



Progressive cerebral large-vessel vasculitis in a patient with Sjögren's syndrome

Woo Jin Kim, MD¹; Jin San Lee, MD, PhD²; Seong-il Oh, MD, PhD^{2,3}

¹Department of Otorhinolaryngology-Head and Neck Surgery, Inje University Busan Paik Hospital, Busan, Korea

²Department of Neurology, Kyung Hee University Hospital, Seoul, Korea

³Department of Neurology, Inje University Busan Paik Hospital, Busan, Korea

IMAGES IN NEUROCRITICAL CARE

Received: December 12, 2022

Revised: February 17, 2023

Accepted: February 26, 2023

Corresponding Author:

Seong-il Oh, MD, PhD

Department of Neurology, Kyung Hee University Hospital, 23 Kyungheedaero, Dongdaemun-gu, Seoul 02447, Korea

Tel: +82-2-958-8499

Fax: +82-2-958-8490

E-mail: seongil.oh@gmail.com

Extraglandular involvement in Sjögren's syndrome (SS) can manifest in the central or peripheral nervous system. Changes in major cerebral vessels in SS are rarely reported [1,2]. Pathomechanisms in SS are known to be mainly associated with small- or medium-sized vasculopathy [1,3]. A 59-year-old woman who presented with left transient hemiparesis a few days prior was diagnosed with SS with dry mouth. Anti-SS-related antigen A (SSA/Ro) and B (SSB/La) had shown strong positivity on previous evaluations. Brain magnetic resonance imaging (MRI) showed multiple diffusion-restricted lesions in the right middle cerebral artery border zone area, suggesting acute infarctions (Fig. 1). Magnetic resonance angiography (MRA) upon admission showed moderate-to-severe stenosis of the right distal internal carotid artery (ICA) (Fig. 2). The patient reported transient right hemi hypesthesia following admission. On MR carotid plaque imaging performed 4 days later, there was focal concentric wall thickening with enhancement in the right distal ICA stenosis at the location of the lesion shown on the previous MRA. Additionally, concentric wall thickening with enhancement was observed in the left distal ICA (Fig. 2). Bilateral distal ICA stenosis with wall thickening and enhancement occurred consecutively. Therefore, it was likely caused by vasculitic stenosis associated with SS.

This report showed that a patient with high disease activity de-

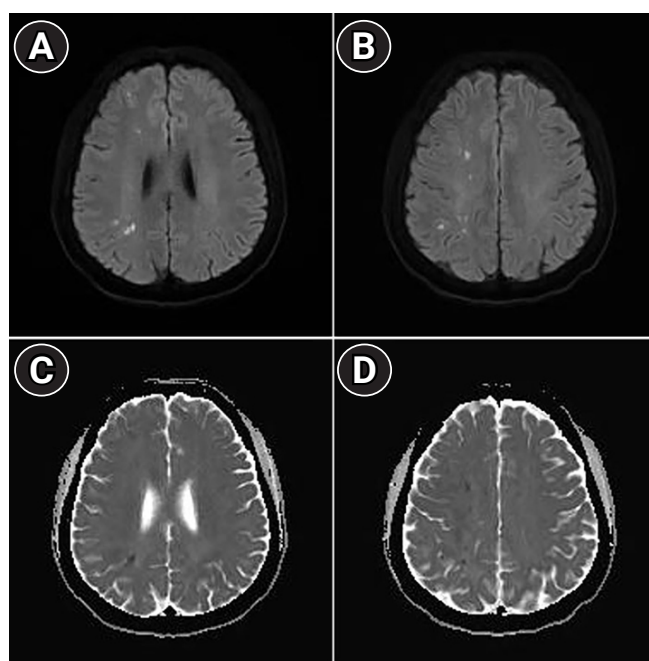


Fig. 1. Brain magnetic resonance imaging scans upon admission. (A, B) There were high signal-intensity lesions on diffusion-weighted images in the right middle cerebral artery borderzone area at the right frontal and parietal lobes, and (C, D) relevant lesions were noted on the apparent diffusion coefficient map.

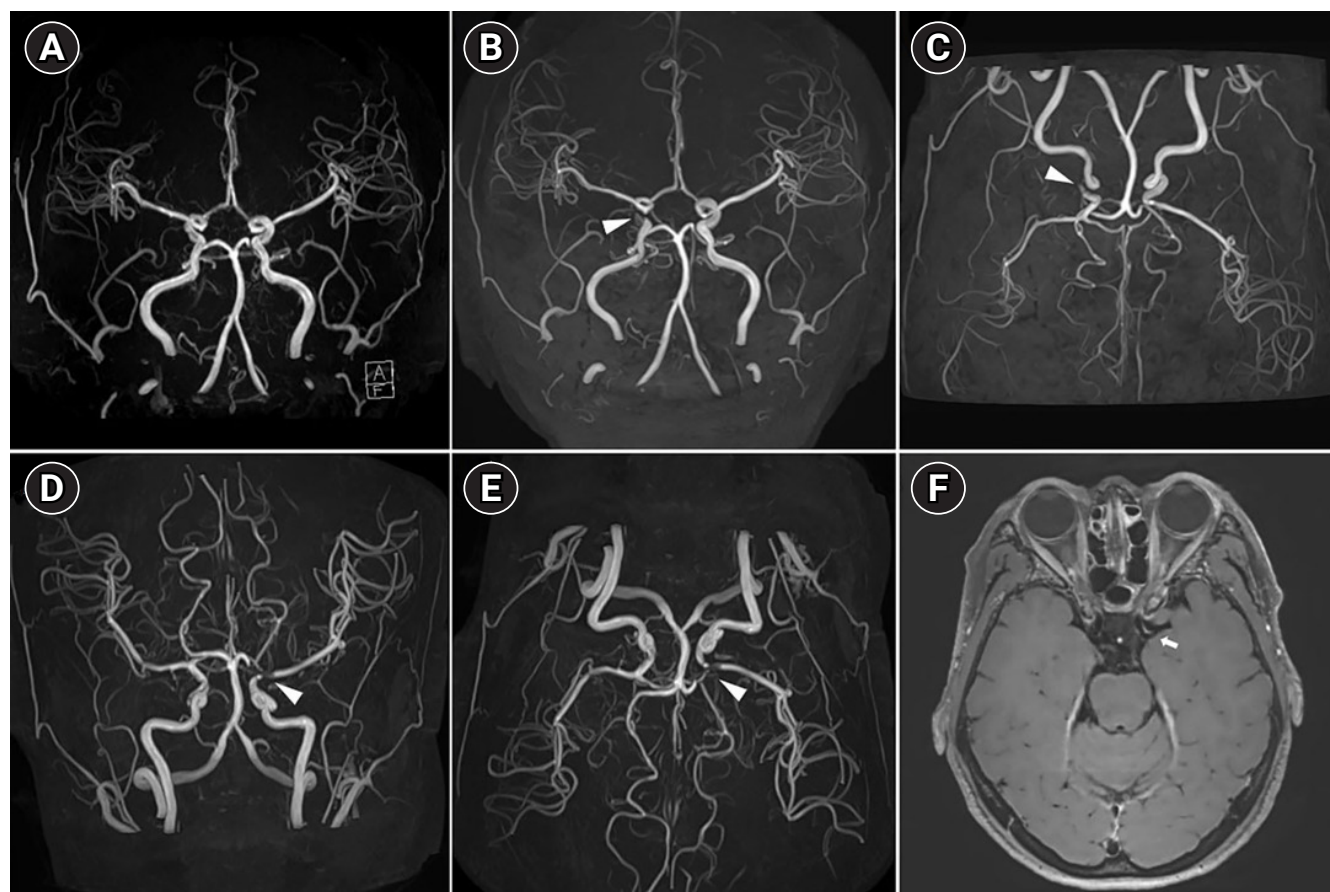


Fig. 2. Brain magnetic resonance angiography (MRA) and high-resolution magnetic resonance imaging (MRI) of the carotid plaque. (A) MRA taken 1 year before admission showed no stenosis in both distal internal carotid arteries (ICAs). (B, C) MRA taken upon admission showed moderate to severe stenosis in the right distal ICA (arrowhead). (D, E) On MRA taken 4 days later, severe stenosis was newly discovered in the left distal ICA, proximal M1, and proximal A1 (arrowhead), and (F) focal concentric wall thickening with enhancement at the left distal ICA was confirmed on high-resolution MRI (arrow).

veloped severe vasculitic stenosis that occurred consecutively within a few days. A high-resolution MRI of the carotid plaque indicated the presence of vasculitis with large-artery involvement, which could be helpful when considering the pathology.

ARTICLE INFORMATION

Ethics statement

This work was approved by the Ethics Committees of the Inje University Busan Paik Hospital (No. 2015-01-271), and written informed consent was obtained from the patient.

Conflict of interest

No potential conflict of interest relevant to this article.

Acknowledgments

This study was supported by the National Research Foundation

of Korea (NRF) grant funded by the Korea government (MIST) (No. 2020R1G1A1008446).

ORCID

Woo Jin Kim

<https://orcid.org/0000-0002-8895-1083>

Jin San Lee

<https://orcid.org/0000-0002-5017-854X>

Seong-il Oh

<https://orcid.org/0000-0002-8067-2135>

Author contributions

Conceptualization: WJK, SIO. Methodology: WJK, SIO. Formal analysis: WJK, SIO. Data curation: all authors. Visualization: all authors. Project administration: SIO. Writing—original draft: all authors. Writing—review and editing: WJK, SIO.

REFERENCES

1. Fox RI. Sjögren's syndrome. *Lancet* 2005;366:321-31.

2. Tobón GJ, Pers JO, Devauchelle-Pensec V, Youinou P. Neurological disorders in primary Sjögren's syndrome. *Autoimmune Dis* 2012;2012:645967.
3. Li JA, Meng HM, Cui ZT, Wang X, Miao J. Recurrent cerebral infarctions in primary Sjögren syndrome: a case report and literature review. *Front Neurol* 2018;9:865.