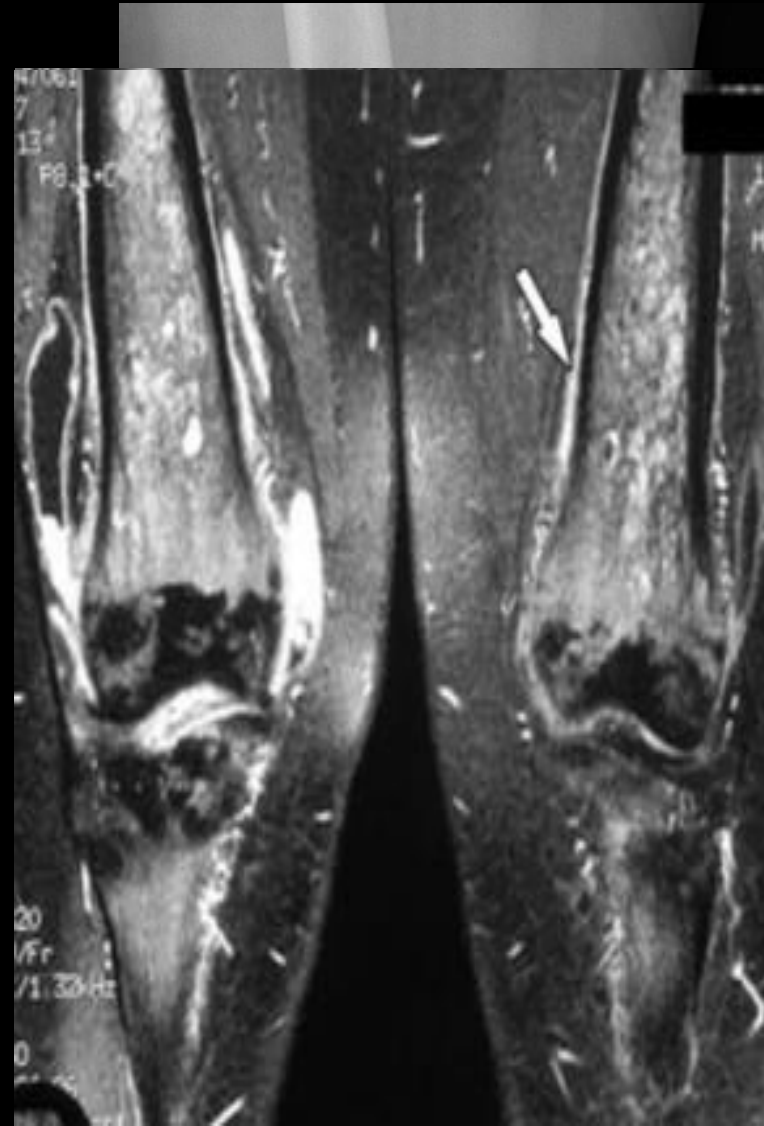
ECD: Extraosseous Involvement

- CNS
 - pituitary defects (**diabetes insipidus**, panhypopituitarism)
 - retro-orbital lesions (**exophthalmos**)
 - cerebellar lesions (ataxia)
 - extra-axial masses
- Cardiovascular
 - pericardial infiltration/effusion
 - periaortic fibrosis
- Pulmonary
 - interstitial lung disease
- Renal/Retroperitoneal
 - perirenal infiltration (may cause renal obstruction or renal artery stenosis)
 - soft tissue thickening around abdominal aorta



ECD: Bone Involvement

- Radiographs
 - bilateral symmetric patchy or diffuse intramedullary sclerosis
 - cortical thickening: periostitis, endosteal thickening
 - relative sparing of axial skeleton and epiphyses, may have metaepiphyseal lucent line
- Scintigraphy
 - bilateral, symmetric increased uptake in tubular bones
- CT
 - help confirm cortical thickening and sclerosis of the cancellous bone
- MR
 - replacement of the normal bone marrow of the diaphyseal and metaphyseal bone segments
 - T1W: heterogenous low signal compared to muscle
 - T2W FS: heterogenous intermediate to high signal compared to muscle
 - T1W FS C+: heterogenous, intense enhancement



ECD: Diagnosis & Treatment

- clinical
- diagnostic imaging
- histology
 - non Langerhans cell histiocytes with foamy or eosinophilic cytoplasm
 - lack of Birbeck granules
 - CD68 (+), CD1a (-), S100 (-/low)
- surgical debulking
- interferon- α
- chemotherapy
- corticosteroids
- bisphosphonates
- anti IL-1, anti IL-6, anti TNF α

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