IMAGE OF INTEREST

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A case of atypical hemolytic uremic syndrome presenting intermittent thrombocytopenia and hemolysis with fatal acute coronary syndrome

Hyun-Je Kim[1] and Young-Hoon Hong[2]

[1]Division of Rheumatology, Department of Internal Medicine, CHA Gumi Medical Center, CHA University, Gumi; [2]Division of Rheumatology, Department of Internal Medicine, Yeungnam University College of Medicine, Daegu, Korea

A 41-year-old female visited our emergency room for recurrent epigastric pain and diarrhea in the last 3 months after a general weakness and vague feeling of illness. The pain lasted for several hours to days and improved spontaneously. The patient had no history of spontaneous abortion or vaccination recently. She had normal blood pressure (110/70 mmHg), pulse (88/min), respiration (20/min), and body temperature (37.1°C). Her mentality was alert without focal neurologic deficit. Her hemoglobin (10.6 g/dL) and platelet count (9,000/μL) were decreased, but recovered completely to normal levels (15.2 g/dL, 255,000/mL) within several days, as abdominal pain disappeared. Serum haptoglobin was undetectable. Lactate dyhy-

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Correspondence to
Young-Hoon Hong, M.D.
Tel: +82-53-620-3841
Fax: +82-53-654-8386
E-mail: yhhong@med.yu.ac.kr
https://orcid.org/0000-0001-8119-0464

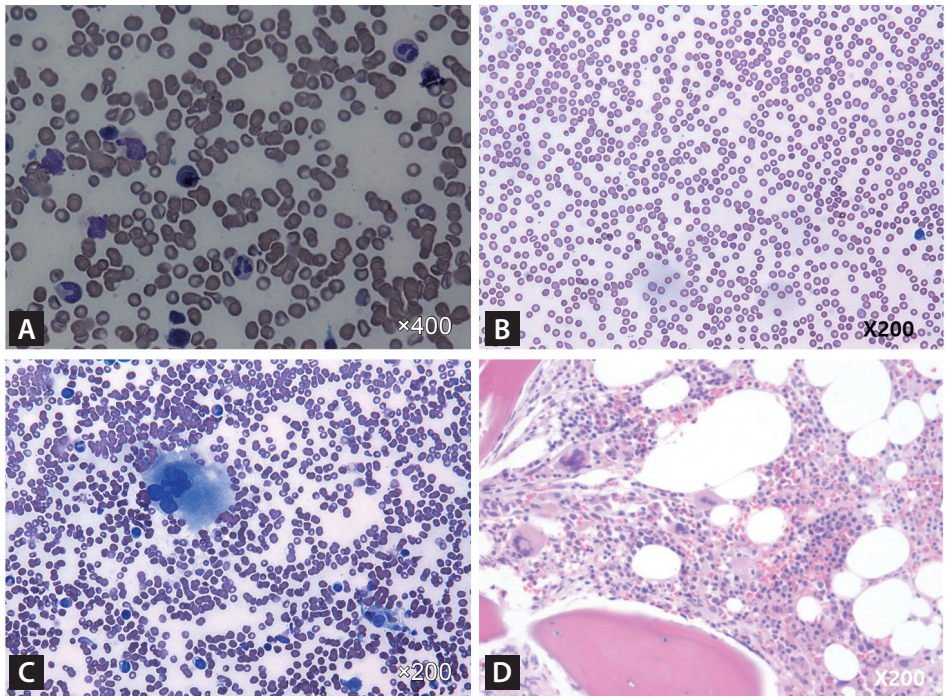

Figure 1. (A) Peripheral blood smear showing white blood cells (WBCs) shifting to the left of maturation (segment, 48%; band form, 23%; metamyelocyte, 3%; myelocyte, 2%) red blood cells (RBCs) were macrocytic hypochromic anemia with moderate poikilocytosis and RBC remnant (Wright's stain, ×400). (B) Platelets were markedly decreased in numbers (Wright's stain, ×200). (C) Bone marrow needle aspiration showing normocellular marrow particles. The granulocytic series was orderly matured. Erythroid series showed that some dysplastic feature such as karyorrhexis and megakaryocytes were increased in numbers with nuclear hyper-lobulation (Wright's stain, ×200). (D) Bone marrow needle biopsy revealing normocellular marrow with increased megakaryocytes (hematoxylin and eosin, ×200).

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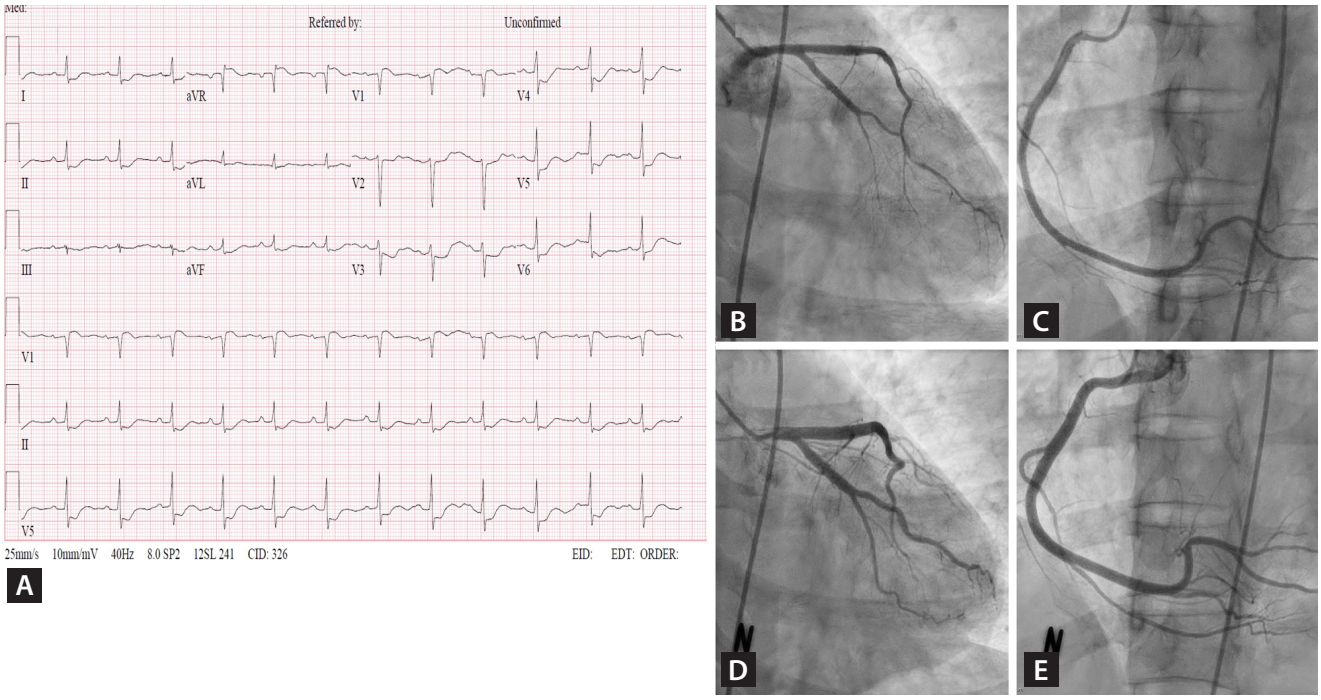

Figure 2. (A) Electrocardiogram showing ST segment elevation at lead aVR and V1 and ST segment depression at lead I, II, and lead V3 to V6. (B) Coronary angiography showing a diffuse irregular stenosis up to 80% at the distal left anterior descending (LAD) artery without septal branch, a diffuse irregular stenosis up to 90%–95% at the mid to distal left circumflex artery (LCX), and (C) a diffuse irregular stenosis up to 90% at the posterior descending artery (PDA) of the right coronary artery. Diffuse stenoses were completely recovered with intra-coronary nitrate injections. (D) Distal LAD was fully dilated and septal branch appeared. The mid to distal LCX was fully dilated. (E) PDA of right coronary artery was also fully dilated.

drogenase level was high (851 IU/L) with poikylocytosis and red blood cell remnants on peripheral blood smears (Fig. 1A and 1B). Anti-Ro antibody (Ab) was positive. Prothrombin time, activated partial thromboplastin time, fibrinogen, fibrinogen degradation production, D-dimer for disseminated intravascular coagulation, FANA, anti-dsDNA/anti-Smith/anti-phospholipid Abs, anti-neutrophil cytoplasmic antibody for autoimmune diseases, direct/indirect Coomb's tests for immune cytopenia, flow cytometry for paroxysmal nocturnal hemoglobinuria, human immunodeficiency virus tests, blood urea nitrogen/creatinine (9.9/0.59 mg/dL), C3/C4 (123.6/30.8 mg/dL), procalcitonin, C-reactive protein, a disintegrin and metalloproteinase with thrombospondin type 1 motif, 13 (ADAMTS-13), computed tomography, and positron emission tomography scans were all normal. Her stool cultures were negative for salmonella, shigella, and shiga toxin producing *Escherichia coli*. Bone marrow studies demonstrated erythroid karyorrhexis and increased megakaryocytes with hyperlobulated nuclei (Fig. 1C and 1D). With symptom free intervals, sudden bouts of abdominal pain and thrombocytopenia developed recurrently which recovered spontaneously. Under the diagnosis of atypical hemolytic uremic syndrome (HUS), associated genetic changes were tested but not identified (Table 1). During conservative treatment with glucocorticoids (20 mg/day) and hydroxychloroquine (400 mg/day), sudden onset of chest pain and cardiac arrest followed nausea and diarrhea. Coronary angiography showed diffuse stenosis of coronary arteries which recovered fully with intra-coronary nitrate injection (Fig. 2). Acute coronary syndrome of vasospastic variant angina was diagnosed, which was accompanied by acutely precipitating thrombocytopenia and anemia without haptoglobin as a HUS. Refractory to β-blocker and vasodilators, her acute coronary attacks occurred repeatedly which resulted in cardiac death.

This study was approved by the Institutional Review Board (IRB) of Gumi CHA Medical Center, CHA University (IRB No: GM 21-06). Written informed consent from the patient was waived by the IRB.

Conflict of interest

No potential conflict of interest relevant to this article was reported.

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