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ABSTRACT N. PO14

SEARCHING FOR SAFER RESCUE IMMUNOSUPPRESSIVE THERAPY PROTOCOLS IN ACQUIRED HEMOPHILIA A: TWO CASE REPORTS.

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BACKGROUND

Eradication of autoantibodies (inhibitors) against Factor VIII (FVIII) by immunosuppressive therapy (IST) is a cornerstone of treatment of acquired hemophilia A (AHA). First-line IST usually relies on corticosteroids, while cyclophosphamide or rituximab are used/combined as rescue treatment. IST is associated with significant risk of adverse events, in particular infections/sepsis. We describe 2 cases of successful alternative rescue IST, aimed at minimizing toxicity in very elderly patients.

CASE REPORTS

1. A 81-yrs old lady with history of rheumatoid arthritis (RA) and Evan's syndrome on prednisone 5 mg presented to the emergency room (ER) with large hematomas. Prolonged APTT, severe isolated FVIII deficiency (0.02 IU/mL) and high-titer FVIII inhibitor (16.7 BU) led to diagnose AHA. The patient received hemostatic therapy and prednisone 1 mg/kg/-day, without any FVIII improvement after 2 weeks. In the light of the relevant autoimmune background, rituximab was given according to the RA schedule (1000 mg infusions, day 1 and 14), in parallel with prednisone tapering. FVIII increased to 0.16 IU/ml on day 15, 0.46 on day 21 and 0.63 IU/mL on day 30. The patient did not show any adverse event, including signs/symptoms of infection or laboratory abnormality.

2. A 90-yrs old lady was diagnosed idiopathic AHA in August 2024 (FVIII 0.001 IU/mL, inhibitor 10 BU). Prolonged first-line IST (prednisone 1 mg/Kg) resulted in a slow increase of FVIII levels (0.3 IU/mL after 6 weeks, peaking 0.7 IU/mL 4 weeks later), while metabolic side effects were experienced. On prednisone tapering, FVIII reduction and inhibitor relapse occurred (0.12 IU/mL and 1.5 BU, respectively, January 2025). Few days later, the patient presented with left iliopsoas muscle hematoma, promptly treated. A recently reported off-label rescue IST combined schedule (CyDRi) was given: cyclophosphamide (1000 mg) on days 1 and 22, dexamethasone (40 mg) and low-dose rituximab (100 mg) on days 1, 8, 15, 22. A prompt response was observed, FVIII being 0.23 IU/mL on day 5, 0.79 IU/mL on day 16 and 1.11 IU/mL on day 32. Prednisone was tapered and withdrawn. The patient showed asymptomatic leucopenia, lymphopenia and severe neutropenia (1030, 430 and and 190/mmc, respectively). Two filgastrim administrations led to white blood cell recovery and acyclovir prophylaxis was associated for 4 weeks. No further complications occurred.

CONCLUSIONS

In these patients experiencing inhibitor relapse or failure after first-line steroid therapy, IST rescue schedules not usually administered in AHA were successfully given. Bolus high-dose administration of immunosuppressive drugs may reduce toxicity associated with chronic treatment and combined drug schedules allow lower doses, as proposed for rituximab in the CyDRi. Alternative IST schedules are crucial to balance efficacy and safety of IST in this setting of elderly patients often with multiple comorbidities.

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