The NEW ENGLAND JOURNAL of MEDICINE

CLINICAL PRACTICE

Caren G. Solomon, M.D., M.P.H., *Editor*

Immune Thrombocytopenia

Nichola Cooper, M.D., M.B., B.S., and Waleed Ghanima, M.B., Ch.B., Ph.D.

This Journal feature begins with a case vignette highlighting a common clinical problem. Evidence supporting various strategies is then presented, followed by a review of formal guidelines, when they exist. The article ends with the authors' clinical recommendations.

A 72-year-old woman who is receiving apixaban for atrial fibrillation but otherwise does not have a clinically significant medical history presents to the hospital with lower gastrointestinal bleeding. On admission, her hemoglobin level is 8.5 g per deciliter, platelet count 2000 per cubic millimeter, and white-cell count 5300 per cubic millimeter. She receives a transfusion of packed red cells and platelets that results in an increase in the hemoglobin level and a decrease in bleeding, but only a transient increase in the platelet count. The examination is unremarkable. A peripheral-blood smear shows no abnormalities other than thrombocytopenia; these findings are consistent with a diagnosis of immune thrombocytopenia. How should this case be managed?

THE CLINICAL PROBLEM

Immune thrombocytopenia (ITP) is an autoimmune disease characterized by isolated thrombocytopenia. Patients may be asymptomatic at presentation or they may present with mild mucocutaneous to life-threatening bleeding. Although only 5% of patients with ITP present with severe bleeding,[1] bleeding leading to hospital admission within 5 years after diagnosis develops in approximately 15%.[2] Irrespective of bleeding problems, patients with ITP often report fatigue and impaired health-related quality of life.[3] The risk of venous thromboembolism is twice as high among patients with ITP as among persons in the general population; the management of venous thromboembolism may be especially problematic given the concomitant risk of bleeding.[4]

ITP may be a primary condition or it may be caused by other diseases. The differential diagnosis of thrombocytopenia and the potential secondary causes of ITP are outlined in Table 1. Overall, the incidence of ITP ranges from 2 to 4 cases per 100,000 person-years, with two peaks: one between 20 and 30 years of age with a slight female predominance and a larger one after 60 years of age with equal sex distribution.[5,6] Although some patients have one episode of ITP followed by an immediate remission, chronic ITP develops in up to 70% of adults with this condition. Both spontaneous and treatment-induced remission can occur many years after diagnosis.

The pathophysiology of ITP is complex and remains incompletely understood (Fig. 1). The traditional concept is that antibody-coated platelets are prematurely destroyed in the spleen, liver, or both through interaction with Fcγ receptors.[7] Autoantibodies can also induce complement-mediated or desialylation-induced

From the Department of Medicine, Hammersmith Hospital, Imperial College, London (N.C.); and the Departments of Medicine, Hematology–Oncology, and Research, Østfold Hospital Trust, Kalnes, and the Department of Hematology, Oslo University Hospital and Institute of Clinical Medicine, University of Oslo, Oslo (W.G.) — both in Norway. Address reprint requests to Dr. Cooper at the Department of Medicine, Hammersmith Hospital, Imperial College, Du Cane Rd., London W12 0HS, United Kingdom, or at n.cooper@imperial.ac.uk.

This article was updated on September 6, 2019, at NEJM.org.

N Engl J Med 2019;381:945-55.
DOI: 10.1056/NEJMcp1810479

An audio version of this article is available at NEJM.org