

Case Report

Internal Carotid Artery Occlusion in Systemic Lupus Erythematosus as a Potential Mimicker of Multiple Sclerosis

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Keywords

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Abstract

The diagnosis of neurological symptoms in patients with systemic lupus erythematosus (SLE) is often challenging, in part because they sometimes mimic features of multiple sclerosis (MS). Herein we report a case of a young female who presented with relapsing-remitting symptoms of unilateral visual loss and motor aphasia. Additionally, radiological findings revealed multiple white matter lesions on her brain that led to an initial diagnosis of MS based on the established diagnostic criteria. However, she was eventually diagnosed with neuropsychiatric SLE (NPSLE) presenting with extracranial internal carotid artery (ICA) occlusion. Her ICA occlusion had not been detected for 2 months until she underwent magnetic resonance angiography. Although exact underlying pathological mechanisms are unclear, both the ICA occlusion and MS-like brain white matter lesions could be attributed to SLE. This case demonstrated that both of these lesions can coexist in the same patient, suggesting that NPSLE with ICA occlusion can be a potential mimicker of MS, and vice versa. Care must be paid to avoid delay in the diagnosis.

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Introduction

The diagnosis of neurological symptoms in patients with systemic lupus erythematosus (SLE) is often challenging [1, 2] because the SLE sometimes mimics multiple sclerosis (MS) in their clinical and radiological features [3]. Herein we report a case of a young female who was initially suspected of having MS based on the established diagnostic criteria; however, she was eventually diagnosed with neuropsychiatric SLE (NPSLE) presenting with extracranial internal carotid artery (ICA) occlusion.

Case Presentation

A 37-year-old female with 1 child and no history of abortion presented with an acute-onset of decreased vision in her right eye (first attack). A brain magnetic resonance imaging (MRI) performed 4 weeks later revealed no abnormal findings, and the visual symptoms partially remitted within 1 month.

Seven months later, she presented with a symptom of acute-onset motor aphasia (second attack). She did not complain of other symptoms such as hemiparesis, dysarthria, head/neck pain, fever, dizziness, or fatigue. A second brain MRI revealed multiple white matter lesions on the right hemisphere of her brain (Fig. 1a), while there were no hyperintensity lesions on diffusion-weighted imaging. A spinal MRI and a contrast-enhanced brain MRI revealed no abnormal findings. Combined with the relapsing-remitting neurological attacks (visual decrease and aphasia) and the compatible white matter lesions on MRI, she had been at first suspected of MS. At this point, MR angiography was not yet performed because of the initial diagnosis.

However, although the clinical presentation of this case had 2 or more attacks and 1 objective clinical lesion, it was controversial if dissemination in space was surely demonstrated (Fig. 1a, b). So, according to the 2010 McDonald criteria [4], it was difficult to make a diagnosis of MS.

In addition, further laboratory testing revealed inconsistency with the diagnosis of MS: significantly elevated levels of anti-nuclear and anti-double strand DNA antibodies, lowered levels of C3/C4 complements, and decreased white blood cell count. Taken together, these symptoms met the American College of Rheumatology (ACR) SLE criteria, 2012 (meeting 3 of the 11 ACR criteria, 1997) [5]. Her renal function was normal as indicated by the absence of proteins in her urine. The C-reactive protein level was normal (0.03 mg/dL), and the erythrocyte sedimentation rate was slightly high (21 mm/h, normal <15 mm/h). Anticardiolipin IgG antibody, anticardiolipin/beta 2 glycoprotein inhibitor antibody, protein C, protein S, antineutrophil cytoplasmic antibody levels and routine coagulation tests (prothrombin time-international normalized ratio, activated partial thromboplastin time, and D-dimer) were all within normal limits. The lupus anticoagulant test, which was performed using the dilute Russell viper venom time method and the diluted activated partial thromboplastin time method, was negative. Her cerebrospinal fluid test was also normal with a cell count of 0/ μ L, a total protein count of 48 mg/dL, and negative oligoclonal bands.

These radiological and laboratory findings do not support the diagnosis of MS, and instead she was diagnosed with SLE and could be classified as having NPSLE according to the ACR classification criteria, 1999 [1, 2]. Although her clinical symptoms seemed to be confined to neurological ones, actually she had transient mild hair loss and an itchy scalp during

the first and second attacks. These symptoms were resolved through treatment with topical lotion dexamethasone, and being missed in the diagnosis.

Her symptoms gradually resolved without the use of standard immunotherapy for SLE, which the patient refused to undergo. Two months after the onset of the second attack, MR angiography was performed for the first time and revealed signal loss in her right ICA (Fig. 1c, d). An echogram of her CA revealed an occlusion in her right extracranial ICA immediately above its origin, with a gradual narrowing of the vessel's internal cavity (Fig. 1e), suggesting that a thrombosis was the cause of ICA occlusion [6]. There were no abnormal findings in her brachiocephalic trunk and common carotid arteries that suggested Takayasu arteritis (TA). Taken together, she was eventually diagnosed with NPSLE accompanied with an ICA occlusion.

Discussion

We consider that SLE was the underlying cause of both ICA occlusion and MS-like lesions, which coexisted during the same clinical course. This suggests that NPSLE with ICA occlusion can be a mimicker of MS, and vice versa. In this case, detailed ICA evaluation was recommended to investigate potential vascular abnormalities. Although the exact time of occurrence of extracranial ICA occlusion is unclear, the delay in its diagnosis was approximately 2 months, which could have resulted in the poorer prognosis depending on the state of collateral blood flow. Additionally, this case did not seem to show any significant radiological and clinical red flags that suggested an alternative diagnosis to MS [3], with the exception of the transient skin rash and hair loss that had been missed prior to the identification of ICA occlusion. Including the unilateral distribution of white matter lesions in the list of potential MS, red flags may help reduce the risk of future misdiagnosis.

The ICA occlusion observed in the present case should be attributed to the comorbid NPSLE, while the exact underlying mechanisms of ICA occlusion in this case are unclear. Several possible pathophysiological mechanisms could be raised, such as premature atherosclerosis, thrombosis, vasculopathy, emboli, dissection of the carotid artery, and antiphospholipid syndrome [1, 2, 4, 6–10]. There would have been atherosclerosis and thrombosis process following the chronic inflammation, and rarely, these pathological changes can result in ICA dissection [1, 8, 9]. The absence of significant pain in her neck or head does not exclude the possibility of an ICA dissection [9]. There was no evidence to support a diagnosis of antiphospholipid syndrome, which could potentially cause arterial stenosis or occlusion [4, 6, 10]. A similar previously reported case of SLE with ICA occlusion presented with positive antiphosphatidylserine/prothrombin antibodies [6]; however, we could not evaluate these antibodies and cannot rule out their presence in the present case [10]. Although TA can rarely coexist with SLE [7], we consider the diagnosis of TA unlikely because of its characteristic distribution of large arterial lesions and rare involvement of ICA. Furthermore, the patient had little clinical and laboratory signs of systemic inflammation, and there was no lesion found on the common carotid arteries and brachiocephalic trunk.

The white matter lesions can also be attributed to SLE pathology and may be related to the ICA occlusion. Indeed, the visual decrease, white matter lesions, and ICA occlusion were all observed on the ipsilateral side. However, it remains unclear how the ICA occlusion could have contributed to the white matter lesions. One possibility is that the ICA occlusion could have caused an artery-to-artery embolism or a hemodynamic hypoperfusion with an acute cerebral infarction, but there was no evidence of diffusion-weighted imaging hyperintense

lesions to support this hypothesis. Alternatively, local cerebral microangiopathy or demyelination of the brain under SLE pathology may be the cause of the white matter lesions [1, 4].

To conclude, we presented the case of a young female with NPSLE, ICA occlusion, and MS-like lesions. This case suggests that NPSLE presenting with ICA occlusion can be a mimicker of MS, and vice versa. Care must be paid to avoid delay in the appropriate diagnosis.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors' financial relationships from the past 2 years are as follows: Dr. Sato reports no disclosures. Dr. Kanzaki reports no disclosures. Dr. Ubara reports no disclosures. Dr. Uesaka served as a primary investigator, received institutional support from the Oki-naka Memorial Institute for Medical Research, and is funded by the Ministry of Health, Labour and Welfare (MHLW) of Japan as a member of "Wake-up stroke" and "Subacute myelo-optico-neuropathy" research.

References

- Jennekens FG, Kater L: The central nervous system in systemic lupus erythematosus. Part 2. Pathogenetic mechanisms of clinical syndromes: a literature investigation. *Rheumatology (Oxford)* 2002;41:619–630.
- Luyendijk J, Steens SC, Ouwendijk WJ, Steup-Beekman GM, Bollen EL, van der Grond J, Huizinga TW, Emmer BJ, van Buchem MA: Neuropsychiatric systemic lupus erythematosus: lessons learned from magnetic resonance imaging. *Arthritis Rheum* 2011;63:722–732.
- Miller DH, Weinshenker BG, Filippi M, Banwell BL, Cohen JA, Freedman MS, Galetta SL, Hutchinson M, Johnson RT, Kappos L, Kira J, Lublin FD, McFarland HF, Montalban X, Panitch H, Richert JR, Reingold SC, Polman CH: Differential diagnosis of suspected multiple sclerosis: a consensus approach. *Mult Scler* 2008;14:1157–1174.
- Uthman I, Noureldine MH, Berjawi A, Skaf M, Haydar AA, Merashli M, Hughes GR: Hughes syndrome and multiple sclerosis. *Lupus* 2015;24:115–121.
- Petri M, Orbai AM, Alarcón GS, Gordon C, Merrill JT, Fortin PR, Bruce IN, Isenberg D, Wallace DJ, Nived O, Sturfelt G, Ramsey-Goldman R, Bae SC, Hanly JG, Sánchez-Guerrero J, Clarke A, Aranow C, Manzi S, Urowitz M, Gladman D, Kalunian K, Costner M, Werth VP, Zoma A, Bernatsky S, Ruiz-Irastorza G, Khamashta MA, Jacobsen S, Buyon JP, Maddison P, Dooley MA, van Vollenhoven RF, Ginzler E, Stoll T, Peschken C, Jorizzo JL, Callen JP, Lim SS, Fessler BJ, Inanc M, Kamen DL, Rahman A, Steinsson K, Franks AG Jr, Sigler L, Hameed S, Fang H, Pham N, Brey R, Weisman MH, McGwin G Jr, Magder LS: Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. *Arthritis Rheum* 2012;64:2677–2686.
- Naoi T, Kameda T, Oku K, Ando A, Hayashi Y, Miyamoto M, Suzuki H, Kawakami T: Internal carotid artery occlusion and cerebral infarction in a case of juvenile systemic lupus erythematosus and positive for phosphatidylserine dependent antiprothrombin antibody (aPS/PT). *Neurol Clin Neurosci* 2016, DOI:10.1111/ncn3.12112.
- Saxe PA, Altman RD: Takayasu's arteritis syndrome associated with systemic lupus erythematosus. *Semin Arthritis Rheum* 1992;21:295–305.
- Lucas C, Moulin T, Deplanque D, Tatu L, Chavot D: Stroke patterns of internal carotid artery dissection in 40 patients. *Stroke* 1998;29:2646–2648.
- Zetterling M, Carlström C, Konrad P: Internal carotid artery dissection. *Acta Neurol Scand* 2000;101:1–7.

10 Devreese K, Hoylaerts MF: Challenges in the diagnosis of the antiphospholipid syndrome. Clin Chem 2010;56:930–940.

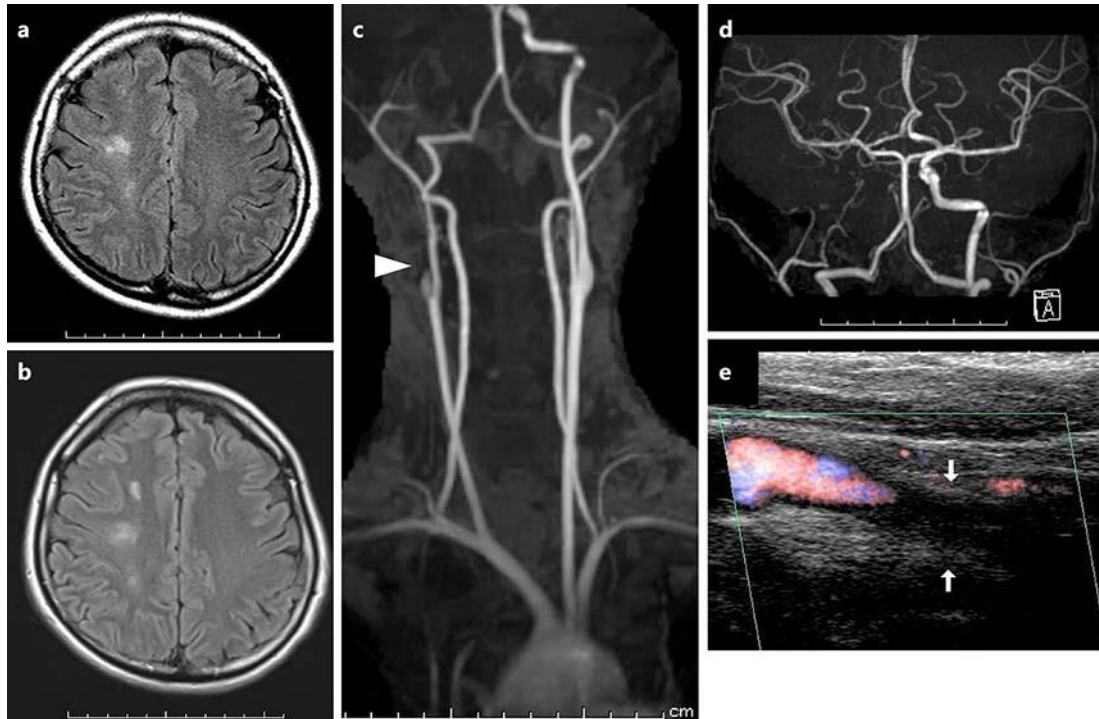


Fig. 1. Brain magnetic resonance imaging at the time of the second attack, revealing multiple white matter lesions on a fluid-attenuated inversion recovery image (a). Follow-up brain magnetic resonance imaging performed 2 months later, revealing similar multiple white matter lesions on a fluid-attenuated inversion recovery image imaging (b) and a magnetic resonance angiography signal loss in her right extracranial internal carotid artery (c [white arrowhead], d). An echogram of her carotid artery, revealing an occlusion of her right extracranial internal carotid artery (e, white arrows) immediately above its origin with a gradual narrowing of the vessel's internal cavity, suggesting thrombosis as the cause of the internal carotid artery occlusion.