CASE REPORT

Narcolepsy in a Pre-teen Girl: A Case Report

W.A.Sheikh[1*], R.C. Fubisha[2]

[1]Department of Psychiatry, Livingstone Central Hospital
[2]Department of Pediatrics & Child Health, Livingstone Central Hospital

ABSTRACT

Background: Narcolepsy is a neurological disorder, the main classic feature of which is excessive day time sleepiness (EDS), with recurrent episodes of irresistible sleep (sleep attacks). It is thought to result from genetic predisposition, abnormal neurotransmitter functioning and sensitivity, and abnormal immune modulation. The onset of narcolepsy symptoms usually starts in late 2nd or 3rd decade of life. Rare cases of narcolepsy have been reported to occur in the first decade of life, even in infants. We are presenting a case of narcolepsy in a 10 years old girl, who presented at Livingstone Central Hospital. This case has been presented due to rarity of this condition in this age group. To the best of our knowledge, this is the first documented case of narcolepsy in a pre-teenaged child in Zambia.

Objective: To familiarize the health workers about narcolepsy and diagnostic challenges it pose in resource poor Zambian health settings.

Case History: A 10 years old girl was brought to the pediatrics department of Livingstone Central Hospital by her mother with complaints of having episodes of EDS for the past three years. The onset was insidious and episodes had increased in frequency in past 3 years. She used to sleep while in the class, during talking and sometimes during eating. There was no history of having cataplexy, hallucinations or sleep paralysis. Clinical diagnosis of narcolepsy without cataplexy was made and she was treated with oral fluoxetine and modafanil. She was discharged after staying in the hospital for 6 days. At her follow up after 3weeks, the patient had responded well to treatment and was free of those episodes of day time sleepiness. Currently the patient is doing fine and still on the same treatment.Her FBC, DLC, ESR, LFTs, Urine routine & microscopy, HIV test, all came out normal. She was weighing 43 Kg and was 1.43m tall. Her CT scan of brain didn't show any abnormality. Her physical & neurological examinations were unremarkable.

Conclusion: Narcolepsy in children is a rare condition but once it presents, it poses diagnostic challenges in Zambia due to unavailability of sleep laboratories and lack of knowledge among health workers about this disorder.

INTRODUCTION & LITERATURE REVIEW

Narcolepsy is a neurological disorder, the main classic feature of which is excessive day time sleepiness (EDS), with recurrent episodes of irresistible sleep (sleep attacks).[1] In its fully developed form the "narcolepsy syndrome" also includes sudden loss of muscle tone in response to strong emotion (cataplexy), vivid dream-like experiences before falling asleep (hypnagogic hallucinations) or on waking (hypnopompic hallucinations), and episodes of inability to move after waking in the morning (sleep paralysis).[1]5% of the adults are excessively sleepy to a clinically relevant extent.[2]The comparable figure in children is

Corresponding Author*
W.A.Sheikh
Department of Psychiatry, Livingstone Central Hospital
Phone number: +260977796947
E. Mail: sheikhdr@live.com

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not well known but sleepiness is associated with many different conditions. Narcolepsy in children is a cause of excessive sleepiness during day time. It has been a topic which has been neglected and not very well studied but it can be the cause of serious psychological and social disadvantage.[3] The onset of narcolepsy symptoms usually starts in late 2nd or 3rd decade of life.[4, 5] Rare cases of narcolepsy have been reported to occur in the first decade of life, even in infants.[6,7] Secondary form of narcolepsy, due to cerebral disease can occur at any age.[8, 9] Cataplexy usually follows the onset of day time sleepiness. Cataplexy, hypnagogic hallucinations, and sleep paralysis, may develop slowly, suddenly or not at all.[10]

Narcolepsy affects men and women equally. Because Narcolepsy may be mistaken for depression, epilepsy or psychiatric illness, an accurate diagnosis is often delayed for many years after the onset of symptoms. It is estimated that fewer than 50% of the patients with narcolepsy have been diagnosed.[10] Narcolepsy is affecting approximately 1 in 2000 people in United States.[10]

There are three main types of narcolepsy.[10]

1. Narcolepsy with cataplexy
2. Narcolepsy without cataplexy
3. Secondary narcolepsy

According to the American Academy of Sleep Medicine, the symptoms must be present for at least three months to make the diagnosis of narcolepsy.[11] The diagnosis can be confirmed by the occurrence of typical polysomnographic features of narcolepsy seen on overnight sleep studies.[10] In Zambia currently there is no sleep laboratory to conduct polysomnographic studies. In the absence of polysomnographic studies it is difficult to make a definitive diagnosis of narcolepsy so one can only make a clinical diagnosis. Rarity of narcolepsy cases, scarcity of neurologists in Zambia, and lack of knowledge among health workers about this condition further makes it difficult for the patients with narcolepsy.

Stimulants like methylphenidate, dextroamphetamine have been used to treat excessive day time sleepiness in narcoleptic patients but in recent times, modafanil has become the first-line treatment.[10] Modafanil has low abuse potential and has no association with rebound hyper somnolence, which usually occurs with other stimulants. Usual dose of modafanil is about 200-400mg/day and it can be conveniently administered once daily due to its long elimination half-life (10-14 hours). TCAs and SSRIs have also been used to treat narcoleptic patients.[10] TCAs use is limited due to side effects like dry mouth, urinary retention, tachycardia and sexual dysfunction. The adverse effects profile of SSRIs is superior to TCAs but they can still cause sexual dysfunction and disturbed nocturnal sleep. Sodium oxybate has beneficial effects on both daytime sleepiness and cataplexy.[12] Sodium oxybate can be used in the dose ranging from 4.5g/day to 9g/day. It can cause side effects like nausea and headaches.[12]

Narcolepsy is a chronic neurological disorder. Knowledge about narcolepsy, however, often remains limited in a country like Zambia, where there are very few neurologists. We are presenting a case of 10years old girl who presented with symptoms of narcolepsy at pediatrics department of Livingstone Central Hospital.

CASE PRESENTATION

A 10 years old girl was brought to the pediatrics department of Livingstone Central Hospital by her mother with complaints of having episodes of excessive day time sleepiness for the past three years. The onset was insidious and episodes had increased in frequency in past 3 years. She used to sleep while in the class, during talking and sometimes during eating. She used to sleep for about 15-20 minutes during episodes and there were about 3-4 episodes each day. She used to feel fresh after those episodes. She had disturbed sleep at night. There was no history of having cataplexy, hallucinations or sleep paralysis. There was no history of alcohol or drugs use. Her illness started about 3 years ago while she was student

in a school at Nakonde, which is a border town between Zambia and Tanzania about 1500 Km away from Livingstone. She was taken to different clinics and hospitals since she started the illness but nobody was able to diagnose her. Sometimes clinicians thought of epilepsy and at times she was just given paracetamol tablets. Patient used to feel embarrass at school due to being stigmatized by fellow pupils at school because of her illness.

Patient came from a family of eight. Two of her siblings died at young age during attacks of status epilepticus. Patient was born as spontaneous vaginal delivery at a local clinic. Her mother was normal during pregnancy. Patient was growing normally and all her milestones were normal. Patient was in grade 3 at school and was among top five pupils in her class.

Patient was admitted for about 6 days at Livingstone Central Hospital where her basic laboratory investigations were carried out. Her FBC, DLC, ESR, LFTs, Urine routine & microscopy, HIV test, all came out normal. She was weighing 43 Kg and was 1.43m tall. Her CT scan of brain didn't show any abnormality. Her physical & neurological examinations were unremarkable.

At Livingstone Central Hospital she was managed by both pediatric and psychiatry departments. She was diagnosed as having narcolepsy and was commenced on tab fluoxetine 20mg/day and tab modafanil 100mg/day. She was discharged after staying in the hospital for 6 days. At her follow up after 3weeks, the patient had responded well to treatment and was free of those episodes of day time sleepiness. Currently the patient is doing fine and still on the same treatment.

DISCUSSION

Our patient was a 10 year old pre-teen aged girl who presented with episodes of day time somnolence lasting 15-20 minutes, since past three years. She didn't have cataplexy, hypnagogic or hypnopompic hallucinations or sleep paralysis. Even though patient had not developed cataplexy but there are chances that she could develop it in future as in the study reported by young et al, that cataplexy usually follows 4-5 years after the onset of day time somnolence.[4] Continuum, lifelong learning in neurology, on sleep disorders[10] reported that Cataplexy usually follows the onset of day time sleepiness. Cataplexy, hypnagogic hallucinations, and sleep paralysis, may develop slowly, suddenly or not at all.[10] In the studies by Young et al and Yoss & Daly, it was reported that narcolepsy usually starts in 2nd or 3rd decade of life[4, 5] but studies done by Hayes and Stores reported that rare cases of narcolepsy can start in the first decade of life.[6, 7] The onset of narcolepsy in our patient was around 7 years of age which is similar to what was reported in studies by Hayes and Stores.[6, 7] Kotagal et al reported that the definitive diagnosis of narcolepsy can be made with polysomnographic studies and multiple sleep latency test in sleep laboratories.[5] According to American academy of Sleep Medicine, the symptoms of excessive day time sleepiness must be present for at least 3 months to make a diagnosis of narcolepsy.[11] In Zambia in resource poor health settings, unavailability of sleep laboratories and scarcity of neurologists makes it impossible to confirm the diagnosis of narcolepsy by polysomnographic studies. We can only make a clinical diagnosis which was done in this case. Continuum, lifelong learning in neurology, on sleep disorders[10] reported that narcolepsy may be mistaken for depression, epilepsy or psychiatric illness, an accurate diagnosis is often delayed for many years after the onset of symptoms. It is estimated that fewer than 50% of the patients with narcolepsy have been diagnosed.[10] The patient we are presenting was in a similar situation as she went to many health centers where they were unable to make a correct diagnosis and treat her condition until she arrived at Livingstone Central Hospital. Even in Livingstone Central Hospital, initially the patient was suspected to have epilepsy but after getting detailed subjective and collateral history, the diagnosis of narcolepsy was made.After the diagnosis the patient responded well to the treatment with SSRI fluoxetine and modafanil.

In the study done by Anders et al, it was reported that narcolepsy in a child is a cause of serious psychological and social disadvantage.[3] The patient presented here, reported of being stigmatized by her fellow pupils at her school due to her being sleepy in the classroom. She used to feel sad and embarrassed due to being stigmatized by fellow pupils at school. The patient was noticed to be very relaxed and happy during her follow up visit due to her episodes of daytime sleepiness being stopped due to the treatment given to her. The patient is currently doing fine at her school and is regularly taking her prescribed treatment, fluoxetine and modafanil tablets.

CONCLUSION

Narcolepsy in children is a rare condition but once it presents, it poses diagnostic challenges in Zambia due to unavailability of sleep laboratories and lack of knowledge among health workers about this disorder. Zambia should have a sleep laboratory so that sleep disorders like narcolepsy can be diagnosed and managed effectively.

ABBREVIATIONS

EDS: Excessive Daytime Sleepiness,FBC: Full Blood Count, DLC: Differential Leukocyte Count, ESR: Erythrocytes Sedimentation Rate, LFTs: Liver Function Tests, TCAs: Tricyclic Antidepressants, SSRIs: Selective Serotonin Re-uptake Inhibitors, Tab: Tablet

DECLARATIONS

Ethical Approval

Not applicable in case report.

Consent

Informed consent to publish the case report was obtained from the patient.

Competing Interest

The authors declare that they have no competing interests.

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Authors Contribution

Both authors contributed in the management of patient and revision and approval of the final manuscript.

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