

An Important Factor in Preoperative Screening

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A previously healthy 25-year-old Guatemalan man presented to the emergency department with 1 day of fever, nausea, vomiting, and right lower quadrant abdominal pain. A computed tomography (CT) scan revealed acute appendicitis. The patient underwent an uncomplicated laparoscopic appendectomy and was discharged in stable condition after 48 hours.

Five days after the operation he returned to the emergency department with abdominal pain, nausea, vomiting, and lightheadedness. He was tachycardic, and his hemoglobin was 9.5 g/dL (normal, 13.3–17.7 g/dL), decreased from 14.4 g/dL prior to his appendectomy. A CT scan showed intraperitoneal blood with active extravasation of contrast at the site of the appendectomy.

Additional laboratory testing revealed an activated partial thromboplastin time (aPTT) of 52 seconds (normal, <37 seconds) and protime (also prothrombin time [PT]) of 14 seconds (normal, <14.1 seconds). The platelet count was 449,000/ μ L (normal, 150–400,000/ μ L) and the fibrinogen level was 337 mg/dL (normal, 170–440 mg/dL). Crystalloid and packed red blood cells were administered. Since further laboratory evaluation of the prolonged aPTT was not immediately available, the patient was empirically treated with fresh frozen plasma (FFP), cryoprecipitate, and Factor VIII/von Willebrand factor concentrate. At laparotomy, bleeding was observed at the previous operative site, and 2 L of intraperitoneal blood was evacuated.

The next morning, Factor VIII and Factor IX (FIX) activities and the ristocetin cofactor study performed on specimens obtained immediately prior to the second operation were normal, but the FIX activity was 5% of normal. The diagnosis of FXI deficiency was made and 2 to 3 units of FFP (the amount necessary to maintain the patient's measured FIX activity near 20% of normal) were transfused daily. Nine days of FFP infusions were required to achieve complete wound hemostasis. The patient had no further bleeding episodes after discharge.

Upon further interviewing, the patient revealed that 2 months prior he sustained a small laceration on his arm

that “bled for a long time” and that his brother had experienced prolonged bleeding after a dental extraction.

Commentary

Routine performance of preprocedural laboratory testing, and complete reliance on the results as a means of excluding a propensity to bleeding, may not only lead to excessive testing and delayed procedures, but also provides false reassurance because normal routine laboratory studies cannot be used to exclude some bleeding disorders (Table 1).

Most studies evaluating routine laboratory testing of hemostatic variables prior to invasive procedures come from patients undergoing elective general surgery. A 1988 study concluded that there is no benefit in the routine preoperative use of the PT, aPTT, platelet count, and bleeding time in the absence of clinical evidence of a hemostatic defect, as assessed by a patient questionnaire and a thorough physical examination.¹ A subsequent European, prospective, multicenter study confirmed that abnormalities of preoperative laboratory screening in the absence of a history of bleeding or clinical abnormality were not associated with worse surgical morbidity or mortality, compared to patients with normal screening laboratory studies.² A recent systematic review has also confirmed the poor positive predictive value of screening tests when used in isolation, and recommended a history-based and physical exam-based approach.³ Questionnaires have been shown to be particularly important tools for eliciting clinically significant bleeding disorders that may require revision of the surgical plan.^{1,4}

FXI is a serine protease whose activity is crucial for robust fibrin clot formation and inhibition of fibrinolysis at sites of vascular injury.⁵ FXI deficiency is an autosomal recessive disorder with an incidence of 1 per 1,000,000 in the general population, with a significantly higher incidence in the Ashkenazi Jewish population. While the risk of spontaneous hemorrhage is typically low, life-threatening bleeding may occur after surgery or trauma. The severity of the

TABLE 1. Disorders of Hemostasis Not Detected Routinely by the Activated Partial Thromboplastin Time, Protome, or Platelet Count

Von Willebrand disease
Mild hemophilia A (Factor VIII deficiency)
Mild hemophilia B (Factor IX deficiency)
Mild hemophilia C (Factor XI deficiency)
Qualitative platelet disorders (congenital or acquired)
Factor XIII deficiency
Disorders of fibrinolysis (eg, antiplasmin deficiency, plasminogen activator inhibitor type 1 deficiency)
Disorders of the vasculature or integument (hereditary hemorrhagic telangiectasia, Ehlers-Danlos syndrome)

measured FXI level deficiency does not always correlate with risk of bleeding. Periprocedural prophylaxis and treatment of bleeding aim to replace FXI to the low-normal range by administering FXI concentrate, (not available in the United States) or FFP. Antifibrinolytic agents such as tranexamic acid or ϵ -aminocaproic acid may be used adjunctively in cases of mucosal bleeding.⁵

In this case, preoperative screening, either using a questionnaire or careful history-taking, would have identified the patient's personal and family history of bleeding and prompted appropriate preoperative coagulation testing, which could have exposed the hemostatic defect, allowing for modification of the perioperative medical plan.

In summary, preoperative bleeding evaluations should be performed routinely and should begin with a careful history (use of a questionnaire may be considered) and physical examination. Excessive bleeding after prior surgery, trauma,

dental extractions, parturition, or circumcision; bleeding tendency in family members; current use of medications that may increase bleeding risk (such as anticoagulants or aspirin); and physical signs associated with bleeding should be assessed. If clinical details fail to expose a potential bleeding disorder, it is safe and cost-effective¹ to proceed with surgery without performing additional laboratory testing. In contrast, any abnormality on the clinical assessment should trigger preoperative laboratory analysis of basic hemostatic parameters, which may prompt further testing or hematology consultation.

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