Case Report :

Narcolepsy type 1 with partial cataplectic features: a case report

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ABSTRACT

Though being the second most common cause of disabling daytime sleepiness in the world narcolepsy in India is seldom diagnosed (1). So far only a few cases have been reported. This is a case of narcolepsy to be reported from India in a 15 year old female student struggling in her scholastic pursuit because of the disease.

Key Words:

Narcolepsy, Cataplexy, Epworth Sleepiness Scale (ESS), Multiple Sleep Latency Test (MSLT)

INTRODUCTION :

Narcolepsy is a clinical syndrome of daytime sleepiness with cataplexy, hypnagogic hallucinations, and sleep paralysis. Narcolepsy with cataplexy is called type 1 Narcolepsy while that without cataplexy is Narcolepsy -type 2. Narcolepsy (type-1) has a prevalence of 25 to 50 per 100,000 people according to western data (2). If prevalence is considered same in India, there is a lot of under diagnosis in reality. Here, we have described a girl student of 15 years of age who presented with type-1narcolepsy and the experience of her improvement following use of Armodanafil.

CASE HISTORY :

A 15-year-old girl was referred for evaluation of excessive daytime sleepiness for the past three years. She was mainly concerned for uncontrollable urges of sleep during lectures classes. Her academic track record has been excellent and as an aspirant for studying medicine, she had to attend roughly five hours of lecture classes for six days a week and study for a similar duration a day at her home. While on this routine she had noted sudden spells of strong urge for sleep that forced her unable to concentrate in the classes. To prevent the sleepiness some times, she even tried to remain standing during the class hours but even then could not resist from falling asleep. At occasions, it happened that she fell on the benches while standing even though she was aware of the circumstances on those moments.

This abnormal sleepiness was first noted three years ago during her ninth grade of schooling and it had gradually worsened over time. She used to have refreshing sleep of 7-8 hours in a night without snoring or awakening when she used to go to bed following studying for a few hours. She has no history of consumption of coffee or watching television before sleep. Her mother use to wake her up daily morning as a matter of habit. The excessive sleepiness used to begin after a few hours of getting up from refreshing sleep and, on an average; she used to get 2-3 urges of excessive sleepiness a day. She generally used to takes a short nap of one hour in the afternoons and had noticed some shadow or an image in room occasionally in the beginning of such naps. She had transient weakness and tingling sensation in her arms and legs often on waking up with her movements turning a little slow and clumsy after the spells of sleep. However, she had not experienced any weakness or tingling sensation in any other situation. She has no problems in

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falling asleep that occur usually within 5 minutes of going to bed. The patient didn't complain of any snoring problem

She remained emotionally stable but had experienced several episodes of forgetfulness, which was affecting her academic performances. She doesn't drink coffee or tea. She denied use of illicit drugs, smoking, and consuming alcohol. She was otherwise healthy and was not taking any medicine for any reason. There was no family history of excessive sleepiness and there was no history of seizures or any psychotic disorder or history of any vaccination.

On evaluation with the Epworth Sleepiness Scale (ESS) questionnaire, she scored 19. The physical examination was unremarkable. She weighed 48 kg with BMI of 17.5. Full night polysomnography was done by Philips Alice 6 LDE 16 channel polysomnogram using Sleepware G 3 diagnostic software.Total duration of sleep evaluated was of 437 minutes. The report ruled out obstructive sleep apnoea as cause for excessive daytime sleepiness. A reduced sleep efficiency of 65% was seen with presence of spontaneous awakenings and onset of REM sleep of 1.5 minutes

The Multiple Sleep Latency Test (MSLT) was performed on the very next day 4 hours after waking up in the morning with 2 hour intervals until patient was given 4 opportunities to nap and monitored for at least 15-20 minutes after onset of sleep. The MSLT revealed onset of sleep within two minutes. The monitored naps consisted of REM sleep with its onset in less than 7 minutes of falling asleep in all the four naps (SOREMPs). MRI Brain study was interpreted as normal.

Hence, a diagnosis of narcolepsy was made based on following clinical parameters as a) excessive and irresistible urges of sleep several times a day with b) history suggestive of cataplexy and sleep paralysis and c) history of hypnopompic and hypnogogic hallucinations. This diagnosis was supported and confirmed by a) the presence of early onset REM sleep, multiple awakening during sleep in polysomnography and b) early onset of REM sleep with SOREM in all four tests on MSLT.

The patient was started on armodanafil (Wakert) 150 mg once daily orally at morning and followed up. She was also counseled on sleep hygiene and was told to get at least 8 hours of sleep at night and take a short afternoon nap of 15-30 minutes. After two months of pharmacotherapy and adherence to the advice of sleeping hygiene, the patient had significant improvement in wakefulness, memory and attention. Epworth Sleepiness Scale (ESS) was again looked for and the score came down to 7.

DISCUSSION :

Etiology of narcolepsy has been discovered recently as loss of orexin (3, 4), a neuropeptide neurotransmitter made by neurons in the lateral hypothalamus.

Other rare causes include brain lesions in the posterior hypothalamus andmidbrain, Prader-Willi syndrome, autoimmunity to HLA haplotype DQB1*0602. Some researchers have mentioned its association with 2009 H1N1 influenza vaccine Pandemrix (5).

Classic clinical features of narcolepsy are chronic daytime sleepiness, cataplexy, and hypnagogic hallucinations and sleep paralysis. Only 33%ofpatients will have all of these classical findings. If a diagnosis is suspected then ESS (Epworth Sleepiness Scale) should be administered. ESS score more than 15 highly suggestive of narcolepsy (6)and should be confirmed with Polysomnogram (PSG) and Multiple sleep latency test (MSLT). While PSG rules out other causes of excessive daytime sleepiness, MSLT confirms the diagnosis of narcolepsy. MSLT has high specificity for narcolepsy, but can be only valid if PSG shows a minimum of 6 hours of sleep at preceding night of the day on which MSLT has been decided to be performed. The classical findings on the MSLT confirming narcolepsy are sleep latency less than eight minutes and two or more sleep onset rapid eye movement periods (SOREMPs). (7) Two other tests with very limited significance are Human Leukocyte Antigen (HLA) typing (HLA DQB1*0602 and HLA DQA1*0102) and measurement of cerebrospinal fluid (CSF) hypo cretin level.

Narcolepsy is managed with a combination of nonpharmacologic and pharmacologic therapy. Maintaining good sleep hygiene, avoiding sedatives and psychological support constitute nonpharmacologic approach.

Managing narcolepsy is a combination of nonpharmacologic and pharmacologic therapy.

Behavioral therapy includes career counseling regarding the jobs; the patient should avoid such as shift work, on call schedules etc. In addition to scheduled naps other behavioral approaches such as good sleep hygiene, avoidance of frequent changes in time zone, avoidance of sedatives and hypnotics, avoiding heavy meals should be observed.

Medical management involves medications for sleepiness and medications for cataplexy (8-16). Medications for sleepiness include modafinil, methylephenidate, amphetamine analogues, and dextroamphitamine sulfate. The latter is associated with rebound hypersomnia. The medications for cataplexy include gammahydroxybutyrate (being used at a doses of 6-9g/day), antidepressant drugs such as fluoxetine , venlafaxine, viloxazine, and other drugs as protryiptyline, imipramine, clomipramine, desipramine. The last category of drugs is known to have atropinic side effects.

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