Navigating the Challenges: Management of Life-Threatening Pure Red Cell Aplastic Anemia Without Transfusion

Ellen Choi, DO; Yu-Han Chen, MD; Todd Lavine, MD; **Vivienne Qie, MD**; Tanganyika Barnes, DO

Introduction

Pure red cell aplasia (PRCA) is a rare disorder marked by the bone marrow's inability to produce mature red blood cells.

Causes of PRCA include various autoimmune, lymphoproliferative disorders, viruses, solid tumors (strong association with thymoma), and idiopathic in nature.

Most common presentation is isolated anemia with reticulocytopenia, leading to symptoms such as fatigue, shortness of breath, and pallor.

Case Presentation

We present a 64-year-old Jehovah's Witness female with stage IV thymoma who was referred to the emergency department by her oncologist after routine laboratory tests revealed a critically low hemoglobin level. She had completed chemotherapy, including cisplatin, doxorubicin, and cyclophosphamide, two months prior to presentation.

(-) Denied shortness of breath, fatigue, palpitations, or weakness.

(+) Upon arrival, tachycardic with a pulse of 122 bpm.

Initial labs: Hemoglobin 4.1 g/dL, reticulocytes 0.2%, white blood cell count 20 k/μL

Work-up:

Bone marrow biopsy on Day 4 confirmed PRCA, showing mildly hypercellular marrow, mild myeloid hyperplasia, increased T cell infiltrate, decreased erythroid precursors on CD71 immunostaining.

Fluorescence in situ hybridization and next-generation sequencing ruled out myelodysplastic syndrome.

Management Strategy

Day of admission

- Outpatient labs showed Hgb of 3.9, sent into ED

Admitted for bloodless management protocol:

- Retacrit 20,000u IV Q12h x 5 days
- IV iron daily x 10 days
- Folic acid 1mg daily
- Vitamin B12 IM x 1 dose
- 100% O2 therapy
- Strict bed rest
- Daily 12-lead ECG
- Propranolol (to minimize O2 consumption)
- Palliative care consulted

Bone marrow biopsy

- Hgb 3.0
- Empiric prednisone 1mg/kg started for suspected PRCA

Biopsy results

- Hgb 2.6
- Bone marrow biopsy results confirm PRCA, ruled out myelodysplastic syndrome
- Started on **Cyclosporine** 3mg/kg oral twice daily

Increase in O2

- Hgb 2.4
- Lactate 4.2, likely due to tissue dysoxia
- Transitioned to high flow nasal cannula
- Patient remains asymptomatic, no shortness of breath, fatigue

Uptick in hemoglobin

- Hgb 3.6, reticulocytes are responding, now >20%
- Switched back to nasal cannula

Discharge!

- Hgb 5.9
- Discharged to subacute rehab

Medications:

- Cyclosporine 125mg twice daily
- Retacrit 40,000u once weekly
- Folic acid 1mg daily
- Prednisone 40mg daily

Follow-up labs in 1 month: Hgb 13.0

Results

Day	Hemoglobin (g/dL)	Reticulocyte %	Absolute Reticulocyte Count (m/μL)
1	4.1	0.2	0
4	3.0	0.2	0
13	2.6	0.2	0
20	**2.1**	0.3	0
23	2.4	7.9	0.05
26	3.6	>20.0	0.37
29	5.9	>20.0	0.63
1-month follow-up	13.0	1.5	0.06

Discussion

This case illustrates successful **bloodless** management of severe anemia due to PRCA in a Jehovah's Witness patient without the use of blood transfusion.

Early identification of the underlying cause of anemia was important in preventing fatal complications such as multi-organ failure and death.

Diagnoses of PRCA can be complicated by chemotherapy-induced anemia or cancer-related anemia.

In this case, supportive bloodless measures were essential in stabilizing the patient until immunosuppressive therapy took effect.

Despite challenges in patient declining transfusion due to religious beliefs, respecting patient's wishes is important in treatment decisions.

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1. Badireddy, M., & Baradhi, K. M. (2018). Chronic anemia.
2. Mangla, A., & Hamad, H. (2019). Pure red cell aplasia.
3. Means Jr, R. T. (2016). Pure red cell aplasia. *Blood, The Journal of the American Society of Hematology*, *128*(21), 2504-2509.
4. Turner, J., Parsi, M., & Badireddy, M. (2023). Anemia. In *StatPearls [Internet]*. StatPearls Publishing.
5. Xavier, R. D., Devaraj, S., Sadasivam, V., Prakasam, O., Menon, N., Hariharan, A., & Sundaramoorthi, T. (2020). Thymoma associated with pure red cell aplasia: a case report and literature review. *Indian Journal of Thoracic and Cardiovascular Surgery*, *36*(4), 404-408.