10/28/24 Morning Report with @CPSolvers

"One life, so many dreams" Case Presenter: Dr. Ann Kim (@annskimMD) Case Discussants: Dr. Jerry Lee (@TheBloodDoc)

CC: 66 yo M SOB with exertion

HPI: developed progressive SOB with exertion over the past month. Increasing fatigue, worsening chest tightness. Referred for outpatient stress TTE - but unable to complete due to severe dyspnea, shakiness. EKG - ST depressions. Echo - wall motion abnormalities. Sent to Ed for worku.
ROS: Palpitations, cough, 10lb weight loss, canker sores for past few years.. No fevers, chills or bleeding.

PMH: Hepatic steatosis, essential tremors, sciatica

Meds: Vit D

Fam Hx: Breast cancer - mother

Soc Hx: non significant

Health-Related Behaviors: No smoking, no alcohol or drugs

Allergies: no

Vitals: T: nl**HR:** 70s **BP:nl RR:** nl
Exam:
Gen: pale, no cervical or axillary lymphadenopathy
CV, Pulm: nl
Abd: splenomegaly present

Notable Labs & Imaging:
Hematology:
WBC: 5,35 Hgb:2,5 Plt:180 MCV 96, RDW 64,1, ANC 0,87 - low, ALC 3,85 - high, ARC 0,02 - low
CMP - normal
Smear - after transfusion - hypochromic RBCs with variation in size, no schistocytes. Lymphocyte predominance, some with large granules. No blasts.
Normal CBC a year ago, except Hb 10.
hsTrop 143
Ferritin 1160, Tsat 94% - high, haptoglobin nl 100, B12 nl 890
CT Scan: splenomegaly and hepatic steatosis.
Bone marrow biopsy - flow cytometry: abnormal T cell, +CD2, CD3, CD7, CD8, CD57, CD 16 monotypic TRBC1 staining = 16% lymphocyte population. Large granular lymphocytes.
Other variants with non clinical significance were also present.
Myeloid NGS - STAT3 mutation, but VUS

Dx: T cell large granular lymphocyte leukemia

Problem Representation: 66 yo M presents with subacute/ chronic SOB, fatigue and chest tightness. PE: splenomegaly. Labs low Hb, smear-large granules. Bone marrow biopsy flow cytometry +CD57.

Teaching Points(Parisa):
SOB Framework→r/o non-hematologic causes (cardiac ECG; pulmonary)→ anemia(chronic blood loss; inflammation); cancer related(unintentional weight loss/fatigue/dyspnea on exertion/night sweats/risk factors: being smoker)
Family history of cancer is significant when happens in a younger age and primary family members (genetic predisposition)
Neutropenia + normocytic anemia→ primary marrow involvement
Splenomegaly→ cancer; extramedullary hematopoiesis → PBS shows lymphocyte predominance and lacks schistocyte r/o TTP; MAHA
Low retic → shows chronic process + no compensation
Immunophenotyping lymphocyte (CLL; lymphoma) could be paraneoplastic picture of underlying malignancy leading to anemia → looking for hemolysis footprint (haptoglobin; LDH)
High Ferritin/ Transferrin → iron overload (Hemochromatosis; thalassemia)
Leukemia → myeloid(myelodysplastic); lymphoid (CLL;CML) → how sick is the patient→ Tempo
Both CLL and SLL are essentially the same disease, differing mainly in the site of lymphocyte accumulation; CML involves proliferation across the myeloid lineage (basophils; neutrophils)
T LGL: pancytopenia; lymphocytic predominant; large granular lymphocyte found on PBS and BM; mutation on STAT3/5; Tx oral MTX;(develops in the context of rheumatologic dx RA/lupus; being on immunosuppression; it might be presented w/o this PMH); splenomegaly is common
Immune cell population markers→T cell CD2-8; CD19-20 B cell
Natural killer cells lymphoma→ marker CD56; aggressive type; asso EBV/co present with HLH; asian population; nasal phenotype; nasal mass
Other Dx T LGL → T cell leukemia; T cell lymphoma; NK lymphoma
Tx of hematologic malignancies : allogeneic Tx(unclear here); autotransplant; immunosuppression(Prednisolone + oral MTX; cyclophosphamide; cyclosporine) more significant role rather than chemotherapy; response to tx is slow esp anemia; shrinkage of splenomegaly should be observed → Long Term outcome → Infection d/t immunosuppression