Dear editor,

This is a case of a 68-year-old right-handed man admitted to the hospital with 2 days of acute onset of slurred speech and unsteady gait. He denied any other neurological complaints. His past medical history was significant for alcohol dependence with daily alcohol use and 1 day a week of binge drinking for >10 years. He denied any history of alcohol-related seizures, DTs, psychiatric diagnosis or alcohol-related legal or interpersonal problems. The patient was oriented, able to name, read, repeat and follow commands. There was impaired short-term memory. He had severe dysarthria. Cranial nerves were intact. There was decreased vibration at toes with hyporeflexia at ankles; the remaining reflexes and motor exam were normal. There was dysmetria in all limbs and a wide-based ataxic gait. The differential diagnosis was a vascular event, alcohol-related cerebellar degeneration, Wernicke’s encephalopathy, Marchiafava–Bignami disease, paraneoplastic cerebellar degeneration or a posterior fossa mass. The baseline CBC and comprehensive metabolic panel were within normal limits except for an elevated white blood cell count and mildly elevated AST. Baseline vitamin levels were not obtained. The MRI-brain showed abnormal high DWI (diffusion-weighted imaging) and T2 signal in the splenium of the corpus callosum with no mass effect or edema and scattered T2 signal hyperintensities within the bilateral hemispheric white matter (Fig. 1). A diagnosis of Marchiafava–Bignami disease (MBD) was made. The patient was started on high-dose thiamine intravenously for 5 days (500 mg three times a day) and oral vitamin B
complex. Seven days after the symptom onset there was significant improvement, the patient was able to ambulate unassisted and the dysarthria was improved. After discharge the patient was lost to follow-up.

MBD is known to be associated with chronic alcoholism and malnutrition. It used to be considered rare and fatal and diagnosed post mortem but imaging has allowed for early detection and intervention. It is characterized by demyelination and necrosis of the corpus callosum. Its pathogenesis is still unclear but it is attributed to deficiency of the B-complex group of vitamins (Bhat et al., 2014; Wenz et al., 2014).

The mean age of onset of MBD is 46 years. The most common manifestations are: cognitive impairment, dysarthria, disconnection syndromes, seizures and pyramidal tract symptoms (Heinrich et al., 2004). There are two acute/subacute types. Type A with major impairment of consciousness, seizures, dysarthria, hemiparesis and Type B presents dysarthria, gait disturbance, signs of interhemispheric disconnection and less impairment of consciousness. In Type A the entire corpus callosum is affected (100 vs. 10% in Type B) and more frequently presents with extracallosal lesions (47 vs. 17% in Type B). Type A has a worse prognosis (Heinrich et al., 2004). A chronic course has been described with progressive dementia, behavioral abnormalities and signs of interhemispheric disconnection (Menegon et al., 2005). The case presented was most likely Type B as there was no impairment of consciousness, only the splenium of the corpus callosum was affected and the prognosis was good.

The diagnosis is made with MR imaging of the brain, which shows lesions involving the central portion of the body of corpus callosum with sparing of dorsal and ventral layer. Lesions are hypointense on T1, hyperintense on T2/FLAIR showing diffusion restriction on DWI and variable reduction in ADC (apparent diffusion coefficient). The lesions lack contrast enhancement and do not show mass effect. In the chronic state, atrophy and cavitation of the corpus callosum is seen (Menegon et al., 2005; Kakkar et al., 2014).

There is no proven treatment. Folate, thiamine, vitamin B12 and other B vitamins are commonly used but the response to treatment is variable. Several case reports have shown improvement with high-dose corticosteroid therapy.

MBD should be considered in the evaluation of alcoholic or malnourished patients that present with acute or subacute neurological symptoms. Early diagnosis and treatment may prevent progression to irreversible corpus callosum damage.

REFERENCES