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Neuromyelitis Optica area Postrema Syndrome(NMOSD-APS) in a Fifteen-year-old Girl: A Case Report

Authors : Merilin Ivanova Ivanova, Kalin Dimitrov Atanasov, Stefan Petrov Enchev

Abstract : Backgroud: Neuromyelitis optica spectrum disorder, also known as Devic's disease, is a relapsing demyelinating autoimmune inflammatory disorder of the central nervous system associated with anti-aquaporin 4 (AQP4) antibodies that can manifest with devastating secondary neurological deficits. Most commonly affected are the optic nerves and the spinal cord-clinically this is often presented with optic neuritis (loss of vision), transverse myelitis(weakness or paralysis of extremities),lack of bladder and bowel control, numbness. APS is a core clinical entity of NMOSD and adds to the clinical representation the following symptoms: intractable nausea, vomiting and hiccup, it usually occurs isolated at onset, and can lead to a significant delay in the diagnosis. The condition may have features similar to multiple sclerosis (MS) but the episodes are worse in NMO and it is treated differently. It could be relapsing or monophasic. Possible complications are visual field defects and motor impairment, with potential blindness and irreversible motor deficits. In severe cases, myogenic respiratory failure ensues. The incidence of reported cases is approximately 0.3–4.4 per 100,000. Paediatric cases of NMOSD are rare but have been reported occasionally, comprising less than 5% of the reported cases. Objective: The case serves to show the difficulty when it comes to the diagnostic processes regarding a rare autoimmune disease with non- specific symptoms, taking large interval of rimes to reveal as complete clinical manifestation of the aforementioned syndrome, as well as the necessity of multidisciplinary approach in the setting of a general paediatric department in an emergency hospital. Methods: itpatient's history, clinical presentation, and information from the used diagnostic tools(MRI with contrast of the central nervous system) lead us to the conclusion .This was later on confirmed by the positive results from the anti-aquaporin 4 (AQP4) antibody serology test. Conclusion: APS is a common symptom of NMOSD and is considered a challenge in a differential-diagnostic plan. Gaining an increased awareness of this disease/syndrome, obtaining a detailed patient history, and performing thorough physical examinations are essential if we are to reduce and avoid misdiagnosis.

Keywords : neuromyelitis, devic's disease, hiccup, autoimmune, MRI
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