7/18/24 Morning Report with @CPSolvers

"One life, so many dreams" Case Presenter:Vijay (@) Case Discussants: Rabih(@) and Mukund (@)

CC: 30 yr F with bilateral limb pain and headache

HPI:
Limb pain: Developed the pain gradually progressive, over 3 months difficulty in walking and calf pain, can now only walk 100m

Simultaneous headache, diffuse, dull aching, no aggravating factors, no photophobia or phonophobia

Blurry vision, can perceive color well, no diplopia, no pain during movement of eyes

ROS (-): fevers, rash, join pain, pet exposure

PMH: Acute pancreatitis, negative w/u for gallstones, drugs, Ca, TGL

Meds:
-

Fam Hx: -

Soc Hx: -

Health-Related Behaviors:

Allergies:

Vitals: T: PR: 76 **BP:** upper limb unrecordable-feeble peripheries, lower limb SBP 70 mmHg

Exam:
Gen: xanthelasma
CV: no murmur, HS heard
Pulm: fine crackles bl
Abd: hepatosplenomegaly
Neuro: Grade 3 papilledema, no neuro deficits
Extremities/skin: absent peripheral pulses

Notable Labs & Imaging:
Hematology:
WBC: 3200 (normal diff) Hgb: 8.6 (Base 10) Plt: 1.4 (1-1.5 baseline)
Ferritin 55 TGL 128
Viral serologies: HIV HCV HBV neg
Echo moderate pericardial effusion w/ normal biventricular function
ESR 98, **CRP** 56
CT: encasement of peri-aortic and perirenal soft tissue, interlobular septal thickening bl lung fields, moderate pericardial diffusion, ill defined mass of the pancreas, peritoneal nodularities
MRI: partial empty sella, optic nerve tortuosity, pachymeningeal enhancement, SOL, osteosclerosis in the clivus
BMA: normal marrow, occasional hemophagocytosis, no atypical cells/eosinophilia, ANA, ANCA, APS, IgG4 wnl
Biopsy from peri-renal soft tissue suggestive of non-specific inflammation/ no atypical cells
TFT - nl
Repeated CD68+, CD1a -, BRAF: neg

Dx: Erdheim Chester disease

Problem Representation:
A 30 y/o male with a PMH of pancreatitis p/w bilateral claudication, papillary edema and headache was found to have xanthelasmen w/o marked lipid panel, a high ferritin, pericardial effusion and pachymeningeal enhancement and osteosclerosis on imaging w/o lymphadenopathy.

Teaching Points (Julia):

Lower limb pain:
- A) **hypoperfusion** *(clot, claudication, vasculitis)*
  - a) Claudication (*vascular* or *neurogenic*) →vulnerable to movement
  - b) **Neurogenic** = Slowly progressive form of cauda equina; dynamic rather than continuous symptoms, increased pain with extension
  - c) Distinguish from **vascular**→ position and timing (ultrasound can help)
- B) **B MSK** consider when pain do not go back to zero after rest

Blurry vision (cave DD visual loss)
is usual non-neurologic
- Neurological causes = pupillary issue (dense lense) or papillary edema

Pulsnessless: where is the lesion? Pan-aortic lesion vs. diffuse vascular disease (medium vessel vasculitis)

Papillary edema: brain, vascular (obstruction) or CSF (drainage problem) → MRI, CT with delayed phase (venous sinus thrombosis)

Dural mass: 1. Meningioma 2. chronic subdural bleeding

Overlap of dural mass + dural thickening → infiltration: **histiocytic** disease & **sarcoid**

Disconnection: **abnormal lipid profile w/o** marked **hypertriglyceridemia** → Macrophage problem (pericardial perfusion, ferritin↑)

Histiocytic disease (BRAF pos? => tissue is the issue)
1. Langerhans cell histiocytosis (lung focus cystic lesions with micro)
2. Erdheim Chester (bony lesions 99% sclerotoc lower limb lesions, pararenal, pericardial)
3. Rosai–Dorfman disease (lymphadenopathy)

Cave: Can coexist, myeloproliferative or autoimmune disease in the background?

Keep pituitary consequence of histiocytic disease in mind