Introduction

Acquired factor V (FV) inhibitor is a rare cause of FV deficiency, with less than 200 cases reported in the literature, and with an estimated incidence is 0.09-0.29 per million person-years [1]. Factor V inhibitor has been associated with a number of conditions, including antibiotic use, sepsis, autoimmune diseases and malignancies. In particular among patients with FV inhibitors, cancer was detected in 9-16% of cases [2]. In addition, factor V inhibitors have been described after exposure to bovine thrombin, used as topical hemostatic agent in surgery [1].

The overall prognosis is good, even if the clinical spectrum varies widely from the more common occurrence of mild bleedings or absence of symptoms to the less frequent severe bleeding events. The diagnosis of FV inhibitor is based on increased prothrombin time (PT), activated partial thromboplastin time (aPTT), decreased FV levels, and confirmed by the presence of FV inhibitors titrated using the Bethesda method. The prognosis is closely related to the underlying disease and the eradication of the triggering factor should be a first goal of the treatment. There is scarce information on treatment, and recommendations are extrapolated from the more common management of factor VIII inhibitors, which is mainly based on immunosuppressive agents. In the last few years the use of the anti-CD20 monoclonal antibody rituximab has been demonstrated to correct coagulopathy in all the reported cases. However, due to the rarity of the disease, no clinical trials are available to establish the efficacy and safety of this treatment, and clinicians have to rely only on case reports [6]. We describe here the case of a patient with acquired FV inhibitor, who failed to respond to rituximab treatment.
At diagnosis the patient was asymptomatic for bleeding, hemoglobin levels were stable, and treatment with prednisone 1mg/Kg/day of body weight was started. After 2 weeks of treatment laboratory data were unchanged, the patient was clinically stable with the exception of rare cutaneous bruises. Treatment with cyclophosphamide 50mg/day was added, but 2 weeks later laboratory findings were still unchanged. Treatment with prednisone 1mg/kg/day was maintained and cyclophosphamide dosage was increased up to 200mg/day. Few days later the patient complained a back pain with diffusion to the right leg. A tomography showed a large hematoma of the obturator muscle. PT and aPTT levels persisted unmodified, factor V inhibitor was still undetectable, and factor V inhibitor title was reduced to 195 Bethesda Units/mL.

In contrast with other coagulation factors inhibitors, particularly those of factor VIII, the level of factor V inhibitor does not correlate with the severity of clinical bleeding [1]. In our case, as frequently reported in these patients, the clinical features of the disease were mild, notwithstanding the severe prolongation of both basal plasma coagulation tests PT and aPTT. As a matter of fact, it is quite common that the severity of laboratory findings rises clinical alarm in contrast to the relatively asymptomatic patients, confirming that the risk of bleeding does not correlate with the entity of PT and aPTT prolongation, nor with factor V and factor V inhibitor levels, with a relative discrepancy between laboratory and clinical manifestations. In this clinical contest the use of aggressive immunosuppressive treatment should be carefully evaluated, bearing in mind the large spectrum of associated adverse events. In our patient, after 8 months of steroidal treatment, the use of immunosuppressive therapy should be carefully evaluated, bearing in mind the large spectrum of associated adverse events.
a severe osteoporosis was developed, and the patient experienced a spontaneous vertebral fracture. Therefore, case reports suggest that rituximab treatment could be useful in patients with factor V inhibitors; however, due to the usually mild clinical manifestations, the use of aggressive immunosuppressive regimens should be carefully evaluated.

References


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