Autoimmune Hemolytic Anemia In Chronic Lymphocytic Leukemia

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Autoimmune Hemolytic Anemia in Chronic Lymphocytic Leukemia

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Introduction

Patients with chronic lymphocytic leukemia have a 5-10% risk of developing autoimmune complications, the most common of which is autoimmune hemolytic anemia. Here we present a case of AIHA in a patient with suspected CLL that underscores several of the diagnostic and treatment challenges unique to this process.

Case Presentation

History of Present Illness

- 79 year old man with progressive dyspnea, exertional chest pressure x 3 weeks; worse over the past few days
- Has chronic stable pedal edema
- No PND, orthopnea, palpitations, syncope
- Went to PCP noted to be jaundiced with mild confusion; sent to ED for ACS rule out and further workup

Past Medical History

- Persistent leukocytosis with flow cytometry suspicious for CLL but no formal diagnosis
- Paroxysmal atrial fibrillation
- Hypertension
- Hypothyroidism
- Type 2 Diabetes Mellitus
- Gout

Physical Exam

- VS: HR 74 RR 16 Temp 36.8 BP 107/51 O2 100% RA
- G: Elderly man in no acute distress, mildly jaundiced
- Pulm: No increased WOB, CTAB
- Cardio: RRR w/ 2/6 systolic murmur best heard at the LUSB.
- Bilateral pedal edema with venous stasis changes
- Neuro: Intermittently confused but oriented x4. CN2-12 intact. No focal findings

Labs and Imaging

- Leukocytosis with WBC 20.8, 80% lymphocytes, 3% atypical lymphocytes
- Severe anemia with Hgb 5.1, MCV 115. Normal platelets.
- Total bilirubin 4.1, normal LFTs
- Haptoglobin < 30, LDH 235, DAT +complement/IgG, retic count 7.3%
- Negative troponin, BNP 172, EKG NSR with no significant changes
- CTA negative for PE
- CT abd/pelvis with cholecystitis and splenomegaly

Hospital Course

- Given 95 mg of prednisone in the ED. PRBC ordered
- Lab was unable to find compatible blood; was given 3 units ABO and RhD matched blood over 4 hours with premedication and careful monitoring for transfusion reaction
- Hgb responded appropriately to transfusion
- Started on 80 mg Prednisone daily
- LDH, reticulocyte count, Tbili downtrended
- Hgb remained stable
- 1 episode of recurrent chest discomfort while straining to have a bowel moment but otherwise asymptomatic for remainder of hospital course
- Discharged on 80 mg qd prednisone with outpatient oncology follow up

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Outpatient Course

• Now following with oncology
• Flow cytometry and FISH analysis confirmed CLL diagnosis; monotypic B-cell population
• FISH positive for deletion of 13q14 with no other abnormalities
• This is the most common deletion in CLL (reported in 10-20% of cases with conventional cytogenetics and 64% by FISH analysis)
• When present as sole abnormality this deletion is associated with a good prognosis with median survival longer than for CLL patients with normal karyotype
• Started on Ibrutinib
• Will continue prednione until Hgb >10, then began slow taper over course of 4-6 months
• PIP prophylaxis with Bactrim while on high dose steroids; also Folic acid, Protonix

Key Points

In virtually all cases of warm AIHA cross-matches will be incompatible

- Auto-antibodies will recognize blood group antigens and react to the vast majority of donor red cells. Consequently, ABO and RhD matched blood should be given in severe anemia requiring urgent blood transfusion.
- The blood should be given slowly with careful monitoring for transfusion reaction, though the risk of this is low (approaching zero without previous sensitization to foreign antigens; less than 10% with previous transfusion or pregnancy

There is a substantial risk of VTE in patients with AIHA, 15-30%

- It is important to evaluate for concomitant PE in patients presenting with AIHA, especially as these patients also frequently present with dyspnea, hypoxia and tachycardia

AIHA is often a chronic, relapsing condition.

- Steroid courses of at least 6 months as well as the addition of Rituximab can decrease recurrences of AIHA

Lymphoproliferative disorders, and specifically CLL, are the most common cause of secondary warm AIHA.

- Given relatively high rate of AIHA in CLL workup for hematologic malignancy should be considered in patients presenting with apparently idiopathic AIHA.

References