

SAMMC

SAN ANTONIO MILITARY MEDICAL CENTER



Society of Skeletal Radiology Case of the Day

03/28/2020 @ 1200

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Disclosure Statements

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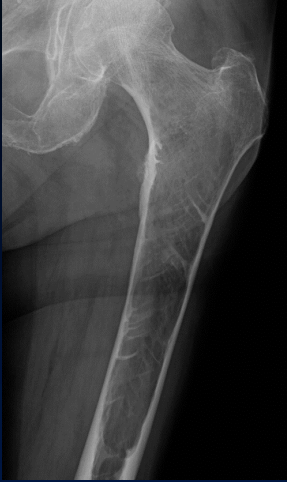
History

- 84 year old with bilateral hip pain

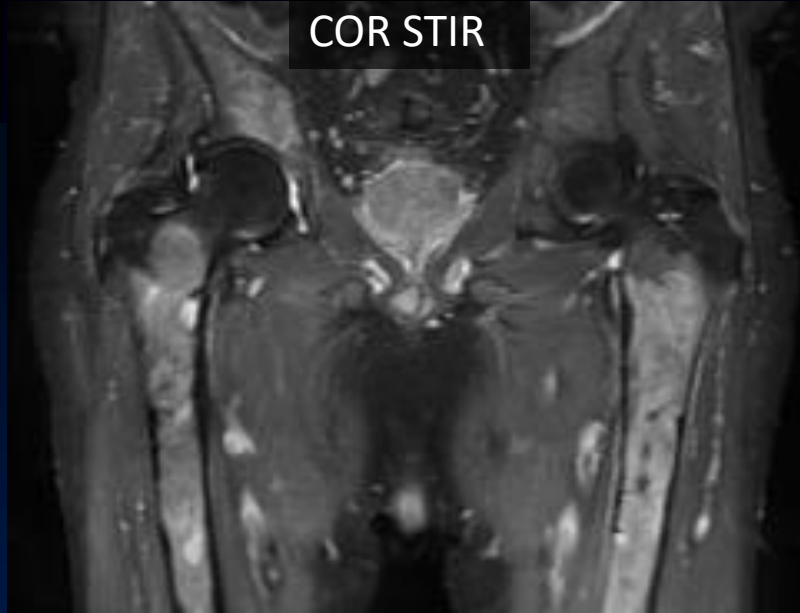
AP Radiograph
Right hip



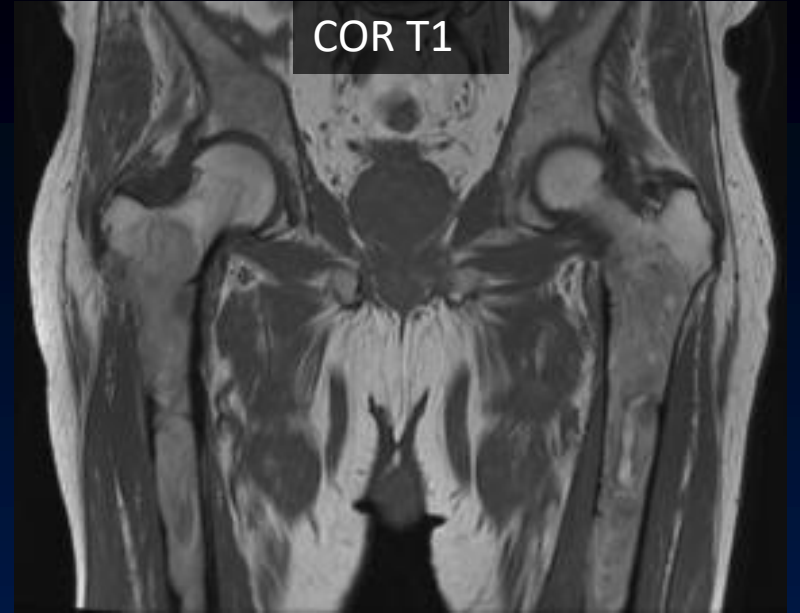
AP Radiograph
Left hip



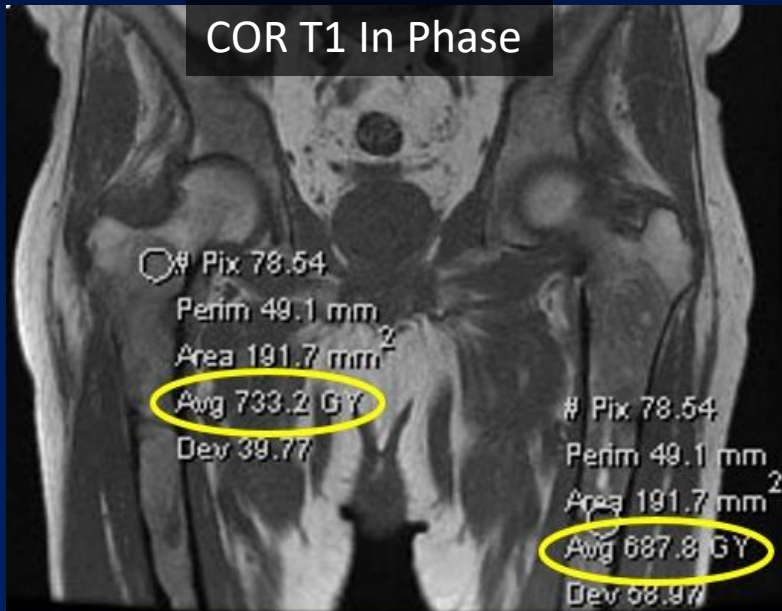
COR STIR



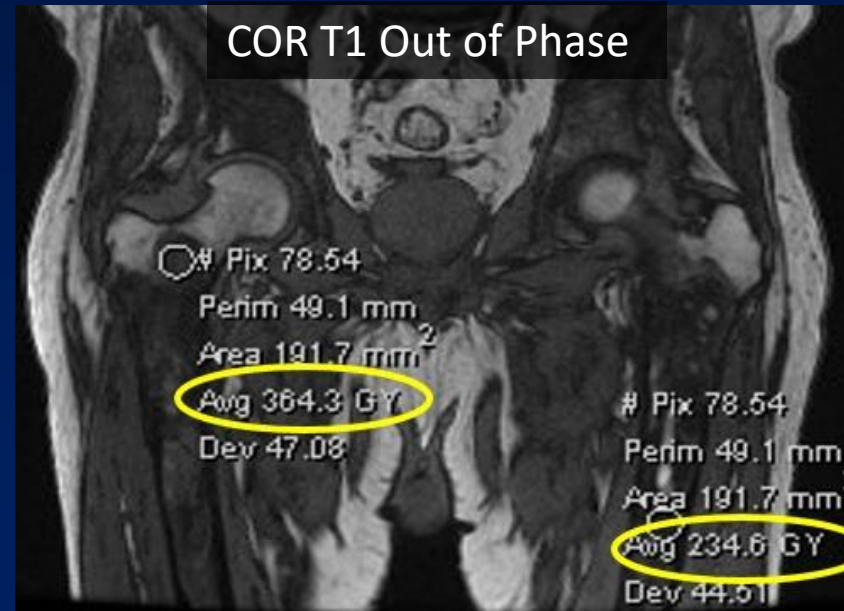
COR T1



COR T1 In Phase



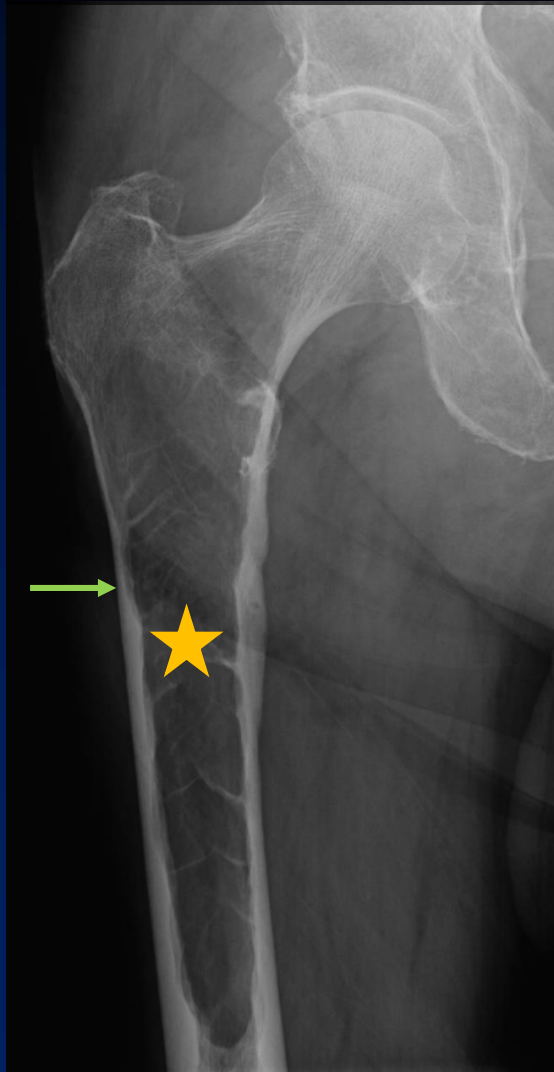
COR T1 Out of Phase



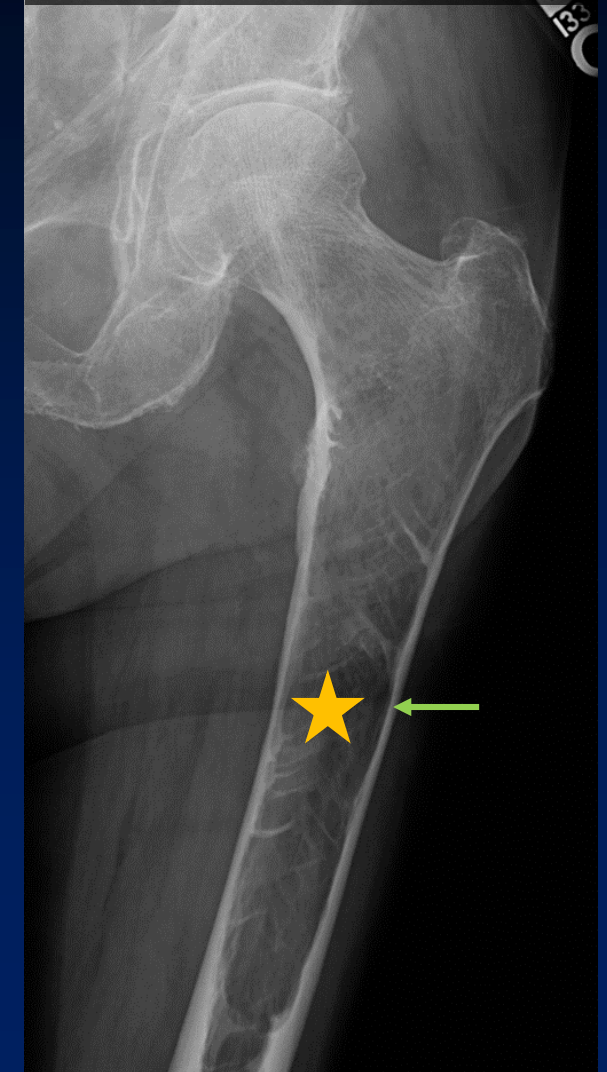
Findings - Radiographs

- Symmetric lytic lesions of both proximal femoral metadiaphyses and diaphyses
- Circumferential endosteal scalloping with narrowing the bony cortex

AP Radiograph Right hip

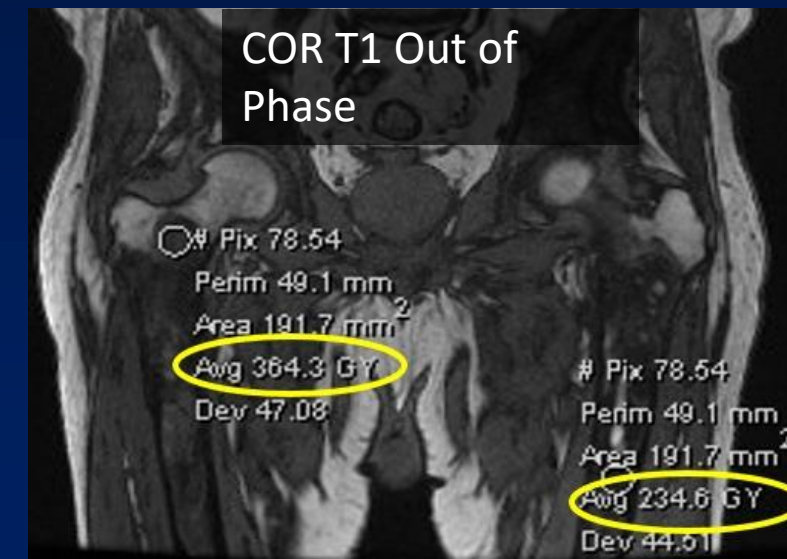
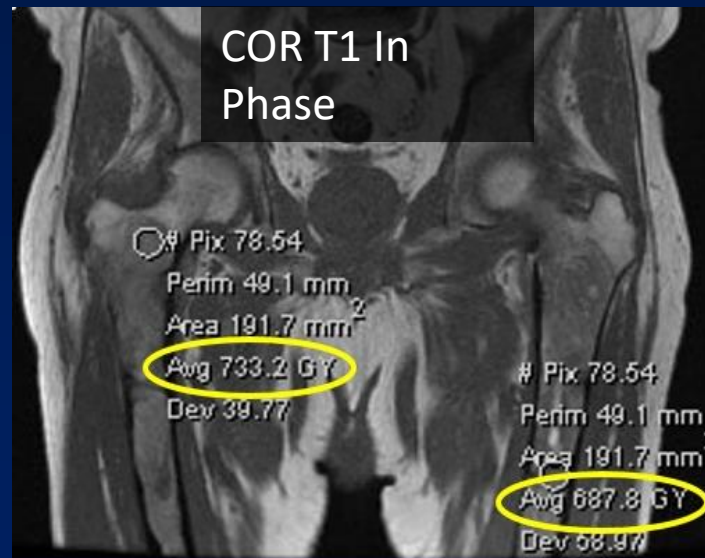
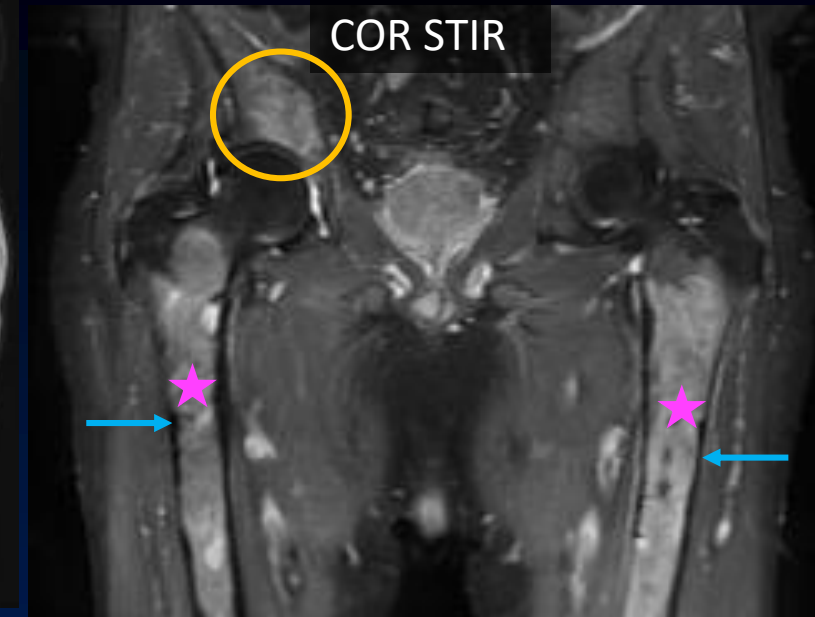
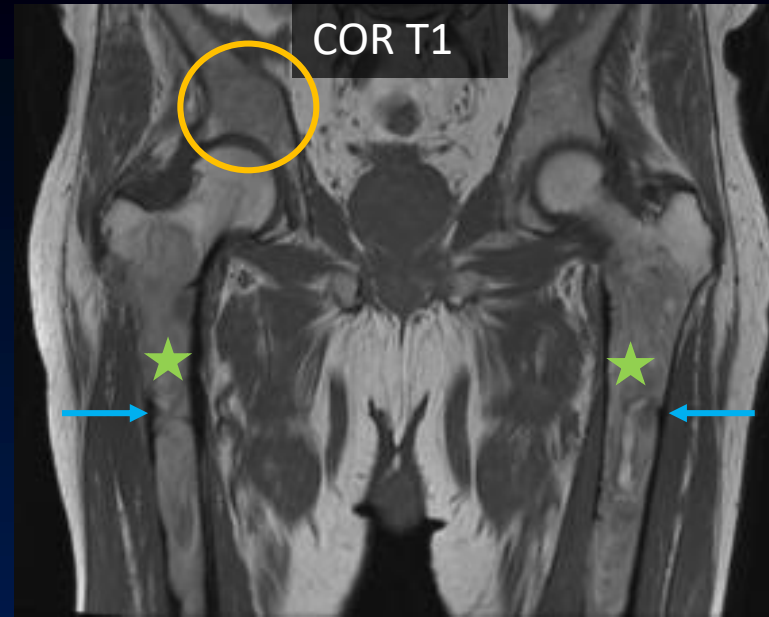


AP Radiograph Left hip



Findings - MRI

- T1 – Heterogeneous intermediate signal with areas of internal hypointense signal
- STIR - Heterogeneous intermediate signal with areas of internal hypointense signal
- Chemical Shift Imaging (CSI) – **>20% signal loss on out of phase imaging**, indicative of intracellular fat (retained hematopoietic marrow elements). This is suggestive of marrow infiltration or deposition rather than replacement



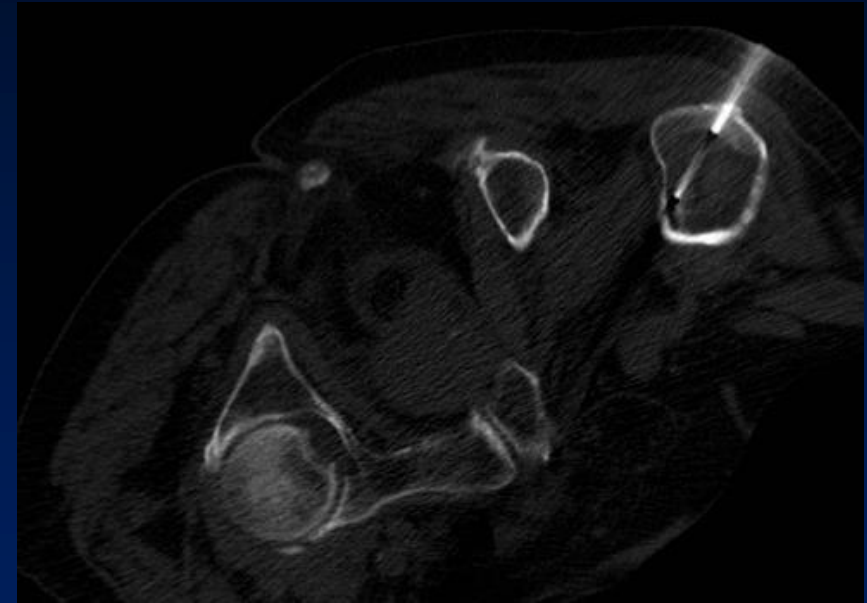
Right acetabular lesion with similar imaging characteristics (not appreciated Radiographically)

DDX

- Brown tumor of chronic hyperparathyroidism
- Myeloproliferative disorders (i.e. polycythemia vera or myelofibrosis)
- Deposition disease (such as amyloidosis)
- Malignancy: multiple myeloma, leukemia, lymphoma

Diagnosis - Primary AL Amyloidosis

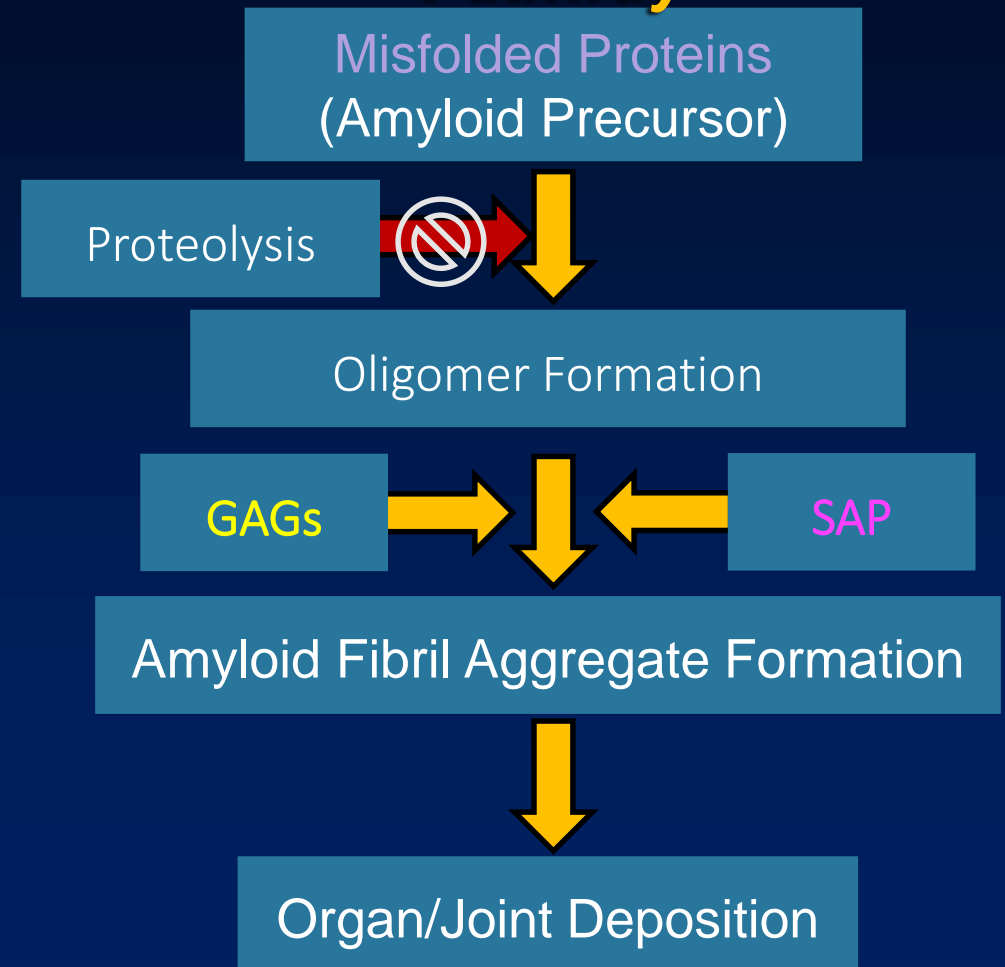
- Pathology Results:
 - Increased serum kappa light chains and B2 microg
 - Positive on Congo Red Stain
 - Renal failure caused by amyloidosis
- Imaging Pearls:
 - T2 hypointense elements are helpful in making DX
 - Marrow deposition and infiltration rather than replacement (preserved marrow elements on chemical shift imaging)



Case Discussion - Amyloidosis

- Amyloidosis is a disease of extracellular protein deposition.
- Misfolded proteins **avoid proteolysis** and aggregate to form oligomers.
- These oligomers are then stabilized by **glycosaminoglycans (GAGs)** and **serum amyloid P (SAP)** to form stable aggregates called amyloid fibrils.
- These fibrils adopt a beta-pleated-sheet rich configuration that is reflected in the histologic changes.
- Amyloid deposits can subsequently occur in the heart, musculoskeletal system, kidney, gastrointestinal tract, and nervous system.
- These deposits are insoluble and are resistant to macrophage degradation.

Amyloidosis Pathophysiology Pathway



Case Discussion - Amyloidosis

- Classification

- Distribution – Systemic vs Localized

- Systemic

- Primary - associated with plasma cell dyscrasias.

- Secondary - association with chronic infections and inflammatory disorders.

- Localized

- Protein Type – Complicated biochemical classification with > two dozen subtypes. Fibrillar proteins - amyloid light chain (AL) and serum amyloid A (AA) are present in the vast majority of cases.

- AL fibrillar protein is derived from the abnormal breakdown of normal immunoglobulin light chains in patients with an underlying plasma cell dyscrasia.

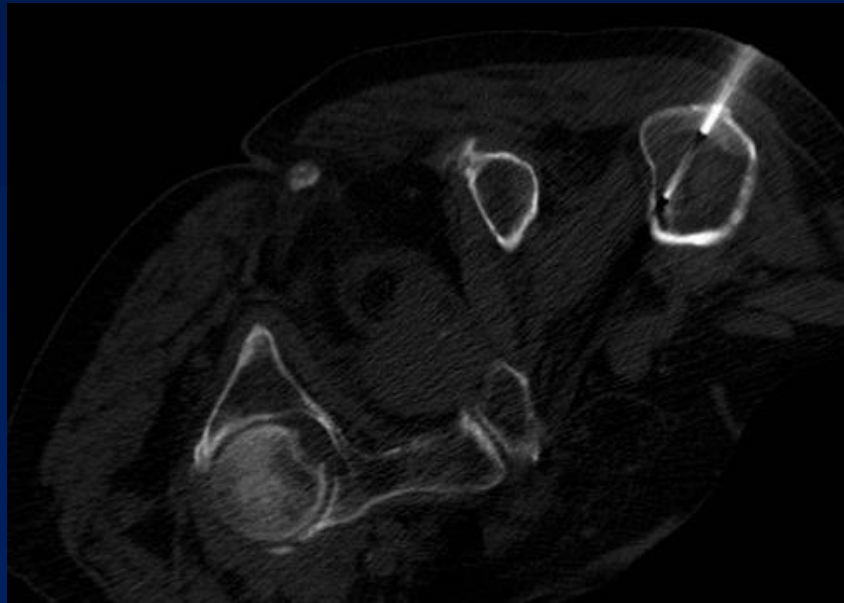
- AA fibrillar protein is produced by the liver as an acute phase reactant as is seen in a variety of systemic infections and inflammatory disorders (eg, rheumatoid arthritis, chronic bronchitis, osteomyelitis). AA production can be seen in patients with and without amyloidosis. However, in a subset of patients these abnormally folded AA deposits lead to systemic amyloidosis.

Case Discussion - Amyloidosis

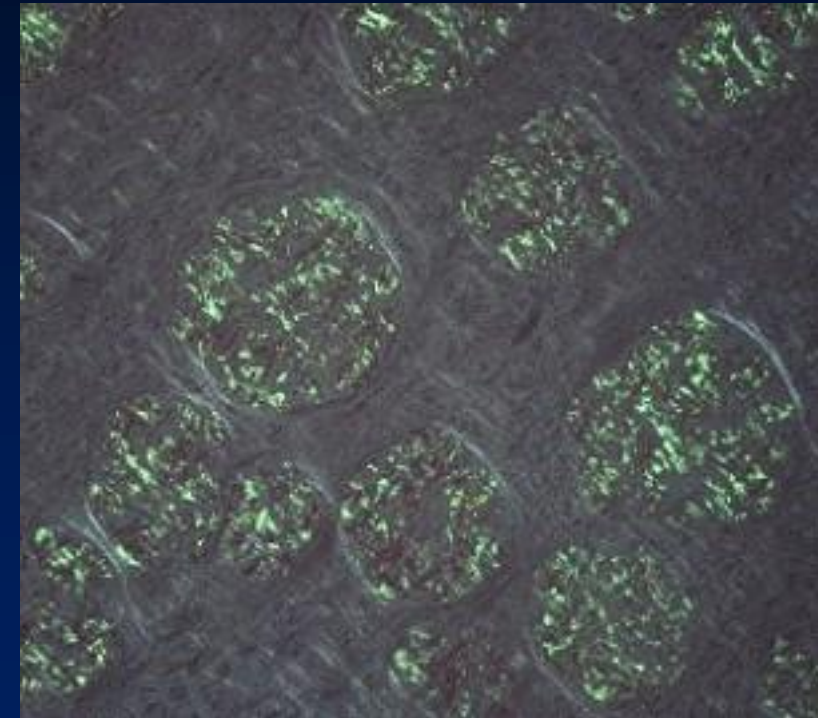
Amyloid Biochemical Tests

Biochemical Test	Abnormal Lab Values
SERUM FREE LIGHT CHAIN ASSAY (FLC)	KAPPA:LAMBDA RATIO <0.26 OR >1.65

Biopsy



Congo Red Staining with Polarized Light

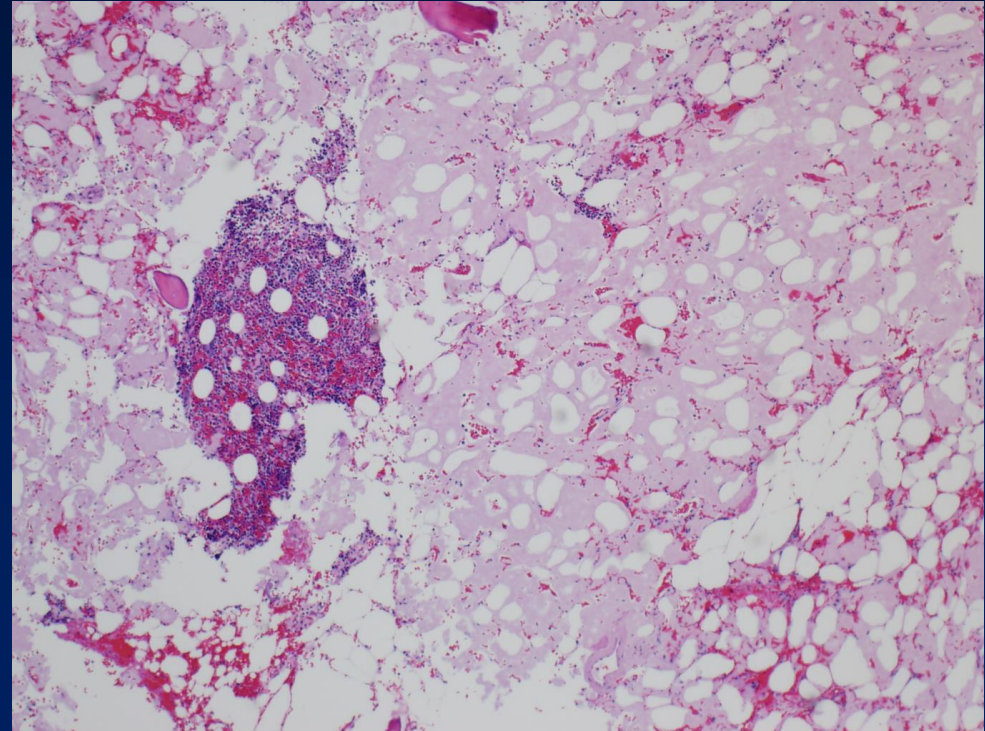
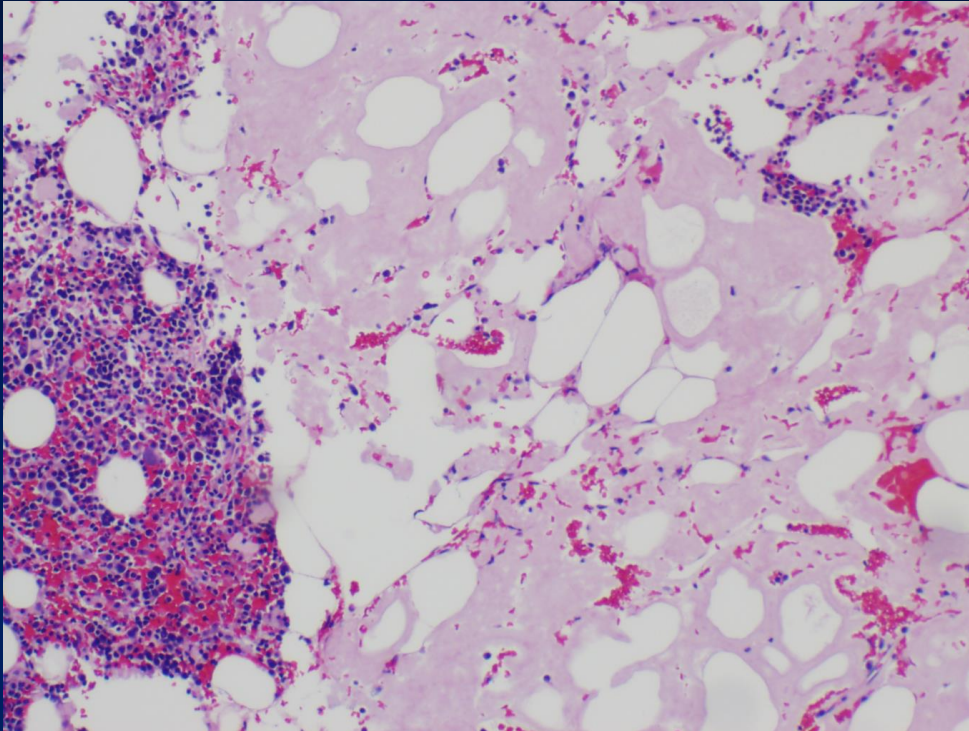


PC: Int J Biochem Cell, Dec 2003

Polarized light analysis of biopsied tissue stained with Congo Red reveal apple-green birefringence

Case Discussion

- Histopathology



Case Discussion - Amyloidosis

- Musculoskeletal Findings
 - Muscle Involvement
 - Muscle enlargement (pseudohypertrophy)
 - Myopathy
 - Arthropathy
 - Low grade symmetric arthropathy in AL Amyloid
 - Usually involves shoulders, knees, wrists, metacarpophalangeal, and proximal interphalangeal joints
 - Osteopathy
 - Solitary or multiple
 - Can lead to pathologic fractures

Case Discussion - Treatment

- AL Systemic – Treated with systemic chemotherapy.
- AA Systemic – Treat the underlying cause.
- If there is extensive femoral or tibial disease, prophylactic fixation should be considered (as in our case).



1 week after biopsy patient had pathologic fracture
ORIF of right fracture and prophylactic left ORIF

References

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