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ACQUIRED IDIOPATHIC HEMOPHILIA A IN PATIENT IN THE ELDERLY: A RARE CASE REPORT AND BRIEF DISCUSSION WITH A FOCUS ON RHEUMATIC DISEASES.

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BACKGROUND

Acquired Hemophilia A (AHA) is a rare secondary hemostasis disorder characterized by the development of anti-factor VIII autoantibodies. The pattern of bleeding is different from that seen in congenital hemophilia A: most patients have substantial skin, cutaneous mucosal and deep tissues, as muscle, bleeding. Its incidence is approximately 1/1.000.000, affecting mainly men and women over 60 years, equally, and the mortality varies from 7% to 22%. About half of the cases are idiopathic, but underlying conditions such as rheumatic diseases, malignancy and gestation may be associated. The clinical presentation is often obscure and requires a high degree of suspicion, since all laboratory tests are normal, except for anemia, hematocrit drop without visible bleeding site and an isolated enlargement of activated partial thromboplastin time (aPTT). The aim of the present study is to describe a case report of idiopathic AHA in an elderly patient.

CASE REPORT

A 84-years-old man with a history of falling from his own height, 15 days ago, developing ecchymosis in the left lower limb (LLL), and continuous falls of hematocrit. On physical examination, weight loss, pallor, tachycardia and tachypnoea were noted, as well as extensive ecchymosis on the posterior face of LLL and ipsilateral gluteus. Laboratory tests indicated anemia (Hb 7.0 g / dL, Ht 22.5%); and activated partial thromboplastin time (aPTT) of 128 seconds. Abdominal computed tomography showed extensive hematoma in the left iliopsoas muscle with adjacent free fluid (see in picture). AHA hypothesis was proposed, and factor VIII activity (FVIIIA) was requested, whose result was 0.4% (range value> 50%). A negative lupus anticoagulant and aPPT with AHA positive pool were performed. Prednisone 1mg/kg/day, was initiated and, after 4 weeks, FVIIIA was normalized and anemia was progressively regressing.

CONCLUSION

AHA is a rare autoimmune disease with significant potential for severity and mortality. General practitioners and emergency physicians are often the first to address these patients and therefore it is imperative that they know the disease. The clinical-laboratory peculiarities build a challenging diagnostic scenario and may motivate the investigation of underlying conditions, especially in the elderly, such as malignancy and even autoimmune diseases such as Systemic Lupus Erythematosus, Rheumatoid Arthritis, Inflammatory Bowel Disease, among others. Treatment of HAA should include the approach of comorbidities, if present and, when early onset, favors a better prognosis.