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EMICIZUMAB IN ACQUIRED HAEMOPHILIA A.

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Background: Acquired hemophilia A (AHA) is a rare bleeding disorder caused by autoantibodies against factor VIII (FVIII), leading to a decrease in its plasma concentration. Bleeding occurs in 70-90% of patients. Treatment for bleeding episodes typically involves activated prothrombin complex concentrates (APCCs) and recombinant activated factor VII (r-FVIIa). Factor VIII concentrates are effective only with low inhibitor titers (<5 Bethesda Units). For patients who do not exhibit cross-reactivity, recombinant porcine FVIII concentrates (rpFVIII) present a potential therapeutic option. The strategy for eradicating inhibitors relies on immunosuppressive therapy. However, the time frame required for inhibitor eradication can be significantly extended in patients with high initial inhibitor levels, putting them at risk of bleeding due to the lack of effective preventive measures during this period. Emicizumab, a bispecific antibody mimicking FVIIIa, has shown efficacy in congenital hemophilia A prophylaxis and has been reported in AHA cases.

Case Report: We present the case of a 73-year-old male with a history of treatment-refractory AHA who presented to our attention. His prior treatment included corticosteroids (1 mg/kg/day) and cyclophosphamide (100 mg/day), followed by four weekly infusions of rituximab (375 mg/m^2) due to a poor response and ongoing multiple bleeding episodes. Initially, the patient received treatment with rpFVIII. However, the subsequent development of a porcine inhibitor necessitated a change in therapy to rhFVIIa. The patient had been diagnosed six weeks prior to our evaluation with a high inhibitor titer of 34 B.U., an activated partial thromboplastin time (aPTT) ratio of 2.37, and undetectable levels of factor VIII. He presented with several recurrent hematomas on his arm, lower right limb, and rectus abdominis muscle. These episodes required transfusions of packed red blood cells and treatment with rpFVIII at a dose of 100 U/kg of body weight. However, the subsequent onset of cross-reactivity to rpFVIII, with an inhibitor titer of 100 B.U., ruled out the use of susoctocog alfa as a treatment option. Consequently, the patient was treated with rhFVIIa at 90 ug/kg every 4-6 hours.

Given the poor and slow response to immunosuppressive therapy and the emergence of new hemorrhages, a decision was made to initiate prophylactic treatment with emicizumab. The regimen consisted of a subcutaneous injection of 6 mg/kg on the first day, followed by 3 mg/kg on the second day, and then a maintenance dose of 1.5 mg/kg weekly. Within two days of commencing emicizumab therapy, the patient no longer required antihemorrhagic treatment and was discharged from the hospital. Notably, after four weeks of emicizumab treatment, the inhibitor against factor VIII was eradicated.

Conclusions: Acquired hemophilia A can present significant challenges, even for experienced clinicians. The commonly used bypassing agents or rpFVIII in AHA bleeding patients, necessitate very close monitoring and often require hospitalization. We describe the successful treatment of a refractory patient who had been hospitalized for an extended period and was discharged after just one week following the initiation of Emicizumab. Emicizumab prophylaxis allowed for the achievement of remission without further bleeding.

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