Wydawnictwo UR 2022
ISSN 2544-1361 (online)
doi: 10.15584/ejcem.2022.1.18

http://www.ejcem.ur.edu.pl
European Journal of Clinical and Experimental Medicine
Eur J Clin Exp Med 2022; 20 (1): 126–128

CASUISTIC PAPER

Naciye Kış, Berrin Erok, Harun Kılıç, Hakan Önder

A benign entity – cerebral multinodular and vacuolating neuronal tumor

Department of Radiology, University of Medical Sciences, Prof Dr Cemil Tascıoglu City Hospital, İstanbul, Turkey

ABSTRACT

Introduction and aim. Multinodular and vacuolating neuronal tumor (MVNT) of the cerebrum is a rare benign, mixed glial/neuronal lesion which has been included in the recent (2016) World Health Organization (WHO) Classification of the central nervous system tumors.Most of the reported cases are remarkable with adult onset seizure in the literature.They can also be found incidentally in nonepilepsy patients with or without headache We aimed to present this unique entity with its typical magnetic resonance imaging (MRI) features.

Description of the case. A 21-year old man presented with complaint of headache that increased in frequency within the last few months.No relevant seizure or any other signs of note.He was diagnosed with MVNT by imaging andstarted to be followed-up.The repeat MRI 6 months later showed no interval changes.

Conclusion. Clinicians should be aware of that it is a do not touch lesion in asymptomatic patients with no need for biopsy or surgery and follow up imaging is sufficient when presented with the typical MRI manifestations. Surgical resection may be required for seizure control and was reported in few cases with no tumoral regrowth in the literature.

Keywords. do not touch brain tumors, magnetic resonance imaging, multinodular and vacuolating neuronal tumor

Introduction

Multinodular and vacuolating neuronal tumor (MVNT) of the cerebrum is a rare benign, mixed glial/neuronal lesion which has been included in the recent (2016) World Health Organization (WHO) Classification of the central nervous system tumors (WHO grade I).[1] However, pathological characteristics are thought to be more closer to a developmental malformation.[2] In addition, lesions with identical imaging features have been described in the cerebellum but have not been histologically confirmed and have, therefore, prudently been named as multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS).[3] Most of the reported cases are remarkable with adult onset seizure in the literature.They can also be found incidentally in nonepilepsy patients with or without headache.[4,5]

Aim

We aimed to present this unique entity with its typical MRI features detected in a 21-year old man.

Description of the case

A 21-year old male patient presented with complaint of headache with increased frequency within the last few months. No relevant seizure or any other signs of note. Brain MRI revealed innumerable, small, well-defined, coalescent nodules extending from the juxtacortical white matter to the juxtaventricular area of the left frontal lobe sparing the overlying cortex. They were T2w/Flair hyper-

Corresponding author: Berrin Erok, e-mail: drberrinerok@hotmail.com

Received: 18.11.2021 / Revised: 17.12.2021 / Accepted: 30.12.2021 / Published: 30.03.2022

Kış N, Erok B, Kılıç H, Önder H. *A benign entity – cerebral multinodular and vacuolating neuronal tumor.* Eur J Clin Exp Med. 2022;20(1):126–128. doi: 10.15584/ejcem.2022.1.18

intense without restricted diffusion and blooming on susceptibility weighted imaging (SWI) (Fig.1).

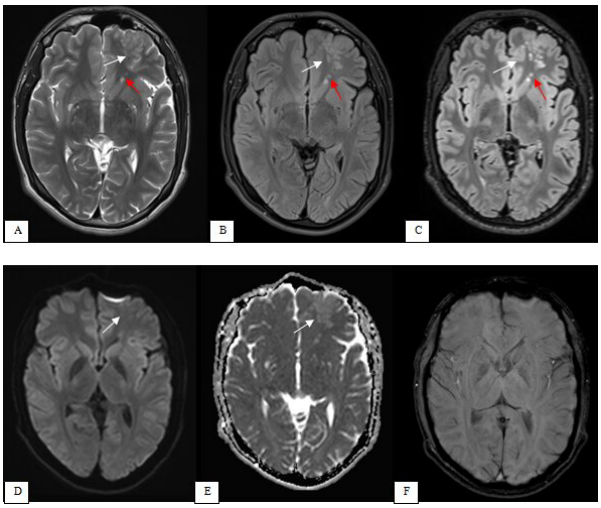

Fig. 1. Axial T2w (A), Flair (B), Flair 3D reconstruction (C) images showing a left frontal intraaxial subcortical lesion consisting of multiple, well marginated coalescent hyperintense nodules (A,B,C, white arrows). Note the lower signal intensity of the lesions in comparison to CSF on T2w images (A) and the absence of surrounding white matter hyperintensity on T2w/Flair images (A,B,C). Axial DWI (D) and ADC mapping (E) showing no restricted diffusion (D,E, arrows). Axial SWI (F) image shows no blooming. Note the superficial subcortical localization of the lesions following the gyral contour with extention to the deeper white matter areas (A,B,C, red arrow)

The lesions were isointense to gray matter on T1w images with no pathological enhancement following contrast administration and no increased perfusion on cerebral blood flow (CBV) map on perfusion imaging (Fig. 2).

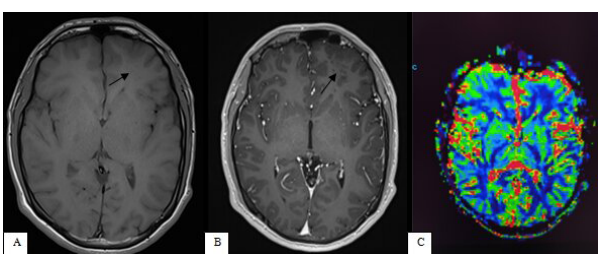

Fig. 2. Axial T1w (A) and postcontrast T1w (B) images showing the lesions are isointense to the gray matter (A, black arrow) with no pathological enhancement (B, black arrow). No elevated CBV was depicted on CBV perfusion image (C)

MR spectroscopy showed no obvious abnormal peaks (Fig. 3). With the typical radiological manifestations MNVT was considered and he was decided to be followed up without surgery. The repeat MRI 6 months later showed no interval changes.

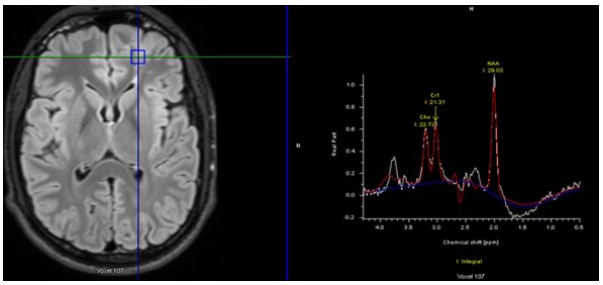

Fig. 3. The MRI Spectroscopy image shows no abnormal peaks in association with the lesion

Discussion

MVNT of cerebrum was first described with a case series of 10 patients by Huse et al. as non-neurocytic, purely neuronal tumors most commonly located in the temporal lobes (7 cases of 10) with presentation of adult onset seizure.[5] In the study of Nunes et al. that retrospectively evaluated 33 MVNT cases with compatible imaging features, 4 of which proven by biopsy and the remaining without interval change for at least 24 months follow up, the mean age of diagnosis was found to be 39 years with slight female predominance 1.4/1. In this study, 9 (27%) lesions were in the parietal lobe, 8 (24%) were in the frontal lobe, 6 (18%) were in the temporal lobe and 2 (4%) lesions were located in the occipital lobe with 8 (24%) involving more than one lobes.[6] On radiological imaging, this entity is characterized with a cluster of supratentorial, intraaxial, multiple rounded/ovoid nodules without mass effect located principally in the subcortical/juxtacortical white matter with usually normal overlying cortex.[4,6] However, there are also some reports showing that the overlying cortex may also be involved.[5,6] In our patient the lesions were also located predominantly in this characteristic location but also had extention to the deeper white matter areas as opposed to the more confined lesions reported in the literature, usually located within the deep cortical ribbon and the superficial white matter. The frequent presentation with late onset epilepsy can be explained with the juxtacortical location, but incidental detection is also common. Our patient was presented with non-focal headache as reported in one case having also left frontal MVNT in the case series of Nunes et al.[6] The signal characteristics include T2w hyperintensity but less than that of CSF with no suppression on Flair images. In our patient there was no surrounding white matter hyperintensity on T2W/Flair images as commonly reported in the literature but, confluent T2/FLAIR hyperintensity sparing the cortex may be present.[6] DWI shows increased signal intensity with increased corresponding ADC value in relation with T2-shine through effect

rather than true restricted diffusion. In addition, there is no blooming on SWI images. Following contrast administration they show no enhancement as in our case, but some faint focus of enhancement was demonstrated in a few cases.[5,6] In the qualitative evaluation of the CBV map, there was no increased perfusion compared to the contrlateral normal parenchyma. On MRS evaluation although the contrlateral normal parenchyma was not sampled there was no obvious pathological peaks in our patient. Furthermore, in some studies in the literature mildly increased choline/creatine ratio was also demonstrated.[4]

Although MVNT of cerebrum is considered as a unique cytoarchitectural pattern of gangliocytoma (WHO grade I), it is very different from gangliocytomas on imaging which had both cystic and enhancing solid components with cortical involvement indistinguihable from ganglioglioma.[1,7] The imaging differentials may include dilated perivascular spaces which are usually more elongated along vessel long axis and are differentiated with attenuation on Flair images. Dysembryoplastic neuroepethelial tumor (DNET) may also be considered with its peripheral location frequently in the frontotemporal lobes.However, DNET is generally cortical rather than subcortical and is associated with gyral expansion.In addition, there is usually some suppression on Flair images with frequent well defined bright FLAIR rim.[8] Focal cortical dysplasia may also have T2w hyperintensity deep to the cortex but is associated with overlying cortical thickening.[9]

In the management of MVNT in asymptomatic patients follow up radiological imaging is sufficient without need for biopsy. Surgical resection may be required for seizure control and was reported in few cases with no tumoral regrowth in the literature.[5,6,10]

Conclusion

MVNT is a benign entity that may be detected in patients presenting with seizure or may be found incidentally in patients with or without headache. Clinicians should be aware of that this is a don't touch lesion in asymptomatic patients with no need for biopsy or surgery when presented with the typical MRI manifestations.

Declarations

Funding

This research received no external funding.

Author contributions

Conceptualization, N.K. and B.E.; Methodology, N.K., B.E. and H.Ö.; Formal Analysis, B.E. and N.K.; Investigation, B.E., H.K. and N.K.; Writing – Original Draft Preparation, B.E. and N.K.; Writing – Review & Editing, B.E., N.K. and H.K.; Supervision, HÖ

Conflicts of interest

The authors declare no conflict of interest.

Data availability

The data sets used and/or analysed during the current study are available from the corresponding author upon reasonable request.

Ethics approval

Not applicable.

References

1. Louis DN, Perry A, Reifenberger G, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol*. 2016;131(6):803-820.
2. Thom M, Liu J, Bongaarts A, et al. Multinodular and vacuolating neuronal tumors in epilepsy: dysplasia or neoplasia? *Brain Pathology*. 2018;28(2):155.
3. Lecler A, Bailleux J, Carsin B, et al. Multinodular and Vacuolating Posterior Fossa Lesions of Unknown Significance. *AJNR Am J Neuroradiol*. 2019;40(10):1689-1694.
4. Makrakis D, Veneris S, Papadaki E. Multinodular and vacuolating neuronal tumor incidentally discovered in a young man: Conventional and advanced MRI features. *Radiology Case Reports*. 2018;13(5):960-964.
5. Huse J, Edgar M, Halliday J, Mikolaenko I, Lavi E, Rosenblum M. Multinodular and vacuolating neuronal tumors of the cerebrum: 10 cases of a distinctive seizure-associated lesion. *Brain Pathol.* 2013;23(5):515-524.
6. Nunes RH, Hsu CC, da Rocha AJ, et al. Multinodular and Vacuolating Neuronal Tumor of the Cerebrum: A New "Leave Me Alone" Lesion with a Characteristic Imaging Pattern. *AJNR A. J Neuroradiol.* 2017;38(10):1899.
7. Louis DN, Perry A, Wesseling P, et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. *Neuro Oncol.* 2021;23(8):1231-1251.
8. Parmar HA, Hawkins C, Ozelame R, Chuang S, Rutka J, Blaser S. Fluid-attenuated inversion recovery ring sign as a marker of dysembryoplastic neuroepithelial tumors. *J Comput Assist Tomogr.* 2007;31(3):348-353.
9. Kabat J, Król P. Focal cortical dysplasia - review. *Pol J Radiol*. 2012;77(2):35-43.
10. Fukushima S, Yoshida A, Narita Y, et al. Multinodular and vacuolating neuronal tumor of the cerebrum. *Brain Tumor Pathol*. 2015;32(2):131-136.