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Case Report

Case Report of An Intriguing Brain Tumor – Mvnt (Multinodular and Vacuolating Neuronal Tumor)

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Abstract:

Background

Multinodular and vacuolating neuronal tumor (MVNT) is an uncommon mixed glial neuronal lesion that is benign and recently identified. It was first included in 2016 WHO Classification of Tumors of the Central Nervous System. Here in, we report a case of MVNT in a 19-year-old female patient and was advised only regular follow up with imaging without the need for biopsy.

Case Description

A 19-year-old female patient presented with complaints of headache for which was evaluated with imaging. The MRI brain showed non contrast enhancing, cluster of well-defined nodules in the subcortical and periventricular white matter of the right temporal lobes, suggestive of multinodular vacuolating neuronal tumor (MVNT).

Conclusion

This case report emphasizes that these lesions are benign, CNS WHO Grade 1 tumors, that usually do not require biopsy in asymptomatic cases and further studies will be required for better understanding of the tumor and as a result, we will be able to prevent misdiagnosis and aggressive treatment.

Key words: biopsy; brain tumors; misdiagnosis

Abbreviations

MVNT - Multinodular and Vacuolating Neuronal Tumor

WHO - World Health Organization

MRI – Magnetic Resonance Imaging

FLAIR – Fluid attenuated inversion recovery

CNS – Central Nervous System

CT – Computed Tomography

Cho - Choline

NAA - N-Acetyl Aspartate

DNET Dysembryoplastic Neuro Epithelial Tumor

Introduction

MVNT (multinodular and vacuolating neuronal tumor) was first described a decade earlier as a benign neuronal tumor in 2013 by Huse et [1] al and has been included in the World Health Organization classification of tumors of the central nervous system in 2016 [2].

It typically occurs in middle aged adults, predominantly in the cerebral subcortical region, and are mostly present incidentally on imaging or in symptomatic patients with seizures. Most of the lesions reported in the literature are seen in the temporal lobe. Recently, a similar pattern of imaging findings was described in the posterior fossa. Although this lesion is rare, one should be aware with the presentation and imaging findings.

Here we present a case of 19-year-old female patient with incidental finding of an MVNT with no neurologic deficits and the treatment protocol.

Case Report

A 19-year-old female patient presented to the neurosurgery department, with headache, involving the frontal region, each episode lasting for 30 to 40 minutes and radiating to the cheek and was associated with nausea.

The headache aggravated on exposure to bright light or watching television and relieved with medications and sitting in dark room or switching off the television.

She had no history of seizure, loss of consciousness, vomiting, blurring of vision or weakness. Her past medical history was not significant and clinically her neurological examination was normal.

MRI brain with contrast (Figure.1)

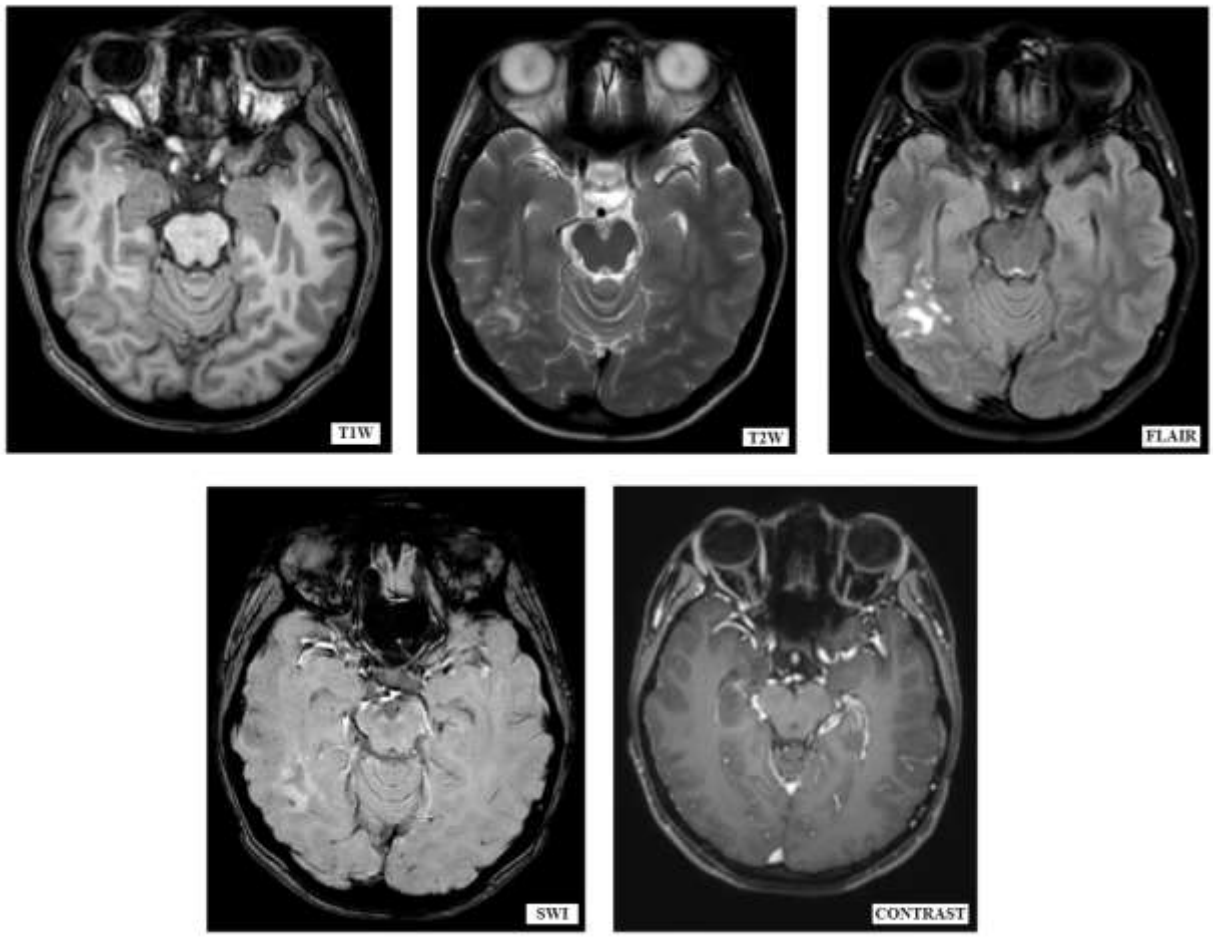

Figure 1: MRI Brain plain and contrast of the patient that shows a cluster of coalescent nodules in the posterior aspect of right temporal and right tempero-parietal lobes.

Was done, that revealed a small cluster of well defined, high T2W/ FLAIR weighted coalescent nodules in the subcortical and periventricular white matter of the posterior aspect of right temporal and right tempero-parietal lobes. Overlying cortex is not thickened which was suggestive of multinodular vacuolating neuronal tumor.

Since the patient had no symptoms or findings except for headache that was not attributed to the lesion and based on the MRI findings, surgical intervention was not recommended. Repeat imaging on follow up was advised after 6 months and 1 year to assess if there is any interval change in the characteristics of the lesion.

Discussion

Huse et al[1] in a case series of 10 patients described these distinctive neuronal lesions first in 2013. It was included in the 2016 WHO classification[2] of CNS neoplasms under the group of neuronal and mixed glial-neuronal tumors. They are usually low grade (CNS WHO Grade 1) tumors[2]. These tumors usually do not show gender predilection and are rare benign, multinodular lesion with a unique membrane-bound appearance of the neuronal cytoplasm. Lesions are usually between 1 mm to 5 mm in size and can be found as separate entities or in clusters.

Most MVNTs are usually asymptomatic and are an incidental finding on imaging studies[3]. In symptomatic patients, the most common presentation is seizures.

On CT imaging, they are difficult to identify, yet as described by Nagaishi et al[4]., in some patients they are seen as hypo dense and non-enhancing lesion in the subcortical white matter. MRI is considered as gold standard imaging for these lesions. They are seen as a group of nodules on the inner surface of the cortex. Nodules are hyperintense to the adjacent white matter on T2W and FLAİR images. They are isointense or mildly hypointense on T1W. They typically exhibit no diffusion restriction or contrast enhancement. They demonstrate no blooming on susceptibility-weighted imaging.

On MR spectroscopy, as described by Lecler et al[5]., there is no Cho peak with mild decrease in NAA. However, a few other reports in literature did show increased Cho peak with decreased NAA.

MVNTs have been shown to harbor the clonal MAPK pathway-activating genetic abnormality.

Histologically, they are well-defined, multiple, oval to round nodules that exhibits coalescence with vacuolating degeneration. The cells do not show mitosis or atypia. The area surrounding the brain parenchyma is usually normal but can also be gliotic on rare occasions.[1,6]

The most common differential diagnosis includes DNET, gliomas and less commonly enlarged perivascular spaces and focal cortical dysplasia.

These lesions usually do not require a biopsy or surgical excision in asymptomatic patients. Surgical excision of the tumor is considered only when it is associated with a seizure. No tumor recurrence is reported regardless of the extent of the excision.

Conclusion

It is essential to recognize the unique appearance of MVNTs and should be considered in the differential diagnosis of a multi nodular lesion in the sub cortical location without any surrounding edema on imaging. Identifying these tumors on imaging studies prevents unnecessary surgical excision or biopsy and in asymptomatic patients, it is enough to perform follow-up images.

Acknowledgements

None

Conflicts Of Interest

None

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