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Case Report

Radiology Section

Hyperglycemic Nonketotic Hemichorea-Hemiballismus- A Case Report with Review of Literature

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ABSTRACT

Hemichorea-hemiballismus can rarely present in non-ketotic hyperglycemia. Elderly aged women after attaining menopause can have this as a first presentation. We reported a case of 70-year-old women presented with loss of consciousness, hemiballismus and hemichorea associated with non-ketotic hyperglycemia. CT imaging of brain reveals no abnormality. MRI was done when patient was having the hemichorea and hemiballismus which it showed hyperintense signal on T1WI, hypointense signal on T2WI involving the putamen region of lentiform nucleus on left side. Her symptoms were not seen after correction of serum glucose value to normal level. In our case we can notice the significance of detecting rare findings which shows in diabetic patients and to differentiate by using MRI from other common pathologies causing neurological symptoms.

Keywords: Diabetes, Hyperintensity on T1WI, Unilateral basal ganglia

CASE REPORT

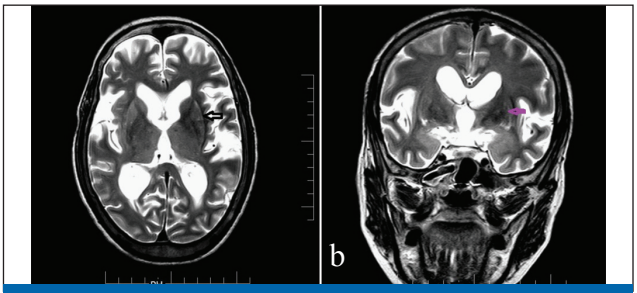
A 70-year-old female presented to our casualty in unconscious state and had a history of 18-20 episodes of vomiting previous day following history of jerking movements of right upper and lower limb movements with deviation of face.

There was no history of preceding trauma. Patient was known case of diabetes mellitus and hypertension and was not on regular treatment for the past 6 months.

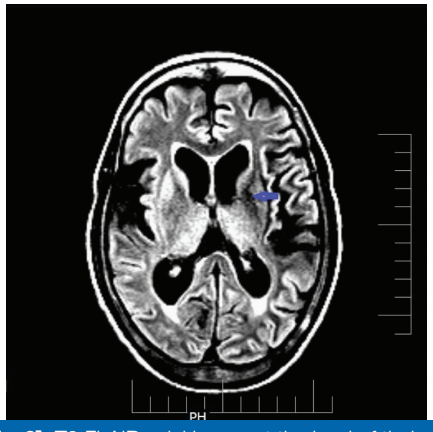
Her respiratory, cardiovascular and abdominal examinations were normal. Neurological examination was notable with Glasgow Coma Scale 10/15 and anisocoria not reacting pupil to light. Patient was responding to painful stimuli but was unable to move right side extremities. Bilateral plantar reflexes were increased.

Initial serum blood glucose values were grossly elevated which is 456mg/dl. Ketone bodies were not seen in urine. A Vascular lesion cause was suspected as it is a most common cause in this presentation scenario.

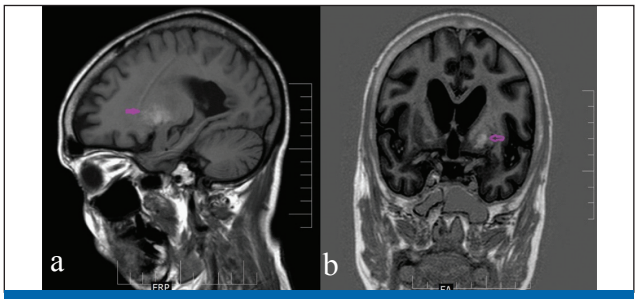
Imaging findings: Computerized tomography (CT) of brain revealed no significant abnormality. Magnetic resonance imaging (MRI) of brain was done. Left lentiform nucleus appeared hypointense on T2-weighted spin echo sequence and FLAIR (Fluid attenuated inversion recovery) sequence images [Table/Fig-1,2]. The same areas showed hyperintensity on T1WI [Table/Fig-3a,b]. No evidence of any cerebral vascular abnormality was detected. On susceptibility weighted (SWI)



[Table/Fig-1]: (a) Axial T2WI at the level of thalamus shows hypointensities at the Left lentiform nucleus
(b) Coronal T2WI at the level of third ventricle shows hypointensities at the Left lentiform nucleus.



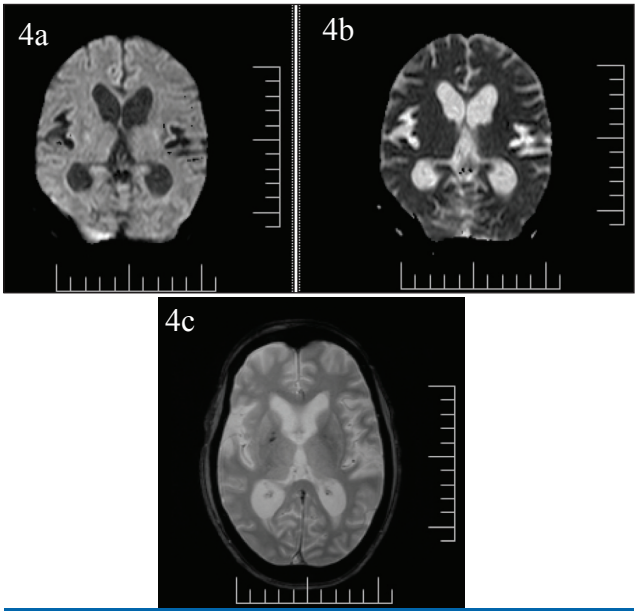
[Table/Fig-2]: T2 FLAIR axial image at the level of thalamus shows hypointense signal at left lentiform nucleus involving putamen area.



[Table/Fig-3]: (a) Saggital T1WI shows Left lentiform nucleus hyperintense signal.
(b) coronal T1WI inversion recovery image at the level of third ventricle shows hyperintense signal in the left lentiform nucleus.

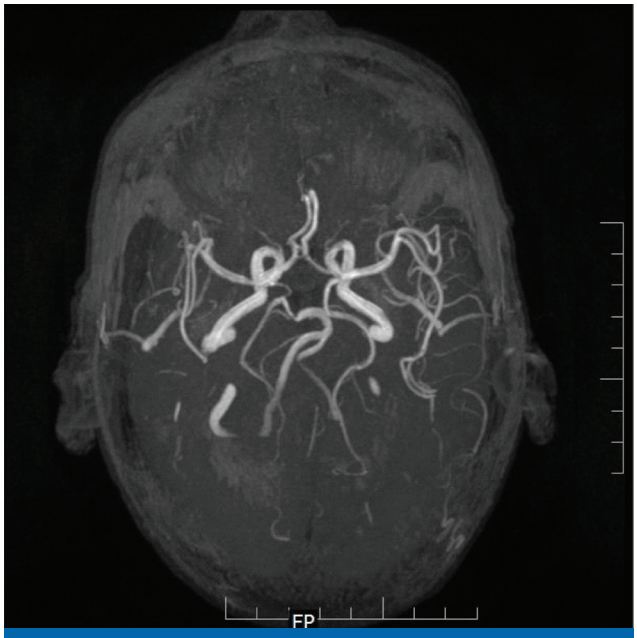
and diffuse weighted (DWI) images no abnormal signal intensity is seen [Table/Fig-4a-c]. No evidence of any cerebral vascular abnormality was detected [Table/Fig-5].

Diagnosis with treatment and follow-up: The imaging findings are not consistent with any hemorrhage, calcifications or any sort of infarct by the appearance of hypointense signal in lentiform nucleus at putamen area on T2-weighted and FLAIR sequence images which showing hyper intensity on T1WI.Other MRI sequences SWI and DWI with corresponding ADC (apparent diffusion coefficient) mapping images show no abnormal signal intensity and no vascular abnormality was found in MR angiography. Other causes of hemi chorea were ruled out like not taking any neurological medicines and thyroid disorders. Hemiballismus was her first episode of presentation and no family history was noted.



[Table/Fig-4]: (a) and (b) Axial DWI and corresponding ADC mapping images at the level of thalamus shows no abnormal signal intensity. (c) Axial SWI image at the level of thalamus shows no blooming at left basal ganglia.

Her symptoms diminished after correction of serum glucose value to normal level by hyperglycemic control medication like Actrapid which contain human insulin. The patient was followed for a period of two months after discharge and no significant hemichorea and hemiballismus symptoms were seen. Hence the diagnosis was made out to be as rare manifestation hyperglycemic non-ketotic hemichorea-hemiballismus.



[Table/Fig-5]: MR Cerebral Angiography- TOF image-showing normal intracerebral vessels. No vascular abnormality detected.

DISCUSSION

Syndrome in Patients with uncontrolled diabetes having nonketotic hyperglycemia and presenting with acute symptoms of hemiballismus-hemichorea i.e. involuntary movements on one side of total limb or of both limbs and a hyper kinetic movement disorder which is rare [1,2].

Involuntary movements are caused by various reasons like hereditary, any medications, infections, perinatal hypoxia, immunological disorders and several metabolic causes of thyroid or parathyroid disorders and disorders like diabetes mellitus causing hyper glycemia in our case [3].

Hemichorea-hemiballismus can rarely present in non-ketotic hyperglycemia. Elderly aged women after attaining menopause can have this as a first presentation [4]. Recent studies of case series in asian population of hemichorea hemiballismus with hyperglycemia of non ketotic type shows typical MR findings [5].

In diagnostic workup of Hemichorea-hemiballismus cases normal Computed tomography (CT) findings can be seen which is evident in this case. Sometimes CT-scan show the subtle basal ganglion high-density in unilateral and contralateral side

of symptoms seen. In MRI it is seen well with characteristic unilateral basal ganglia hyper intensity signal. Three "H" of hemichorea and hemiballismus in non-ketotic hyperglycemia are hyperintense signal in T1, Hyperglycemia with or without coma and hemichorea-hemiballismus [2].

There were reported cases early on diabetic non-ketotic patients with hemichorea hemiballismus who are hyperglycemic showing basal ganglia hyperdensity and T1WI hyper intensity on MR imaging on contralateral side [2].

Study of Chang and his colleagues described petechial hemorrhage as a likely cause two of his three patients were post menopausal and hypersensitivity of dopamine receptor causing movement disorder was likely pathogenesis postulated in his study [6].

Histopathology and Biopsy from striatum demonstrated brain tissue which is gliotic and abundant gemistocytes where seen suggesting the MR imaging T1 hyperintensities could be because of protein hydrated layer seen inside swollen gemistocytes cytoplasam. Gemistocytes which are present abundantly in basal ganglia causing neuronal activity excessively and this could be the likely possibility of generating hemichorea-hemiballlsimus [7]. The hyperintensities in basal ganglia will resolves by few months and very few cases were reported which it persist for years [8].

Other basal ganglion T1 hyperintensities causes are because of deposition of elements in basal ganglia like intracranial hemorrhage containing methemoglobin, hemorrhage transformed from infarct, idiopathic calcification, hamartomas in NF (Neurofibromatosis) type1, toxicity like carbon monoxide and manganese, copper related Wilsons disease, global hypoxia, chorea or ballsimus associated with hyperglycemia, non ketotic hyperglycemic hemi chorea and other causes [9].

T1 hyperintensites in unilateral basal ganglia are seen very rarely which are unique. CT /MRI abnormal signal in Unilateral basal ganglia mostly seen in putaminal area who are having non-ketotic hyperglycemia and other acute conditions should be ruled out like vascular lesions and infarct having neurological symptoms.

Very few cases has been published on non-ketotic hyperglycemia MRI image findings in sequences diffuse weighted images (DWI) and susceptibility weighted images (SWI) which showed mild and some showing moderate restricted diffusion which corresponds to T1WI (T1weighted imaging) hyper intensities in respective lesions of basal ganglia and no notable abnormality which is evident in our case [1,10,11]. On SWI, results have been mixed with some of these cases demonstrating susceptibility while some cases have not shown any abnormality like in our case [12,13].

Metabolic disorders leading to movement disorders or chorea are easily treated and can be reverted back with prompt correction of hyperglycemia [14]. So, hemichorea-hemiballismus occurring in diabetes mellitus owing to non-ketotic hyperglycemia is a rather benign condition with a good prognostic outcome but provided the syndrome is recognized early and corrected like in our case.

Imaging findings and clinical presentation can mimic stroke and pathophysiology of non-ketotic hyperglycemia is not ischemic clearly [10]. So radiologists and physicians should also keep this entity as a possible cause which it can be easily reversible by appropriate treatment.

CONCLUSION

Non-ketotic hyperglycemia can sometimes present with normal CT brain findings and we should not conclude to a normal and MRI can help us in differentiating the cause. Hemichorea-hemiballismus is clinico-radiological diagnosis and proper history of the patient is needed to get a prompt diagnosis. Non-ketotic hyperglycemias, even though are not common the other possible diagnosis in cases presenting acute symptoms to be ruled out and treating the hyperglycemia by correcting to normal glucose levels in hemichorea-hemiballismus will have the better results. Follow-up of such patients on regular medications for diabetes will make the patient to relieve from again occurrence of the hemichorea-hemiballsimus.

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