A CASE OF SPONTANEOUS PONTINE HEMORRHAGE ASSOCIATED WITH NEUROMYELITIS OPTICA SPECTRUM DISORDER

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Introduction: Neuromyelitis optica spectrum disorder (NMOSD) is an inflammatory demyelinating disease of the central nervous system and its pathophysiology is not completely understood. Intracerebral hemorrhage (ICH) in demyelinating disease has been rarely reported and not in NMO. We describe a case of ICH into pre-existing demyelinating lesion in an anti-aquaporin-4 antibody (AQP4-Ab) positive patient.

Case report: A 42-year-old male developed right arm and facial paresthesia at 35 years old. Brain magnetic resonance imaging (MRI) showed hyperintense signals at T2 and fluid attenuated inversion recovery (FLAIR) in bilateral pons, midbrain, cerebellum, and periventricular white matter with gadolinium enhancement. After steroid therapy, symptoms were improved and follow-up MRI showed almost resolution.

Five years later, he complained left side motor weakness and paraesthesia for 2 months. Brain MRI revealed multiple hyperintensities at similar areas. Symptoms were improved with steroid treatment; however, some abnormal signal intensities were remained.

Seven months later, sudden onset left extremities paresthesia was developed. Brain MRI and computed tomography revealed spontaneous ICH at right pons, corresponding to the previously demonstrated several demyelinating lesion. The AQP4-Ab was positive. We diagnosed NMOSD and the treatment was started with azathioprine and oral prednisolone.

Conclusion: Our patient developed ICH in corresponding to the location of previously demyelinating lesion: The association between demyelination and hemorrhage remains unclear. However, AQP4-Ab may compromise the integrity of the blood brain barrier and facilitate the perivascular inflammation and consequently lead to ICH. This indicates the possibility that ICH in NMOSD is associated with AQP4-Ab vascular pathogenesis.