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# A Case Report of Cerebral Atrophy at Hyperostosis Frontalis Interna Associated with Hypoglycemia and Hyperthermia in Huntingston's Disease in a 57 Years Aged Women

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**Abstract:** ***An unusual case of Huntingston's disease (HD) was reported due to cerebral atrophy at Hyperostosis Frontal Interna region of brain in a 57 years female patient. Patient came to hospital with complaints of mild slurred speech, tremors in right hand, difficulty in walking, generalized myalgia, on and off fever with chills, abnormal movements of lower jaw and urinary incontinence. Her family history was known with HD in grandfather. Her past medical history was known to be with Post menopausal state. Her physical examination reported with high temperature-101$^0$ F. Her abnormal blood reports were mild depletions of Hb-10.3 g/dl (), PCV-31 g%, & RBC Count-3.6 ml/m$^3$ and very low RBS levels- 105mg/dl. Brain CT scan report reveals diffuse cerebral atrophy at hyperostosis frontalis interna. Patient scored low in her emotion recognition tests and cognitive ability tests and finally diagnosed as HD associated with hypogycemia and hyperthermia.***

**Keywords:** Huntingston disease, cerebral atrophy, hyperthermia, hyperostosis frontalis interna.

## 1. Introduction

HD is a progressive and an inherited neurodegenerative brain disorder [1]. It is caused by abnormal expansion of a trinucleotide (CAG) which repeats in the huntingtons gene on chromosome 4 [2, 3] results in misfolded protein (mutant protein) [4, 5] and makes the death of nerve cells. This defect is dominant who inherits it from parent gene of HD. HD damages some specific areas of brain which results in difficult in movements as well as behavioral and cognitive changes. HD occurs in the middle age between 30-50 years, but in rare cases it can also occur in juvenile life [6].

HD motor symptoms include chorea of the arms, legs, head, face and upper body and also include jerks twitches, muscle spasms, fidgeting, speech difficulties, dystonia [7]. Its cognitive symptoms are communication, executive functions, memory, awareness, organization, regulation [8]. Its behavioural symptoms are apathy, depression, aggression, disinhibition, repetition, anxiety, hallucinations [9].

It is important to identify the triggering areas of brain which shows its effect on behavioural aspects in order to minimize the difficult behaviours by providing a safe environment [10]. Some studies reveals there is a characteristic decrease in glucose utilization in the caudate nuclei and putamen and this makes hypometabolism, this appears as early and bulk tissue loss which lead to cerebral atrophy in HD [11, 12]. Some other studies reports there is a robust decrease in grey matter density i.e atrophy were founded bilaterally in striatal areas as well as in the hypothalamus and in the opercular cortex and unilaterally in the right paracentral lobule which leads to neuronal loss in HD [13, 14]. Some says striking losses in cerebral cortex, white matter, caudate, putamen and thalamus at percentage levels demonstrating a severe atrophy of the neostriatum [15]. Elevated body temperature is common in HD but its relation is unclear.

Diagnosis can be done by physical examination of symptoms, emotion recognition test, and cognition test. CT scan, MRI scan reveals the affected area of brain. Therapy can be done with Tetrabenazine, Trihexypenidyl medications, additionally symptomatic drug therapy can be suggested.

## 2. Case Presentation

An unusual case of HD with acute confusional state and fever was reported in a 57 years female patient due to cerebral atrophy at Hyperostosis Frontal Interna region of Brain associated with hypoglycemia and hyperthermia. Patient was admitted in tertiary care hospital in Hyderabad with a chief complaints of mild slurred speech, tremors in right hand, difficulty in walking, generalized myalgia, on and off fever with chills, abnormal movements of lower jaw. She also suffers from urinary incontinence.

Her family history was known with HD in grandfather. Her past medical history was known to be with Post menopausal state and no known history of diabetes, hypertension, COPD and coronary artery disease. She has no social history of smoking, alcohol drinking and tobacco chewing. Her physical examination was reported with high temperature 101$^0$ F and CNS was slurred speech but conscious. Her abnormal blood reports were mild depletions of Hb-10.3 g/dl, PCV-31 g%, & RBC Count-3.6 ml/m$^3$ and very low RBS levels- 105mg/dl. Brain CT Scan report reveals diffuse cerebral atrophy at hyperostosis frontalis interna. Patient scored low in her emotion recognition tests and cognitive

ability tests. Finally her diagnosis was understood and confirmed with HD with acute confusional state and fever.

She was treated with IM Haloperidol 2.5 mg BD, Tab Trihexyphenidyl 1mg OD, IV Cefoperazone 1.5mg and got a symptomatic treatment with acetaminophen.

## 3. Discussion

A female patient with 57 years reported with HD due to cerebral atrophy at hyperostosis frontalis interna with hypoglycemia and hyperthermia. The patient's family is known with HD, so it can be causative factor to inherit from parent gene.

Patient came to hospital with complaints of mild slurred speech, tremors in right hand, difficulty in walking, generalized myalgia, on and off fever with chills, abnormal movements of lower jaw, and urinary incontinence. By considering the patients symptoms, some studies reveal striking losses in cerebral cortex, white matter, caudate, putamen and thalamus at a low percentage levels demonstrating a severe atrophy of the neostriatum. Some other studies say decrease in density of grey matter in stiatal areas, hypothalamus, opercular cortex and paracentral lobule leads to neuronal loss in HD. These abnormal conditions make motor, cognitive and behavioral symptoms.

The patient is under postmenopausal condition, but its relation with HD is not known. Patient also suffers from on and off fever, where some studies discuss it as common in HD but its mechanism is unclear.

Patient's RBS was very low; previous studies discuss hypoglycemia leads to decrease in glucose utilization to brain which makes hypometabolism. This appears bulk tissue loss and results cerebral atrophy in HD. Elevated body temperature is common but its relation with HD is unclear.

Because of 3 reasons, it can be considered as unsual case report

1) Cerebral atrophy in HD was detected at region of hyperostosis frontal interna region of brain, as no of the study was previously detected.
2) HD was associated with both hypoglycemia and hyperthermia conditions.
3) Mostly HD occurs in middle age of 30-50 years, but this case was diagnosed with early or start of HD at 57 years in female with a postmenopausal condition.

Patient was under the medications of IM Haloperidol 2.5 mg BD, Tab Trihexyphenidyl 1mg OD, IV Cefoperazone 1.5mg and got a symptomatic treatment with acetaminophen and responded well from fever, urinary incontinence, and myalgia.

## 4. Conclusion

It is most important to identify the triggering areas of brain which shows its effect on behavioural aspects in order to minimize the difficult behaviours. Although HD is a progressive disease, early diagnosis and early treatment is essential to prevent more complications. More studies are needed to know HD association with hyperthermia.

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