



Apathy - Forme Fruste of Autoimmune Encephalitis

Ananthanarayanan Kasinathan¹ · Razia K. Adam² · Naveen Sankhyan¹  · Pratibha Singhi³

Received: 29 December 2017 / Accepted: 27 February 2018 / Published online: 13 March 2018
© Dr. K C Chaudhuri Foundation 2018

To the Editor: A previously healthy, 2-y-old boy presented with new-onset behavioral change for the past two months. Parents noticed that he became quiet and distant, indifferent with unusual or minimal response to the questions posed. He soon preferred to stay in bed and with little impetus to eat, drink or speak. His sleep cycle was, however, not altered. His interaction with parents and peers had gradually reduced. For the fifteen days prior to admission, he developed peculiar, repetitive, nose picking behavior and minimal intermittent twisting movements of the limbs. On examination, he was self-absorbed, indifferent, socially withdrawn and lacked all emotional reactions. He was visually attentive and mute. Repetitive nose picking and intermittent dystonia were observed. Cranial nerves, fundus and rest of the neurological examination were normal. Magnetic Resonance Imaging of the brain was unremarkable. Cerebrospinal fluid analysis was acellular with mildly elevated protein (62 mg%), normal sugar (68 mg%) and strongly reactive anti-NMDAR antibodies. He was initiated on a combination therapy with Intravenous Immunoglobulin and pulse methyl prednisolone. At 4 wk follow up, the motor stereotypies and dystonia resolved with marked reduction in apathy. However, the child was hyperactive, had an altered sleep-wake cycle, and persistent speech deficits. He was given Rituximab weekly (375 mg/m²) for four weeks. He showed complete resolution of all symptoms at 8 wk follow-up. At the two-year follow-up, the child was symptom free; was in first standard; had good scholastic skills; normal peer relationship; and no residual motor, speech or behavioral deficits.

Anti-NMDAR encephalitis is a neuroimmune syndrome characterized by profound psychiatric and cognitive deficits [1]. Unlike adults, where psychotic symptoms are dominant, manic features inclusive of irritability; behavioral outburst and hyperactivity are commoner in children [2]. Light-switch mental status with rapid fluctuations in sensorium is another salient presenting feature [3]. The index child merits discussion because of the prominent apathetic features at onset. Apathy as a feature of Limbic encephalitis and Hashimoto thyroiditis has been well reported in adults. However, literature review of the three largest published series in children did not yield any mention [2, 4, 5]. The added findings of motor stereotypies; mild dystonia; and normal neuroimaging were pointers to the underlying diagnosis. Our case highlights the relatively rare apathetic presentation of pediatric Anti-NMDAR encephalitis.

Compliance with Ethical Standards

Conflict of Interest None.

References

1. Graus F, Titulaer MJ, Balu R, et al. A clinical approach to diagnosis of autoimmune encephalitis. *Lancet Neurol.* 2016;15:391–404.
2. Florance NR, Davis RL, Lam C, et al. Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis in children and adolescents. *Ann Neurol.* 2009;66:11–8.
3. DeSena AD, Greenberg BM, Graves D. Light switch mental status changes and irritable insomnia are two particularly salient features of anti-NMDA receptor antibody encephalitis. *Pediatr Neurol.* 2014;51:151–3.
4. Armangue T, Titulaer MJ, Malaga I, et al. Pediatric anti-N-methyl-D-aspartate receptor encephalitis-clinical analysis and novel findings in a series of 20 patients. *J Pediatr.* 2013;162:850–6.e2.
5. Sartori S, Nosadini M, Cesaroni E, et al. Paediatric anti-N-methyl-D-aspartate receptor encephalitis: the first Italian multicenter case series. *Eur J Paediatr Neurol.* 2015;19:453–63.

✉ Naveen Sankhyan
dmsankhyan@yahoo.co.in

¹ Pediatric Neurology and Neurodevelopment Unit, Department of Pediatrics, Postgraduate Institute of Medical Education and Research, Chandigarh 160012, India

² Department of Pediatrics, Super Speciality Pediatric Hospital and Postgraduate Teaching Institute, Noida 201310, India

³ Department of Pediatrics, Medanta, The Medicity, Gurgaon, India