CASE REPORT

Multinodular and vacuolating neuronal tumor of the cerebrum: three case reports

Tumor neuronal multinodular e vacuolizante do cérebro: relato de três casos

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ABSTRACT

Multinodular and vacuolating neuronal tumor (MVNT) of the cerebrum is a seizure-associated rare lesion that currently appears to be a malformative lesion or hamartoma rather than a low-grade neoplastic lesion. MVNT pathognomonic magnetic resonance imaging (MRI) features allow for diagnostic accuracy in the differential diagnosis. Lesions are usually asymptomatic, nonprogressive and incidentally found requiring only imaging monitoring over time. Although uncommon, it is important to be familiar with MVNT clinical presentation and key imaging features. We here present a series of three cases with imaging findings similar to those described in the literature. These patients were followed with serial neuroimaging.

Keywords: Magnetic resonance imaging. Epilepsy. Central nervous system. Brain neoplasms.

RESUMO

Os tumores neuronais multinodulares e vacuolizantes (MVNT) do cérebro são lesões raras associadas a crises epilépticas. Acredita-se atualmente que se situem mais provavelmente entre as entidades malformativas ou hamartomas que entre as neoplasias de baixo grau. Apresentam características patognomônicas à Ressonância Magnética Nuclear (RMN) o que os distinguem com grande precisão de seus potenciais diagnósticos diferenciais. Pelo fato de permanecerem estáveis ao longo do tempo e a maioria serem assintomáticas ou achados incidentais, adota-se nestes casos uma postura apenas de vigilância radiológica. Embora incomuns é importante a familiaridade com sua apresentação clínica e os seus característicos aspectos radiológicos. Na série aqui apresentada os achados de imagem foram idênticos aos descritos na literatura para os MVNT's e a conduta final foi a de supervisão seriada com neuroimagem cerebral.

Palavras-chave: Imagem por ressonância magnética. Epilepsia. Sistema nervoso central. Neoplasias encefálicas.

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INTRODUCTION

Multinodular and vacuolating neuronal tumor (MVNT) of the cerebrum was first described in the neuropathology literature (Huse et al.) in 2013[1] in a small case series as being a purely neuronal lesion with grade I benign behavior according to the World Health Organization (WHO) classification.[1,2] The lesion is commonly found in the temporal lobe and is associated with seizures affecting both sexes equally.[1] In a recent historic cohort study, approximately 19% of patients presented epileptic seizures, a large proportion had nonspecific neurological complaints and most were asymptomatic, incidentally discovered cases and were not biopsied or resected.[3] MVNT is included in the 2016 WHO Tumor Classification as a lesion with a unique new cytoarchitectural pattern related to gangliocytoma.[2] Histopathologically, it consists of well-defined, coalescent and discrete nodules of non-neurocytic neuroepithelial cells demonstrating a unique membrane-bound appearance and coarse vacuolization of the neuronal cytoplasm, arranged within the deep cortical ribbon, juxtacortical area and superficial subcortical white matter.[1,4] The nodules form curvilinear groupings within the cortex with vacuolization of neuroparenchyma showing a cystic pattern.[3] Immunohistochemical studies reveal immunopositive dysplastic cells that are indicative of an earlier stage of neuronal development (positive for HuC/HuD and OLIG2), which suggests a malformative lesion.[3] Markers of more advanced neuronal maturation, including synaptophysin, neurofilament and neuronal nuclear antigen (NeuN), are either negative or weakly positive. Genetic testing has not demonstrated a particular type of tumor.[1,2] It is histologically distinct from dysembryoplastic neuroepithelial tumor (DNET) and ganglioglioma.[1,5] Magnetic resonance imaging (MRI) is the method of choice for assessing MVNT lesions, and computed tomography (CT) scans may not show abnormalities. MRI usually assesses lesion location, size, pattern, and signal intensity on T1, T2, FLAIR and DWI. Characteristic imaging features include multiple small cystic or solid lesions with discrete, clustered and/or coalescent arrangement of variable sizes (1–5 mm).[5,6] The typical foci have a round to ovoid shape and are located in the cortical ribbon and juxtacortical white matter. Nodular foci show T1 hypointensity and T2 hyperintensity with no suppression on T2 FLAIR, no diffusion restriction, no mass effect, no vasogenic edema, no visible calcification and rare contrast enhancement.[3,4] In all case series reported in the literature, MVNT lesions were of supratentorial location, deep inside the brain sulci. Potential differential diagnoses on brain imaging include DNET, focal cortical dysplasia, nodular neuronal heterotopias, focal cerebritis, gliomas, hamartomas and enlarged perivascular spaces. However, MVNT imaging findings are pathognomonic, which allows for diagnostic accuracy in most cases.[3,4]

METHODS

This is a descriptive case report study comprising three cases of patients presenting with diagnostic imaging features consistent with probable MVNT. We here present their clinical characteristics and key pathology and imaging features, and discuss these findings supported by a literature review. Ethics committee approval has been obtained for this report at Hospital Universitário Walter Cantídio (HUWC).

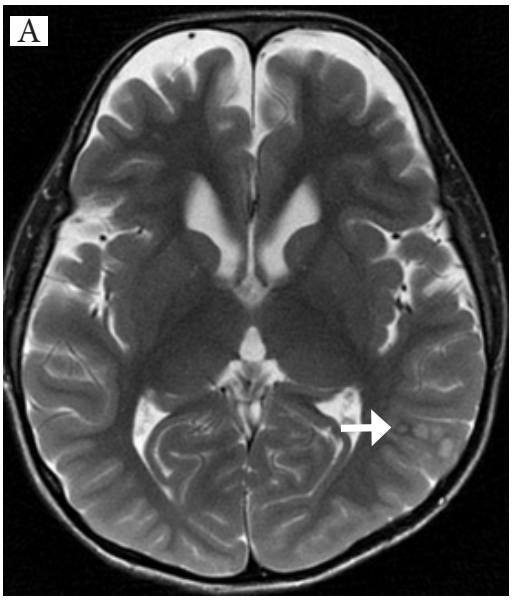
CASE REPORTS

Patient #1: 8-year-old female, presenting with spastic cerebral palsy and neuropsychomotor development delay with no history of epileptic seizures or any other prior neurological complications. An MRI study revealed subcortical lesions in the left temporal lobe consisting of multiple nodule groupings, hyperintense on FLAIR and T2, without DWI hyperintensity or gadolinium enhancement (Figure 1).

Patient #2: 25-year-old male with a history of epilepsy and no other relevant symptoms or clinical findings. MRI was performed and revealed a lesion in the parasagittal area of the right frontal lobe consisting of multiple coalescent nodules in the deep cortical ribbon, hyperintense on T2WI and FLAIR and no gadolinium enhancement (Figure 2).

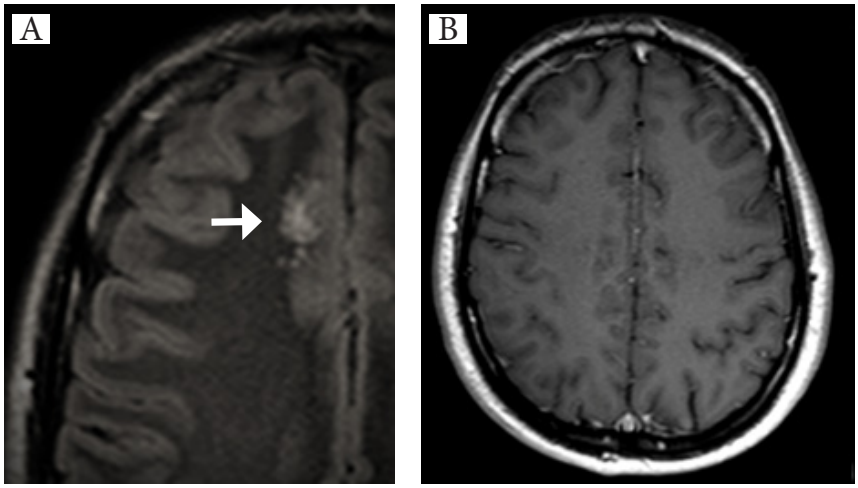
Patient #3: 52-year-old male who underwent brain MRI for non-focal chronic headaches. He had no other relevant symptoms, signs, medical, surgical nor drug history of note. Imaging showed extensive supratentorial, intra-axial left temporoparietal lesions consisting of groupings of multiple nodules with ill-defined margins located in the juxtacortical ribbon and superficial subcortical white matter following the gyral contour with high signal intensity on T2WI and short tau inversion recovery (STIR) imaging and no DWI hyperintensity or gadolinium enhancement (Figure 3).

Figure 1. Patient #1: 8-year-old female, asymptomatic with development delay.



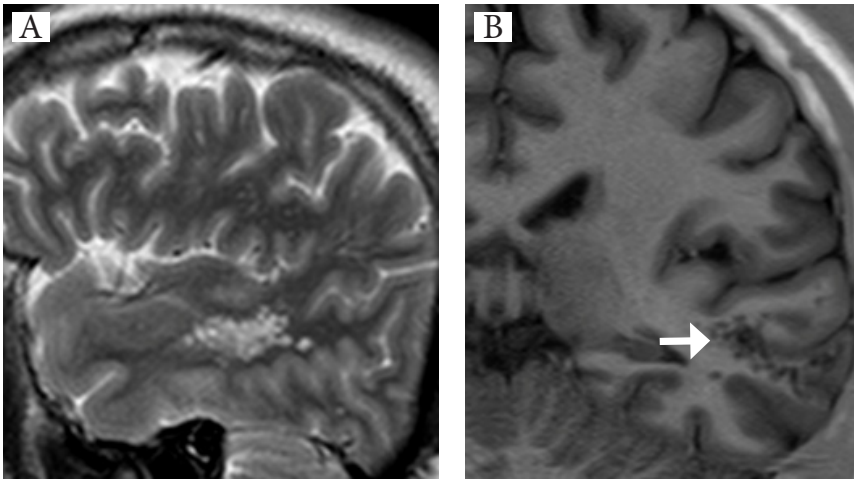
Key imaging features are subcortical lesions in the temporal lobe consisting of multiple nodule groupings, hyperintense on T2WI (A). Evidence of groupings of multiple coalescent intralesional and perilesional nodules are better depicted.

Figure 2. Patient #2: 25-year-old male presenting with seizures.



Hyperintense lesion in the right frontal parasagittal lobe consisting of multiple coalescent nodules in the deep cortical ribbon, hyperintense on FLAIR (A) and no gadolinium enhancement (B).

Figure 3. Patient #3: 52-year-old male with nonfocal headaches.



Characteristic imaging features are high signal intensity on sagittal T2WI (A) and hypointensity in T1WI (B); These selected images of a presumed MVNT demonstrate extensive supratentorial, intra-axial left temporoparietal hyperintense lesions consisting of groupings of multiple nodules with ill-defined margins located in the juxtacortical ribbon and superficial subcortical white matter following the gyral contour, without mass effect or vasogenic edema.

DISCUSSION

MVNT of the cerebrum is included in the 2016 WHO classification of tumors of the central nervous system as an entity with a distinct pattern of cytoarchitecture.[1,2] However, its histological features are indeterminate and MVNT characterization is difficult because it shares features that are common to both neoplastic and malformative lesions — though studies suggest malformation.[2,3] Key imaging features are characteristic, showing multiple small nodules grouped in subcortical or juxtacortical areas, T2WI and FLAIR hyperintensity, no diffusion restriction and no gadolinium enhancement and allows to rule out other diagnoses in the differential diagnosis.[1,3] In our case series, two of the three cases were incidentally discovered. Two of the three patients had lesions located in the temporal lobe, but all showed homogeneous imaging patterns identical to those features described in MVNT cases[3,4]: multiple small cystic or solid lesions in the juxtacortical area and superficial subcortical white matter, T2 hyperintensity, no signal suppression on T2 FLAIR, no diffusion restriction and no contrast enhancement.[5] (Figures 1, 2 and 3). These characteristic imaging features helped support a diagnosis of presumed MVNT. Our cases were discussed with both radiology and neurology teams at our center and surveillance neuroimaging was considered an adequate management since these are nonprogressive lesions with a benign behavior over time. The largest case series in the literature reported a 24-month patient follow-up and most lesions did not require biopsy or resection.[3,5]

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