Hemophilia

Mild hemophilia without spontaneous bleeding can remain undiagnosed for a lifetime. However, intracranial hemorrhage is one of the most serious complications for patients with hemophilia. In addition, hemorrhagic complications after emergency surgery tend to arise from coagulopathy.

In people with hemophilia type A and B who have a deficiency of factors VIII and IX, these two factors are administered for controlling bleeding or as prophylaxis medication before starting surgeries. However, in some cases they subsequently develop neutralizing antibodies (NAb) against the drug. These NAbs increase over time and inhibit the action of (coagulation) in the persons body. rFVIIa, which is activated form factor VII, bypasses factors VIII and IX and causes coagulation of blood without the need for factors VIII and IX. This is important for some patients to shift to proper blood factors according to their level of NAbs. Other indications include use for patients with acquired hemophilia, people born with a deficiency of factor VII, and people with Glanzmann's thrombasthenia.

Desmopressin causes an increase in factor III coagulant activity and von Willebrand factor which helps coagulation and platelet activity in mild hemophilia type A and in von Willebrand’s disease Type I (where the factors are normal in makeup but low in concentration, ✖ but may cause thrombocytopenia in von Willebrand’s disease Type IIB where factors may be abnormal or missing). May prevent abnormal bleeding in minor procedures. Not all patients with mild hemophilia A or von Willebrand disease respond to DDAVP. ℞ 0.3 mcg/kg (use 50 ml of diluent for doses ≤ 3 mcg, use 10 ml for doses > 3 mcg) given over 15– 30 minutes, 30 minutes prior to a surgical procedure.

Case reports

Not uncommonly, intracranial bleeding is the first sign of a severe inherited coagulation disorder. In the presence of an unexpected intracranial bleeding after a minor trauma or without a clear history of the related events, physicians and caregivers may be confronted to the dilemma of a possible child abuse. It must be bear in mind that physical abuse and bleeding disorders can co-exist in the same child.

Hinojosa et al. reported the case of two siblings in whom a diagnosis of hemophilia coexisted with the presumption of a non-accidental head trauma. Child abuses were inflicted in both children with a spare time of 2 years. A diagnosis of mild hemophilia was prompted in the first sibling after initial NAHT, while inflicted trauma was evident in the second sibling after neuroimaging findings and concomitant lesions. Lessons from this case in co-existing bleeding disorders and inflicted trauma and legal implications derived will be discussed thereafter.The possibility of a bleeding disorder should be considered in all children presenting with unexplained bleeding at a critical site in the setting of suspected physical maltreatment, particularly intracranial hemorrhage (ICH) [1].

An 80-year-old man was admitted with left hemiparesis and disturbed consciousness. He had no history of trauma, fever, or drug and alcohol intake. Computed tomography imaging upon admission

disclosed a hemispheric subdural hematoma with a midline shift. No vascular abnormalities were identified as a source of the hemorrhage. The hematoma was removed on an emergency basis with external decompression. However, a large subcutaneous hematoma was again evident on the following day. Insufficient hemostatic maneuvers during surgery were considered the cause of this hemorrhagic complication. A second operation was performed to achieve hemostasis of the subcutaneous and muscle tissue. Thereafter, he was rehabilitated without treatment for hemophilia as he had no bleeding episodes. Cranioplasty proceeded using artificial bone at 40 days after the first operation. However, epidural hematoma developed again on postoperative day 1. His neurological status did not worsen so a repeat procedure was unnecessary. Close scrutiny uncovered a diagnosis of mild hemophilia A.

Accurate diagnosis is important for the management of postoperative hemorrhagic complications caused by pathologies of the coagulation system. Sufficient hemostasis of hemorrhage from subcutaneous and muscle tissue is essential even during emergency surgery to avoid postoperative complications. A diagnosis of hemophilia should be considered in the face of prolonged activated partial thromboplastin time (APTT) [2].

1)
Hinojosa J, Simó M, Armero G, Becerra MV, Alamar M, Candela S, Culebras D, Muchart J, Berrueco R. Hemophilia and non-accidental head trauma in two siblings: lessons and legal implications. Childs Nerv Syst. 2022 Oct 28. doi: 10.1007/s00381-022-05713-2. Epub ahead of print. PMID: 36303077.
2)
Ono H, Sase T, Takasuna H, Tanaka Y. Mild hemophilia A presaged by recurrent postoperative hemorrhagic complications in an elderly patient. Surg Neurol Int. 2017 Sep 6;8:205. doi: 10.4103/sni.sni_235_17. eCollection 2017. PubMed PMID: 28966812; PubMed Central PMCID: PMC5609363.