

Anti-aquaporin-4 antibody positive Sjogren syndrome presenting with recurrent brain involvement

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Sjogren syndrome can affect the central nervous system and sometimes shows the manifestations of neuromyelitis optica (NMO) with optic neuritis or myelitis. Whereas it used to be said that NMO does not show brain lesions at the onset of the disease, recently a NMO case with preceding brain demyelinating episode was described.^{1,2} We report a patient with anti-aquaporin-4 antibody positive Sjogren syndrome, presenting with recurrent brain involvement.

Case Report: A 35-year old woman visited our clinic due to right-sided hemiparesis and sensory change. A T2-weighted image showed high signal intensities at the left internal capsule. On laboratory evaluation, she showed positive anti-SSA/SSB antibodies and Schirmer test. Salivary gland biopsy was compatible to Sjogren syndrome. She was diagnosed as a primary Sjogren syndrome and treated with prednisolone. Three years later, she developed left-sided hemiparesis with a lesion in the right side, from the internal capsule to pons. Cerebrospinal fluid study showed increased IgG index (0.8) and positive oligoclonal band. After another year, sensory disturbance of the right side developed with large and extensive (over 3 cm) left frontoparietal lobes lesion (Figure 1). During the course of the illness, she did not have optic neuritis or myelitis. Anti-aquaporin-4 antibody was found to be positive in the serum.

Discussion and Conclusion: NMO-IgG or anti-aquaporin-4 antibody was originally associated with long cord myelopathy and complete blindness, clinically manifesting as severe optic-spinal disease. Sjogren syndrome patients without symptoms or signs of NMO are found to be seronegative for NMO-IgG. This patient with Sjogren syndrome showed recurrent brain involvement of up to three times. She was also found to have positive anti-aquaporin-4 antibody, and she did not develop optic neuritis or myelitis. Our patient demonstrated that it is plausible to have brain involvement accompanied by MRI findings developing as the initial manifestation of NMO in Sjogren syndrome. Latent risk of NMO should be looked for in Sjogren syndrome, even though there is only recurrent brain involvement clinically.

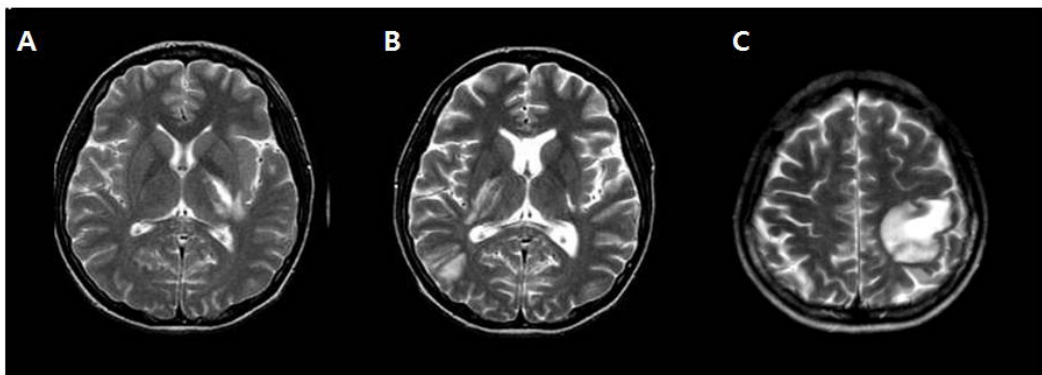


Figure 1. T2-weighted images showed brain abnormalities which are known as characteristic for neuromyelitis optica; the longitudinal internal capsular lesions (A and B) and large lesions over 3cm (C).

References

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2. Amemiya S, Hamamoto M, Kumagai T, Ueda M, Katayama Y, Tanaka K. Neuromyelitis optica preceded by brain demyelinating episode. *J Neuroimaging* (in press)