

CHOLECYSTECTOMY IN A MAN WITH HEMOPHILIA A AND INHIBITOR ON EMICIZUMAB PROPHYLAXIS: A CASE REPORT

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There are few reports about the hemostatic effectiveness and safety of emicizumab on major surgeries, especially non-orthopedic ones. We described a man with severe hemophilia A and high-responding inhibitor under emicizumab prophylaxis who was submitted to an open cholecystectomy due to acute-on-chronic calculous cholecystitis. A 32-year-old man first visited the Emergency Department complaining severe diffuse abdominal colic unresponsive to oral analgesics, in the last 7h. He denied fever, nausea, or diarrhea. He had severe hemophilia A with high-response inhibitor, receiving prophylaxis with emicizumab (1.5 mg/kg weekly). He also had been submitted to several abdominal surgeries (appendectomy, midgut volvulus, and abdominal trauma). Physical examination was unremarkable. Aspartate and alanine aminotransferases were mildly elevated (56 and 83 U/L, respectively). He was discharged home after ameliorating with dipyrone, tramadol, and bromopride. He came back 4 days later, with the same complaints. He had a positive Murphy's sign and a negative Blumberg's sign. Blood tests were unremarkable. The gallbladder had a normal wall thickness (0.6 cm) with several mobile calculi (from 0.9 to 1.8 cm) by abdominal ultrasound, without dilation of the biliary tract, alteration of the pancreas/pancreatic duct, or lymphadenomegaly. He declined hospitalization and returned 5 days later, after having discussed with his Hematologist about the surgical procedure. Emicizumab was maintained throughout the described period. Before the procedure, he received intravenous rFVIIa 93 µg/kg and tranexamic acid (TxA) 1 g. rFVIIa was maintained each 2h, during the first 24h. The procedure was performed under general anesthesia. Videolaparoscopic cholecystectomy was withheld, due to adhesions precluding peritoneal inflation. Upon a right subcostal incision, a hydropic gallbladder was removed (anatomopathological analysis confirmed acute-on-chronic calculous cholecystitis).



A purified pigskin sterile absorbable gelatin sponge was placed on his hepatic bed, before closing the surgical wound. The surgeon reported an above-the-usual bleeding during the procedure (estimated blood loss of 500 mL against usually minimal blood loss). Ceftriaxone and metronidazole were prescribed throughout the hospitalization. rFVIIa 77 µg/kg was scattered on a daily base: every 2h until every 12h along 1 week (total 2,400 µg/kg). TxA 1 g every 8h was prescribed during the same period. He had mild hematic drainage (less than 30 mL on the post-operative day and decreasing in the following days). This was judged as good hemostasis by the surgeon. The drain was removed in the 4th post-operative day. Hemoglobin decreased from 13.7 g/dL at the day of the procedure until 9.4 g/dL the day before discharge (lowest 8.9 g/dL). No blood transfusion was required throughout the hospitalization. He was discharged home 1 week after surgery. No bleeding event was reported during the outpatient follow-up within 3 weeks. No symptoms nor signs of thrombosis were reported. In conclusion, we would like to reinforce the importance for the surgeons (a) to know that people with hemophilia A, with or without inhibitors, are susceptible to the same risks of developing gastrointestinal diseases as individuals without hemophilia; (b) in such cases, an interdisciplinary approach is always the best option; and (c) there is a biopharmaceutical which may effectively and safely reduce the bleeding risk in the periprocedural period.

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DEFICIÊNCIA COMBINADA DE FATOR V DE LEIDEN + FATOR VIII: RELATO DE CASO

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Introdução: A deficiência combinada de fator V e de fator VIII é um distúrbio hemorrágico genético autossômico recessivo, que cursa com reduções simultâneas dos níveis plasmáticos de ambos fatores de coagulação. A prevalência estimada é de um caso por milhão, sendo extremamente raro. **Objetivo:** Relatar caso de uma paciente que apresentou deficiência combinada de fator V e fator VIII. **Relato de caso:** Paciente feminina, 21 anos, previamente hígida, com histórico de menarca aos 12 anos e colocação de dispositivo intrauterino com levonorgestrel aos 16 anos. Encaminhada ao hematologista para avaliação de menometrorragia com até 20 dias de duração por ciclo menstrual. Refere ainda quadro de equimoses frequentes e sangramento gengival por 36 horas depois de um procedimento dentário. Foram realizados exames para investigação que evidenciaram hemograma e provas de coagulação normais, mas com alteração nos fatores de coagulação V (62,7%) e VIII (60,9%). O diagnóstico final foi de deficiência combinada do fator V e do fator VIII, e a conduta instituída foi de orientações e transfusão de plasma fresco congelado em caso de sangramento, de trauma ou de necessidade de cirurgia. **Discussão:** A deficiência combinada de fator V e fator VIII

