

Case Report

Hemorrhagic Stroke as the Manifestation of Neuropsychiatric Systemic Lupus Erythematosus

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ABSTRACT

Stroke is a major cause of mortality and disability. It can be classified into ischemic stroke and hemorrhagic stroke. Systemic Lupus Erythematosus (SLE) is a systemic autoimmune disease that can cause neuropsychiatric systemic lupus erythematosus (NPSLE) complications, one of those manifests as stroke, which happens often in young adults. Ischemic stroke is more common in SLE patient, but hemorrhagic stroke can also ensue. We present a case of SLE in a young woman with left-sided hemiparesis and convulsion. After undergoing clinical evaluation, laboratory testing, and radiological imaging, she was diagnosed with hemorrhagic stroke. She was treated conservatively; blood pressure control, steroid administration to improve the condition, and anticonvulsants.

Keywords: neuropsychiatric systemic lupus erythematosus; hemorrhagic stroke; stroke management

INTRODUCTION

Stroke is a major health problem that causes high mortality and morbidity rates both in the world and in Indonesia. Based on the pathology, stroke is classified into ischemic and hemorrhagic stroke. Several studies show that the mortality rate of hemorrhagic stroke in the first 30 days is 40%-50%, twice as high as in ischemic strokes, with only 27% of patients achieving independent functional outcome in 90 days after the stroke.¹

SLE is a systemic autoimmune inflammatory disease which characterized by autoantibodies and immune complexes in tissues, resulting in clinical manifestations in various organ systems including NPSLE. Cerebrovascular disease or stroke, neuropathy, acute confusional state, seizure, and psychosis are the common manifestations in NPSLE.²

The most common type of stroke in SLE is ischemic stroke, which is associated with comorbid conditions such as vasculitis, antiphospholipid

syndrome, hypertension, premature atherosclerosis, and complications from the use of immunosuppressants.³

Hemorrhagic stroke occurs as a result of cerebral blood vessels rupture, it can happen as a complication of hypertension, cerebral amyloid angiopathy, as well as abnormalities of the brain blood vessel such as an aneurysm and arteriovenous malformation.⁴ The risk of hemorrhage stroke in SLE increases due to the presence of endothelial dysfunction that raises the likelihood of vascular rupture in the brain.^{3,4}

This paper attempts to report hemorrhagic stroke that is relatively rare in SLE patients, to understand the diagnosis and the management of a hemorrhageal stroke on SLE.

CASE REPORT

An 18-years-old young woman came to emergency room with sudden left-sided weakness one hour before admitted. She also had fever, pulsing headache with numeric rating scale of 4. There was no loss of consciousness, face asymmetry, dysarthria, blurred or double vision, vertigo, paresthesia, or seizure.

She had history of SLE, nephrotic syndrome, and hypertension since she was thirteen years-old. She also had chronic kidney disease and underwent routine dialysis. She consumed clonidine 0,075 mg twice daily, candesartan 160 mg once daily, amlodipine 10 mg once daily, calcium carbonate three times daily, sodium bicarbonate three times daily, methylprednisolone 16 mg three times daily, and mycophenolic acid 360 mg three times daily.

On physical examination she had blood pressure of 150/90 mmHg, heart rate of 96 bpm, respiratory rate of 20 x/minutes, temperature of 38.4⁰C, and oxygen saturation of 98%. Neurological examination showed Glasgow Coma Scale (GCS) of 15, left 7th and 12th cranial nerve palsy, left-sided hemiparesis with manual muscle test (MMT) of 5/5/5/5/4/4/4/4 on both upper and lower extremities, increased physiological reflexes on the left side, and positive Babinski's sign on her left side.

Laboratory testing showed anemia (Hb 7.09 g/dL and hematocrit 22,6%), leucopenia (3830 mg/dL), thrombocytopenia (46600/uL),

increased creatinin in serum (6.513 mg/dL), and high d-dimer level (100 mg/dL). Chest x-ray showed cardiomegaly without pulmonary abnormality. The patient underwent non contrast head Computed Tomography (CT) and it showed right frontal lobe intraparenchymal bleeding with an estimated volume of 8.22 mL with perifocal edema which narrowed the right lateral ventricle and caused a 0.7 cm subfalcine herniation to the left.



Figure 1. Axial slice of non contrast CT

Based on the anamnesis, physical, laboratory, and radiological examinations, she was diagnosed with right frontal lobe hemorrhagic stroke which caused by SLE-induced thrombocytopenia and end stage chronic kidney disease on hemodialysis.

On the fifth day of hospitalization, the patient had a seizure with unknown pre-ictal. During ictal phase, her mouth was pulled left side and eyes gazed left. She had her consciousness intact during seizure and it lasted for two minutes. Post ictal the patient was confused but regained her consciousness shortly. On the seventh day of hospitalization, during her routine dialysis, she had repeated seizures with the same pattern as before, so the dialysis was stopped. She was given topiramate as antiepileptic drug, antihypertensive drugs to maintain her blood pressure, steroid, and packed red cells and platelet transfusion as the advice from hematologic division.

After one month of hospitalization, she was discharged with controlled seizure, although there were minor sequelae of left 7th and 12th cranial nerve palsy and slight left-sided hemiparesis.

DISCUSSION

The patient in this case was an 18 years-old female with a five years history of SLE came to the hospital for a sudden left-sided weakness one hour prior. The neurological deficits

in patient indicated central nervous system disturbance in form of NPSLE. The sudden onset suggested a vascular origin, which counted for 8-15% of NPSLE cases.² History of SLE, hypertension, nephrotic syndrome, and chronic kidney disease were risk factors for cerebrovascular disease or stroke in young age.^{5,6}

Neurological examination showed Glasgow Coma Scale (GCS) of 15, left 7th and 12th cranial nerve palsy, left-sided hemiparesis with manual muscle test (MMT) of 5555/4444 on both upper and lower extremities, increased physiological reflexes on the left side, and positive Babinski's sign on her left side. Sensory and autonomic functions were within normal limits. From physical examinations indicated there was an intracranial focal lesion which needed to be proven by neuroimaging, to determine whether it was ischemic or hemorrhagic lesion. Laboratory testing showed anemia (Hb 7.09 g/dL and hematocrit 22,6%), leucopenia (3830 mg/dL), and trombocytopenia (46600/uL). Low platelet count (<50000/uL) can induce bleeding, including a brain hemorrhage.⁷ The trombocytopenia in patient could be

caused by SLE, through autoimmune platelet surface glycoproteins antibody, or the side effects of azathioprine, methotrexate dan hydroxychloroquine therapy.⁸

Non contrast head CT showed intraparenchymal bleeding in right frontal lobe with estimated volume of 8.22 mL with perifocal edema which narrowed the right lateral ventricle and caused a 0.7 cm subfalcine herniation to the left

During hospitalization, the patient had repeated seizures. Seizure in SLE patient was one of clinical manifestations in NPSLE, occurs in 2-8% patients especially in female. Seizure occurrences in SLE can be caused for various reasons ranging from antiphospholipid autoantibodies to cerebral focal lesion.⁹ In our case, the seizure was caused by irritative cerebral focal lesion in frontal cortex, which caused spontaneous electrical activity of cortical neurons in the form of seizures.

The patient was treated conservatively. There was no indication for surgical intervention because she had good clinical feature and there was no sign of increased

intracranial pressure. To prevent bleeding recurrence, the blood pressure was maintained with antihypertensive agents. Considering the patient's kidney condition, the patient was given with angiotensin II receptor antagonist Candesartan 160 mg once daily, calcium channel antagonists Amlodipine 10mg once daily, and an alpha blocker Clonidine 0.075 mg twice daily, which were safe and recommended antihypertensive agents for patients with renal impairment.¹⁰ In addition, correcting the thrombocytopenia was also done to prevent recurrent bleeding with platelet transfusion. The patient continued to be given steroid that was beneficial in reducing inflammation in the brain as well as the therapy for immune mediated thrombocytopenia and anemia that occurred.¹¹ Topiramate was given as anti-epileptic drug to control her seizures, which was a safe anti-seizure option in cases of seizure with pancytopenia and multiorgan problems.^{12,13}

The patient's outcomes were categorized into three. *Ad vitam* prognostic had a good outcome because the stable hemodynamic

and neurologic conditions in time of discharge. *Ad functionam* prognostic was relatively good, the patient started to do her activity daily living independently by the time of discharge and it was expected to improve as she underwent neurorehabilitation. *Ad sanationam* prognostic had relatively poor outcome, the underlying diseases SLE, hypertension, nephrotic syndrome, and CKD on dialysis; those increase the risk of recurrent stroke, both hemorrhagic and ischemic stroke.¹⁴

CONCLUSION

Although the prevalence is not as frequent as ischemic stroke, hemorrhagic strokes can be one of the clinical manifestations of NPSLE. This case report showed a case of hemorrhagic stroke in young woman with multiple risk factors such as SLE with severe thrombocytopenia, chronic kidney disease on dialysis, and hypertension. The clinical manifestations that occurred in the patient were left-sided hemiparesis and seizure due to lobar hemorrhage in the right frontal cortex. Treatment was done conservatively by managing

risk factors with steroid administration, maintaining blood pressure, and platelet transfusion. In addition, the patient was also treated according to her symptoms using anticonvulsant and motoric rehabilitation to improve motor strength. The risk of recurrence of stroke in patients with SLE is high, especially if it is accompanied by other risk factors such as hematologic complication and renal impairment.

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