Autoimmune Hemolytic Anemia Mimicking a Delayed Hemolytic Transfusion Reaction in a Patient with Primary Myelofibrosis: A Diagnostic Challenge

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BACKGROUND

AIHA is an uncommon complication in primary myelofibrosis (PMF). In transfusion-dependent patients, AIHA and delayed hemolytic transfusion reaction (DHTR) may both present with post-transfusion anemia and laboratory evidence of hemolysis, making differentiation difficult.

CASE PRESENTATION

- 54-year-old woman with JAK2-negative PMF; DIPSS Intermediate risk 2.
- Treated with erythropoietin, ruxolitinib and eltrombopag; subsequently became transfusion dependent.
- Two weeks after a red-cell transfusion, presented with severe symptomatic anemia.
- FBC: Hb 4.8 g/dL with thrombocytopenia and a leucoerythroblastic blood picture.
- Hemolysis: bilirubin 46.8 µmol/L; LDH 818.5 U/L,Retic count 3.5%

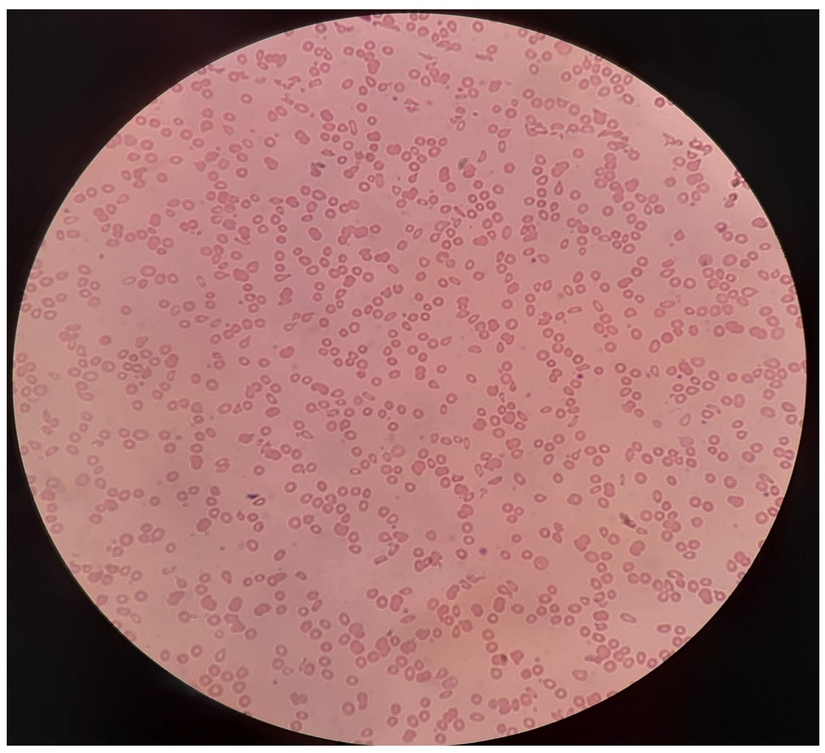
IMMUNOHAEMATOLOGY FINDINGS

DAT: Polyspecific 4+, C3d 4+, IgG +

ANTIBODY SCREEN: Pan reactive auto antibodies.

AUTOCONTROL : Positive

ELUATE: No allo antibody detected



Peripheral Blood Smear: Evidence of Hemolysis.

DIAGNOSTIC CHALLENGE

2 weeks post-transfusion → **Severe anemia + hemolysis** → **DHTR suspected** → **Pan reactive Auto Antibodies +** → **Eluate :No allo antibodies**

AIHA SUSPECTED

Why the distinction mattered: the temporal relationship to transfusion and poor hemoglobin increment suggested DHTR, but the serological pattern and absence of an identifiable alloantibody supported autoimmune hemolysis.

MANAGEMENT & RESPONSE

1) Corticosteroids
Partial improvement

2) Persistent hemolysis
IVIG required

3) After IVIG
Clinical improvement; Hb 8.8 g/dl

4) Oral prednisolone
Further Hb improvement

5) Latest status
Hb 11 g/dL; steroids tapering

KEY LEARNING POINTS

- Post-transfusion hemolysis in PMF should not automatically be attributed to DHTR.
- DAT, antibody screening, auto control and eluate studies are central to the differential diagnosis.
- Failure to identify an underlying alloantibody, together with the serological pattern, supported AIHA in this case.
- Early recognition allowed prompt immunomodulatory therapy and subsequent hematological improvement.

CONCLUSION

AIHA should be considered in transfusion-dependent PMF patients presenting with post-transfusion hemolysis. Comprehensive immunohaematological evaluation is essential to distinguish AIHA from DHTR and guide appropriate therapy.

Key words: Primary myelofibrosis • AIHA • DHTR • DAT • Transfusion medicine