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Case Report

Multinodular and vacuolating neuronal tumor incidentally discovered in a young man: Conventional and advanced MRI features

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ABSTRACT

Multinodular and Vacuolating Neuronal Tumor (MVNT) has been included in the most recent (2016) *World Health Organization Classification of Tumors of the Central Nervous System* as unique cytoarchitectural pattern of gangliocytoma. We present a case of a MVNT incidentally discovered in a 22-year old male, who presented with seizures after a head injury. Conventional MRI revealed a left parietal lesion with characteristic tiny, coalescent, well-defined, non-enhancing nodules, located in the juxtacortical white matter with partial involvement of an otherwise normal adjacent cortex and characterized by slight relative increase of the cerebral blood volume (CBV), compared to the contralateral white matter (lesional CBV/contralateral CBV = 1.112) and mild increase of choline and reduction of NAA (lesional choline/creatine ratio =1.36 and choline/NAA ratio=0.77, compared to 0.87 and 0.51, respectively). The patient fully responded to treatment with phenytoin and a follow-up MRI, six months later, showed the lesion without any substantial difference.

Introduction

Multinodular and vacuolating neuronal tumor (MVNT) has been included in the most recent (2016) *World Health Organization Classification of Tumors of the Central Nervous System* as a unique cytoarchitectural pattern of gangliocytoma, although it remains unclear whether it is a true neoplastic process or a dysplastic hamartomatous/malformative lesion [1–11]. Magnetic resonance imaging (MRI) findings are rather characteristic showing multiple, tiny, discrete or coalescent, sharply marginated, round or ovoid, nonenhancing nodules, located within the deep cortical ribbon and the superficial white matter, without remarkable mass effect [1–11]. We present a case of a patient with a MVNT tumor discovered incidentally after head injury and discuss its morphologic, diffusion, hemodynamic, and metabolic properties.

Competing Interests: The authors have declared that no competing interests exist.

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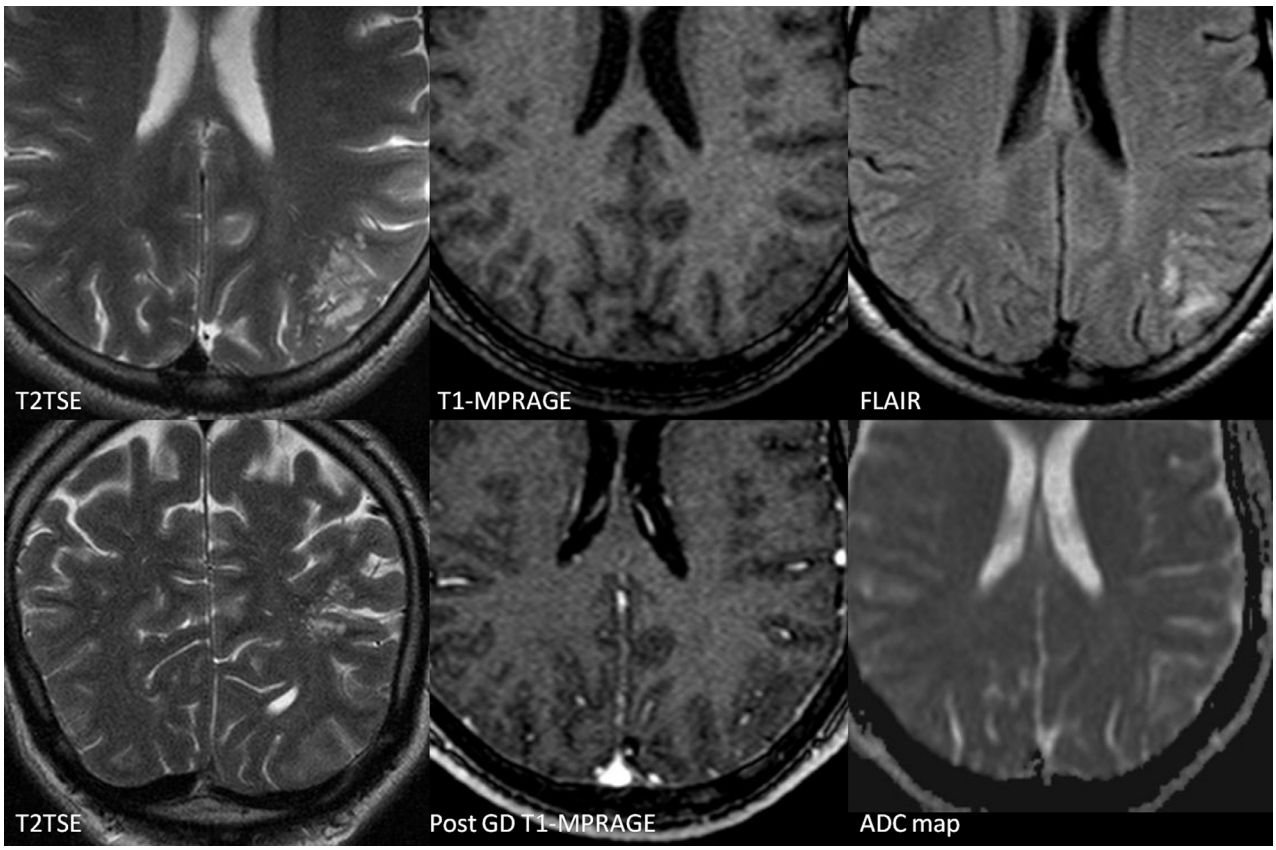

Fig. 1 – Conventional MRI revealed a left juxtacortical parietal lesion, with partial involvement of the adjacent cortex, which comprised of tiny, coalescent, well-defined nodules, hyperintense on T2 and FLAIR and hypointense on T1 sequences, with increased diffusivity and lack of gadolinium enhancement.

Case report

A 22-year-old male presented with a 2-month history of seizures, started immediately after a head injury. His previous history was seizure free. Neurologic evaluation, along with electroencephalogram, indicated left temporal epilepsy. Conventional MRI revealed a left parietal lesion comprised of tiny, coalescent, well-defined nodules, hyperintense on T2 and FLAIR, and hypointense on T1 sequences, located in the juxtacortical white matter with partial involvement of an otherwise normal adjacent cortex. The lesion was characterized by increased diffusivity and lack of hemorrhagic elements or gadolinium enhancement (Fig. 1). Dynamic susceptibility contrast MR perfusion revealed a slight relative increase of the lesional cerebral blood volume (CBV) compared to the contralateral white matter (lesional CBV/contralateral CBV = 1.112; Fig. 2). MR spectroscopy showed mild increase of choline and reduction of NAA leading to increase in choline/creatine ratio (1.36) and choline/NAA ratio (0.77), compared to 0.87 and 0.51 (respective values of the contralateral brain)(Fig. 3). Furthermore, head CT, at the time of the traumatic insult, had revealed a left temporal hemorrhagic contusion, which appeared as a gliotic area with hemorrhagic deposition on the subsequent conventional MRI and considered to be responsible for the epileptic episodes (Fig. 4). The patient fully responded to treatment with phenytoin and has been seizure free ever since. Six months later a follow-up MRI was performed and showed the left parietal lesion, without any substantial structural, diffusion, hemodynamic, or metabolic changes (Fig. 5). A written informed consent was obtained from the patient after being briefed on study and publication details.

Discussion

MNVTs are rare entities that have been recently recognized, by WHO, as distinctive neuronal lesions, although they were first reported by Huse et al, in 10 patients, in 2013 [1]. Since then, 51 additional cases have been reported worldwide [2–11], including a case series of 33 patients by Nunes et al [9]. MVNT is frequently associated with seizures or seizure equivalents and occur mainly in adults older than 35 years. It presents as a cluster of nodules, normally numerous and of small size, located juxtacortically on the inner surface of an otherwise normal-appearing cortex and most frequently on the parietal or temporal lobe. Gadolinium enhancement, restricted diffusion, diffuse infiltration, mass effect, or edema is not normally observed [1–11].