

Narcolepsy - A Rare but under-recognized Problem in Children

Narcolepsy is a rare but potentially debilitating, rapid eye movement (REM) sleep disorder that causes significant developmental morbidity [1]. It is characterized by the classic tetrad of disproportionate daytime sleepiness with irrepressible sleep attacks, cataplexy, hypnagogic hallucination, and sleep paralysis [2]. Narcolepsy has an early onset but is associated with significant delay in – symptom recognition and diagnosis [2,3]. Nocturnal polysomnography (PSG) followed by a daytime multiple sleep latency test (MSLT) is the diagnostic standard.

A 4-year-8-month old school-going boy, presented with complaints of excessive daytime sleepiness and intense urge to sleep for last one year, and drop attacks for last six months. There was no significant family history of seizures or neurological disorder. In view of recurrent/persistent symptoms, he was extensively evaluated for seizure disorder/epilepsy syndrome/Wilson's disease. In view of inconclusive results and persistent symptoms, he was started on anti-epileptics but later referred to us due to persistence of symptoms.

On examination, he was neuro-developmentally normal with both general physical and systemic examination within normal limits. During conversation with the parents, the child not only had intense urge to sleep but also had a drop attack. Considering the symptoms and chronicity of presentation in otherwise developmentally normal child, a provisional diagnosis of narcolepsy with cataplexy was considered. Child underwent a nocturnal polysomnography (PSG) followed by daytime multiple sleep latency test (MSLT). Nocturnal PSG was within normal limits and MSLT showed, a mean sleep latency of 4 minutes with average REM latency of 1.4 minutes, thus diagnostic of Narcolepsy. A genetic test (HLA B1*0602) to support the diagnosis was positive,

thus confirming the diagnosis. He was treated with Modafanil (50 mg/day) with good clinical response with respect to both excessive daytime sleepiness and cataplexy.

Patients with narcolepsy are usually initially investigated for epilepsy, encephalopathy, and psychiatric disease [3,4]. A lack of awareness of the condition, delay in symptom recognition and absence of all the characteristic features are the primary reason for missed diagnosis of narcolepsy in children [3,4].

Administering daytime MSLT after overnight PSG is the primary modality of choice for diagnosis of narcolepsy [3,5]. Presence of both: (a) a mean sleep latency (MSL) of <8 minutes and (b) two or more sleep onset REM periods (SOREMPs) on MSLT performed after at least six hours of sleep during the previous night confirms the diagnosis [3,5]. Management of narcolepsy is multimodal and involves Sleep hygiene, frequent daytime naps, Diet recommendations, Medications, and Caregiver counseling. Though lifestyle modifications are important in management, medical treatment is the cornerstone and should be initiated as early as possible after confirming the diagnosis.

JT SRIKANTA* AND KM CHANDAN KUMAR

Institute of Pulmonology, Apollo Hospitals, Bengaluru, India.

**Shrek_jt@yahoo.com*

REFERENCES

1. Thorpy MJ, Krieger AC. Delayed diagnosis of narcolepsy: Characterization and impact. *Sleep Med.* 2014;15:502-7.
2. Daniels LE. Narcolepsy. *Medicine.* 1934;13:1-122.
3. Thorpy MJ. Diagnostic criteria and delay in diagnosis of narcolepsy. *In: Goswami M, Thorpy M, Pandi-Perumal S (eds). Narcolepsy.* Springer Cham. 2016.
4. Morrish E, King MA, Smith IE, Shneerson JM. Factors associated with a delay in the diagnosis of narcolepsy. *Sleep Med.* 2004;5:37-41.
5. American Academy of Sleep Medicine. *The International Classification of Sleep Disorders, Diagnostic and Coding Manual.* 2nd ed. Westchester, IL: American Academy of Sleep Medicine. 2005. p.298-9.