Case Report

Hirayama disease either leads to atypical psychiatric presentation or atypical psychiatric presentation can occur in Hirayama disease or atypical psychiatric presentation just comorbid with Hirayama disease

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Abstract:
Various psychiatric symptoms have been reported in neurological disorders but no case is reported regarding psychiatric symptoms co-existing with HIRAYAMA DISEASE. We report a patient with HIRAYAMA DISEASE who had psychiatric symptoms but not mounting to specific psychiatric disorder and had atypical presentation.

Key words:
Hirayama disease, psychiatric symptoms, atypical presentation

Introduction:
Hirayama disease is also known as juvenile muscular atrophy of the distal part of upper limb. It is a rare slowly progressive neurological disorder usually affecting young men in 2nd and 3rd decade of life 1, 2. The muscular atrophy establishes between 2 and 5 Years of onset of illness. After this period of time, the muscle atrophy does not improve and does worsen either. Patients usually do not complain any pain or sensory changes 1, 2.

The patient usually present with wasting and weakness of the distal part of upper limb mainly hand or forearm and rarely may progress to opposite side. Person experiences certain level of disability due to the disease. It is believed that it is a neck flexion induced cervical myelopathy that leads to progressive degeneration and loss of motor neurons 2, 7.

Diagnosis of the disease is made mainly by history and physical examination. Use of Electromyography may show loss in nerve supply of the affected limb. MRI or CT scan can be done to show muscle atrophy.

Yet there is no cure of this disease. Treatment approaches includes muscle strengthening exercise and training related to hand coordination. Cervical collar may be useful. 8 . Sometimes surgical intervention needed 9.

Till now no case report is available describing psychiatric disorder co-morbid with Hirayama disease or Hirayama disease leads to such type of psychiatric manifestation. Clinical symptoms pattern of this patient does not meet either bipolar disorder or non-organic psychotic disorder. So we are reporting this case.

The case
A 26 years old male, intermediate, unemployed, was referred by general practitioner with the chief complaint of gradual weakness of right upper limb for the last 8 years and multiple brief episodes of abnormal behavior in the form of disorganization, irritability, decreased self care, absconding tendency and decreased sleep for the last seven and half years.

The patient was apparently asymptomatic about 8 years ago when he reported weakness of the distal part
of right upper limb to the family members. The patient was then taken to a physician. As per his advice the MRI of brain and cervical spine was done. The MRI finding was within normal limit. The physician referred the patient to a tertiary care centre, where he was hospitalized and diagnosed as suffering from a neurological disorder known as 'Hirayama Disease'. They advised physiotherapy and to wear cervical collar to the patient.

Two month after discharge from that hospital he developed abnormalities in behavior. He would urinate in the cloth in front of the family members and guest. When the patient was asked about this he did not give any rationale and he started roaming here there and some time visit to the relative home with informing family member on asking he would not give any reason behind this behavior the patient also used to steal the money and jewelry, mobile and would sell them in the market and purchase items containing psychoactive substance like nicotine. He would become very irritable during these days. He would become aggressive and use to abuse and beat family members on trivial issues. His psychomotor activity was also increased. He would roam in house and village throughout day and night without any specific reason. The sleep of the patients was also decreased. He would sleep for only 3-4 hours in 24 hours. His self-care was also decreased. He would not take bath used to wear dirty clothes. He was forced by the family members. His appetite was normal during this period; he would take all three meals as before.

The symptoms were episodic and last for 4-5 days and would occur in 15 days to 2 months period randomly. In between the episodes the symptoms were subside but decreased sleep (about 3-4 hours) persisted.

The neurological problem in the form of weakness in right fore arm was progressive and gradually involved the left fore arm and he was unable to hold objects properly by hands. He did not report any pain in upper limb or anywhere in the body. The above mentioned course of illness continued till 1st week of June 2009.

The patient was not taking medication regularly although he was shown to many psychiatrists. Mean time he was also shown in the semi-government tertiary care centre for 2-3 times. The patient did not use cervical collar and did not cooperate for physiotherapy. The details of treatment were not available. For the last two month before reporting psychiatric department of tertiary care center, he again developed similar symptoms as describe in episode but this time the symptoms were last for longer time and the patient was unmanageable at home. He was hospitalized in the department of psychiatry.

There is no evidence of elevated mood, cheerfulness, grandiose ideas, increased religiosity, increased sexuality, increased goal directed activities, peripatetic sadness of mood, hopelessness, worthlessness, death wishes, suicidal ideas, posturing, mutism, holding of saliva, muttering to self, suspiciousness and fearfulness. No history of head injury, unconsciousness, seizure, high grade fever or any other medical and surgical illness.

Past history and family history are not contributory. He belongs to low socio-economic status. His family is joint family.

The developmental history of the patient was normal. He is educated up to 12th class and currently, not employed. The patient used to consume tobacco in the form of Gutkha about 3-4 pouch per day and 5-6 Bidli per day for the last 10 years.

Pre-morbidly he was social; enjoyed the company of his friends, a cool and calm person. He generally avoided to take responsibility but able to maintain good relationship with family member.
Treatment:

The patient was hospitalized in psychiatry department for management and he was put on Tab Olanzapine 10mg 1 Tab BD and Tab lorazepam 2mg 1 Tab BD he was not showing appropriate response so the dose was increased upto Tab olanzapine 10 mg TDS and tab lorazepam 2 mg TDS. He showed response on these medications. The patient needed relatively higher dose of medication than usual. For his neurological problem a opinion was sought from department of neurology, they advised cervical collar and physiotherapy but he did not cooperate for physiotherapy.

Discussion:

Various psychiatric symptoms have been reported in neurological disorders for example manic symptoms, depressive symptoms, psychotic symptoms, anxiety symptoms, obsessive symptoms and personality change. Most of these neurological disorders have simultaneous evidence of morphological changes in the brain but Hirayama disease per se usually does not have any neurological changes in the brain. In this patient also there is no neurological changes has been found in MRI examination and the presentation of psychiatric symptoms were not classical to the any of the major psychiatric disorder either classical psychotic disorder or affective disorder, the patient had psychiatric symptoms in the form of multiple brief episodes of behavior change like irritability, aimless wondering, decreased self care, decreased sleep for the last 7 years. In between the episodes he had persistently decreased sleep and no other symptoms. Since psychiatric symptoms started after the onset of Hirayama disease it is not sure whether this disorder lead to such type of psychiatric manifestation or such atypical presentation of psychiatric disorder can occur in patient with Hirayama disease or these psychiatric manifestation are just co-morbid with HIRAYAMA DISEASE. This is open for us.

References:


