12/24/21 Morning Report with @CPSolvers

Case Presenter: *Kirtan Patolia(@kirtanpatolia)* ***Case Discussants***: *Rabih (@rabihmgeha) and Reza (@DrRxEdu)*

CC: **"Translucent diagnosis"**
Bilateral lower extremity swelling

HPI: 63yoF w/ PMH of hypothyroidism and faty liver disease p/w complaint of lower extremity edema , heaviness, and fatigue for 6 months. The swelling had been persistent and increase since last 1 month. She had seen many health care providers and told her protein levels were low. She denied having any improvement in swelling after making dietary changes.
ROS: denies swelling of arms, face,SOB, orthopnea, polyuria, any rashes or pruritus

PMH:
HTN, hypothroidism

Meds:
Chlorthalidone, telmisartan, amlodipine, levothyroxine

Fam Hx:
none

Soc Hx:
Homemaker. Denies use of alcohol, illicit drugs

Health-Related Behaviors:

Allergies:
none

Vitals: **T**: **37 HR**: 78 **BP**: **160/98 RR**: 16 **SpO$_2$**: 99 on RA
Exam:
Gen: appeared well and was in good spirits she
HEENT: normal
CV: normal S1 and S2. No murmurs.
Pulm: BLAE, no crackles
Abd and Neuro: nl
Extremities/Skin: symmetric bilateral pitting edema up to knees

Notable Labs & Imaging:
Hematology:
WBC: 8k w/ normal differentiation Hgb: 13 (MCV=86) Plt:253k
Chemistry:
Na/K/Ca/P / Mg within normal limits
Cl: CO2: BUN: 18 Cr: 0.8 glucose: Ca: Phos: Mag:
AST: 34 ALT: 32 Alk-P: T. Bili: Albumin: 2.41 globulin 2.12 A :G ratio: 1.14 Cholesterol 235 Direct LDL 172.6 TGL: 108.2 INR/aPTT nl
TSH/T3/T4 nl ESR nl
ECHO: nl LVEF No RWMA Grade 1 diastolic dysfunction , Mild TR, MR, PAH, RVSP: 31
USG: bilateral bulky kidneys w/ attenuated corticomedullary differentiation
UA: pH 6, SG: 1.02, urine protein 3+, no casts or RBCs or pus cells
Glucose 24h protein: 13g/day
HIV/HBV//HCV neg CEA 1.92 CA:1245:11.9 ANA and C3 and C4 nl Anti PLA2R antibodies nl SPEP with IgA, IgG, and IgM nl
FLC: free kappa and lambda chain and ratio normal NT pro BNP and troponin nl
Renal biopsy: acellular hyaline material in mesangium and capillary BM. Congo red + for amyloid IF study: neg for IgS, kappa, lambda, chain, C3, C1q, fibrinogen, IHC for SAA protein neg.
Bone marrow biopsy: normocellular marrow w/ no evidence to suggest plasma cell neoplasm. No evidence of increased plasma cell.
UPEP and UIA: definite evidence of IgG lambda M band
Final dx: primary AL amyloidosis of kidneys
(Translucent as labs don't fit transparently)

Problem Representation: 63 yoF w/PMH of hypothyroidism and HTN p/w lower extremity edema. Found to have evidence of IgG lambda M band on UPEP and UIA.

Teaching Points (Sukriti):
Investigating the Sx:
NAFLD exists in a spectrum w/ NASH, Cirrhosis
Law of proportionality: The presence of edema in the context of NAFLD makes one think of the more severe forms of this dz i.e Cirrhosis
- Use stigmata of CLD, elevation of LFT's and evidence on imaging to clue you into this Dx
- Remember it is a Dx of exclusion

Edema - Heart, Kidney, Liver
Others: Non cirrhotic portal HTN - schistosomiasis, Nephrotic syndrome
Medications (Amlodipine), Endocrinopathy (Cushing's), IVC dz
IVC dz & Obesity: increased abdominal pressure -> IVC obstruction

Collecting the clues: Kidney + Heart
CRP: Most pt w/ B/L LE are minimally symptomatic w/ no extension beyond these dimensions, the presence of Sx should clue you into a more concerning Dx and warrants further work up
When do you suspect Nephrotic syndrome? Hypoalbuminemia (<3), nephrotic range proteinuria, edema
- The work-up for nephrotic syndrome is informed by the histopathology, but must be initiated before the Bx results
- The four most common types: FSGS, membranous nephropathy >> amyloidosis, Minimal change dz

CRP: Diabetes in a majority of pts. does not result in overt nephrotic syndrome, although commonly results in nephrotic range proteinuria

Framing the Hypothesis: Heart + Kidney + Amyloidosis = AA, AL
Immunohistochemistry is a good test for AA, but often misses the other types, mass spectroscopy has greater sensitivity