Neuromyelitis optica spectrum disorder mimicking Wernicke's encephalopathy

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A 23-year-old female presented to a local physician with multiple episodes of vomiting. She had persistence of symptoms despite symptomatic treatment. Magnetic resonance imaging (MRI) brain plain showed hyperintensities in bilateral dorsomedial thalamus, the periaqueductal gray area and tectum of the midbrain in T2-weighted and fluid-attenuated inversion recovery (FLAIR) images. (Figure 1 A,B,C,D). The findings were typical of Vitamin B1 (thiamine) deficiency (Wernicke's encephalopathy). Intravenous thiamine was given and her symptoms resolved. Ten days later she presented with recurrence with worsening associated with hiccups and giddiness. Clinically she had left eye rapid afferent pupillary defect; the rest of the neurological examination was normal. Repeat MRI after ten days showed persistent findings of hyperintensities in the periaqueductal region and bilateral thalami (Figure 1 E, F, G, H) with no diffusion restriction and no contrast enhancement.

On further probing, her history revealed that she had left eye ocular pain with diminution of vision four months previously. She was evaluated elsewhere and diagnosed with papillitis. Her brain and spine imaging were normal. Her symptoms resolved a week after being treated with intravenous and oral tapering steroids. She had a similar episode one month previously, where she was diagnosed with left eye optic neuritis and started on intravenous pulse steroids followed by oral steroid therapy.

With the history of two episodes of optic neuritis in the past, and with the current clinical event of area prostrema

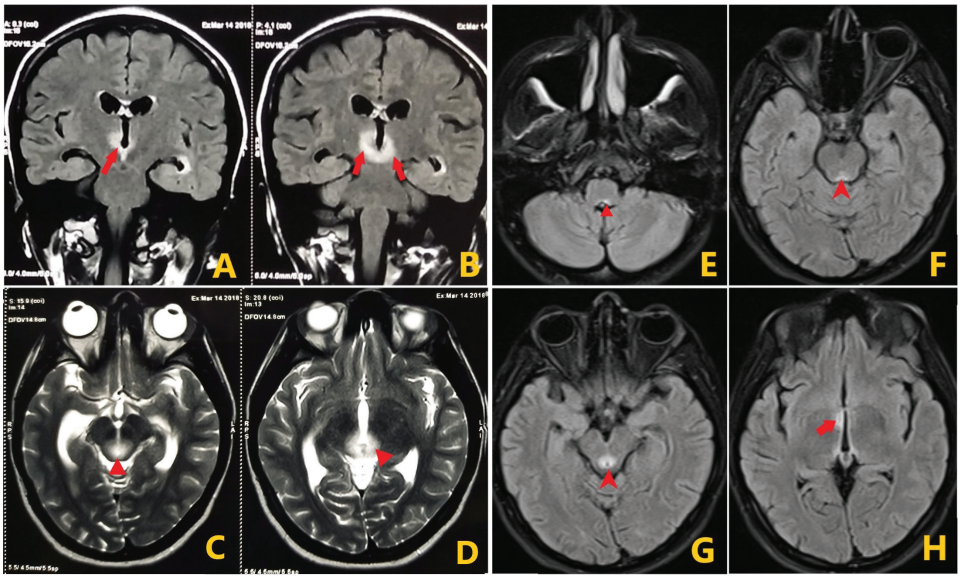

Figure 1 Magnetic resonance images: T2 fluid-attenuated inversion recovery (FLAIR) images (A, B) showing hyperintensities in bilateral dorsomedial thalamus (arrows) and T2-weighted images (C, D) showed hyperintensities in periaqueductal regions (triangular heads). Magnetic resonance images: T2 FLAIR images showing hyperintensities in the area postrema (E) (triangular head) and periaqueductal region (F, G) (arrowheads) and thalamus (H) (arrow).

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Table 1 Similarities and differences between neuromyelitis optica and Wernicke's encephalopathy

	Similarities	**Differences**	
		Neuromyelitis optica	Wernicke's encephalopathy
Clinical features	Encephalopathy or somnolence or confusion Ataxia Vomiting Vertigo or giddiness	Optic neuritis Transverse myelitis	Ophthalmoplegia
Radiological	Around the ventricles (more commonly third and fourth) Periaqueductal area Thalamus or medial thalamus Cerebellum	Hypothalamus Brainstem Corpus callosum Deep white matter Spinal cord Cloud-like enhancement	Mamillary bodies Tectal plate No enhancement
Histopathological	Astrocyte damage	Demyelination	Haemorrhage and cytotoxic oedema in astrocytes and neurons Break of blood-brain barriers
Diagnostic		Aquaporin 4 antibody	Serum thiamine levels
Treatment		Intravenous pulse steroids, oral steroids, immunosuppressants, rituximab	Intravenous or oral thiamine

syndrome, she was evaluated for demyelinating illness, with neuromyelitis optica in particular. Her complete blood picture, blood glucose, electrolytes, renal and liver functions were normal. Her vitamin B1, B6, and B12 levels were normal. She was positive for IgG antibodies against aquaporin 4 (AQP4 autoantibodies). Cerebrospinal fluid (CSF) analysis was negative for oligoclonal bands and infections.

Brain involvement in neuromyelitis optica spectrum disorder (NMOSD) is known to have a variable prevalence ranging from 25% to 84%, and most of them are clinically silent.[1] They typically involve periependymal regions, circumventricular regions, corpus callosum, brainstem, hypothalamus and optic chiasm.[2] In this case, the radiological picture was atypical and rare for NMOSD (Table 1). These imaging findings were characteristic of thiamine deficiency but not specific for the same. Other conditions with similar imaging findings include multiple sclerosis, Miller-Fisher syndrome, Behçet's disease, Leigh's disease, primary cerebral lymphoma, paraneoplastic encephalitis and metabolic disorders like severe hypophosphataemia and acute methyl bromide intoxication.[3] Hence, a detailed history with the clinical picture together with imaging findings is vital to make a concordant diagnosis.

References:

1 Kim HJ, Paul F, Lana-Peixoto MA et al. MRI characteristics of neuromyelitis optica spectrum disorder: An international update. *Neurology* 2015; 84: 1165–73.

2 Dutra BG, Da Rocha AJ, Nunes RH et al.Neuromyelitis optica spectrum disorders: Spectrum of MR imaging findings and their differential diagnosis. *Radiographics* 2018; 38: 662.

3 Sechi G Pietro, Serra A. Wernicke's encephalopathy: new clinical settings and recent advances in diagnosis and management. *Lancet Neurol* 2007; 6: 442–55.