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PSYCHIATRIC PRESENTATION OF AKINETIC RIGID SYNDROME

Neurology

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ABSTRACT

Akinetic Rigid Syndrome has been reported to be caused by a diversity of etiologies which include inter alia idiopathic Parkinsonism, anoxia, trauma, Wilson's disease, Huntington's disease, and may involve the basal ganglia. The entity is known to be characterized by akinesia and rigidity-lead pipe or cogwheel type, accompanied by slowness of movement (bradykinesia) and thought (bradyphrenia), diminishing amplitude of repetitive alternating movements with or without tremor at rest and postural instability. However, an evocative history, symptomatology and neurological findings signifying that of catatonia, along with lack of significant findings on radiological assessment and lack of suspicion may land up the patient in a psychiatric setting, and further to be misdiagnosed and hence mistreated. This is a case report of a 16 years old male presented with decreased speech output, difficulty in initiation and slowness of movement, inability to maintain posture and sleep disturbance of acute onset with past history of suicidal attempt by hanging and other significant findings suggestive of a diagnosis of Akinetic Rigid Syndrome. Diagnostic implications and management of this unique presentation are discussed in the case report.

KEYWORDS

Basal ganglia, akinesia, anoxia, catatonia.

INTRODUCTION:

A rare, but often debilitating, consequence of hypoxic-ischemic injury is the development of movement disorders.[1] Similar to the development of movement disorders in cardiac-arrest survivors, due to metabolic disturbances resulting from hypoxic-ischemic damage to the liver or kidney, from medications administered to treat the complications of cardiac arrest, or from cardioembolic ischemic stroke; many movement disorders are described after hypoxic-ischemic brain injury, including parkinsonism, dystonia, chorea, tics, athetosis, tremor, and myoclonus.[2]

Dystonic and akinetic-rigid syndromes, alone or in combination, represent a sizeable proportion of the post-hypoxic movement disorders described in the literature.[3] These syndromes may occur acutely, either at the time the hypoxic insult occurs or shortly thereafter, or more commonly in delayed fashion, months to years after the initial hypoxic insult.[4]

It is usually a symmetric condition characterized by various combinations of bradykinesia, micrographia, axial and appendicular rigidity, resting or postural tremor, and marked postural instability.[5]

The literature shows development of akinetic-rigid syndrome, typically within 3 months of the hypoxic event; after a rapid evolution, the majority of patients remained clinically stable for many subsequent years. In contrast, reports of development of the pure dystonic syndrome, on average, 10 months after the hypoxic event, and progressing gradually over several years, are also to be seen.[6] The majority of such patients had visible lesions in the basal ganglia on brain CT or MR imaging. Treatment of akinetic-rigid symptoms with levodopa or dopamine agonists and administration of high-dose anticholinergic drugs for dystonic symptoms conferred little benefit to these patients.[5]

Case History:

A16 years old boy, studied till class 9, unemployed, from rural background, with insignificant birth and developmental history, was brought to PGIMS Rohtak, Haryana, India by his mother with a history of decreased interaction with family members for six months and decreased activity, poor sleep and poor appetite for past three months. Patient had alleged history of suicidal attempt about seven months ago by hanging, which had led to him being admitted at Department of Surgeryfor about one week. On detailed assessment the act was found to be impulsive in nature after a dispute with his sister, with no mood symptoms being appreciated.

However, upon returning home after discharge, the patient was noticed to be forgetful about the suicidal attempt he had made, and was informed by his family members about the same. As per patient's mother, he was behaving normally at that time. He used to take care of himself and would interact with friends, family and acquaintances like his normal self. Although his sleep was disturbed with difficulty in initiation.

After about one month of suicidal attempt, his mother started noticing changes in his behavior. He was not initiating talks with his mother like he usually did, had to be engaged in a conversation and upon being enquired by his mother would deny any issue.

Since then his condition continued to deteriorate, and within next two months, his interaction so poor as to reply to his family members only in a word or two that too upon being asked multiple times, his sleep was disturbed with problems in both initiation as well as maintenance of sleep and would sleep for about 2-3 hours in a day only. He would remain confined to his bed for most of the day and night, stopped going to meet his friends, his appetite decreased substantially and self-care dropped.

As such the patient was brought to the Department of Psychiatry, PGIMS Rohtak and was advised admission for detailed assessment and management but was refused by the family members. He was given Inj. Lorazepam in the emergency and T. Lorazepam was prescribed with an advice of short follow up to family members.

While the compliance was good, only his sleep pattern ameliorated with slight improvement being noticed in other symptoms. With inability of the family members to follow up due to financial restraints, treatment was left over within 15 days of its inception.

For last one month before admission, in addition to his exiting symptoms, patient had developed frequent falls while making any attempt to walk. Patient had frequent blackouts while walking, that make him loose his posture and lead to him sustain injuries on to his head, face, chest and palms.

There was no history of seizure, head trauma, high grade fever, suspiciousness, hearing of voices/ seeing things in clear consciousness or substance abuse. No history of sadness of mood, ideas of hopelessness, worthlessness, helplessness was elicited.

On general physical examination, his body temperature, pulse rate, and blood pressure were 99.2°F, regular at 92 beats/min, and 100/70 mm Hg, respectively. On CNS Examination, patient was oriented, his attention being arousable but not sustained, speech- only in response to question with decreased rate, tone, volume and output, cranial nerve examination was normal, on motor system examination- increased tone and cogwheel rigidity in bilateral upper and lower limb along with axial rigidity was appreciated, no sensory impairment was appreciated, among signs of cerebellar dysfunction, finger nose test was found to be positive along with the presence of Romberg's sign and intentional tremors, gait was ataxic, while no signs of meningeal irritation could be elicited.

On mental status examination revealed poor eye contact, decreased psychomotor activity with non-spontaneous speech with decreased rate, tone, volume and only in response to question. His affect was inappropriate. No thought or perceptual disorder could be elicited.

PGI Battery of Brain Dysfunction was administered and was suggestive of organicity.

Laboratory analyses showed that blood cell count (Hb- 14.2, TLC-6500/mm^3, DLC-60/30/8/2/0, Platelets- 1.24L/mm^3), biochemical profile (B. Urea-26 mg/dL, S. Creatinine- 1.0 mg/dL, S. Uric Acid- 5.6 mg/dL, S. Calcium- 11.0 mg/dL, S. Phosphorus- 3.5 mg/dL, AST- 26 U, ALT- 32 U, ALP- 96 U, S. Protein- 8.3 mg/dL, S. Albumin- 5.0 mg/dL, Total S. Bilirubin- 0.3 mg/dL, S. Triglyceride- 125 mg/dL, S. Cholesterol- 140 mg/Dl, S. HDL- 29 mg/dL, S. LDL- 86 mg/dL, S. VLDL- 25 mg/dL, S. Sodium- 141 mEq/L, S. Potassium- 3.9 mEq/L, S. Amylase- 2 U), viral markers for HIV, HBV, HCV were non-reactive and total CPK levels (60 U) was within the normal range.

An MRI of brain and spine was obtained and found to be within normal limits, while the EEG was suggestive of post-anoxic burst suppression. Consultation with the neurology department was obtained and CSF for anti-measles antibodies and serum ceruloplasmin along with 24 hour urinary copper level were done to rule out SSPE and Wilson's disease respectively (This was done to rule out impulsive behavior in Wilson's disease and cognitive affect in SSPE). Both were ruled out after a negative report.

Based on history, clinical and neuro-psychological assessment, diagnosis of "Akinetic Rigid Syndrome due to Post-Anoxic Etiology" was kept. Patient was started on Tab Sodium Valproate 1000 mg and T. Lorazepam 2 mg in divided doses. He showed good response with medications in symptoms of frequent falls and sleep pattern, while modest improvement was noticed in his interaction with family members and rigidity.

DISCUSSION:

The literature on Akinetic Rigid Syndrome points towards varied etiologies of idiopathic, vascular, hereditary, autoimmune nature as well as those precipitated by drugs (haloperidol, flupenthixol, chlorpromazine, sulpride, etc.), toxins (heavy metals, MPTP, cyanide, carbon monoxide), metabolic and autoimmune (Wilson's, Hashimoto's encephalopathy, anti-phospholipid syndrome, etc.), anoxia, trauma; all of which have neuro-degeneration in common.[5]

Unavailability of proper history may make matter difficult. Clues to etiology like anoxia and trauma may alone be there. Several studies have reported differential diagnosis for such a presentation and suggest the high probability of missing this diagnosis in the absence of significant suspicion, in case history does not offer anything substantial.

The confirmatory diagnosis is by clinical examination alone as in the case of Parkinson's disease in which all tests are normal. Sodium Valproate in this case has served to control the abnormal impulses in brain activity and has helped in symptomatic improvement in the patient in a modest way along with saving the patient from sustaining repeated physical trauma due to frequent falls. However, the overall prognosis remains to be poor.

REFERENCES:

1. Allen KA, Brandon DH. Hypoxic ischemic encephalopathy: pathophysiology and experimental treatments. Newborn and Infant Nursing Reviews. 2011;11(3):125-33.
2. Venkatesan A, Frucht S. Movement disorders after resuscitation from cardiac arrest. Neurologic clinics. 2006;24(1):123-32.
3. Bhatt MH, Obeso JA, Marsden CD. Time course of postanoxic akinetic-rigidand dystonic syndromes. Neurology 1993;43: 314-7.
4. Zhang YX, Liu JR, Jiang B, Liu HQ, Ding MP, Song SJ, Zhang BR, Zhang H, Xu B, Chen HH, Wang ZJ. Lance-Adams syndrome: a report of two cases. Journal of Zhejiang University SCIENCE B. 2007;8(10):715-20.
5. Gupta P, Behari M. Akinetic rigid syndrome: an overview. Ann Indian Acad Neurol. 2007;10:21–30.
6. Kuoppamäki M, Bhatia KP, Quinn N. Progressive delayed-onset dystonia after cerebral anoxic insult in adults. Movement disorders: official journal of the Movement Disorder Society. 2002;17(6):1345-9.