

ANSWER: 3) MarchiafavaBignami Disease

DISCUSSION

These classical neuro-radiological features are suggestive of Marchiafava Bignami Disease (MBD), a rare condition mostly seen in alcoholics and malnourished states. Marchiafava and Bignami in 1903 described toxic degeneration of corpus callosum in chronic alcoholics, who consumed inexpensive Chianti red wine. [1]Most MBD patients are 40 to 60 year old males with history of chronic alcohol consumption. [2]MBD can present as acute (catastrophic seizures, coma and death), subacute (variable mental changes, gait disturbances, confusion, memory deficits) or chronic (progressive dementia) forms. On imaging, typically corpus callosum body is affected first, followed by genu and lastly splenium. Low signal intensity in the corpus callosum on T1 weighted imaging and high signal intensity on T2 weighted imaging is classical, but the lesions can extend into the cortical deep white matter and pericallosal and peri ventricular areas also, in case of severe or chronic MBD. [3]Flair images reveal central hypointensities corresponding peripherally hyperintense rim forming the 'Sandwich Sign', a rare, but characteristic diagnostic sign in MBD. The hypointensity represents necrosis and hyperintensity represents gliosis. [4]This patient died within 10 hours of presentation following uncontrolled seizures and cardio-pulmonary compromise, even after aggressive anti seizure medication use and nutritional corrective efforts were undertaken.

REFERENCES

1. E. Marchiafava, A. Bignami: *Sopraun' alterazione del corpallosoosservata da sogettialcoolisti*. Rivista di patologia nervosa e mentale, 1903, 8 (12): 544-549.
2. Goswami P, Medhi N, Sarma PK, Sarma BJ. MRI findings in marchiafava - bignami disease with central pontinemyelinolysis : A case report. *Neuroradiology*. 2006; 16(4):779-81.
3. Bano S, Mehra S, Yadav SN, Chaudhary V. Marchiafava-Bignami disease: Role of neuroimaging in the diagnosis and management of acute disease. *Neurology India*. 2009; 57(5):649-51.
4. Iwai T, Matsuo K, Mori-Kitani F, Azuma F, Matsuo H et al. Marchiafava-Bignami disease with hyperintensity on late diffusion-weighted imaging *Acta Radiol Short Rep*. 2014 August; 3(7): 2047981614543284.