

Case Report

Immune Thrombocytopenia as a Consequence of Rocky Mountain Spotted Fever

Cherisse Baldeo^a Karan Seegobin^a Lara Zuberi^b

^aDepartment of Medicine, University of Florida, Jacksonville, FL, USA; ^bDepartment of Oncology, University of Florida, Jacksonville, FL, USA

Keywords

Immune thrombocytopenia · Rocky Mountain spotted fever

Abstract

Primary immune thrombocytopenia (ITP) – also called idiopathic thrombocytopenic purpura or immune thrombocytopenic purpura – is an acquired thrombocytopenia caused by autoantibodies against platelet antigens. It is one of the more common causes of thrombocytopenia in otherwise asymptomatic adults. Rocky Mountain spotted fever (RMSF) is a potentially lethal, but curable, tick-borne disease. We present a case of ITP that was triggered by RMSF.

© 2017 The Author(s)
Published by S. Karger AG, Basel

Introduction

Immune thrombocytopenia (ITP) is defined as an isolated platelet count of less than $100 \times 10^9/L$ ($100,000/\mu L$) and usually presents without symptoms [1]. The pathogenesis of ITP is caused by antibodies against platelet glycoproteins. Thrombocytopenia is a common manifestation of all tick-borne diseases which remains poorly understood [2]. ITP associated with infection may arise due to molecular mimicry.

Corticosteroids have been the backbone of initial treatment in ITP [3]. Patients who relapse are traditionally considered for splenectomy. Rituximab, a chimeric anti-CD20 monoclonal antibody that targets B cells, has gained popularity in treating ITP.

Case

A 20-year-old Caucasian male with no significant past medical history was diagnosed with Rocky Mountain spotted fever (RMSF) after a tick bite. He completed treatment with doxycycline for 10 days. However, 2 weeks later routine laboratory tests showed a platelet count of 9,000/mm³. The complete blood count was otherwise normal as well as the chemistry panel. The peripheral blood smear and coagulation panel were also unremarkable. He denied any bleeding. He had no prior history of thrombocytopenia and no family history of any hematological disorders. Vital signs were stable. Notably, he had a diffuse petechial rash. Physical examination was otherwise normal. The platelet count was rechecked with a citrated blood sample and showed that there was no evidence of platelet clumping.

He received platelet transfusions and steroids but his platelet count remained low (<10,000/mm³). He was treated with intravenous immunoglobulin and his platelet count normalized after 2 doses. However, 1 week later his platelet count dropped again to 9,000/mm³. He was transfused with platelets again but had no response.

Given that the platelet count did not show an adequate response to steroids and that he declined splenectomy, he was started on rituximab weekly. Intravenous immunoglobulin and steroids were also continued concomitantly. Fortunately, the platelet count normalized within the next 3 months and remained normal even after a 2-year follow-up.

Discussion

ITP is defined as an isolated platelet count of less than $100 \times 10^9/L$ (100,000/ μ L) and usually presents without symptoms [1]. Patients without symptoms who have a platelet count above $30 \times 10^9/L$ should generally not be treated unless they have an increased risk of bleeding [1]. A low platelet count may be the sole initial manifestation of ITP, as demonstrated in our case. The pathogenesis of ITP is caused by antibodies against platelet glycoproteins, most commonly platelet glycoprotein IIb/IIIa, the platelet fibrinogen receptor [1].

Thrombocytopenia is a common manifestation of all tick-borne diseases [2]. Quantitative changes in platelet counts associated with infection may result from decreased marrow production, hypersplenism, consumption due to widespread endothelial damage, or disseminated intravascular coagulation, as well as immune-mediated platelet destruction [2]. The pathogenesis of thrombocytopenia in many of the tick-borne diseases remains poorly understood, and therapy for this complication has been largely anecdotal and poorly addressed in the literature [2].

ITP associated with infection may arise due to molecular mimicry. That is, infection may result in amino acid sequences that may have structural similarity to regions within platelet glycoproteins. Thus, antibodies directed against the pathogen may cross-react with the glycoprotein, leading to thrombocytopenia [1]. This may perhaps have been the mechanism by which RMSF lead to the development of ITP in our case.

Due to their effectiveness, low cost, and convenience of use, corticosteroids have been the backbone of initial treatment in ITP [3]. However, in most patients the platelet count

decreases once the dose is tapered or stopped; remission is sustained in only 10–30% of cases [3]. Patients who relapse and have a platelet count of less than $20 \times 10^9/L$ are traditionally considered for splenectomy. More than two-thirds of patients respond with no need for further treatment [3]. Although splenectomy has the highest rate of durable platelet response, the risks associated with surgery are an important concern.

Rituximab is a chimeric anti-CD20 monoclonal antibody that targets B cells. Although initially approved for the treatment of lymphomas, rituximab has gained popularity in treating ITP due to its safety profile and ability to deplete CD20+ B cells responsible for antiplatelet antibody production by Fc-mediated cell lysis [3]. Based on results from Godeau et al. [4], rituximab may spare some patients from splenectomy, or at least delay it, as demonstrated in our case.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors declare that there is no conflict of interest regarding the publication of this paper.

References

- 1 Keith McCrae MD: Immune thrombocytopenia: no longer “idiopathic.” *Cleve Clin J Med* 2011;78:358–373.
- 2 Pantanowitz L: Mechanisms of Thrombocytopenia in Tick-Borne Diseases. *Internet J Infect Dis* 2002;2:2.
- 3 Swapna Thota MD, et al: Immune thrombocytopenia in adults: an update. *Cleve Clin J Med* 2012;79:641–650.
- 4 Godeau B, Porcher R, Fain O, et al: Rituximab efficacy and safety in adult splenectomy candidates with chronic immune thrombocytopenic purpura: results of a prospective multicenter phase 2 study. *Blood* 2008;112:999–1004.