Multinodular and Vacuolating Neuronal Tumor Presenting with Neuropsychiatric Decompensation in an Adult with Lennox-Gastaut Syndrome

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Clincal History

A 36 year old woman with a history of Lennox Gastaut syndrome, schizophrenia, and global developmental delay presented with one month of progressive cognitive decline, loss of ambulation, increased dependence in activities of daily living, and diffuse bilateral lower extremity pain and swelling. Family denied recent seizures, fever, or infectious symptoms. Psychiatric medications had been recently adjusted with discontinuation of antipsychotic therapy, raising concern for neuropsychiatric decompensation. Neurologic examination revealed diffuse proximal weakness without focal deficits. Initial laboratory studies were largely unremarkable aside from hypoalbuminemia. EEG demonstrated generalized background slowing without epileptiform discharges. Given subacute neurologic decline, autoimmune and infectious encephalitis were initially considered.

Imaging Findings

MRI brain performed with seizure protocol demonstrated a nonenhancing lesion within the subcortical white matter of the left superior frontal gyrus characterized by clustered nodular T2 and FLAIR hyperintensities with a multinodular "soap bubble" appearance. The lesion showed no diffusion restriction, no surrounding vasogenic edema, and no mass effect. T1 weighted pre and post contrast sequences confirmed absence of enhancement. Imaging characteristics were classic for multinodular and vacuolating neuronal tumor. No additional acute intracranial abnormalities were identified.

Figure 1. *Sagittal T1-weighted pre-contrast MRI* demonstrates hypodense subcortical/cortical lesion located within the superior frontal gyrus.

Figure 2. *Sagittal T1-weighted post-contrast MRI* image confirms the absence of enhancement.
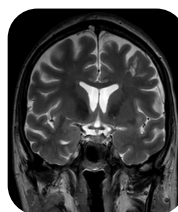

Figure 3. *Coronal T2-weighted MRI* shows T2 hyperintense lesion with cystic/soap-bubble appearance mainly involving the subcortical white matter.
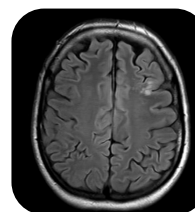

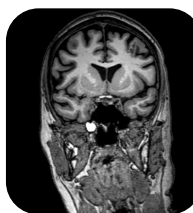**Figure 4.** *Axial FLAIR MRI* reveals mild hyperintensity of the lesion.

Figure 5. *Coronal T1-weighted post-contrast MRI* images also confirm the absence of enhancement.

Teaching Point

Multinodular and vacuolating neuronal tumor should be recognized as a benign WHO grade 1 lesion characterized by clustered nonenhancing T2 and FLAIR hyperintense nodules without diffusion restriction. Awareness of this classic imaging pattern can prevent misdiagnosis as inflammatory or neoplastic disease and avoid unnecessary invasive procedures, particularly in patients presenting with complex neuropsychiatric symptoms.

Discussion

Multinodular and vacuolating neuronal tumor is a WHO grade 1 neuronal neoplasm first described in 2013 and typically discovered incidentally. These lesions are most commonly located in the temporal lobes but may also occur in frontal subcortical regions. Radiographically, MVNT presents as clustered T2 and FLAIR hyperintense nodules without enhancement or diffusion restriction, distinguishing it from neoplasm, abscess, demyelination, or autoimmune encephalitis. Although classically asymptomatic, MVNT has been associated with seizures and focal neurologic symptoms in some patients. In this case, frontal lobe involvement raised concern for contribution to executive dysfunction and neurobehavioral decline, particularly in a patient with complex baseline neurologic and psychiatric disease. Extensive evaluation including lumbar puncture, infectious and autoimmune panels, and EEG was unrevealing, supporting radiographic diagnosis and conservative management. Recognition of MVNT imaging patterns is critical to prevent unnecessary biopsy or aggressive intervention.

References

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Osborn AG, et al. **The 2021 WHO Classification of CNS Tumors: What the Radiologist Needs to Know.** *AJNR.* 2022. Practical radiology summary; notes MVNT as nonprogressive WHO grade 1 with characteristic MRI pattern.