

AMSER Case of the Month

October 2023

HPI: 55 y/o male with abdominal pain and bloody stool

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Patient Presentation

- **HPI:** 55 y/o male who presented to Hershey ED with COVID, cyanotic extremities, and worsening diffuse, cramping, nonfocal abdominal pain associated with loose stools and small volume bright red blood per rectum. Endorses mild cough, shortness of breath, and sore throat. Denies flank tenderness.
- **PMHx/PSHx:** Bilateral knee pain (7 years), Hyperlipidemia, Kidney Lesions, Nephrolithiasis, Hernia Repair
- **Family Hx:** Mother with Lupus. Three healthy children
- **Social Hx:** Smokes cigars occasionally. No alcohol use.

Patient Presentation

- **Vitals:** T 37.4C, HR 92, RR 21, BP 150/120, SpO2 96% on RA
- **Physical Examination:** Nondistended abdomen with normoactive bowel sounds. Mild diffuse abdominal tenderness without organomegaly. Unremarkable otherwise
- **Pertinent Labs:** Na 134, CO2 21, WBC 11.60, Hct 51.2, RBC 6.20
- **Studies:** Urinalysis negative for leukocytes/RBCs

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

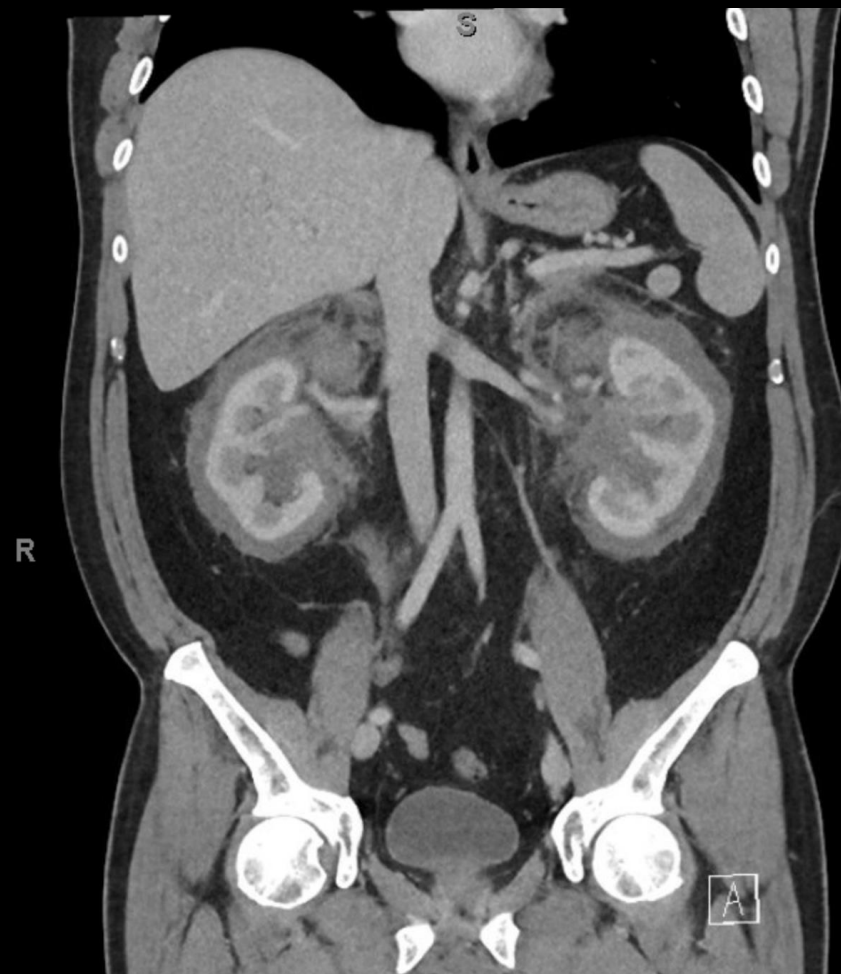
Variant 1: Acute nonlocalized abdominal pain and fever. No recent surgery. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	⊕⊕⊕
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate	○
US abdomen	May Be Appropriate	○
CT abdomen and pelvis without IV contrast	May Be Appropriate	⊕⊕⊕
MRI abdomen and pelvis without IV contrast	May Be Appropriate	○
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕
Radiography abdomen	May Be Appropriate	⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕
WBC scan abdomen and pelvis	Usually Not Appropriate	⊕⊕⊕⊕
Nuclear medicine scan gallbladder	Usually Not Appropriate	⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	⊕⊕⊕

CT Abdomen was ordered by the ED Physician to evaluate for acute pathology within the intestinal system contributing to bloody stools.



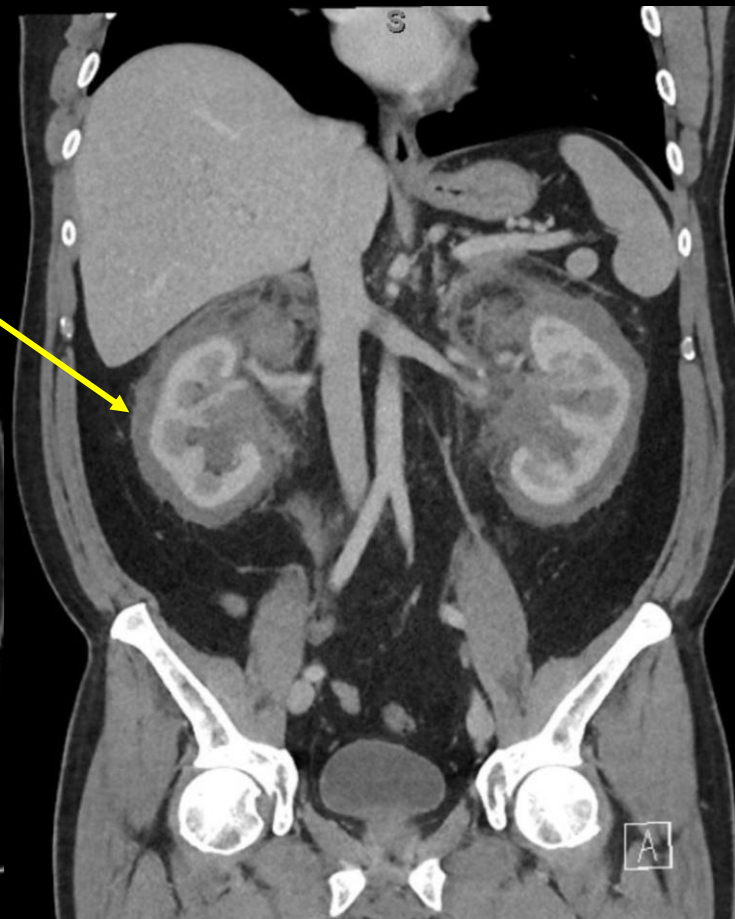
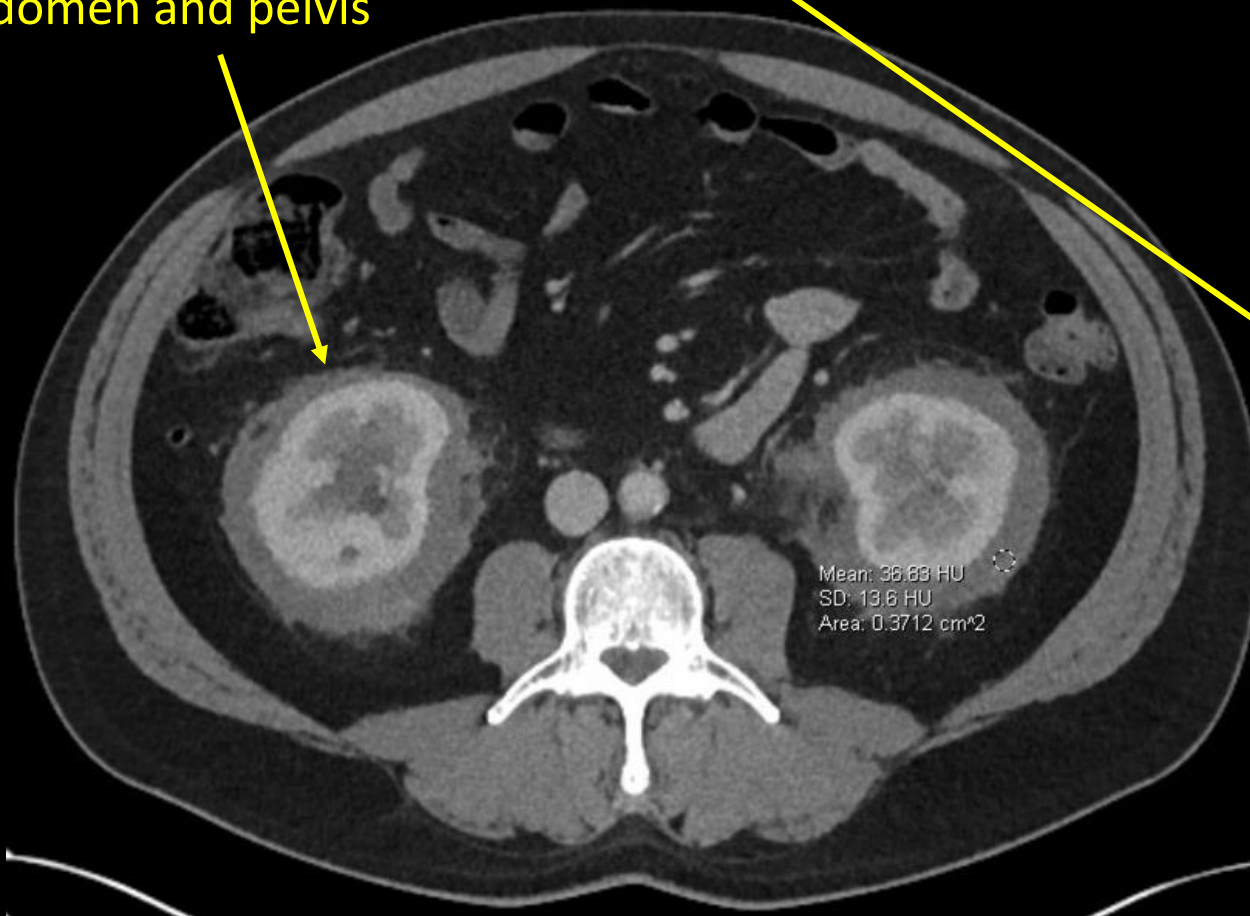
Findings (unlabeled)



CT AP + Coronal w/ Contrast

Findings (labeled)

Rind-like tissue surrounding the bilateral kidneys (37 HU) possibly representing retroperitoneal fibrosis or isolated lymphoma. Otherwise unremarkable CT of the abdomen and pelvis



Percutaneous biopsy was recommended to exclude lymphoma and multifocal fibrosclerosis. He followed with outpatient Urology, Rheumatology, and Nephrology.

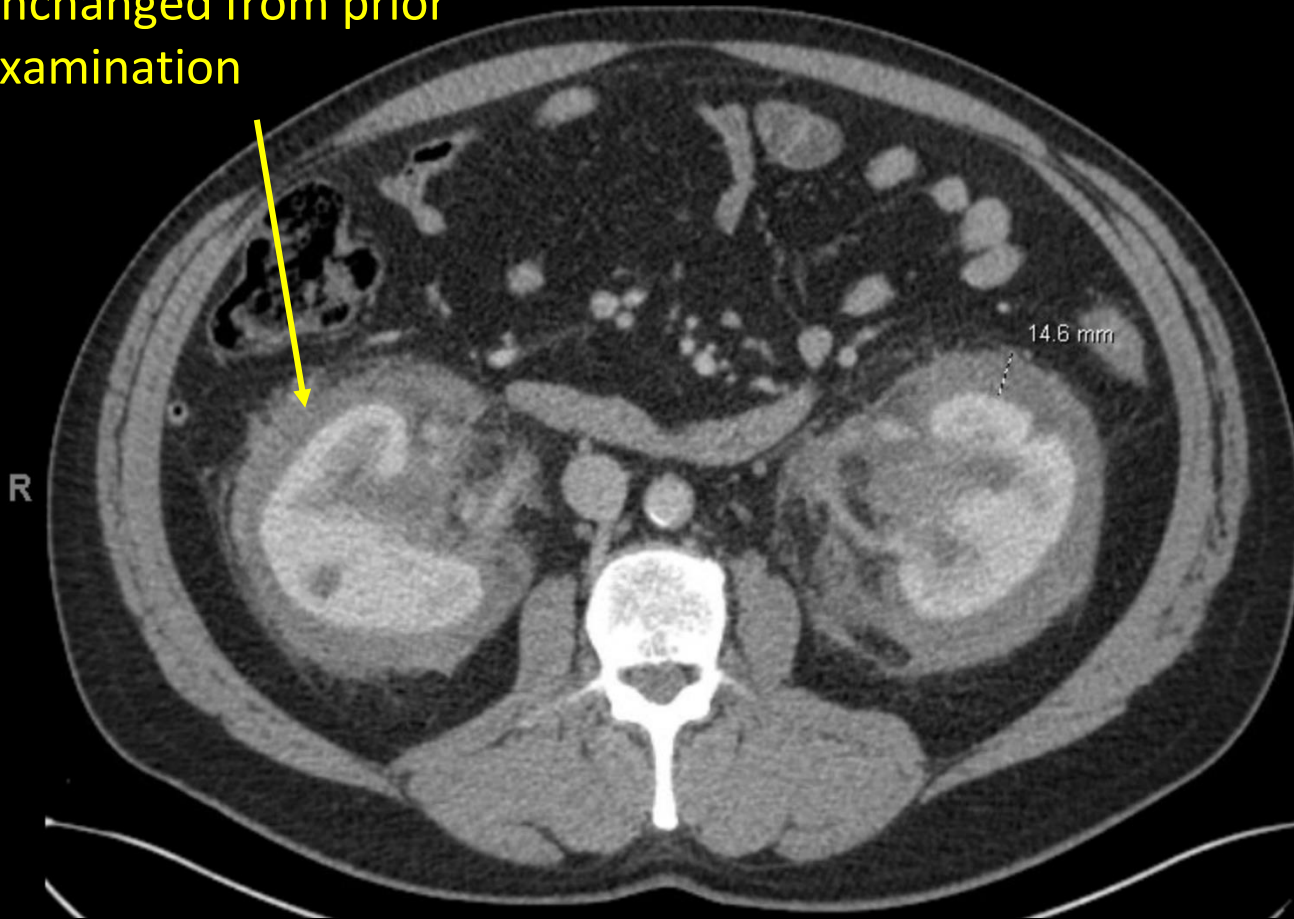
CT AP + Coronal w/ Contrast

Follow-up

- **Nephrology:** Biopsy negative for immune complex/paraprotein mediated disease
- **Urology:** ANA negative, dsDNA negative, RF negative, Lyme negative, ESR 17 (normal 0-15), WBC 7, Hgb 16, Plt 300, Cret 0.8.
 - Evidence of infiltrative process bilaterally, recommended referral to Oncology
- **MSK:** Was following with ortho for long-standing bilateral knee pain at outside hospital. MRI Knee (R) demonstrated diffuse abnormal marrow signal and periosteal edema of the distal femur, suspicious for pathologic marrow infiltration/myeloproliferative disorder
 - R knee bone biopsy: CD68+ Histiocytoma, foamy macrophages, negative gram stain/AFB/CD34/SMA/Desmin/S100
 - Heme/Onc referral recommended
- **Heme/Onc:** IgG Kappa gammopathy, negative SPEP/UPEP
 - Recommended skeletal survey

Repeat Imaging 6 months later

Infiltrative perirenal soft tissue density essentially unchanged from prior examination



CT AP + Coronal w/ Contrast

Select the applicable ACR Appropriateness Criteria

Variant 6: "Incidental" osseous lesion on MRI or CT scan for unrelated indication. Suspect primary bone tumor. Not clearly benign. Next imaging study.

Procedure	Appropriateness Category	Relative Radiation Level
Radiography area of interest	Usually Appropriate	Varies
MRI area of interest without and with IV contrast	May Be Appropriate	0
CT area of interest without and with IV contrast	May Be Appropriate (Disagreement)	Varies
CT area of interest without IV contrast	May Be Appropriate	Varies
MRI area of interest without IV contrast	May Be Appropriate	0
Bone scan whole body	May Be Appropriate	⊕⊕⊕
FDG-PET/CT whole body	Usually Not Appropriate	⊕⊕⊕⊕
CT area of interest with IV contrast	Usually Not Appropriate	Varies
US area of interest	Usually Not Appropriate	0

Osseous survey was ordered based on biopsy showing bony histiocytoma with foamy macrophages to evaluate for additional lesions

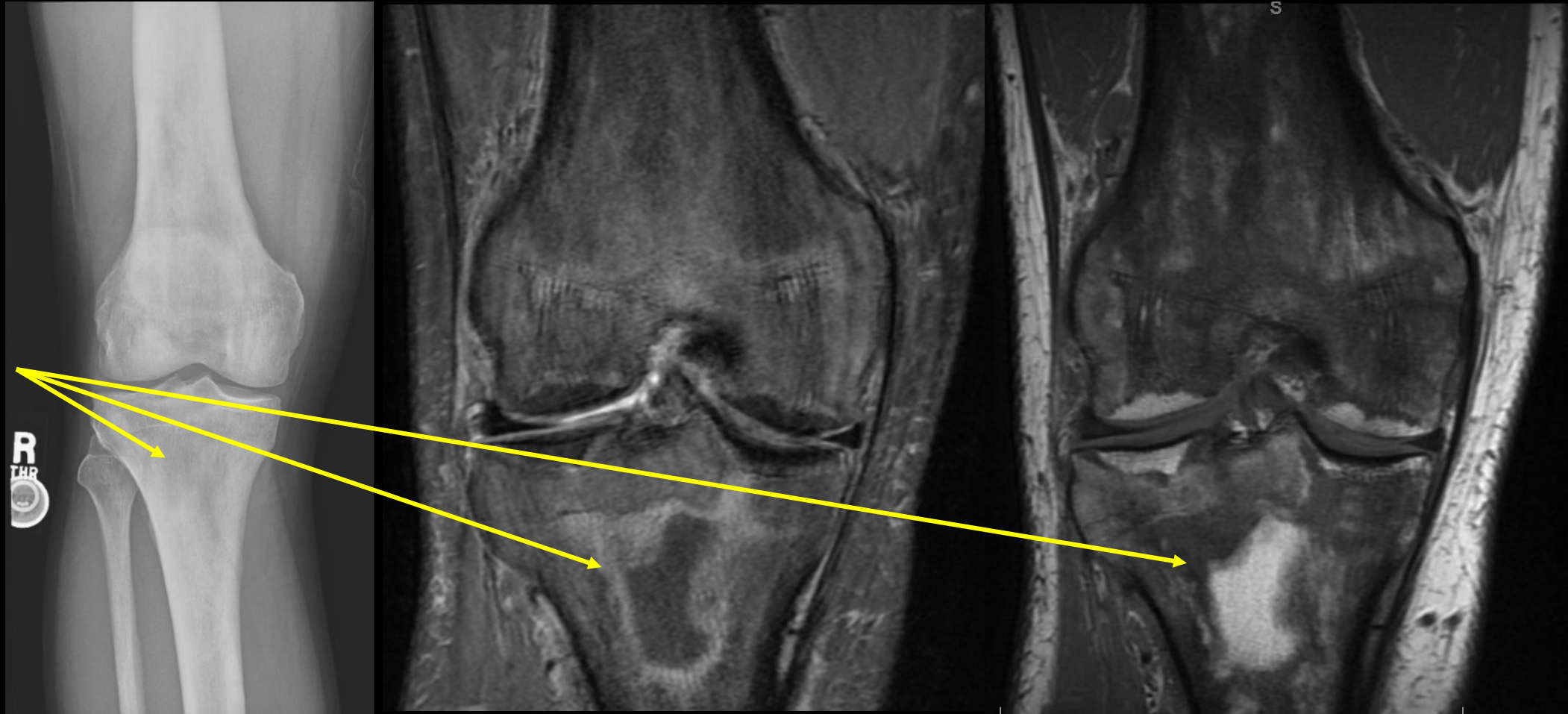
Findings (unlabeled)



Osseous XR, Proton-Density, and T1 MRI

Findings (labeled)

On XR, poorly marginated sclerotic pattern with cortical thickening. On MR, Diffuse marrow signal abnormality consistent with red marrow conversion, related to underlying myeloproliferative disorder



Osseous XR, Proton-Density, and T1 MRI

Final Dx:

Erdheim-Chester Disease

Case Discussion

- Erdheim-Chester Disease
 - Uncommon non-Langerhans Cell Histiocytic proliferative disorder with multisystem involvement
 - Peak incidence is in 5th-7th decade of life with male predilection
 - Given rarity, exact etiopathogenesis is unclear and no definite genetic base has been established
 - Theories include immune-mediated phenomenon from exaggerated proliferation of helper-T cells and subsequent release of interferons that recruit and activate mast cells at site of involvement
 - Symptoms include fever, weight loss, night sweats are common. Most common is bone pain, implicated in majority of cases
 - Extraosseous manifestations include central diabetes insipidus and exophthalmos
 - Biopsy reveals lipid-laden histiocytes, CD68 positivity, and lack of CD1a and Birbeck granules.
 - CNS and cardiovascular involvement worsens prognosis given poorer response to chemotherapy

Case Discussion

- Management
 - There is strong consensus (>95%) that treatment is indicated for most Erdheim-Chester Disease patients except for asymptomatic minimal burden disease, which is monitored closely
 - Systemic corticosteroids, surgery, and radiation therapy may be used to relieve edema or acute symptoms, but are not recommended as monotherapies
 - For patients with cardiac/neurologic disease or end-organ dysfunction, BRAF-inhibitor therapy should be implemented
 - Optimal duration and dosing of targeted therapies is unknown, although relapse is observed in majority of cases following cessation of BRAF-inhibitor
 - IFN- α /PEG-IFN- α or cladribine may also be considered

References:

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