

Case report / Olgu sunumu**An under-recognized case of childhood narcolepsy with cataplexy****Serkan GÜNEŞ,¹ Mehmet Akif CAMKURT²****ABSTRACT**

Narcolepsy is a type of dyssomnia characterized by excessive daytime sleepiness, cataplexy, sleep paralysis, and hypnagogic hallucinations. The signs of narcolepsy usually begin during the teenage years and it persists throughout the lifetime. Diagnosing narcolepsy in children can be challenging and the disorder can remain undiagnosed until adulthood. We describe here a case of childhood narcolepsy with cataplexy who was not diagnosed accurately during several admissions. The patient's signs were initially related to conversion disorder and she took inappropriate medications. However, after a polysomnography, the diagnosis of narcolepsy was established and she was treated appropriately with stimulant medications. (*Anatolian Journal of Psychiatry* 2017; 18(4):407-409)

Keywords: cataplexy, childhood, narcolepsy

Tanı konamamış katapleksi ile giden çocukluk çağı narkolepsi olgusu

ÖZ

Narkolepsi gün içinde aşırı uykulu olma, katapleksi, uyku paralizi ve hipnagogik varsanılarla karakterize bir uyku bozukluğudur. Narkolepsi belirtileri genellikle çocukluk çağında başlayıp yaşam boyu sürmektedir. Çocuklarda narkolepsi tanısı koymak çoğu zaman zor olabilmekte ve bu bozukluk tanı konulamadan yetişkinliğe kadar sürebilmektedir. Bu yazıda, birçok kliniğe başvurmasına rağmen kesin tanısı konulamayan bir çocukluk çağı narkolepsi olgusu sunulmuştur. Hastanın belirtileri ilk olarak konversiyon bozukluğu ile ilişkilendirilmiş ve bu nedenle hastaya uygunsuz bir tedavi başlanmıştır. Ancak, polisomnografi uygulandıktan sonra narkolepsi tanısı konulmuş ve stimulan tedavisine geçilmiştir. (*Anadolu Psikiyatri Derg* 2017; 18(4):407-409)

Anahtar sözcükler: Katapleksi, çocukluk çağı, narkolepsi

INTRODUCTION

Narcolepsy is a chronic neurological disease usually occurring from childhood and persisting through adolescence and adulthood.¹ It is believed that narcolepsy results from autoimmune destruction of hypocretin-producing neurons and dysfunction of hypocretin-1 and hypocretin-2 (orexin-A and orexin-B) hormones.² These hormones are known as sleep-wake and rapid eye

movement sleep-regulating neuropeptides.³ Patients with narcolepsy show excessive daytime sleepiness, sleep attacks, dyssomnia, metabolic abnormalities including obesity, and problems in social interaction and academic performance.¹ The prevalence of narcolepsy in pediatric population remains unknown but can be estimated to be 20-60 per 100.000 in Western countries. However, recent studies suggest an increase of narcoleptic cases both in children and in adults.¹

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Narcolepsy is frequently under-recognized or under-diagnosed, especially when its signs begin during childhood.⁴ To bring attention to this diagnosis, we report a case of childhood narcolepsy with cataplexy who was under-recognized for several admissions.

CASE

The patient was an 11-year-old girl who developed progressively worsening excessive daytime sleepiness. Initially, she showed light sleep and frequent arousal at night. Then, she started taking naps during the day and fell asleep in class. She eventually could not function well in school. She also reported visual hallucinations before the onset of sleep. During her first evaluation in pediatrics clinic, many diagnoses were considered, but clinicians did not make an accurate diagnosis. So, she was referred to pediatric neurology clinic. Her blood examination, EEG, and brain MRI were normal. She did not show any other neurologic findings and was misdiagnosed with conversion disorder and forwarded to a child and adolescent psychiatrist. She was admitted to a child and adolescent psychiatry clinic for the same problems. Aripiprazole 5 mg/day was initiated, but she did not benefit and her symptoms worsened. When she first came to our psychiatry clinic, we thought it might be a kind of sleep disorders. So, we initiated imipramine 50 mg/day, but she did not benefit enough from the medication. One month after, on the second psychiatric evaluation, she fell to the ground for a few seconds with no stimulus. This definite observation of cataplexy led us to think about narcolepsy. Parents of the patient mentioned that she also showed cataplexy at home and school. After some of these episodes, she fell asleep. But no history of sleep paralysis or weight change was reported. Nocturnal polysomnography showed reduced sleep latency, predominance of sleep stage I and II, increased number of awakenings, and disrupted sleep. She was diagnosed with narcolepsy with cataplexy and began methylphenidate 20 mg/day. On follow-up visits up to 10 months, we observed a reduction in her daytime sleepiness, she was more awake and interactive. She was able to function very well in school and her marks increased significantly. She did not have any cataplectic episodes after initiation of methylphenidate.

DISCUSSION

The diagnosis of narcolepsy in pediatric popula-

tion can be challenging, particularly in younger children.⁵ Key reasons are the unfamiliarity of the disorder among clinicians, negative test results in early stages, and atypical presentations of the features.⁶ The tetrad of symptoms (excessive daytime sleepiness, cataplexy, sleep paralysis, and hypnagogic/hypnopompic hallucinations) occurring together is a rare condition in childhood narcolepsy.⁷ If these symptoms are present, they may show variable presentations compared with adults and make it difficult to recognize. For example, children with narcolepsy may present cataplexy that is not related to emotion.⁸ As a result, diagnoses can be delayed by 10-15 years after the onset of signs and the financial and emotional results of such a long process can be substantial for the patient and family.^{5,8} In our case, we also see cataplexy unrelated to emotion and delayed diagnosis in several visits to various clinicians.

Differential diagnoses of childhood narcolepsy include endocrine problems, epilepsy, attention deficit hyperactivity disorder, depression, obstructive sleep apnea, and other hypersomnias, etc.^{4,5} Unfortunately, when a child presents with features of excessive daytime sleepiness, clinicians often think of tiredness, laziness, inattentiveness, depression, or epilepsy. In addition, cataplexy might be confused with a variety of seizure types.^{4,6} Therefore, narcolepsy is also important to include in this differential diagnosis.

The early recognition and treatment of this disorder allow children to improve their quality of life and achieve normality both academically and socially.⁹ The key point to the early diagnosis and treatment of children with narcolepsy includes regular evaluation for identification, testing, counseling, and medication.^{5,9} Taking detailed medical history and being sensitive for variable presentations of narcolepsy are also important.¹⁰ Lack of clinical awareness about the atypical presentations can result in misdiagnosis and inappropriate medication.⁸ In this context, we see that our case was misdiagnosed with conversion disorder, and so inappropriate medication worsened her symptoms.

In conclusion, this case was reported in order to bring attention to childhood narcolepsy and warn clinicians to be careful about this diagnosis. Because, a misdiagnosis can have negative effects on children, and early diagnosis and treatment can greatly improve their quality of life. When we observe children with excessive daytime sleepiness, we should consider the possibility of narcolepsy with cataplexy.

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