The Immune System Wants What the Immune System Wants

O. McTabi, M. Reichmuth; Pulmonary and Critical Care Fellowship, Parkview Medical Center, Pueblo, CO, United States.

Introduction Primary Immune Deficiency (PID) refers to over 100 groups of disorders that lead to dysfunction of one or more parts of a person’s immune system. The estimated prevalence of these disorders in the United States is approximately 1 in 1200 live births and clinical presentation is variable. Intravenous immunoglobulin (IVIG) therapy can be overwhelming from the perspective of the physician as well as the patient. As of 2017, the Immune Deficiency Foundation lists 13 different products licensed for use in the United States to treat PID. Many variables can play a role in the availability of medications and understanding alternatives will benefit patients. Case Description A 55-year-old female with a history of PID has been on IVIG therapy for over 18 months. She was receiving Gammagard, an IV infusion which she received at home every 4 weeks. A shortage of Gammagard led to a change in her therapy to Gamunex-C. Approximately 2 days after her infusion of Gamunex-C she developed acute shortness of breath with hemoptysis and was admitted to the hospital with a new oxygen requirement of 3 liters per minute and bilateral diffuse ground glass infiltrates on her computed tomography (CT) scan concerning for diffuse alveolar hemorrhage. A vasculitis panel demonstrated elevated myeloperoxidase (anti-MPO) levels and an elevated anti-proteinase 3 (Anti-PR-3). However, her anti-neutrophil cytoplasmic antibodies were within normal limits. A bronchoscopy with serial aliquots confirmed diffuse alveolar hemorrhage. Infection was ruled out, as was a pulmonary embolism. She was placed on high-dose IV steroids and responded to therapy over the course of 3 days at which point she was discharged. Discussion The immune system can be unpredictable. The difference between Gammagard and Gamunex-C is the latter has a lower pH of 4.0 to 4.5, compared to 4.6 to 5.1. During the clinical trials for Gamunex, there were no cases of diffuse alveolar hemorrhage. The most serious reaction was one case of pure auto-immune red cell aplasia. Our patient’s reaction was also an auto-immune process. Our patient has not been able to go back to her Gammagard therapy, and remains on a prolonged oral corticosteroid taper and was able to come off supplemental oxygen. She is scheduled to visit with Immunology on an outpatient basis. This case demonstrated the importance of prescribing physicians being able to discuss the risks of therapies openly with their patients to better manage expectations.

This abstract is funded by: none