Case Report of Secondary Narcolepsy Presenting as Self-Inflicted Genital Injury

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Abstract

Primary Narcolepsy is a sleep disorder with classical presentation showing symptoms of cataplexy, excessive daytime sleepiness, sleep paralysis, and Hypnogogic hallucinations. A rare case of secondary narcolepsy was seen in a patient with self-inflicted genital injury. A 30 year old male was referred to Psychiatry from Surgery for a self-inflicted incised wound on hydrocele. Since last 1 year, he had multiple episodes of -- 1. Sudden falls while working 2. Sleep during daytime often at unusual places 3. Periods of unresponsiveness during which he was aware but unable to move. During hospital stay, all symptoms of Narcolepsy were noted several times, but prominent hallucinations were not consistently reported. Patient's EEG was normal. MRI brain showed Gliosis at cervico-medullary junction.MRI spine was advised to examine the cervico-vertebral junction but patient was lost to follow-up. But from history and investigations, based on DSM-IV-TR, Narcolepsy was diagnosed. It was concluded that he had secondary narcolepsy due to traumatic brain injury. As Self Inflicted Genital injury was done after hallucinations and sleep attack, it was considered to be part of Narcolepsy - a rare presentation. Narcolepsy typically begins in young adulthood mainly 2nd decades of life and causes adverse effects on quality of life of patients. Diagnosis relies on history of patient and objective details obtained from polysomnography and multiple sleep latency testing. Main treatment is symptomatic relief through medication, psycho education, and behavioural modification.

Keywords: Cataplexy, Narcolepsy, Polysomnography, Self-inflicted injury

Introduction

Narcolepsy is neither a type of epilepsy nor a psychogenic disturbance. It is an abnormality of the sleep mechanisms specifically, REM-inhibiting mechanisms. Primary Narcolepsy has been studied in dogs, sheep, and humans. Few conditions that lead to secondary narcolepsy are tumors, traumatic brain injury and stroke [1]. Narcolepsy can occur at any age, but it most frequently begins in adolescence or young adulthood, generally before the age of 30. The disorder either progresses slowly or reaches a plateau that is maintained throughout life [2]. The prevalence of narcolepsy varies across countries and with different ethnic groups, so the exact prevalence is not known. Prevalence has been reported to be between 168 and 799 per 100,000 in most studies, although Japanese studies have indicated a higher prevalence of 100,000 [2,3]. No genetic tests are currently available to make diagnosis of narcolepsy. Genetic tests may correlate best to narcolepsy when there is already established cataplexy [4,5].

Supporting the evidence for an environmental influence is the fact that the disease is not apparent at birth, but commonly has its onset during the second decade of life. There are some precipitating factors like, infections, head trauma, and changes in sleep-waking habits which have been identified in some cases. In many patients with Traumatic Brain Injury (TBI), Chronic daytime sleepiness is a major, disabling symptom, but its etiology is not well understood so far. One possible cause can be extensive loss of the hypothalamic neurons that produce the wake-promoting neuropeptide hypocretin which causes the severe sleepiness of narcolepsy, and partial loss of these cells may contribute to the sleepiness of the disorder. One study has found that the number of hypocretin neurons is significantly reduced in patients with severe TBI. This observation highlights the often overlooked hypothalamic injury in TBI and provides new insights into the causes of chronic sleepiness in patients with TBI [6]. Substances like Amphetamine has been associated with addiction, psychosis and self-injurious behaviour. There are reports on some patients who repeatedly and severely mutilated their own genitalia while intoxicated with amphetamines and consider possible diagnostic aetiologies [7]. Genital mutilation is common in males compared to females [8]. But http://www.omicsgroup.org/journals/clinical-case-reports.php t narcolepsy presenting as self-inflicted genital injury has not been reported so far. That is why this is a rare case.

Case History

A case of secondary narcolepsy was seen in a patient presenting as self-inflicted genital injury. A 30 year old Hindi speaking unmarried illiterate male was referred to Psychiatry Department from Surgical side for a self-inflicted incised wound on swelling of hydrocele. Patient was admitted in surgical ward in emergency for bleeding scrotum. On detailed evaluation it was found that patient himself had cut his scrotal swelling with blade at home and severe bleeds started so he was taken to Hospital urgently (Figure 1) after primary wound closure at surgical side, patient was taken transfer to Psychiatry for detailed assessment for such behaviour. On taking detailed history from his elder brother and father, it was found that since last 1 year, he had multiple episodes of sudden falls while working at kitchen as he was a cook. He used to sleep during daytime often at unusual places like in the courtyard, once...
During hospital stay of about 10 days also, daytime somnolence, sleep paralysis and cataplexy were noted several times, but hallucinations were not consistently reported. Several investigations like complete blood count, blood sugar, liver function tests, renal function tests, cardiogram were carried out to rule out any pathology but all reports were within normal range (Figure 3). Patient was also assessed for Major Depression, Schizophrenia and Borderline Personality disorder by using Hamilton Depression Scale, PANSS and DSM-IV criteria for Borderline Personality Disorder. His EEG was also normal. In history there were multiple episodes of fall without loss of consciousness or bleeding or convulsions. At last Plain MRI Brain was done to find out any organic pathology. On MRI Brain, there was gliosis at cervico-medullary junction (Figure 2). Based on DSM-IV-TR Narcolepsy was diagnosed. MRI spine advised to examine the cervico-vertebral junction but patient was lost to follow-up. But from history and investigations, it was concluded that he had secondary narcolepsy due to traumatic brain injury.

Discussion

Narcolepsy is a condition characterized by excessive sleepiness, as well as auxiliary symptoms that represent the intrusion of aspects of REM sleep into the waking state. The sleep attacks of narcolepsy represent episodes of irresistible sleepiness, leading to perhaps 10 to 20 minutes of sleep, after which the patient feels refreshed, at least briefly. They can occur at inappropriate times (e.g., while eating, talking, or driving and during sex). The REM sleep includes Hypnagogic and Hypnopompic hallucinations, cataplexy, and sleep paralysis. The appearance of REM sleep within 10 minutes of sleep onset (sleep-onset REM periods) is also considered evidence of narcolepsy. The disorder can be dangerous because it can lead to automobile and industrial accidents.

Other symptoms include Hypnagogic or Hypnopompic hallucinations, which are vivid perceptual experiences, either auditory or visual, occurring at sleep onset or on awakening. Patients are often momentarily frightened, but within a minute or two they return to an entirely normal frame of mind and are aware that nothing was actually there. Here patient had symptoms of narcolepsy after head injury and patient himself injured his scrotum with sharp blade for which he had no clear memory and there was no history suggestive of epilepsy. He also reported some unclear hearing of voices before putting incision on scrotum. So diagnosis of secondary narcolepsy presenting as self-inflicted genital injury was considered. In this case, patient showed clinical features of narcolepsy as diagnosed by DSM-IV which was secondary type as there was history of multiple falls and MRI brain showed gliosis. But in this case, unusual presentation was genital self-inflicted injury. To make confirm diagnosis in this case, polysomnography and sleep EEG were needed. Also to find out the

Figure 1: Cut in the scrotal.

Figure 2: MRI Brain, gliosis at cervico-medullary junction.

Figure 3: Types of Narcolepsy.
extent of gliosis in spine, MRI cervical spine was necessary but patient was given discharge on request for his personal reasons, we could not perform confirmatory tests.

Possible reasons for genital injury are:
1. Automatic behavior.
2. Acting on Hypnogogic/Hypnopompic hallucinations.
3. Due to impaired cognitive function/ judgment due to long-standing Narcolepsy.

No cure exists for narcolepsy, but symptom management is possible. A regimen of forced naps at a regular time of day occasionally helps patients with narcolepsy and, in some cases, the regimen alone, without medication, can almost cure the condition. When medication is required, stimulants are most commonly used [9]. Although drug therapy is the treatment of choice, the overall therapeutic approach should include scheduled naps, lifestyle adjustment, psychological counselling, drug holidays to reduce tolerance, and careful monitoring of drug refills, general health, and cardiac status.

References