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Case Report

A Rare Case of Wilsons Disease with Zoophilia

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Abstract: Wilsons Disease also known as hepatolenticular degeneration is a autosomal recessive disorder, characterized by abnormal copper deposition in the liver, brain, and other tissues caused by mutation in the copper transporting gene ATP7B. Patients presents with neurological and hepatic disorder. Psychiatric manifestation are common along with movement disorder but its association with paraphilic disorders like zoophilia is very rare and there is practically no literature regarding this in North-Eatern India. In this article we report the unique case of a 21 year old boy from rural Assam suffering from Wilson's disease with multiple neuropsychiatric manifestations among which zoophilia was noteworthy. This case report highlights a rare and atypical association between an adolescent patient of Wilson disease & zoophilia--- a relatively rare paraphilic disorder. A 21 year old boy presented in the Psychiatry emergency department in a tertiary care hospital with aggressive, disorganized behavior with reported sexual act with a cow. Examination of the patient also revealed massive organomegaly and K-F ring which on further investigation confirmed the diagnosis of Wilsons Disease. The occurrence of zoophilic behavior in Wilsons disease is very rare and we have not come across any such report regarding this. To conclude, the association between copper deposition in specific brain areas due to Wilsons disease and zoophilic behaviour we need further studies with better investigative modalities, which will in turn help in drawing an individualise treatment plan.

Keywords: Wilsons Disease, Hepatolenticular Degeneration, Copper Deposition, Adolescent, Neuropsychiatric, Zoophilia

1. Introduction

Wilsons Disease or Hepatolenticular Degeneration is a rare inherited disorder affecting both the liver and central nervous system. It occurs due to a mutation in the ATP7B gene located on the long arm of chromosome 13. This gene encodes a membrane bound copper binding protein expressed primarily in the liver that is responsible for transporting copper into the secretary pathway for incorporation in apoceruloplasmin and excretion in Bile. A mutation in the gene leads to impaired elimination of copper leading to its accumulation in many organs and tissues giving rise to the various clinical features of the disease.

The illness usually manifest in childhood or adolescence, but may be delayed till 5th decade. The presentation may be typically with hepatic disorder, Neurological disorder or both. Patients also presents in rare cases with psychiatric disorders, gynaecological disorders, cholelithaisis, nephrolithiasisetc. In hepatic manifestation, patient may have hepatitis, cirrhosis or hepatic decompensation. Usually the diagnosis is confirmed by estimation of serum ceruloplasmin level which is decreased and 24 hour urinary copper level, which is increased. Commonly found radiological findings are hepatomegaly, splenomegaly on USG along with double panda sign on MRI Brainwhich is highly significant of Wilson's disease.

The Neurological manifestations (40-50%) [1] which are most commonly manifested as movement disorders namely—dystonia, incoordination and tremor are mainly due to involvement of basal ganglia & occasionally pons, medulla, thalamus, cerebellum and subcortical areas. Dysarthria and Dysphagia are also common. Half of patient with neurological disorders presents with Psychiatric disturbances. In fact, the psychiatric manifestation of WD are so vast that it was popularly termed as "a great masquerade". [2]. Various

epidemiological data suggest that neuropsychiatric symptoms in WD can range upto 30%. [1]. (EASL). Wilson himself in 1912, in his paper, described the disease in 12 people out of which 8 had psychiatric symptoms [3]. Later studies also revealed that along with hepatic and neurological symptoms, psychiatric symptoms can be the primary clinical presentation in WD [1]. The THfeatures are diverse and includes loss of emotional control (temper tantrums, crying bouts), depression, hyperactivity or loss of Sexual inhibition. In this study, we are reporting a case of WD, starting in early second decade of life with predominantly neuropsychiatric symptoms and an unique association with zoophilia.

2. Case History

A 21 year old unmarried muslim male born out of consanguineous marriage, belonging to lower socio economic class hailing from rural residency of NE part of India presented to the emergency department of TCC with chief complaint of aggressive & violent behavior, multiple acts of bizarre sexual behavior, slurring of speech, generalized weakness and gradual decline in academic performance since 6 years, increased for the last one month which was insidious in onset with a gradual progressive course. The patient was apparently well upto 6 years. He was a soft spoken, jovial young boy, academically sound and shared cordial relationship with all family members. But gradually his health started deteriorating. His apetite decreased and he started getting more and more weak with each passing day. He started getting irritable, aggressive and started putting up fights with everyone, every other day without reason or with trivial issues. His speech started getting muffled and most of his words became almost incomprehensible at the time of presentation which was 6 years since onset. He used to secure a good position in class till class 8. Then the illness started and his academic performance started declining, his handwriting started worsening and he used to get easily distracted whenever he tried to study. Ultimately he could not pass class 10, even in multiple attempts. Following that, his father removed him from school and tried to indulge him in minor jobs but he could not sustain any job more than a month and kept changing jobs. At around this time, he was discovered by a neighbour, trying to make a sexual advance towards a young girl in his neighbourhood. Subsequently he was severely rebuked for his actions by his parents and neighbours. On another occasion, several months after the latter, he was found by his own mother, in their stable, trying to engage in sexual act with a cow. Following that incident he was severely beaten and was brought to the emergency department of tertiary care centre, for the treatment of his physical injury as well as aberrant behavior. A detailed physical and mental state examination was done. On examination, his vitals were found within normal limits. He was found to have moderate pallor and massively enlarged spleen. On neurological examination, patient was found to be conscious, alert, well oriented with intact comprehensions. However the speech was dysarthric with interrupted flow but grammatically correct. Deep Tendon Reflexes and Superficial Reflexes were intact and no sensory deficits were noted. Power of all the four limbs were found to be clinically intact though there was generalised muscle mass atrophy. The gait was ataxic but no other cerebellar signs were noted.

Opthalmological investigation was done and it demonstrated the development of K-F ring in the clear anterior segment of the cornea under slit lamp examination. (Figure 1)

Figure 1. *Showing K-F ring under slit lamp examination.*

On Mental state examination, he was found to be of ectomorphic build, cooperative with normal psychomotor activity. Rapport was established, mood was low, affect was appropriate, stable, reactive and of normal range. Though the speech was relevant with normal reaction time it was difficult to understand as the speech was not clear and was muffled at times. It was dysarthric as mentioned earlier. Volume and tone were within normal limits. No formal thought disorder or any perceptual disturbances were noticed. On evaluations of cognitive functions, his comprehension was intact, but his attention and concentration was found to be poor with subaverage intelligence (I.Q-70). Though his personal and test judgement were intact but social judgement was found to be compromised. His abstract thinking was also poor and insight was of grade III. With all these findings the patient was provisionally diagnosed as a case of Wilson disease [E83.01], with associated organic personality disorder [F07.0 according ICD 10 guidelines], and was investigated for the confirmation of the diagnosis.

3. Laboratory Investigation

His biochemical investigations showed Hb to be 8.5gm, TLC to be 1310 cells/ml, platelet count to be 110000/ml. Serum ceruloplasmin was markedly reduced (<0.10mg/dl) (refvalue-16-47mg/dl), 24hrs urinary copper concentration was high at 196.16microgm/24hrs (ref value less than 38microgm/24hrs). However, his LFT and KFT were within normal limits. HisUSG whole abdomen further supported the diagnosis showing massive splenomegaly and changes suggestive of chronic parenchymal liver disease. His MRI

brain revealed hyperintensities in B/L putamen, substantianigra, ventrolateral thalami, tegmental tectal plate of midbrain and pontine tegmentum revealing the face of "double panda" which is classical for WD. Considering his aberrant sexual behavior, MRI spectroscopy was done which revealed neuronal loss in B/L temporal lobe. (Image 3 and 4)

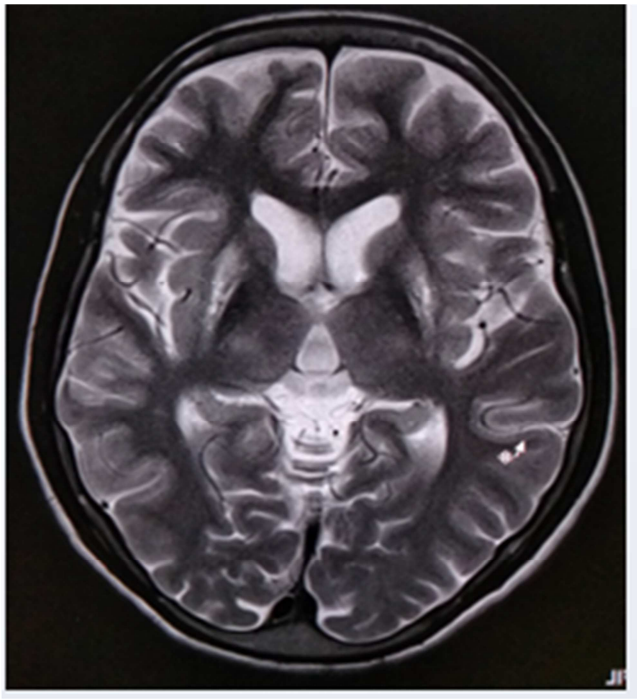

Figure 2. *Axial T2W1 image showing hyperintense signal in bilateral ventrolateral thalami.*

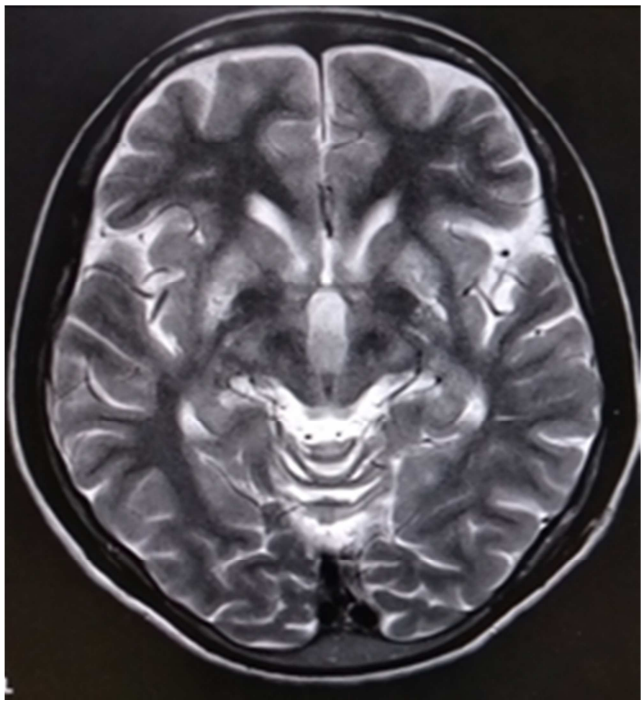

Figure 3. *Axial T2W1 shows 'panda sign' in midbrain at the level of quadrigeminal plate.*

With all these findings the diagnosis was confirmed. The patient was started on d-penicillamine (250mg) and zinc acetate (50mg) on a thrice daily basis and for his behavorial abnormality olanzapine was added. Other symptomatic treatment was also started as required. Patient was stable by 7 days and medicine consultation was done for his other associated physical problems. He was further advised to come for review in psychiatry department after 4 weeks.

On first review after 4 weeks, A significant improvement in his physical health and behavioral abnormalities was noticed and other biochemical parameters also started showing signs of improvement. No more aberrant sexual behavior was reported by family members during this period.

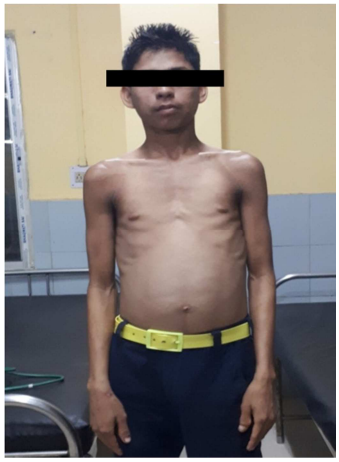

Figure 4. *The patient, frontal view.*

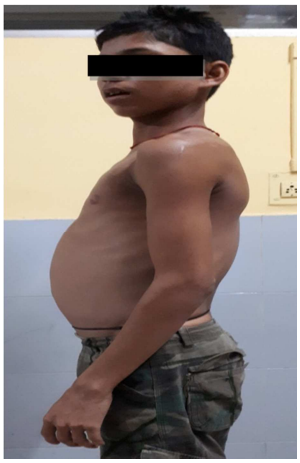

Figure 5. *The patient, lateral view.*

4. Discussion

In Wilson's disease, in addition to neurological and various physical symptoms, we know that a number of psychiatric manifestations are also common. Various studies, like by Svetel and colleagues, 72% of WD had psychiatric symptoms [4] Akil and Brewer 65% [5], Kumar and colleagues 60% [6], Dennig and bettios [7], Portala colleagues 46% [8]. Scheinberg and Sterleib concluded that almost every patient of WD have one or more underlying psychopathology [9]. This psychiatric symptoms, common in elderly patient of

WD, can have varied presentation starting most commonly from mood disorder [10], mainly depression 20%-60% [11] followed by psychosis [12-13], behavioural and personality problem [14], mania and hypomania [15], anxiety disorder [11], inappropriate behavior and impulsivity, gradual decline in academic performance [16], cognitive dysfunction [17], sleep problems [18] and sexual problems including hypersexuality. [19].

Zoophiliais a disorder of sexual preference classified under paraphillias in DSM 5, where an individual develops recurrent, intense sexual fantasy, urges or behavior towards an animal. Human sexual behavior is influenced by external events like stress and drugs and internal events controlled by hypothalamic, limbic system and cortical stimuli. Orbitofrontal cortex is involved in emotions related to sexual behavior. The left anterior cingulate cortex is involved in hormonal control and sexual arousal, and the right caudate nucleus regulates sexual activity followed by arousal. Therefore any dysfunction in the these regions causes various sexual disorders. In this index case, the zoophilic behavior of the patient may be related to the severe emotional disturbances and uncontrolled impulsivity. This may arise because of temporal lobe dysfunctions due to heavy depositions of copper. Though, MRI findings were not conclusive but neuronal loss in bitemporal regions revealed in MRI Spectroscopy, may be a pointer towards this conclusion. However, as we have not come across such case report, final conclusion cannot be drawn but it requires close observations and detailed evaluation of sexual behavior in such cases.

5. Conclusion

The severity of Wilson's disease (WD) is linked to free copper accumulating in the liver and brain. It is hypothesized that it may also enable a good evaluation of extra - hepatic involvement and its severity [22]. Studies shows that if WD is properly treated, in most patients the liver can be stabilized, even severe neurological disability reversed, and patients can resume normal lives [23]. To conclude, the association between copper deposition in specific brain areas due to Wilsons disease and zoophilic behaviour we need further studies with better investigative modalities, which will in turn help in drawing an individualized treatment plan. The addition of antipsychotics or mood stabilizers may curb down the erratic sexual behavior temporarily but with that, the therapy of underlying WD, targeting the copper deposited areas, should be continued for a long time for the overall improvement of his mental and physical condition.

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