2/21/22 Morning Report with @CPSolvers

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CC: Exercise intolerance and SOB

HPI: 52yo M presenting in the clinic with thrombocytosis. Patient reports SOB for past few years. No fever, chills, night sweats, early satiety.

PMH: IDA (started on iron supp.)

Meds: None

Fam Hx: None reported

Soc Hx: None reported

Health-Related Behaviors: None reported

Allergies: None

Vitals: **T**: **HR**: 57 **BP**: **RR**: **SpO_2**: 95% RA
Exam:
Gen: pallor
HEENT: no mouth sores, headache
CV: unremarkable
Pulm: SOB when exercises
Abd: no splenomegaly
Neuro: no focal deficits
Extremities/Skin: unremarkable

Notable Labs & Imaging:
Hematology:
WBC: wnl Hgb: 9 Plt: 650 Ferritin: low B12, LDH: normal
Peripheral smear: No atypical cells
GI bleed workup (i.e. Colonoscopy): Unrevealing
Thrombocytosis persisted despite improvement in IDA with iron supplementation.
Bone marrow biopsy: 50% cellular bone marrow. Megakaryocyte hyperplasia with frequent small clusters. Grade 1/3 fibrosis. Consistent with ET
Molecular testing: BCR-ABL (-), JAK2 detected (V617F mutation)
Mutations found: DNMT3A, ASXL1

Final diagnosis: JAK2 (+) Essential thrombocytosis

Problem Representation: 52 yoM w sx of exercise intolerance and SOB found to have IDA and thrombocytosis, started on iron supplementation, with improvement in anemia though persistent thrombocytosis.

Teaching Points (Andrea):

- Thrombocytosis Approach:
  Important to know the time. Compare with previous CBC
  -Secondary or Reactive (Majority): Common causes iron deficiency anemia, inflammation, sepsis, rheumatologic
  -Primary problem of the BM: myeloproliferative neoplasm
- If patient is admitted to the hospital and had acute events: Thrombocytosis is more likely to be secondary
- Flushing or redness: Thrombocytopenia related to Myeloproliferative Neoplasms (MPN): policitemia vera, essential thrombocytosis, primary myelofibrosis, CML (BCR-ABL fusion gene)
- MPN typically presents with splenomegaly. In physical exam it has to be 14 cm to be palpated. Do not rule out if not palpated. Point of care ultrasound is not helpful
- Anemia has to be significant to show pallor (4 or 5)
- Bone marrow biopsy: Essential for MPN dx. It is difficult to differentiate secondary vs primary trombocitosis at the beginning of dz. Low risk procedure mainly pain. Sterile, no bleeding. Local anesthesia+lorazepam
- Bone marrow biopsy with peripheral smear and 1 cm core tissue that can be stained,
  -Standard cytogenetic (Karyotyping Of 20 cells. To be detected it has to be a large deletion or rearrangement) and consider FISH. Fish has to be specifically ordered. Molecular testing (JACK2, CALR and MPL mutations)
- Myelofibrosis: Drop cells
- Proliferation of Megakariocytes: Essential thrombocythemia
- JAK2 is very common in ET and Primary MyeloFibrosis
- Essential thrombocythemia Treatment. Dz is indolent but can evolve to secondary myelofibrosis (more likely w some mutations). Start Drive down or cytoreduced dependent of risk. Use of IPSET criteria. JAK2 (Complicacionts) and old age are high risk
  -Cytoreduction of ET: Hydroxyurea (worsening anemia), interferon, anagrelide (in pts with anemia)
  -Therapeutic anticoagulation, if severe thrombopheresis (only last a couple of days)
  -Secondary myelofibrosis: Allo genetic BM Transplant
  -Allogeneic Transplant in MPN and leukemia because you cannot take stem cell Dx of pts
  -Apart from coagulation problems, Micro-cutaneous bleeding in TE: High blood count (over 1 M) because there is vWF sequestration