C’est La Vie… de la Valve

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Introduction

When a patient does not improve with initial treatment, me must distinguish between treatment failures versus diagnostic mishaps. Cognitive biases of anchoring and status quo perhaps favor the treatment failure choice, however we must re-visit our diagnostic tests, to evaluate their sensitivity in ruling out alternative etiologies.

Outside Hospital Hemolysis Work-up:

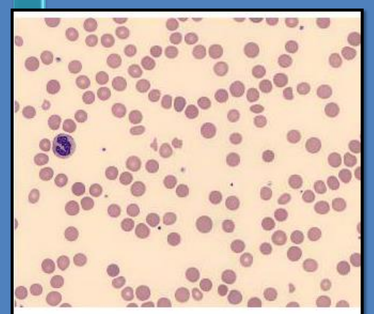
1st Round: Hapto low, High LDH, elevated indirect bili. PNH/DAT negative

2nd Round: low hapto, LDH high, negative DAT, neg: C3/C4, ANA, DsDNA, ANCA, HCV, HIV, blood Cx, anti-GBM, cryo, HBV negative

Total Transfusion Burden: 37 Units

January:

- Hematuria, HH Stable, CT abdo/pelvis nml



February:

- ED: Dark urine
  - Hb: 8.6, UA: large blood CT Abdo/Pelvis: NML
- Contrast Nephropathy: Cr 4.8 *BL 1.5
- Hospitalization w/CC: dark urine
  - negative. TTE: nml
- Rx: High Dose pred @ 80 mg daily

March, continues on Pred

- Bone Marrow: hypercellular
- Re-admit: hematuria:
- Cystoscopy: no stones
- Re-admit: hematuria, anemia.

Notable Labs at OHSU:

Chem: Cr: 1.95 *baseline

CBC: Hb/Hct 7/22, plt: 158 (MCV:94)

INR: 1.09, LDH: 3159, hapto <10, bili:1.5, Retic Index <3

G6PD: NML, Coombs/DAT: negative

PNH: negative

UA: Large Blood (1-2 RBCs), negative Bili, hemosiderin negative

April:

- Rituxamab Initiated
- Admission: Dyspnea, Dx: Lobar PNA
- ED Visit: thrush
- 2nd Ritux Infusion

May:

- 3rd Ritux infusion

June:

- Admission: Anemia:
  - Bone Marrow
  - TTE: nml
  - Rituxamab # 4

July:

- Admission: dyspnea, fatigue
  - Bone Marrow: NML
- Admission: Dyspnea
  - TTE: mild posterior MR

Case

Who: a 75 year old gentleman with notable history of triple bypass with mitral valve annuloplasty and remote lung cancer s/p right pneumonectomy who presented after 6 months of diagnostic uncertainty, after diagnosis of Coombs negative Auto-immune hemolytic anemia.

CC: shortness of breath, chest pain and fatigue

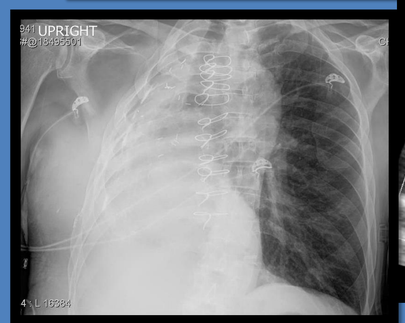
Prior Hospitalizations: See above

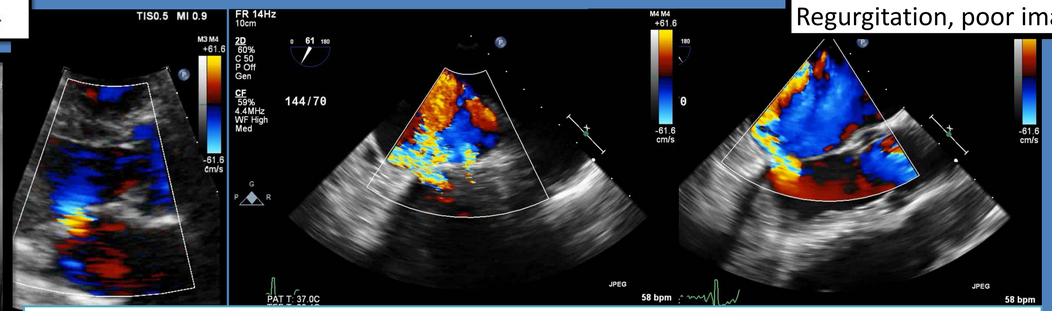
Objective:

Exam: Nml Vitals, 3/6 Holosystolic Murmur at R sternal border, b/l LE edema

Findings:

TTE: “at least moderate eccentric Mitral Regurgitation, poor image quality”





Left TTE: moderate MR, **Middle** TEE: Mid-esophageal, commissural view **Right** Mid-esophageal Aortic Valve Long Axis View Interpretation: dehisced mitral annuloplasty ring with severe regurgitation.

Box 1: Differential diagnosis of hemolysis

Conditions associated with hemolysis and a positive DAT result

- Hemolytic disease of the newborn
- Drug-induced hemolytic anemias
- Acute hemolytic transfusion reaction
- Delayed hemolytic transfusion reaction
- Autoimmune hemolytic anemia (warm autoimmune hemolytic anemia, cold agglutinin syndrome, paroxysmal cold hemoglobinuria, mixed-type autoimmune hemolytic anemia)

Conditions associated with a positive DAT result, with or without hemolysis

- Exogenous immune globulin administration
- Recent hematopoietic stem-cell transplantation
- Recent solid organ transplantation
- Systemic lupus erythematosus
- Infectious mononucleosis
- Some hematologic diseases, including lymphoproliferative diseases

Conditions associated with hemolysis and a negative DAT result

- Microangiopathic hemolytic anemias (thrombotic thrombocytopenic purpura, disseminated intravascular coagulation)
- Hypersplenism
- Liver disease
- Hemoglobinopathies (sickle cell disease, thalassemia)
- Erythrocyte membranopathies (spherocytosis)
- Deficiencies of erythrocyte enzymes (G-6-PD deficiency, pyruvate kinase deficiency)
- Infectious diseases (*Clostridium difficile* infection)
- Erythrocyte trauma (mechanical heart valves, improper use of blood warmers)

Conclusions

- Hemolytic anemia from prosthetic valve frequently occurs at the mitral valve in the setting of para-valvular regurgitation
  - An unexpected 19% of mitral valve repairs performed secondary to hemolysis
- Choose the right diagnostic study:
  - TTE vs TEE demonstrated leak in 31 patients using TEE compared to only 7 with TTE, concluding TEE’s superiority for evaluation of mitral valve pathology.
- Mis-diagnosis occurs when anchoring on results from a less-than-ideal diagnostic study.