NARCOLEPSY IN A YOUNG CHINESE CHILD

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ABSTRACT

Narcolepsy is characterized by narcoleptic sleep attacks, cataplexy, sleep paralysis and hypnagogic hallucinations. It is very rare in children. A 7-year old Chinese boy presenting with typical narcoleptic symptoms is reported. Diagnosis of narcolepsy usually relies on the clinical picture, the presence of sleep-onset REM periods on nocturnal polysomnograph and the Multiple Sleep Latency Test. The strong association with HLA-DR2 may help in diagnosis, especially in children.

Keywords: Narcolepsy, Child , Chinese

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INTRODUCTION

Narcolepsy is characterized by sleep attacks, hypnagogic hallucinations, sleep paralysis and cataplexy. Its occurrence in children is very rare. Yoss and Daly [1], in a review of 400 patients diagnosed as narcoleptics, found that only 4% were 15 years or younger. Kanner [2] reported 2 cases of hypersomnolence in children, and commented that the early onset at age 8 was remarkable in one of them. Salfield [3] described a 6-year old boy diagnosed as narcoleptic. Guilleminault [4] reported that in their sleep disorders clinic at Stanford, over a 10-year period, they had only seen 5 children (aged between 7 and 11 years) referred for suspected narcolepsy and 5 for suspected cataplexy. He concluded that narcolepsy rarely manifests before puberty. We would like to report on a Chinese boy who presented with narcolepsy at the age of six in Hong Kong.

CASE REPORT

C is a 7-year old student and the second of 3 siblings. His birth history and developmental milestones were normal. His family members are all healthy. At the age of 6, he was hit on the head at the right parietal region by a basketball. He looked dazed for a short while but had no loss of consciousness or external injuries

One month later, he developed repeated episodes of falling asleep during the day. The attacks occurred about 10 times a day lasting from 15 minutes to an hour. He would fall asleep in the classroom or during walking when he dropped to sleep on the ground. Despite its long duration, his night sleep was disrupted, and associated with frequent awakenings and excessive bodily movements. There is no history of snoring. Soon he began to experience repeated episodes of hypnagogic hallucinations. While he was dozing off in the sitting room, he was frightened by ghosts or people standing beside him so that he insisted on the company of his mother. There were several episodes when he experienced muscle weakness of the whole body and dropped to the ground and slept. These episodes were not associated with laughter or strong emotions. He never reported sleep paralysis. His character also changed, becoming irritable and quarrelsome, resulting in temper tantrums and fights with his siblings. With an increased appetite, he gained 8 pounds in six months, whereas his previous body weight was 50 pounds.

Physical and ENT examination, complete blood picture, renal function, liver function, blood sugar and thyroid function, brain stem evoked potential, electroencephalogram, and CT scan of brain were all normal. A diagnosis of narcolepsy was made. On HLA typing, he as well as his mother and elder brother were DR2 positive. He was started on methylphenidate 15 mg om and noon. This resulted in significant improvement in his daytime sleepiness but not night sleep.

He was subsequently referred to our sleep disorders clinic. Sleep recording was carried out one month after withdrawal of methylphenidate. On Multiple Sleep Latency Test (MSLT), the mean daily score of sleep latency was 0.8 minutes across 5 daytime naps. There were 5 sleep onset REM (SOREM) episodes in the 5 daytime naps. His mean night time sleep lasted for about 10 hours on 3 nights' recording, with many awakenings and SOREM periods. There was no sleep apnoea. Twenty-four hour EEG did not reveal any seizure activities. Hence the

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diagnosis of narcolepsy was confirmed.

DISCUSSION

A review of the literature shows that narcolepsy is very rare in childhood. The cases reported by Kanner [2] and Salfield[3] were diagnosed clinically, not polysomnographically confirmed, and could have been suffering from other causes of excessive daytime sleepiness instead.

The diagnosis of narcolepsy is usually suggested by the clinical picture but polysomnographic recordings are needed for an objective confirmation. Nocturnal recordings usually reveal the presence of SOREM periods (defined as the appearance of REM sleep during the first 15 minutes of sleep onset), and absence of obstructive sleep apnoea syndrome. The MSLT measures a person's degree of daytime sleepiness and identifies SOREM periods. It measures the subject's sleep latency across 5 naps scheduled 2 hours apart starting from 10 a.m. Pathological sleepiness is revealed by a mean sleep latency of less than 7 minutes [4]. The presence of 2 SOREM episodes out of the 5 naps is proposed as diagnostic of narcolepy in adults [5].

The diagnosis of narcolepsy is difficult before puberty because the initial presentation of daytime sleepiness without cataplexy may lead to a delay in diagnosis especially if school performance is maintained adequately [4]. Morever, SOREM period was present in only one out of 25 teenagers in Guilleminault's series [4] on the MSLT test.

Our patient is probably the first Chinese patient reported to have narcolepsy at such an early age. The early onset and the severe symptoms are remarkable in this case. The presence of HLA-DR2 in this patient is consistent with the findings of Honda [6] who concluded that HLA-DR2 was a prerequisite for the development of narcolepsy. In a child whose clinical picture is suggestive of narcolepsy but without any definite evidence on the MSLT or overnight polysomnograph, the presence of HLA-DR2 may support the diagnosis.

The usual age of onset in narcolepsy is after puberty. It is tempting to speculate that hormonal or other developmental factors during puberty may facilitate the emergence of the symptoms. Whether the minor head injury sustained a month before the onset of symptoms in our patient acts as a precipitating factor is uncertain. In patients who are genetically predisposed to develop narcolepsy, minor insults to the brain may precipitate the illness at an earlier age but this would need to be confirmed in a larger series of patients.

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