Mystery Case: Syndrome of bilateral basal ganglia lesions in uremic encephalopathy

A 38-year-old patient with chronic kidney disease on regular hemodialysis was investigated for complaints of headache and excessive drowsiness. Significant fluctuation of blood glucose levels (predominantly hyperglycemia) along with deranged creatinine levels was reported over the preceding week with no accompanying history of respiratory tract infection, fever, liver disease, hypoxia, or toxic fume exposure. Spot blood urea nitrogen, serum creatinine, sodium, potassium, and blood sugar levels were within normal range. Brain MRI revealed symmetrical T2/fluid-attenuated inversion recovery (FLAIR) hyperintense nonhemorrhagic bilateral basal ganglia lesions showing mild diffusion restriction (figure). Findings were attributed to a combined effect of uremic toxins and metabolic acidosis and a diagnosis of syndrome of bilateral basal ganglia lesions was made in a setting of uremic encephalopathy.1

AUTHOR CONTRIBUTIONS
C.K. Ahuja and M.K. Yadav were responsible for data acquisition and collection. C.K. Ahuja wrote the report. N. Khandelwal and M.K. Yadav made the necessary corrections. N. Khandelwal proposed the final diagnosis.

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REFERENCES

MYSTERY CASE RESPONSES
The Mystery Case series was initiated by the Neurology® Resident & Fellow Section to develop the clinical reasoning skills of trainees. Residency programs, medical student preceptors, and individuals were invited to use this Mystery Case as an educational tool. Responses were solicited through a group e-mail sent to the American Academy of Neurology Consortium of Neurology Residents and Fellows and through social media.

The majority of respondents—68%—suggested this was the lentiform fork sign, associated with metabolic acidosis and uremic encephalopathy. Other suggested diagnoses included manganese toxicity, hypercalcemia, and iron deposition, all of which can cause changes in the basal ganglia visible on MRI. The images in this mystery case are an example of the syndrome of bilateral basal ganglia lesions, caused by uremic encephalopathy. This finding was originally described in end stage renal disease patients with diabetes who developed acute onset of movement disorders in the setting of severe uremia. These reversible lesions are characterized by extensive T2-weighted hyperintensities in the basal ganglia that are hypointense on T1, with significant edema and, in some cases, petechial hemorrhage. The pathophysiology of these findings is posited to be a failure of cellular metabolism and vascular autoregulation when an acute metabolic insult occurs in already chronically damaged tissue.

The lentiform fork sign was first reported by Kumar and Goyal, who described primarily vasogenic edema manifesting as bright T2/FLAIR hyperintense demarcation of the putamen bilaterally, resembling a fork, and without the diffuse hyperintense changes seen in this case. The theorized cause of this sign is the difference in lactate metabolism in the neurons of the basal ganglia vs the astrocytes in the surrounding white matter in the setting of metabolic acidosis.

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