Initial presentation of central nervous system (CNS) demyelinating disease in Thai patients: Retrospective study

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Background

Neuromyelitis optica (NMO) is recognized increasingly in Asia. The prevalence and clinical data have not been investigated systematically in Thailand.

Objective

To describe the demographics and frequency of intractable hiccup or vomiting, or chronic myelopathy without preceding acute myelitis, as initial presentations of NMO in Thai patients.

Method

We retrospectively reviewed clinical records of patients: 1) admitted to or evaluated as outpatients at the Prasat Neurological Institute between November, 2009 - February, 2012, 2) assigned a diagnosis of CNS demyelinating disease, 3) with serum available for AQP4-IgG testing (M1-transfected HEK293 cells). These patients were classified as NMO or NMO spectrum disorder (Wingerchuk criteria 2006, 2007), multiple sclerosis (MS), clinical isolated syndrome (CIS) (McDonald criteria 2010), unclassified (imaging and or laboratory profiles suggesting an inflammatory process, but not fitting diagnostic criteria). The initial presentation was classified as optic neuritis (ON), brainstem dysfunction (BS, includes intractable hiccup and vomiting), acute myelitis (AM), optic neuritis and myelitis (ON+AM; within 2 weeks), periventricular lesion (PV).

Result

125 patients were identified; 98 were female (F: M=3.6: 1). Mean onset age, 39 years (12-73). Diagnostic classification: NMO 36 (28.8%); NMOSD 60 (48%); MS 15 (12%); CIS 12 (9.6%) and unclassified 2 (1.6%). For combined NMO plus NMOSD, F: M was 5.4:1 and AQP4-IgG was positive in 63.5%. Initial clinical presentation in NMO and NMOSD patients: ON, 18%; BS, 21%; SC, 55%; ON+SC, 5%; PV, 1%. Interval to second attack with respect to AQP4-IgG serostatus: did not differ significantly for NMO group (21.8 months [positive] and 20.6 months [negative]); was shorter for NMOSD group (10.3 months [positive] and 16.5 months [negative]). Eight patients presented with intractable hiccup or vomiting (NMO; 1, NMOSD; 7); 7 were female and 75% were AQP4-IgG-positive. The initial clinical presentation in 8 patients was chronic myelopathy (painful tonic spasms without preceding clinically acute myelitis): NMO; 3, NMOSD; 4 and unclassified; 1. All were female and 63% were AQP4-IgG-positive.

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Conclusion

In Thailand, NMO and NMOSD account for more CNS demyelinating disease cases than MS. Acute myelitis is the most common initial presentation. It is noteworthy that 8% of the patients in this study initially presented with isolated chronic myelopathy without an acute clinical episode.