Ref: Ro J Neurol. 2023;22(3)
DOI: 10.37897/RJN.2023.3.6

Pathognomonic neuroimaging of a Multinodular and Vacuolating Neuronal Tumor of the Cerebrum incidentally discovered in an 8-year old female. A case report

Luigi Di Lorenzo[1], Serena Schipani[2], Marina Minichiello[3], Franco Carmine Muccio[2]

[1]Mebic Consorzio, San Raffaele Cassino, Italy
[2]Neuro-Radiology Unit, San Pio Hospital, Benevento, Italy
[3]Neurosurgery Unit, San Pio Hospital, Benevento, Italy

ABSTRACT

Background. The WHO 2016 classification of tumors of the CNS includes Multinodular and Vacuolating Neuronal Tumors (MVNT) as a benign ganglion cell tumor, possibly associated with seizures. MTVN pathologic hallmark to be consisting of a "subcortical cluster of nodular lesions located on the inner surface of an otherwise normal appearing cortex, within the deep cortical ribbon and superficial subcortical white matter". MVNT is mostly present in adults.

Case report. In this case, we report pathognomonic pics of MVNT in a noticeably rare young patient, where it was unclear whether MVNTs should be considered a true neoplasm or malformative lesion, that typically occurs causing seizures

Conclusion. It confirms nowadays that in clinical practice the MRI can help neurosurgeons to confirm the clinical non-surgical diagnosis even if in the lack of more common symptoms such as seizures.

Keywords: Multinodular and Vacuolating Neuronal Tumors, brain tumor MRI, children brain tumor MRI

INTRODUCTION

World Health Organization's (WHO) 2016 classification of central nervous system (CNS) tumors encompasses Multinodular and Vacuolating Neuronal Tumors (MVNT) as benign ganglion cell tumors potentially associated with seizures [1]. In a case series study by Geka et al. in 2021, MVNT pathologic features were defined as a "subcortical cluster of nodular lesions located on the inner surface of an otherwise normal-appearing cortex, within the deep cortical ribbon and superficial subcortical white matter" [2]. Hyperintense nodular lesions on FLAIR and FSE T2-weighted images were observed at 1.5 Tesla MR imaging, devoid of edema or mass effect. Nevertheless, despite the recent classification by WHO, MVNT retains its status as a clinically and pathologically ambiguous entity, with an uncertain categorization. The authors reflect, stating, "It remains unclear whether MVNTs should be considered a true neoplasm or a malformation lesion" [2]. Furthermore, while the epidemiologic data are incomplete, it is acknowledged that MVNT predominantly affects adults and manifests in the cerebral subcortical region, often being linked with seizures or seizure-like manifestations [2,3]. A comprehensive review in 2020 affirms the prevalence of MVNT primarily among adults and underscores its pathologic hallmark of a "subcortical cluster of nodular lesions located on the subcortical white matter" [3]. This extensive review outlines a series of suspected MVNT cases observed within their institution. All patients underwent MRI scans on either 1.5 or 3 Tesla scanners, utilizing sequences including T1, T2, GRE/SWI, T2 FLAIR, and DWI. The imaging substantiated the pathologic hallmark of MVNT – a subcortical cluster of nodular lesions situated on the subcortical white matter [3].

Corresponding author:
Luigi Di Lorenzo
E-mail: drluigidilorenzo@gmail.com

Article History:
Received: 14 September 2023
Accepted: 29 September 2023

While a reported substantial difference exists between 1.5T and 3T field strengths [4], both systems offer compelling imaging capabilities, rendering them viable options. The prevalent use of 1.5T systems underscores their role in delivering safe and high-quality diagnostic images [4-6]. In this case report, we present pathognomonic images acquired using the current 1.5-tesla magnetic resonance (MR) technology – a cornerstone of contemporary diagnostic neuroradiology. These images were captured in an unusually young patient without a history of seizures, a rarity in MVNT cases [2,3].

CASE REPORT

Our case involves an eight-year-old female patient who experienced an accidental trauma approximately about two year ago. The eight-year-old patient, devoid of seizures or focal neurological deficits, had a meticulous monitoring after MVNT diagnosis without epileptic symptoms, and presented with minor symptoms such as persistent headaches. Following the procurement of appropriate consent for examination and data processing, safeguarding privacy, the patient underwent an MRI due to recurring headaches. Subsequent imaging unveiled a lesion within her right pre-rolandic gyrus. The lesion comprised clusters of hyperintense round or ovoid nodules on FLAIR T2 and FSE T2-weighted images, positioned in the subcortical white matter alongside a normal-appearing adjacent cortex. Based on its characteristics and stability, it was identified as an instance of MVNT. No history of epilepsy was reported by the patient. Figure 1 displays our pathognomonic findings, captured using a 1.5-Tesla superconducting Siemens "Aera" system, exhibiting characteristic MVNT lesions that remained unchanged during a follow-up period of 17 months. Diffusion-weighted imaging (DWI) was conducted utilizing an echo-planar spin-echo technique with b values of 0 and 1,000 s/mm^2, revealing an increased apparent diffusion coefficient (ADC) value of 0.861 × 10^{-3} mm^2/s, confirming the solid nature of the tumor.

DISCUSSION

MVNT, a low-grade neuronal tumor, predominantly impacts the cerebral hemispheres, with an affinity for the temporal lobes. Key radiological features include the location and MRI appearance. Typically situated within the superficial cortical layers of the cerebral hemispheres, especially the temporal lobes, MVNT often exhibits bilateral and multifocal nodular and vacuolated areas. In T1-weighted images, these lesions tend to appear isointense to slightly hypointense compared to normal brain tissue. Additionally, on T2-weighted and FLAIR images, heightened water content is evident, while contrast-enhanced T1-weighted images may exhibit subtle to no contrast enhancement, distinguishing MVNT from more aggressive tumors. Notably, vacuoles within lesions may appear hypointense on T2-weighted images due to their fluid-filled nature. MVNTs are typically well-circumscribed, often devoid of significant mass effect or peritumoral edema, distinguishing them from more aggressive neoplasms. Moreover, the absence of significant calcifications or hemorrhagic features is a common characteristic of MVNT. When considering the radiological differential diagnosis for MVNT, it is crucial to differentiate from entities with similar imaging features. Conditions such as Focal Cortical Dysplasia, certain low-grade gliomas like pilocytic astrocytomas or gangliogliomas, and DNET, a type of low-grade neuronal tumor, warrant careful consideration. Inflammatory lesions or granulomatous infections may also present with nodular

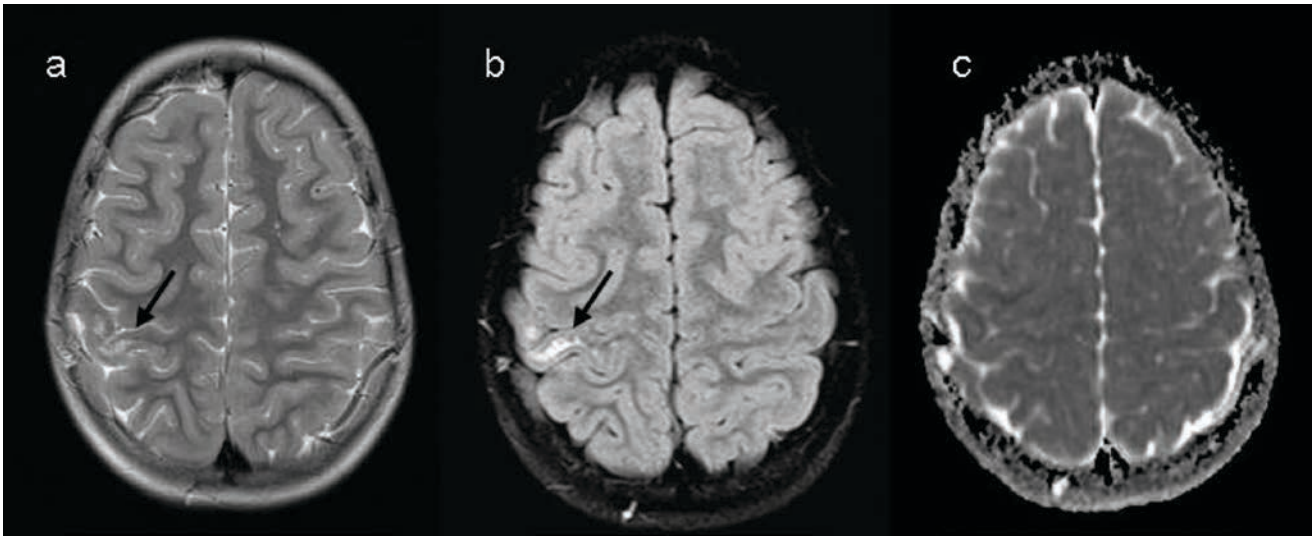

FIGURE 1. Lesion in her right pre rolandic gyrus, consisting in clusters of multiple hyperintense round or ovoid FLAIR T2 and FSE T2-weighted

or vacuolated imaging appearances. Accurate diagnosis necessitates an amalgamation of radiological findings, clinical presentation, and pathological evaluation. Given the rarity of MVNT, an interdisciplinary approach involving neuroradiologists, neuropathologists, and neurosurgeons is pivotal for precise diagnosis and guiding treatment choices. Additionally, consultation of contemporary medical literature and adherence to current guidelines ensures accurate understanding of MVNT and its imaging features, facilitated by 1.5 tesla MRI technology. While the prospect of ultra-high field MRI technology, operating at 7-10 teslas, holds promise for enhanced diagnostic resolution [7], the prevailing 1.5-Tesla and 3-Tesla systems remain indispensable for current clinical practice. These technologies offer substantial improvements over older 0.3 T units. Moreover, ongoing developments in high field MRI systems are poised to revolutionize diagnostic accuracy, particularly in the intricate anatomical structures of the brain stem, bolstering the potential of MRI in diagnostic and clinical applications [8].

CONCLUSION

In conclusion, this paper presents a distinctive case of MVNT in a young patient, underscoring its atypical manifestation in early adulthood and the potential association with epilepsy. Collaborative efforts among neuroradiologists, neuropathologists, and specialists play a pivotal role in ensuring accurate diagnoses. The case of an 8-year-old patient, devoid of seizures or focal neurological deficits, further highlights the diverse clinical spectrum of MVNT. While surgical intervention proves curative in cases with epileptic symptoms, meticulous monitoring emerges as a valuable strategy for managing MVNT in pediatric patients, even when presented with minor symptoms such as persistent headaches. As understanding of MVNT continues to evolve, its recognition as a distinct clinical entity underscores the critical role of advanced imaging techniques, like 1.5-tesla MR, in guiding clinical decisions and enhancing patient care.

Compliance with ethical standards

The study treatments were conducted according to ethical principles and was responsive to all applicable guidelines for good clinical practice

Funding: None

Conflict of interest: None

Informed consent: Obtained

Author contribution:

D.L. and FCM conceived of the presented idea. D.L. developed the theory S.S. and MM followed patient during practical sessions and verified methods. FCM did MRI and supervised the findings of this work helping translation and text revision with DL.

REFERENCES

1. Louis DN, Perry A, Reifenberger G, von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 World Health Organization Classification of Tumors of the Central Nervous System: a summary. *Acta Neuropathol.* 2016 Jun;131(6):803-20. doi: 10.1007/s00401-016-1545-1.
2. Okamoto K, Natsumeda M, Oishi M, Fujii Y. [Multinodular and Vacuolating Neuronal Tumor of the Cerebrum (MVNT)]. *No Shinkei Geka.* 2021 Mar;49(2):383-387. Japanese. doi: 10.11477/mf.1436204402.
3. 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 May;47(3):216-20. doi: 10.1016/j.neurad.2019.05.010. Epub 2019 Jun 20. PMID: 31229580.
4. Huang BY, Castillo M. Neurovascular imaging at 1.5 tesla versus 3.0 tesla. *Magn Reson Imaging Clin N Am.* 2009 Feb;17(1):29-46. doi: 10.1016/j.mric.2008.12.005. PMID: 19364598
5. Evans E. The Pros and Cons of 1.5T V. 3T MRI: One Size Does Not Fit All 14 feb 2018. KMG news (Kings Medical Group) freely retrieved online at. https://www.linkedin.com/pulse/pros-cons-15t-v-3t-mri-one-size-does-fit-all-eric-evans.
6. Potchen MJ, Kampondeni SD, Seydel KB, Haacke EM, Sinyangwe SS, Mwenechanya M, et al. 1.5 Tesla Magnetic Resonance Imaging to Investigate Potential Etiologies of Brain Swelling in Pediatric Cerebral Malaria. *Am J Trop Med Hyg.* 2018 Feb;98(2):497-504. doi: 10.4269/ajtmh.17-0309. Epub 2018 Jan 4. PMID: 29313473; PMCID: PMC5929182.
7. Budinger TF, Bird MD, Frydman L, Long JR, Mareci TH, Rooney WD, et al. Toward 20 T magnetic resonance for human brain studies: opportunities for discovery and neuroscience rationale. *MAGMA.* 2016 Jun;29(3):617-39. doi: 10.1007/s10334-016-0561-4. Epub 2016 May 18. PMID: 27194154; PMCID: PMC5538368.
8. Runge VM, Case RS, Sonnier HL. Advances in clinical 3-tesla neuroimaging. *Invest Radiol.* 2006 Feb;41(2):63-7. doi: 10.1097/01.rli.0000185877.91066.01. PMID: 16428974