



ANS CASE LESSON 01/09/2022

Leave me Alone...!

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Abstract

Multinodular and Vacuolating Neuronal Tumor (MVNT) of the cerebrum are recently described and very rare lesions, that are often found incidentally. From their first description in 2013, there are in total 48 cases reported in pubmed, from which only 19 have a confirmation with biopsy.

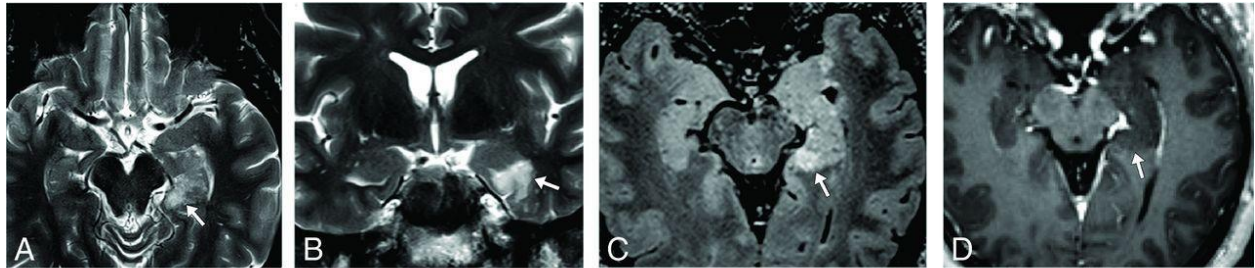
Here we present the case of a 62 years old man with an epileptogenic left parietal MVNT confirmed with biopsy. Our patient showed epileptic seizures two weeks before diagnosis.

In absence of clinical signs, these benign lesions should be followed up and no surgery is recommended.

Introduction

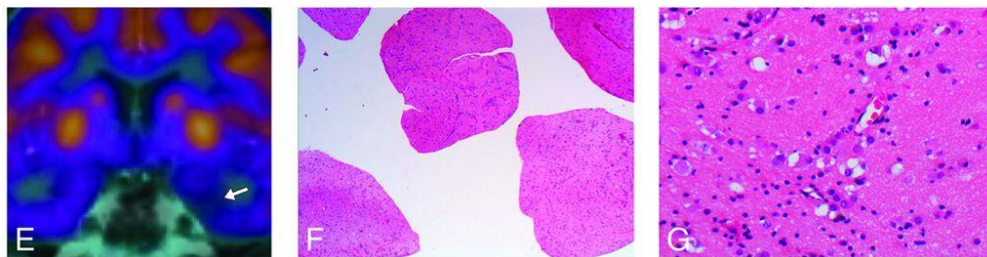
Multinodular and Vacuolating Neuronal Tumor (MVNT) of the cerebrum was first described in 2013 from Huse at al., in a series of 10 cases of a distinctive seizure associated lesions¹ In the 2016 WHO Classification of Tumors MVNT is considered as a unique pattern of gangliocytoma². These lesions are uncommon but fascinating because they are less aggressive than the more common glial tumors and their prognosis is excellent. Neurologic manifestations vary from case to case and include seizures, symptoms of increased intracranial pressure, and neurologic deficits according to tumor location. However, in most cases they do not show up with clinical signs and may be incidental³. Radiological findings as described so far in the literature are: hyper intense lesions on T2 and Flair in more than 1/3 of cases and they very rarely enhance contrast.

In figure 1 we show a case from R.H. Nunes et al. AJNR Am J Neuroradiol 2017;38:1899-1904, recently published, to illustrate the radiologic features of these lesions that commonly are diagnosed on MRI as DNET.



*Fig 1. A case from R.H. Nunes et al. AJNR Am J Neuroradiol 2017;38:1899-1904 (MRI)
A 53-year-old male patient with temporal lobe epilepsy (case 31). A–D, MR imaging demonstrates a probable MVNT in the left parahippocampal gyrus (arrows).*

Studies show that the histopathological features of these tumors are characterized by neuroepithelial cells with conspicuous stromal vacuolation arranged in nodules principally within the deep cortical ribbon and superficial subcortical white matter. These cells typically orient perpendicular to the cortical surface which correlates with clusters in the radiological appearance. These cells typically show immunopositivity for HuC/HuD and Olig 2 (typical neurogenesis markers) but weak positivity for mature neuronal lineage cells. These characteristics suggest that these cells may represent a dysplastic early phenotype and therefore these lesions fall into a category between cortical malformation and hamartoma's lesion, which show a benign behavior³.



*Fig.2 A case from R.H. Nunes et al. AJNR Am J Neuroradiol 2017;38:1899-1904 (histology)
E, FDG-PET MR brain imaging shows focal hypometabolism in the left medial temporal lobe corresponding to the site of suspected lesion. F (20x) and G(200x), Hematoxylin-eosin-stained histopathology slides demonstrate the abnormal clustering and vacuolation of the neuronal cells.*

A graphical illustration representing a schematic coronal view of these lesions, reprinted from Osborn is presented in Figure 3.

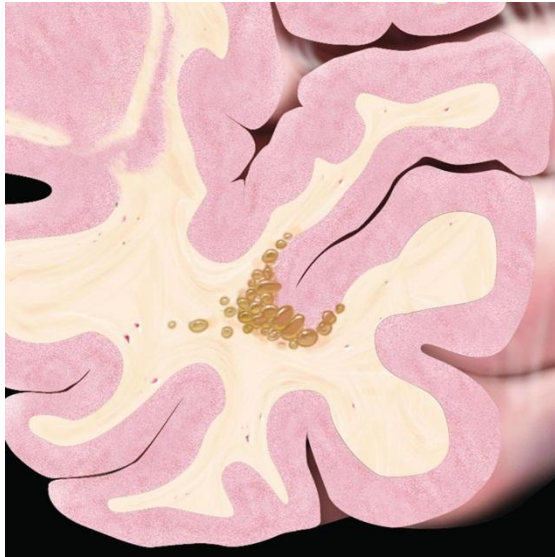


Fig.3 MVNT typical presentation graphic illustration. Schematic coronal view depicts a nodular “bubbly-appearing” lesion located on the inner surface of otherwise normal-appearing left temporal lobe cortex within the deep cortical ribbon and superficial subcortical white matter. Reprinted from *Osborn's Brain*^{9, 3}

From the first description in 2013, there are in total only 48 published cases described in pubmed. Only 19 of them have been confirmed through biopsy¹.

Herein we report a case of MNVT involving a 62 years old man who presented with an epileptogenic, superficial solid lesion at the left parietal lobe in “Mother Teresa Hospital”. MVNT diagnosis was confirmed by the pathologist.

Our Case:

A 62 years old man was referred to “Mother Teresa” University Hospital after several generalized seizures for two weeks, without past history for neurological or other diseases. No focal neurological deficits were present in the objective examination.

MRI showed a round shaped lesion in the left parietal lobe, with dimensions about 1cm diameter, adjacent to the cortical area of the left parietal lobe. The lesion was hyper intense in comparison with white matter on T2 and isointense with gray matter on T1 (Fig. 4) . Radiological diagnosis was DNET.

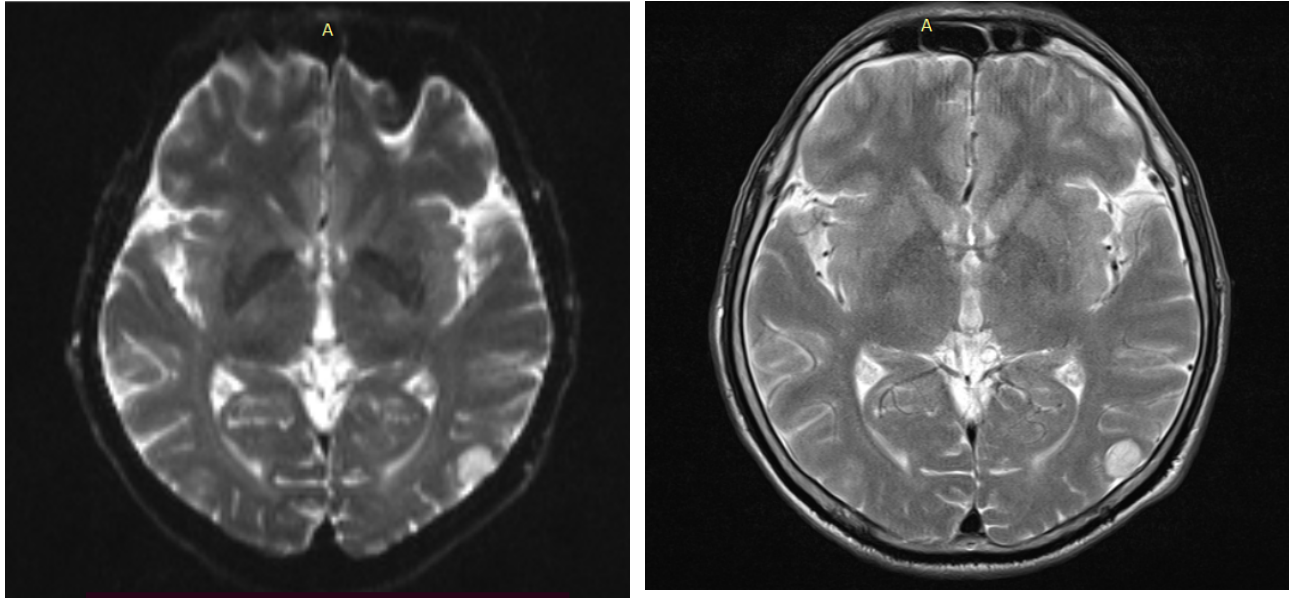


Fig. 4. T2 and flair sequence show both hyper intense subcortical lesions with a well defined border and a diameter about 1cm, with a nodular appearance, without contrast enhancement.

The patient underwent eksjional biopsy and the specimen was sent for examination to Ashford and St. Peter's Hospitals NHS Foundation Trust, Pathology Unit.

As shown in Figure 5 microscopic examination revealed a subcortical lesion with e proliferation of cells resembling ganglion cells, with eccentric round nuclei, foamy, and relatively ample eosinophilic cytoplasm suggestive of multinodular and vacuolating neuronal tumor of the cerebrum, WHO grade 1.

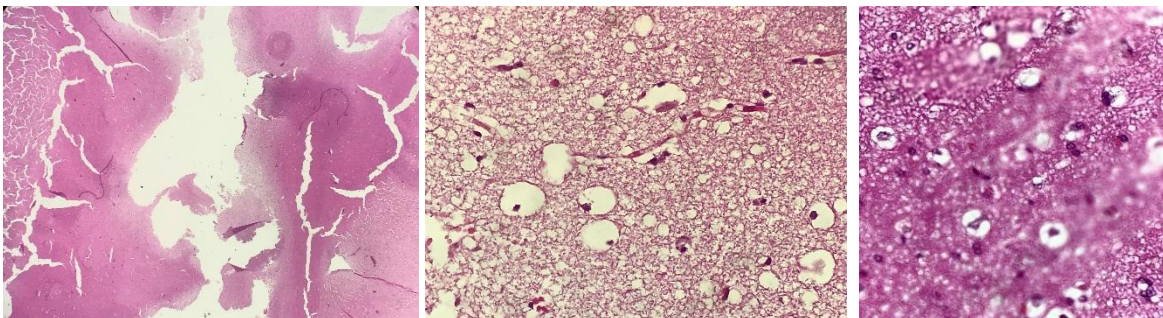


Fig 5. Subcortical lesion ganglion like cells with vacuolated cytoplasm suggesting vacuolated neuronal tumor WHO grade 1



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References

1. Huse JT, Edgar M, Halliday J, et al Multinodular and vacuolating neuronal tumors of the cerebrum: 10 cases of a distinctive seizure-associated lesion. *Brain Pathol* 2013;**23**:515–24 doi:10.1111/bpa.12035 pmid:23324039
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3. Nunes RH, Hsuet CC et al, Multinodular and Vacuolating Neuronal Tumor of the Cerebrum: A New “Leave Me Alone” Lesion with a Characteristic Imaging Pattern: *American Journal of Neuroradiology* October 2017, 38 (10) 1899-1904; doi.org/10.3174/ajnr.A5281.