Non-ketotic Hyperglycemia-induced Hemiballism-Hemichorea

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Case Summary

A middle-aged patient presented to the emergency room complaining of acute onset of involuntary movements in the left upper and lower extremities.

Imaging Findings

Head computed tomography (CT) demonstrated asymmetric hyperdensity in the right basal ganglia involving the caudate and lentiform nuclei (putamen and globus pallidus), (Figure 1). Magnetic resonance imaging (MRI, Figure 2) showed T1 shortening (hyperintensity) involving the right basal ganglia (caudate and lentiform nucleus), whereas postcontrast T1 MRI showed no associated enhancement. Gradient echo sequence revealed no areas of abnormal susceptibility effect. Diffusion weighted imaging did not reveal any regions of restricted diffusion.

Diagnosis

Non-ketotic hyperglycemia-induced hemiballism-hemichorea

Figure 1. Noncontrast axial head CT demonstrates asymmetric hyperdensity in the right basal ganglia involving the caudate and lentiform nucleus (putamen and globus pallidus).

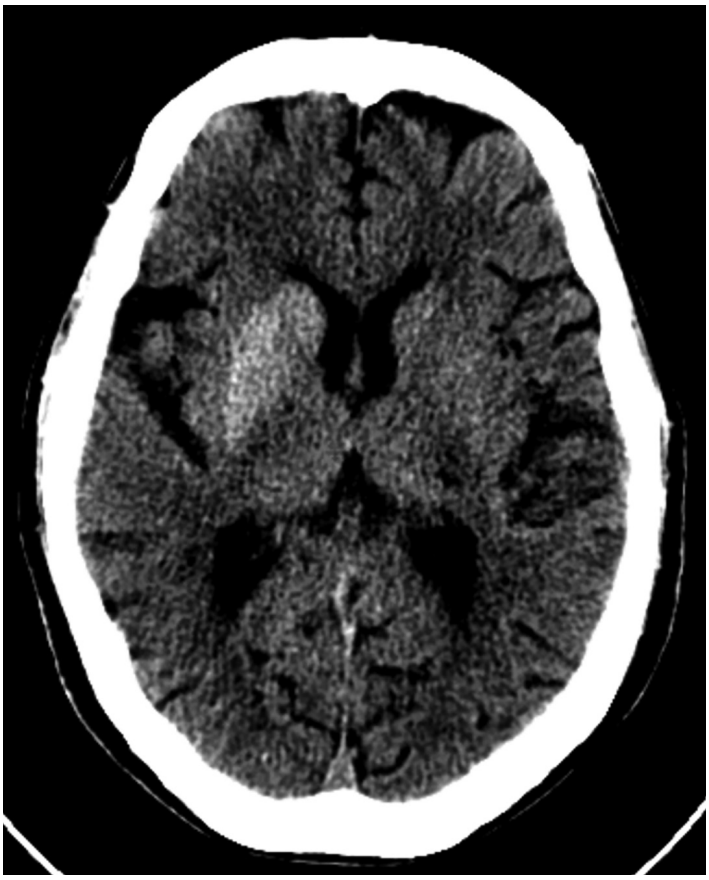

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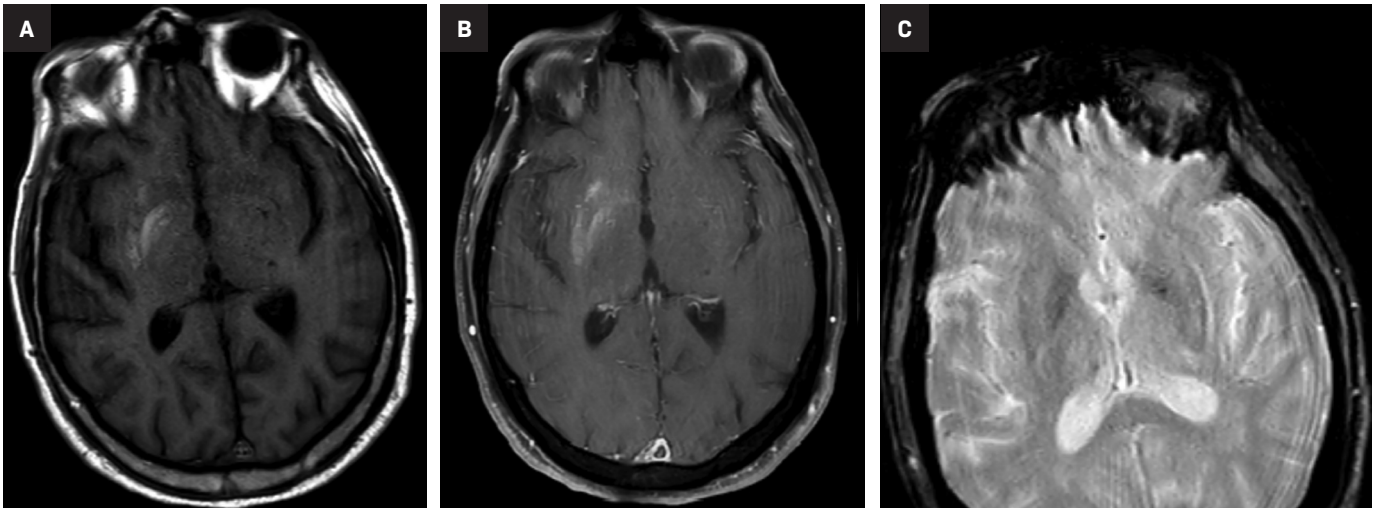
Figure 2. MRI shows T1 shortening (hyperintensity) involving the right basal ganglia (caudate and lentiform nucleus) on axial T1 (A); no significant enhancement on fat-suppressed postcontrast T1 (B); and no abnormal paramagnetic susceptibility effect on a T2 gradient echo sequence (C).



Discussion

Hemiballism-hemichorea manifests through involuntary jerky movements within one limb or two limbs on the same side of the body.[1] The two most common causes of hemiballism-hemichorea are vascular complications and non-ketotic hyperglycemia. Traditional research on the pathophysiology of hemiballism-hemichorea points to the role of lesions in the subthalamic nucleus, while more recent research focuses on lesions in the basal ganglia structures or abnormal neuronal firing patterns in these structures.[2,3] Hemiballism-hemichorea affects female patients more frequently than male, and is usually late onset.[4]

Radiologic exams in these cases typically reveal abnormalities in the basal ganglia, especially the putamen, contralateral to the side experiencing involuntary movement.[1] CT demonstrates high-attenuation changes, T1 MRI shows hyperintensity, and T2 MRI shows hypointensity or isointensity.[1] The intrinsic T1 shortening/hyperintensity may be related to the presence of gemistocytes in the lesion that may be causing a shorter T1 relaxation time.[5] Awareness of this unique set of radiologic findings is important to ensure the correct diagnosis.

The primary treatment for this disorder is clinically correcting and controlling serum glucose levels in the short-term and long-term.[1] Symptoms usually subside after normal metabolism is restored (correction of hyperglycemia).[6] The disorder has a favorable prognosis, although 20% of patients may continue to experience hemiballism-hemichorea following clinical treatment.[1] Pharmacologic treatment in these cases helps mitigate symptoms.

Conclusion

Non-ketotic hyperglycemia-induced hemiballism-hemichorea is a rare hyperkinetic disorder characterized by involuntary jerky movements in one or both limbs on the same side of the body; clinically this can be confused with other disorders. For this reason, recognizing the unique radiologic findings associated with the disorder is important to ensure accurate diagnosis. When promptly identified, it is relatively simple to treat and has a favorable prognosis.

References

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