THE NEW ARMENIAN MEDICAL JOURNAL
Volume17 (2023), Issue 3, p.91-94

DOI: https://doi.org/10.56936/18290825-2023.17.2-91

A RARE CASE OF THROMBOTIC THROMBOCYTOPENIC PURPURA COMPLICATED BY MACROVASCULAR EVENTS

Kumar-Anmol K., Chandrasekaran M.S., Adarsha G.K., Nitin Bhat N., Rao R.

Department of Medicine, Kasturba Medical College, Manipal, Manipal Academy of Higher Education, Manipal, India

Received 9.12.2022; accepted for printing 5.06.2023

Abstract

Thrombotic thrombocytopenic purpura is a rare but serious disease with high rates of mortality. It is a thrombotic microangiopathy associated with fever, thrombocytopenia, haemolytic anaemia and can also present with neurological and renal dysfunctions. It is caused by the reduced activity of ADAMTS13, a metalloprotease which cleaves von Willebrand factor resulting in widespread microvascular occlusion due to platelet rich thrombi. Although small vessel infarcts are common in kidney and brain, large vessel occlusions are rarely seen in thrombotic thrombocytopenic purpura.

We present a case of a 56-year-old woman who presented with symptoms of urinary tract infection subsequently diagnosed with Thrombotic thrombocytopenic purpura. The initial treatment regimen consisted of plasma exchange therapy followed by steroids and cyclophosphamide. She was gradually improving, however her course of disease was complicated by a large cerebral infarct and bilateral pulmonary artery thrombosis resulting in intensive care unit admission and prolonged hospitalization. The patient was subsequently started on anti-coagulant therapy with fondaparinux and monitored continuously due to the increased risks of haemorrhage. The patient gradually improved over time with sustained improvement in laboratory reports. The outcomes of anti-coagulant therapy were favourable in our case and patient was discharged with rituximab therapy on subsequent follow ups.

This case report intends to highlight the macrovascular thrombotic events and the challenge it brings to physicians regarding thrombolysis.

Keywords *TTP, cerebral infarct, pulmonary artery thrombosis, macrovascular.*

Introduction

Thrombotic thrombocytopenic Purpura, a rare life-threatening hematological condition has significant morbidity and mortality unless diagnosed and treated promptly [*Balasubramaniyam N et al., 2021*]. The underlying pathogenesis of thrombotic thrombocytopenic purpura (TTP) is a severe deficiency in ADAMTS13 (A Disintegrin and Metalloprotease with ThromboSpondin type 1 motifs 13) activity, a metalloprotease that cleaves large von Willebrand factor multimers [*Fujikawa K et al. ,2001; Levy G et al., 2001*]. In the absence of ADAMTS13 activity vWF polymers accumulate and cause severe platelet clumping with resultant microthrombi formation. This deficiency is either autoantibody mediated (acquired TTP) or due to deleterious mutations in the gene encoding ADAMTS13 (congenital TTP). It is classically characterized by the pentad of microangiopathic hemo-

Cite This Article as:

Kumar-Anmol K., Chandrasekaran M.S., Adarsha G.K., Nitin Bhat N., Rao R. (2023). A Rare Case Of Thrombotic Thrombocytopenic Purpura Complicated By Macrovascular Events; The New Armenian Medical Journal, vol.17(3), p 91-94; https://doi.org/10.56936/18290825-2023.17.3-91

Address for Correspondence:

Dr. Raghavendra Rao, Associate Professor
Department of Medicine, Kasturba Medical College
Udupi, Karnataka, Manipal 576014, India
Tel.: +91-9901731924
E-mail: ragsmanipal1983@gmail.com