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Multinodular and vacuolating neuronal tumour (MVNT): Case report of a rare epileptogenic brain tumour

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Abstract
Multinodular and Vacuolating Neuronal Tumours (MVNT) are newly described entities with distinct radiological patterns and cytoarchitecture. These are associated with epilepsy although most are usually asymptomatic. We present a case of a 55 years old male with new onset seizures and altered sensorium. No electrolyte imbalances were present or any previous history of seizures given. MRI of the brain revealed subcortical tiny nodular cystic lesions suggesting a probable MVNT rather than a Dysembryoplastic Neuroepithelial Tumour (DNET) due to the subcortical location. No surgical intervention was recommended. The patient was stable on antiepileptics.

Keywords: Neuronal tumour, epilepsy, multinodular and vacuolating neuronal tumour (MVNT), neuroradiology

Introduction
Multinodular and Vacuolating Neouronal Tumours (MVNT) are recently described benign brain tumours that have been included in the new edition in the 2016 WHO classification of the tumours of the central nervous system as an architectural pattern [1] and has been recognized as a distinct entity in the 2021 5th edition [2]. Usually seen in middle age individuals, clinical manifestations include headache and episodes of seizure however can be incidentally detected. They were previously reported as enlarged perivascular spaces or Dysembroplastic Neuroepithelial Tumour (DNET) due to recent knowledge of the entity hence the true prevalence cannot be ascertained. Most of these lesions are described as “leave me alone” lesions and have a benign course and hence biopsy or surgical intervention is not usually recommended [3].

Case presentation
A 55 years old male patient with a past history of hypertension and newly diagnosed type 2 diabetes mellitus presented to the emergency with ~5 episodes of involuntary movements of all the four limbs suggesting seizures followed by loss of consciousness and few episodes of vomiting. The patient was hypertensive (190/110mm Hg) and had a high blood glucose level on presentation (361mg/dl). The patient had normal liver and renal function tests. All the electrolytes including sodium, potassium, chloride and bicarbonate levels were within normal range.

CT scan of the head at the time of presentation revealed no significant abnormality. Following which, MRI brain was obtained on 1.5T including thin sections and three dimensional heavily T2 weighted and FLAIR sequences which showed a cluster of band like tiny nodular cysts along the grey white matter junction/ superficial subcortical white matter in a ribbon like configuration in the right superior frontal gyrus (Figure 1). They were T2 hyperintense/T1 hypointense and most of them did not suppress on FLAIR (Figure 2). No surrounding white matter edema was seen. The lesion showed no diffusion restriction or blooming foci within. No significant abnormality was seen on MR angiogram and venogram. Differential diagnoses of MVNT and DNET were considered. DNET was ruled out due to minimal cortical involvement, lack of mass effect and advanced age of the patient. Radiological features were consistent with multinodular and vacuolating neuronal tumour. EEG was obtained and epileptogenic focus was localized to the right frontal lobe.

The patient was stable on anti-epileptics. Surgical intervention was advised in view of the seizure episodes however the patient was not willing for surgery.

Figures with legends

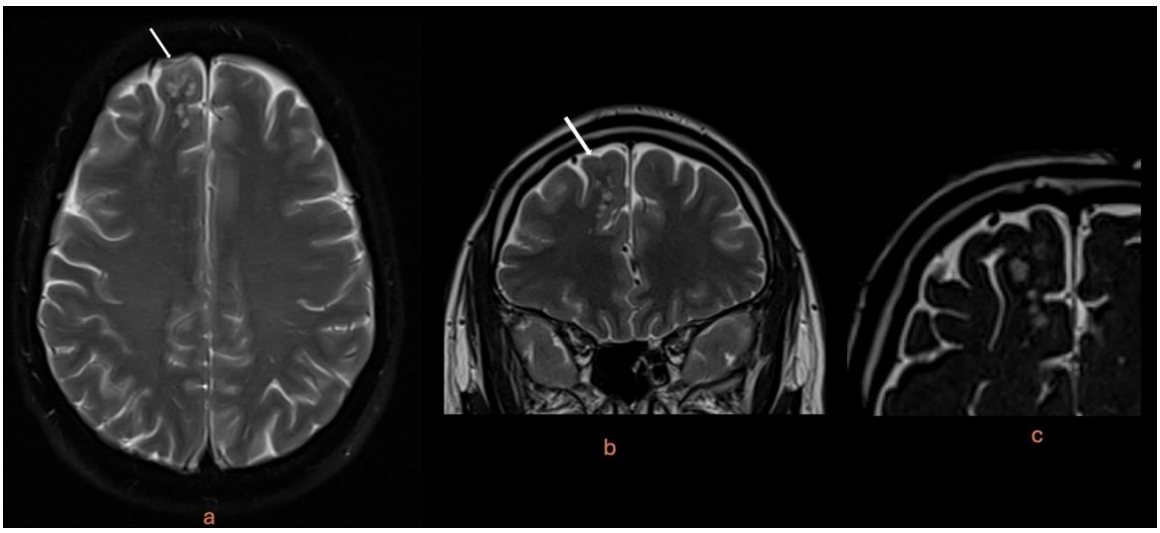

Fig 1: Axial T2 (a) and coronal T2 (b) showing nodular cysts in ribbon configuration along the right superior frontal gyrus (white arrow) along the subcortex without any surrounding edema. (c). Cut image of axial 3D T2 Space focusing on the right superior frontal gyrus highlighting the cystic areas in the subcortex

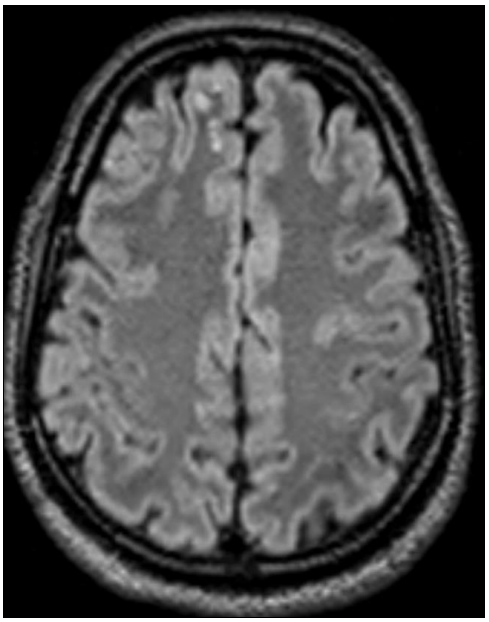

Fig 2: Axial T2 FLAIR showing non suppression of the cystic areas and absence of any white matter edema

Discussion

MVNT is a rare neuronal lesion that is first reported in 2013 by Huse *et al*. [4] and is recently recognized as a separate entity in the 2021 WHO classification of CNS tumours[2]. Some have proposed that MVNT is a malformative lesion rather than a true clonal neoplasm, but the molecular basis of MVNT is uncertain [5].

The majority of MVNT patients are middle-aged, and they may be asymptomatic and diagnosed incidentally or present with headaches or seizures. Most of these lesions exhibit a benign course and remain stable on follow up. They involve the deep cortical layers or the superficial subcortical white matter. Previously published cases have similar histological profile. Dysplastic cells affect the deep cortex and superficial subcortical white matter, are grouped in small to medium sized nodules and show stromal vacuolation and orient perpendicular to the cortical surface [3]. Radiological features correlate well with histology and may be sufficient for a diagnosis. Typical features include cluster of variable sized nodules following the gyral contour exhibiting T2 hyperintensity which do not suppress on FLAIR located along the subcortical ribbon. High protein or solid components within the vacuolations may be the cause of the FLAIR hyperintensity. These typically do not show any adjacent edema or diffusion restriction. These rarely enhance on post gadolinium administration [6].

Usual differential diagnosis includes Dysembryoplastic Neuroepithelial Tumour (DNET), enlarged perivascular spaces and focal cortical dysplasia. DNET is usually cortical based and shows mas effect, post contrast enhancement and usually diagnosed by the second decade. Perivascular spaces suppress on FLAIR and focal cortical dysplasia is characterised by cortical thickening and blurring of grey white junction [7].

Radiological features are highly characteristic; however, a definitive diagnosis can only be made on biopsy. Surgery is

advised in cases where the lesion is definitively established to the be the cause for seizures. Otherwise due to the benign course of the lesion, conservative management is the usual recommendation [3].

Conclusion

MVNT is a recently described and rare brain lesion that is associated with adult epilepsy presenting as ribbon like tiny nodular cysts along the gyrus in the superficial subcortex and do not suppress on FLAIR without any edema or diffusion restriction. Radiological features are characteristic and unnecessary surgical intervention should be avoided to confirm the diagnosis.

Conflict of interest

None to declare.

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