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Radiological and clinical features of multinodular and vacuolating neuronal tumor (MVNT)

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Abstract

Background: To investigate the imaging findings and clinical features of multinodular and vacuolating neuronal tumor (MVNT).

Methods: We retrospectively sought for cases that have suspicious imaging findings for MVNT through the hospital information system. The patients' demographics and clinical symptoms were extracted. All available images were re-examined.

Results: Headache was the most common complaint ($n=7$). Other complaints included seizure, stroke-like symptoms and numbness. Conventional MRI revealed that all lesions consisted of tiny, sharply marginated, round or ovoid nodules following the gyral contour. These nodules were hyperintense on T2 and FLAIR WI, hypointense on T1 WI. All lesions were characterized by a lack of enhancement and diffusion restriction. Mass effect and peripheral edema were not observed. MVNT presented as an incidental finding in one case who complained gynecomastia and had pituitary adenoma on pituitary MRI. All lesions were supratentorial—mostly on the right side (10/11)—and located in subcortical white matter. Follow-up MRI was available for 11 patients with a mean of 14.8 months (3–40 months). No change in lesion size and morphology was observed in these follow-up images.

Conclusions: Radiological and clinical follow-up data suggest MVNT may exhibit indolent behavior. If asymptomatic, patients can be followed by imaging alone. Surgery should be considered for symptomatic patients.

Keywords: Multinodular and vacuolating neuronal tumor, MVNT, Brain tumor, MRI

Background

Multinodular and vacuolating neuronal tumor (MVNT) was first defined as seizure-related lesions in 2013 [1]. It is a rare, recently described (WHO 2016) entity among the large spectrum of glioneuronal tumors of central nervous system (CNS). The frequency and pathophysiology of these lesions are yet to be discovered. Characteristic magnetic resonance imaging (MRI) findings and lack of change in size and morphology in follow-up examinations are highly suggestive for MVNT [2]. The group of nodular lesions is mainly located in deep cortical ribbon and superficial subcortical white matter. These lesions are seen as hyperintense on fluid-attenuated inversion recovery (FLAIR) and T2-weighted image (T2 WI). They do not exhibit edema, mass effect or diffusion restriction on diffusion-weighted image (DWI) and are non-enhancing after IV contrast administration.

In this study, we elaborated the radiological and clinical findings of 11 patients demonstrating imaging features of MVNT observed in our hospital to raise awareness of these rare entity.

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Methods

Eleven cases were found between 2012 and 2018 with suspicious imaging findings that suggest MVNT through the hospital information system. The patients' demographics and clinical symptoms were extracted. All available images were re-examined. Due to the inadequateness to establish a strong relationship between lesions and symptoms, histopathological diagnosis was not made and it was not present in any of the patients. The final diagnosis was made by typical radiological findings and no change in follow-up.

MR imaging examination was performed on either 1.5T or 3T scanners (Magnetom Verio and Avanto Siemens, Erlangen, Germany). The scanning parameters consisting of axial T2-WI (TR/TE,4000/114 ms), FLAIR(TR/TE, 8000/94) and DWI/ADC at b-values 0,1000 (TR/TE 7400/91), pre- and post-contrast T1-WI in axial, coronal and sagittal planes (TR/TE,410/10 ms) were acquired. Contrast-enhanced MRI scans were obtained after intravenous Gd-DTPA administration. Computed tomography (CT) images of only 6 patients were obtained.

The study protocol was approved by the local Institutional Review Board and was performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Results

The patients' demographics and clinical data are presented in Table 1. Mean age was 38 years (16–56 years) with 1,2:1 F/M ratio. Headache was the most common complaint ($n=7$). Other complaints included seizure (one case), stroke-like symptoms (one case), numbness (one case) and gynecomastia (one case). MVNT was detected as an incidental finding in one patient, who complained about gynecomastia and diagnosed with pituitary adenoma on pituitary MRI. One case, who had prior history of lymphoma without CNS involvement, presented with headache.

Conventional MRI revealed that all lesions consisted of clusters of tiny, sharply marginated, round or ovoid nodules following the gyral contour. These nodules were hyperintense on T2 WI and FLAIR, and hypointense on T1 WI. Only one case had a nodular lesion that was characterized by focal signal suppression on FLAIR images suggesting cystic component (Fig. 1). None of the cases showed enhancement or diffusion restriction (Fig. 2). Mass effect and peripheral edema were absent in all cases. However, abnormal hyperintensity on T2/FLAIR in surrounding white matter of the bubble-like lesions was observed in 4 patients.

All lesions were supratentorial—most on the right (10/11)—and located in subcortical white matter.

Cortical involvement was present in 2 (%18) cases. Five (%45) were located in the parietal lobe, whereas 1 (%9) in the occipital lobe, 1 (%9) in the temporal lobe, and 1 (%9) in the frontal lobe. In three cases (%27), the lesion affected more than 1 lobe (parietooccipital). Lesion size was measured on T2 FLAIR sequence which ranged from 15 to 27 mm in maximum axial diameter. Follow-up MRI images were available in 11 patients ranging between 3 and 40 months with a mean of 14.8 months. No change in lesion size or morphology was observed at follow-up examinations of 11 patients. The CT images of five patients were normal. One case had a cystic area, which could be distinguished as a hypodense lesion on CT scan (Fig. 1).

Table 1 Patients demographics, clinical data and imaging findings

Case	Age	M/F	Location	Clinic presentation	MRI follow-up period/change over follow-up	CT
1	47	F	R Temporal	Headache	27 m/No change	
2	42	F	R Parietal	Seizure	40 m/No change	Hypodense cystic lesion
3	56	M	R Parietal	Gynecomastia	12 m/No change	No lesion
4	55	F	R Frontal	Headache	12 m/No change	
5	28	F	R Occipital	Headache	17 m/No change	No lesion
6	25	M	R Parietooccipital	Headache	7 m/No change	
7	35	M	L Parietal	Stroke-like symptoms	7 m/No change	No lesion
8	44	M	R Parietal	Headache	15 m/ No change	No lesion
9	38	F	R Parietal	Headache	3 m/No change	No lesion
10	36	M	R Parietooccipital	Headache	17 m/No change	
11	16	F	R Parietooccipital	Numbness	6 m/No change	

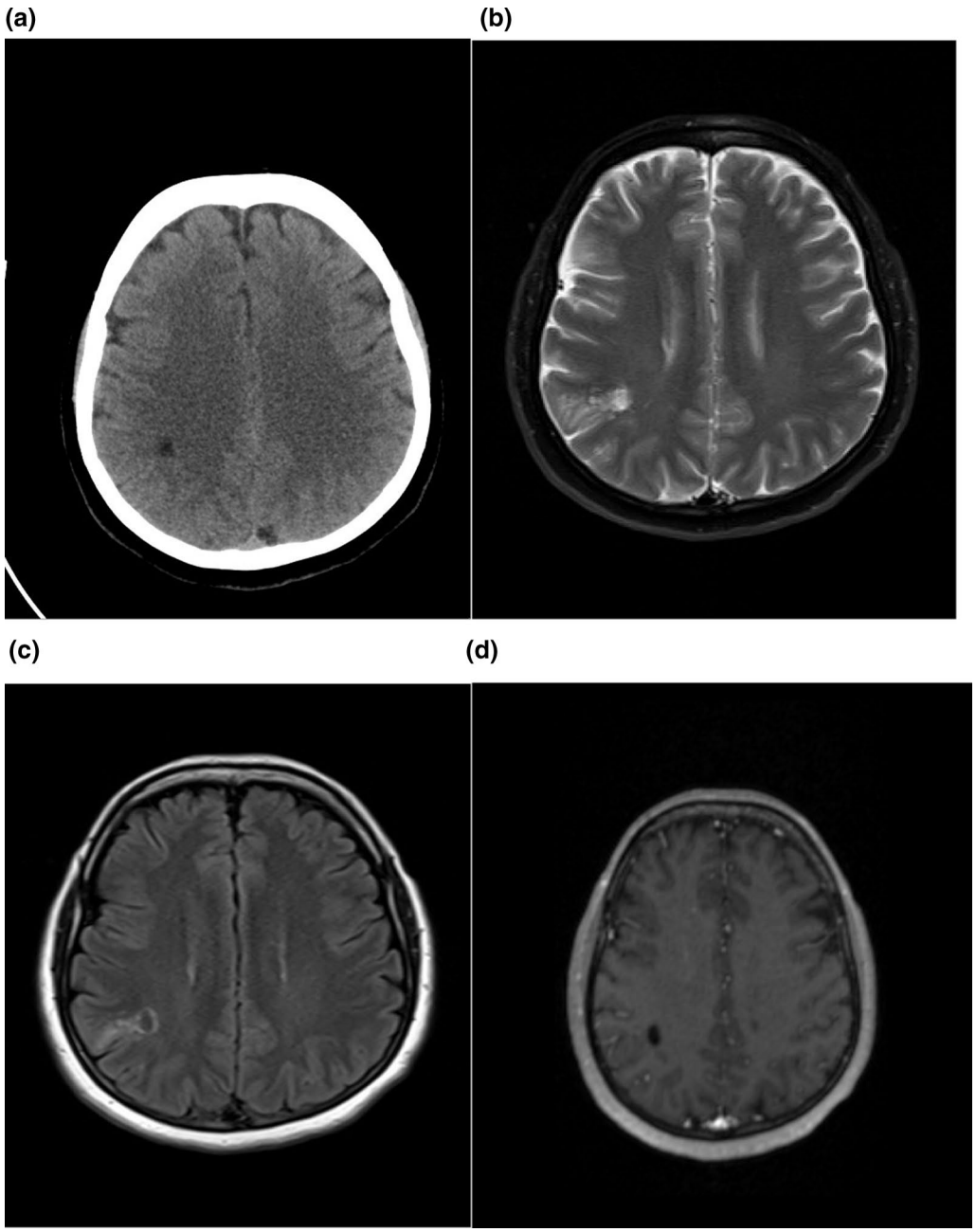

Fig. 1 MVNT with cystic component. Axial non-contrast CT image (**a**) shows low-density area in right parietal lobe white matter. Axial T2 (**b**) and FLAIR (**c**) demonstrate conglomerated hyperintense punctate lesions without contrast enhancement (**d**)

Discussion

MVNT is an unusual, neuronal lesion which was initially described by Huse et al. in 2013 and was later included in the 2016 updated WHO classification of tumors of the CNS [1]. The true epidemiology of these tumors is unknown. Before the introduction of this entity, it is highly likely that these lesions were underrecognized.

Conventional MRI is the primary imaging modality for the diagnosis of MVNT. MVNTs exhibit a characteristic appearance on MRI imaging. Only the imaging feature may be sufficient in the diagnosis. All lesions in our study contain nodules following the gyral contour and these nodules are hyperintense on T2 and T2/FLAIR sequences. They do not enhance and cause mass effect. Diffusion restriction and peripheral edema were absent

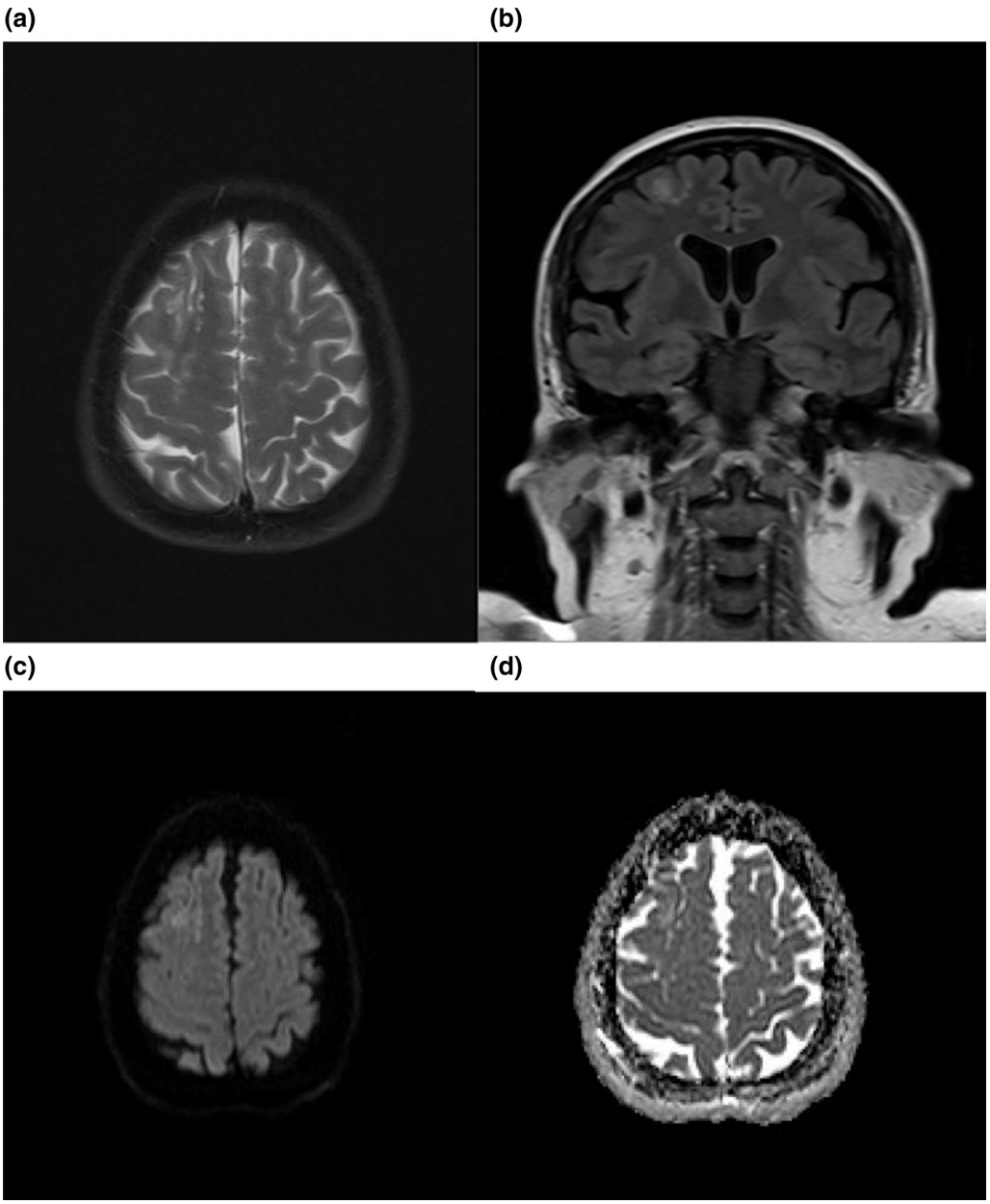

Fig. 2 Axial T2 (**a**) and coronal FLAIR (**b**) images reveal clustered hyperintense lesions in superficial white matter of right frontal lobe. Also, no diffusion restriction was observed (**c**, **d**)

in all cases. Nonsuppression pattern on FLAIR indicated high protein or solid component within vacuoles [3]. These MRI findings are in accordance with the cases in the radiology literature.

Because of its pathognomonic imaging appearance and asymptomatic nature, it was called a 'leave-me-alone' brain lesion by Nunes et al. [3]. Although seizures were expressed as the most common clinical finding by Huse et al. [1], in our study seizure was observed in only one patient. Headache was by far the most common complaint and was seen in seven of 11 patients. Nunes et al. also reported that headache (16/33) and seizure (8/33) were most common clinical findings. Similarly, in the Buffa et al. study seizure or equivalents (5/16), and non-focal headache (5/16), were the most commonly seen complaints [2, 3]. MVNTs are mostly incidental findings on MR imaging studies.

One case in our cohort was a 16-year-old female patient who had a temporary numbness on both hands mostly left. On MR imaging right parietooccipital lesion was demonstrated. No changes in follow-up imaging was observed after 6 months. The patient's symptoms disappeared at follow-up. Alizada et al. reported that a similar patient, who was a 17-year-old female, had numbness and MRI showed the lesion was hyperintense on T2-WI and FLAIR sequences without any contrast enhancement. After surgery, the lesion was diagnosed as MVNT histologically [4].

Alsufayan et al. reported that %20 of MVNT had cystic areas [5], and in our study similar MRI finding was shown in only one case (%9). Our two patients (%18) had cortical and subcortical white matter involvement on imaging which was described before by Alsufayan %57 of patients (17/30) and Lecler %7.8 of patients (5/64) [5, 6]. The white matter hyperintensity on T2/FLAIR adjacent to the bubble-like lesions was found in four of our cases (%36). This finding was reported by Nunes et al. at similar rate (%45) [3].

Based on the literature, we observed that CT was not recommended in the diagnosis or follow-up in these patient groups, and Yamaguchi et al. [7] reported that CT scans showed no abnormality. Nagaishi et al. [8] described CT findings as low-density lesions without calcification or cysts. In our retrospective study, lesion which was described in MRI scan could not be identified on the CT images of 5 patients (%83). One patient (%9) had a cystic area which could be recognized as a hypodense lesion on CT.

In our study, five lesions (%45) were located in the parietal lobe and 3 lesions (%27) affected more than 1 lobe (parietooccipital). When it was first described by Huse et al., 7 of 10 cases were confined to the temporal lobes [1]. Lecler et al. [6] reported that the MVNTs were mostly located in the frontal (20/64) and parietal lobe (26/64). The location of this entity seems to be non-discriminatory.

Surgery can be performed on symptomatic patients. In the literature, the symptoms have mostly regressed following resection without regrowth [1, 9]. None of our patients were undergone surgery, since the exact relationship between symptoms and lesions was not established.

The differential diagnosis of this lesion should include dysembryoplastic neuroepithelial tumor (DNET), enlarged periventricular space (PVS) and focal cortical dysplasia. The absence of enhancement, cortical involvement, mass effect and the presence of clustered appearance help to distinguish MVNT from DNET. In addition, DNET has a bright rim sign on FLAIR sequences and may scallop the inner table of the skull. Hyperintensity on FLAIR excludes the possibility of enlarged perivascular space. Focal cortical dysplasia is characterized by focal cortical thickening and blurring of white–gray matter junction. Some MVNTs may show a T2 FLAIR hyperintense radial like band as in focal cortical hyperplasia (transmantle sign), but the presence of cortical and gray-white matter junction involvement is consistent with focal cortical dysplasia [5].

However, there were some limitations to our study (shortness of mean follow-up duration, absence of histologic proof and MR spectroscopy images and variety of MRI devices used from different models), MVNT seems to be a static lesion, and no progression was detected in our study. According to our experience and point of view, follow-up with MRI is sufficient in the management of patients.

Conclusions

Imaging and clinical follow-up suggest that MVNT may exhibit indolent behavior. If the patients are asymptomatic, they can be followed up by imaging alone. For the symptomatic cases, surgery is the treatment option.

Abbreviations

MVNT: Multinodular and vacuolating neuronal tumor; CNS: Central nervous system; MRI: Magnetic resonance imaging; T2WI: T2-weighted image; FLAIR: Fluid-attenuated inversion recovery; DWI: Diffusion-weighted image; CT: Computed tomography; IV: Intravenous; DNET: Dysembryoplastic neuroepithelial tumor; PVS: Periventricular space.

Author contributions

Conceptualization: EB and FB; Data collection: MK and DA; Data analysis: EB, MK, DA and FB; Investigation: MK, DA and YB; Supervision: EB, YB and FB; Writing original draft: EB, MK and DA; Writing—review and editing: EB, MK, DA, YB and FB. All authors read and approved the final manuscript.

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Availability of data and materials

The datasets used during in this study are available from the corresponding author, and they may be provided when requested.

Declarations

Ethics approval and consent to participate

This study was approved by the Marmara University School of Medicine ethics committee (approval number: 09.2020.1018). The authors declare that the work described has been carried out in accordance with the Declaration of Helsinki of the World Medical Association revised in 2013 for experiments involving humans as well as in accordance with the EU Directive 2010/63/EU for animal experiments.

Consent for publication

This was not required because the data used were obtained retrospectively.

Competing interests

The author declares that he has no competing interests.

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References

1. Huse JT, Edgar M, Halliday J, Mikolaenko I, Lavi E, Rosenblum MK. Multinodular and vacuolating neuronal tumors of the cerebrum: 10 cases of a distinctive seizure-associated lesion. Brain Pathol. 2013;23(5):515–24.
2. Buffa GB, Chaves H, Serra MM, Stefanoff NI, Gagliardo AS, Yañez P. Multinodular and vacuolating neuronal tumor of the cerebrum (MVNT): a case series and review of the literature. J Neuroradiol. 2020;47(3):216–20.
3. Nunes RH, Hsu CC, Da Rocha AJ, Do Amaral LLF, Godoy LFS, Watkins TW, et al. Multinodular and vacuolating neuronal tumor of the cerebrum: a new "leave me alone" lesion with a characteristic imaging pattern. Am J Neuroradiol. 2017;38(10):1899–904.
4. Alizada O, Ayman T, Akgun MY, Sar M, Urganci N, Kemerdere R. Multinodular and vacuolating neuronal tumor of the cerebrum: two cases and review of the literature. Clin Neurol Neurosurg [Internet]. 2020. https://doi.org/10.1016/j.clineuro.2020.106149.
5. Alsufayan R, Alcaide-Leon P, de Tilly LN, Mandell DM, Krings T. Natural history of lesions with the MR imaging appearance of multinodular and vacuolating neuronal tumor. Neuroradiology. 2017;59(9):873–83.
6. Lecler A, Broquet V, Bailleux J, Carsin B, Adle-Biassette H, Baloglu S, et al. Advanced multiparametric magnetic resonance imaging of multinodular and vacuolating neuronal tumor. Eur J Neurol. 2020;27(8):1561–9.
7. Yamaguchi M, Komori T, Nakata Y, Yagishita A, Morino M, Isozaki E. Multinodular and vacuolating neuronal tumor affecting amygdala and hippocampus: a quasi-tumor? Pathol Int. 2016;66(1):34–41.
8. Nagaishi M, Yokoo H, Nobusawa S, Fujii Y, Sugiura Y, Suzuki R, et al. Localized overexpression of alpha-internexin within nodules in multinodular and vacuolating neuronal tumors. Neuropathology. 2015;35(6):561–8.
9. Fukushima S, Yoshida A, Narita Y, Arita H, Ohno M, Miyakita Y, et al. Multinodular and vacuolating neuronal tumor of the cerebrum. Brain Tumor Pathol. 2015;32(2):131–6.

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