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Case Report

Thrombosis of a basilar perforator aneurysm associated with pontine infarction in a patient with systemic lupus erythematosus ☆,☆☆

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ABSTRACT

Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect multiple organ systems. Cerebral aneurysm formation is a rare central nervous system manifestation of SLE and tends to present as subarachnoid hemorrhage. Here, we report a 34-year-old woman with SLE complicated by a thrombosed aneurysm that had arose at the origin of a perforating artery, thereby causing obstruction of the artery and subsequent development of pontine infarction. Detailed examination of thin-slice CT and magnetic resonance imaging scans led to the correct diagnosis of uncommon cause of stroke.

Case report

A 34-year-old woman was admitted to our hospital for evaluation and treatment of systemic lupus erythematosus (SLE). She was given a diagnosis of SLE with arthralgia and leukopenia and exhibited positivity for anti-double-stranded DNA antibodies. Antinuclear antibodies were initially negative, but turned positive on day 13. She was started on oral prednisolone at a dose of 50 mg daily on day 14. She developed dysarthria and ataxia on day 17. Computed tomography (CT) and magnetic resonance imaging (MRI) of the head were performed at 2 hours and 2.5 hours after the onset of symptoms, respectively. Nonenhanced CT revealed that there were no abnormalities except for a small high-density spot close to the distal basilar artery (Fig. 1). The MRI scan revealed that there was a faintly hyperintense area on diffusion-weighted imaging with corresponding low diffusivity in the left side of the pons (Fig. 2A and B). Heparin infusion was started. She was also treated with intravenous methylprednisolone at a dose of 1 g daily for 3 days. Six days later, follow-up MRI revealed that there was clearly defined infarction in the left side of the pons (Fig. 2C and D) as well as a small ovoid lesion on the left side of the basilar artery (Fig. 2E and F), whereas no definite abnormalities were observed using magnetic resonance (MR) angiography (Fig. 2G). The ovoid lesion was considered to

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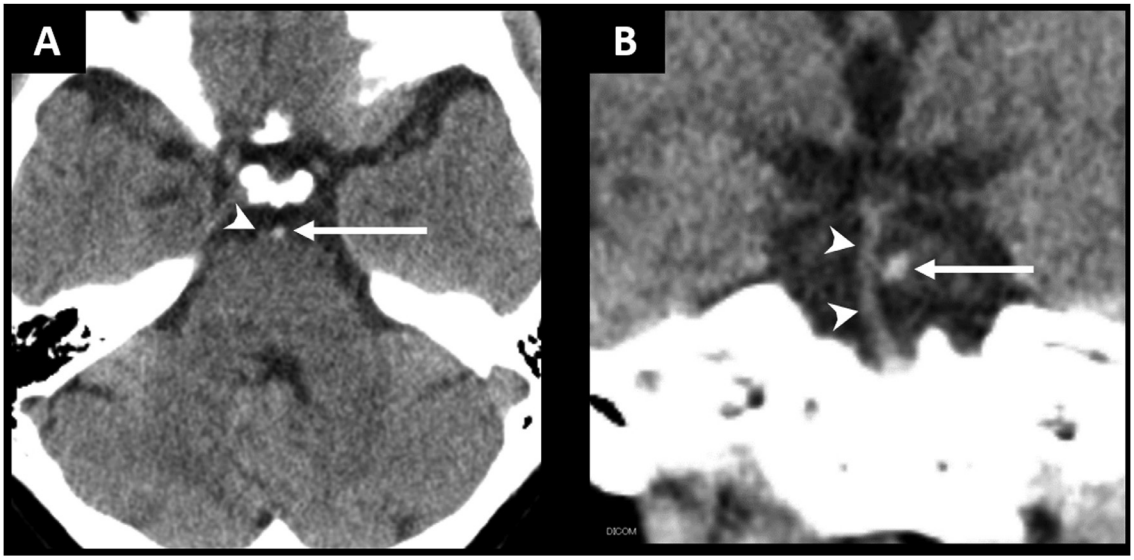

Fig. 1 – Computed tomography (CT) without contrast enhancement was performed at 2 hours after symptom onset. Axial (A) and reformatted (B) coronal images revealed the presence of a small high-density lesion (arrows) close to the basilar artery (arrowheads).

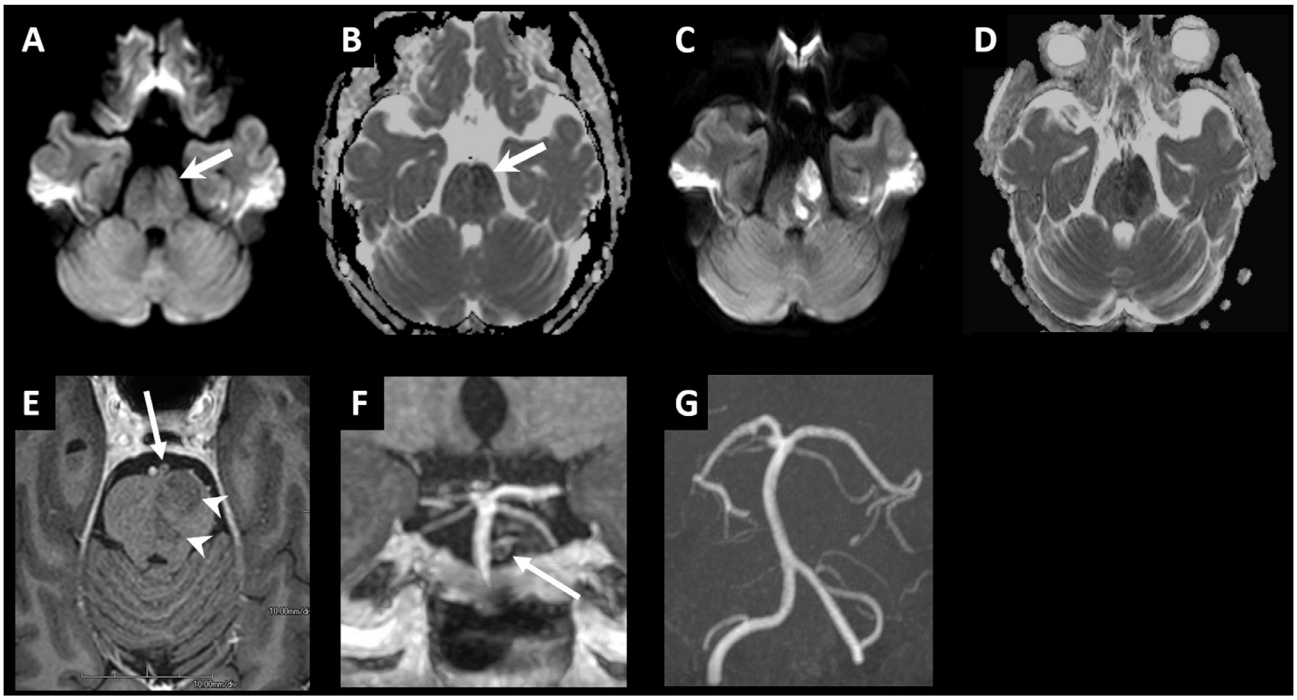

Fig. 2 – Diffusion-weighted imaging (DWI) (A) obtained at 2.5 hours after the onset of neurological symptoms revealed a faintly hyperintense lesion in the left side of the pons with corresponding low diffusivity on apparent diffusion coefficient (ADC) map (B) (arrow). Follow-up DWI (C) and ADC map (D) obtained 6 days later revealed clearly defined infarction in the left side of the pons with decreased diffusivity. Axial (E) and reformatted (F) coronal images from contrast-enhanced magnetization prepared rapid gradient echo imaging revealed the presence of a nonenhanced ovoid lesion (arrow) close to the enhanced basilar artery. Pontine infarction was observed as an area of decreased signal intensity on the same side as the ovoid lesion (arrowheads). The ovoid lesion was not observed using magnetic resonance angiography (G). Note the proximity of the ovoid lesion and pontine infarction and their position on the left side.

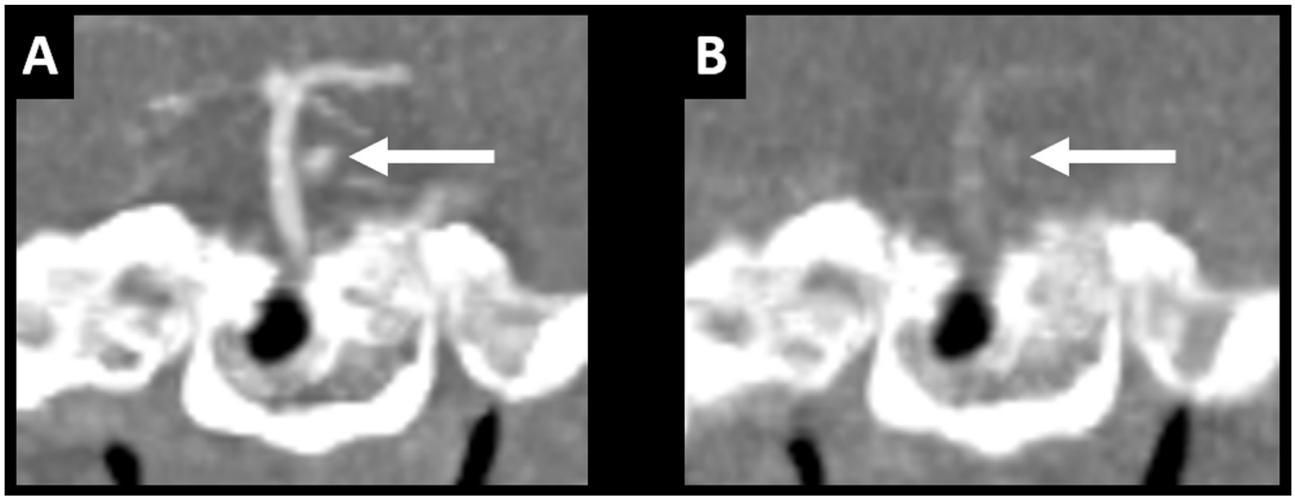

Fig. 3 – Coronal reformatted images from dynamic contrast-enhanced CT obtained at 1 week before the onset of neurological symptoms. The early phase image (A) revealed the presence of an ovoid enhanced lesion close to the basilar artery. The delayed phase image (B) indicated that there was reduced attenuation of both the basilar artery and ovoid lesion. The similar densities as observed in both the early and delayed images indicate that the ovoid lesion was an aneurysm that had not yet thrombosed.

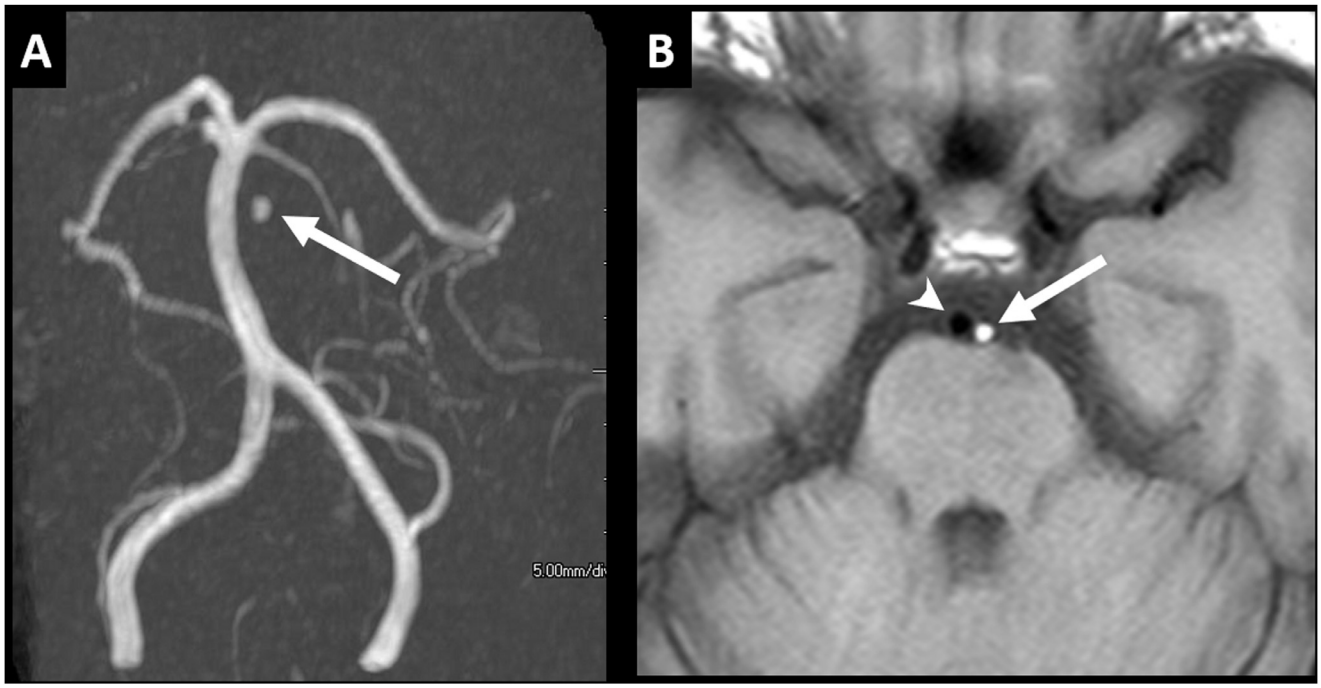

Fig. 4 – Follow-up MRI at about 1 year later. Magnetic resonance angiography (A) revealed the presence of a high-intensity spot close to the basilar artery (arrow). Pre-contrast T1-weighted imaging (B) revealed the presence of a hyperintense nodular lesion (arrow) close to the basilar artery (arrowhead), indicating that the lesion was a basilar perforator aneurysm that still remained thrombosed.

correspond to the high-density spot observed using CT and was located just ventral to the infarcted area of the pons. Considering the proximity of the ovoid lesion and pontine infarction and their position on the left side, we suspected that the ovoid lesion represented a thrombosed aneurysm that had arose at the origin of a perforating artery, thereby causing obstruction of the artery and subsequent development of pontine infarction. To explore this possibility, we reviewed contrast-enhanced CT results that were obtained to investigate systemic vasculitis at 1 week before the onset of neurological symptoms, and found a contrast-enhanced vascular lesion corresponding to the aforementioned ovoid lesion, which

likely indicated the presence of a basilar perforator aneurysm that had not yet thrombosed (Fig. 3). Oral steroid treatment in this patient was continued without any additional stroke events. One year later, follow-up MRI revealed that there was a hyperintense ovoid lesion close to the basilar artery using both MR angiography and precontrast T1-weighted imaging (Fig. 4). This lesion was considered to represent the basilar perforator aneurysm that remained thrombosed.

Discussion

SLE is an autoimmune collagen vascular disease that can affect multiple organ systems. CNS involvement has been reported in up to 50% of patients with SLE. Cerebral aneurysm formation is a rare CNS manifestation of SLE with a reported incidence of less than 4% [1,2]. According to a review of ruptured cerebral aneurysms associated with SLE, 79.1% of aneurysms were located in a major artery, whereas 20.9% were in a small peripheral artery [3]. As the frequency of peripheral cerebral aneurysms in the general population is less than 1%, SLE-associated aneurysms are more likely to be located in distal branches. Although the mechanisms underlying the initiation and development of lupus-associated cerebral aneurysms have not yet been fully elucidated, focal transmural angiitis, fibrinoid degeneration of collagen, and damage to the internal elastic lamina and medial smooth muscle have been observed in histopathological examinations [4–7]. SLE-associated aneurysms usually present as subarachnoid hemorrhage. We found 1 report of a case of CNS lupus with cerebral aneurysms and ischemic stroke [8]; however, there appeared to be no direct association between the aneurysms and the onset of infarction. To our knowledge, this is the first report of an SLE-associated cerebral aneurysm complicated by pontine infarction due to a thrombosed aneurysm arising in one of the pontine perforators. The increase in disease activity in SLE may have been one of the factors related to the thrombosis of the aneurysm in our case, because seroconversion of antinuclear antibodies was observed at 4 days before the onset of infarction.

We reported a very rare case of SLE with a pontine infarction due to thrombosis of basilar perforator aneurysm in the parent artery of the pons. Detailed examination of thin-slice CT and MRI scans led to the correct diagnosis.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

For this type of retrospective study formal consent is not required.

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