

Treatment of bleeding episodes in individuals with a FVIII inhibitor utilises by-passing agents (BPA) such as recombinant activated factor VII and activated prothrombin complex concentrates. Recent studies have reported prevention of bleeds in patients with inhibitors using a novel monoclonal antibody, Emicizumab. We report a case where Emicizumab therapy led to the resolution of a massive retroperitoneal haematoma originating from the iliopsoas muscle, and allowed removal of a port-a-cath.

Methods: Case report.

Results: A 15-year-old male, who had failed immune tolerisation, and had a chronic high titre FVIII inhibitor (maximum 3328 BU/mL) presented in haemodynamic shock with a massive retroperitoneal bleed (estimated 2L, nadir Hb 64 g/L) despite daily BPA prophylaxis. He had a history of recurrent musculoskeletal bleeds (ABR >30), chronic arthropathy (HJHS 25) with subsequent mobility issues, regularly missed school due to bleeds, and previously suffered a pulmonary embolus (PE) during BPA therapy. The acute iliopsoas bleed was controlled with BPA therapy, and a port-a-cath was reinserted (complicated by a large wound haematoma) to facilitate intensified haemostatic therapy. An application for compassionate use Emicizumab was lodged a month before the bleed. Emicizumab therapy (3 mg/kg weekly sc injections for 4 weeks and 1.5 mg/kg weekly injections thereafter) was approved and commenced 14 weeks after the initial bleed. On Emicizumab the TEG normalised, the haematoma (pre-Emicizumab size 6 x 4 x 5 cm) fully resolved over 19 months, the ABR was 0, school attendance increased and the port-a-cath was removed without BPA therapy. No complications or adverse events were encountered on Emicizumab.

Discussion/Conclusion: Emicizumab is a novel monoclonal antibody that bridges FIXa with FX, activating FX and thereby bypassing FVIII. This case demonstrates the safe and effective use of Emicizumab for over 20 months in an individual with a prior history of PE complicating his severe haemophilia A with a chronic high titre FVIII inhibitor.

Disclosure of Interest: None declared.

were collected (pre and post RTX) up to last follow-up (FU): demographic, comorbidities, absolute lymphocyte count (ALC), serum protein electrophoresis (SPEP), antimicrobial prophylaxis, clinical manifestations of *Pneumocystis Jirovecii* (PJ), Herpes Zoster Virus (HZV) and Herpes Simplex Virus (HSV) infections, confirmed diagnosis of any infections.

Results: Overall, we analyzed 20 PTs with ITP treated with RTX (Table 1). All PTs had previously received first-line therapy. RTX was always administered according to "standard regimen" (4 weekly 375-mg/m²). 5 PTs did not receive AP: 2 PTs had the lower median age and 3 presented only hypertension as comorbidity. 2 PTs discontinued AP within 1 week due to allergic reaction. 11 PTs received AP for PJ with trimethoprim/sulfamethoxazole (TMP/SMX) and 10 with acyclovir (AC) for HZV. Therapy was maintained for 1 year(y) following 1st RTX. 3 PTs had ALC < 1000/mm³ at the end of RTX, value turned to normal after a mean of 4 m. SPEP was available in 8 PTs and showed no decrease in g-globulin level up to 1y post RTX. No clinical signs of infection by PJ, HZV and HSV were reported. Hospitalization or specific assay for any suspected infection during FU was never required.

Discussion/Conclusion: In our study, 55% of PTs received AP. PTs not treated with AP had a lower median age and less comorbidities. Assayed parameters did not show any persistent alteration, neither did clinically diagnosed infections occur. This analysis shows that AP could be suitable in a subgroup of PTs with ITP having higher risk of infections as elderly PTs or those with multiple comorbidities. However prospective studies are needed to better define this indication.

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P281 | Antimicrobial prophylaxis in patients with immune thrombocytopenia treated with rituximab: A retrospective analysis

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Introduction: Rituximab (RTX) increases the risk of viral and fungal infections. Although antimicrobial prophylaxis (AP) is used in patients (PTs) with hematological neoplasms receiving RTX, evidence is lacking in the field of immune thrombocytopenia (ITP). We here reported the role of AP in a group of PTs with ITP under RTX.

Methods: PTs with ITP treated at our Centre from January 2013 to January 2019 were retrospectively evaluated. The following data