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"LEAVE ME ALONE" Posterior Fossa Lesion - A Rare Case Report on Multinodular and Vacuolating Tumor of the Posterior Fossa

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ABSTRACT

Multinodular and Vacuolating Neuronal Tumors (MVNT) are a rare benign, mixed glial neuronal lesion usually involving the supra tentorial brain. It is mostly seen in adults and often associated with seizures.[1-3] Similar multinodular lesions in the infratentorial brain have been described in a few studies. They are classified as either Infratentorial MVNT or MVNT-PLUS (Posterior Fossa Lesion of Unknown Significance).[4,5] Due to the absence of malignancy criteria and the lack of evolutivity on follow-up MRIs, it is considered to be a "leave me-alone" lesion. The diagnosis of MVNT is based on the history with characteristic location and appearance, with lack of progressivity. Here, we report a case of a posterior fossa lesion showing striking similarity to Supratentorial MVNT in a young adult patient.

Keywords: Multinodular; Vacuolating Tumor; Brain; Tumor

INTRODUCTION

Multinodular and vacuolating neuronal tumor (MVNT) has been classified under Glioneuronal and Neuronal Tumors in the latest World Health Organization(WHO 2021) classification as a Grade 1 lesion. In the previous classification, it was grouped under Gangliocytoma.[8,9] It is a rare, benign, mixed glioneuronal supratentorial brain tumor that is often associated with seizures and mostly affects adults. It consists of hyperintense clustered nodules in the superficial subcortical white matter, deep cortical ribbon, and juxtacortical areas, with usually rare or no post-contrast enhancement. It can also be incidentally found in patients without epilepsy.

Infratentorial multinodular lesions resembling MVNT (Multinodular and Vacuolating Neuronal Tumor of the cerebrum) are considered extremely rare. In literature, it has been elucidated as belonging to two different pathologies: Infratentorial MVNT thus corresponding to similar lesions in the supratentorial location & Multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS) thus suggesting a new entity. Since only a limited number of cases have been reported and histopathological results are lacking, there is ambiguity regarding the use of denomination.

These cases were not histologically proven because of difficulty in accessing the posterior fossa. However, they exhibit coalescence of small nodules and are non-progressive and strikingly similar to MVNT.[6,7]

CASE REPORT

A 16 year old boy with a history of two episodes of seizures was referred to our Radiology Department. The patient had no history of any trauma. The patient had a similar history for five years, for which no imaging studies had been performed, and the patient was placed on regular antiepileptics by the consulting physician. The patient had a history of discontinuation of antiepileptics, after which the current episode had occurred.

MRI Brain showed multiple well-defined T1 hypointense and T2/FLAIR hyperintense oval-shaped lesion in the right cerebellar hemisphere in a parasagittal location. A few punctate T2/FLAIR hyperintense lesions were also noted in the bilateral cerebellar periventricular region and vermis, as shown in Figure 1 and Figure 2 A-C. No evidence of restricted diffusion or SWI blooming was present, as shown in Figure 2 D-F. Flattening of the posterior disc with an increased perioptic halo was also noted bilaterally. The appearance of multiple coalescent nodules was strikingly similar to that of supratenorial MVNT. Hence, an imaging diagnosis of MVNT-PLUS was established. A characteristic central dot sign was noted in our case, which was a focal area of T2/FLAIR hypointensity within the nodules, as shown in Figure 3. This is because of the high protein content or solid components within the lesion.[6]

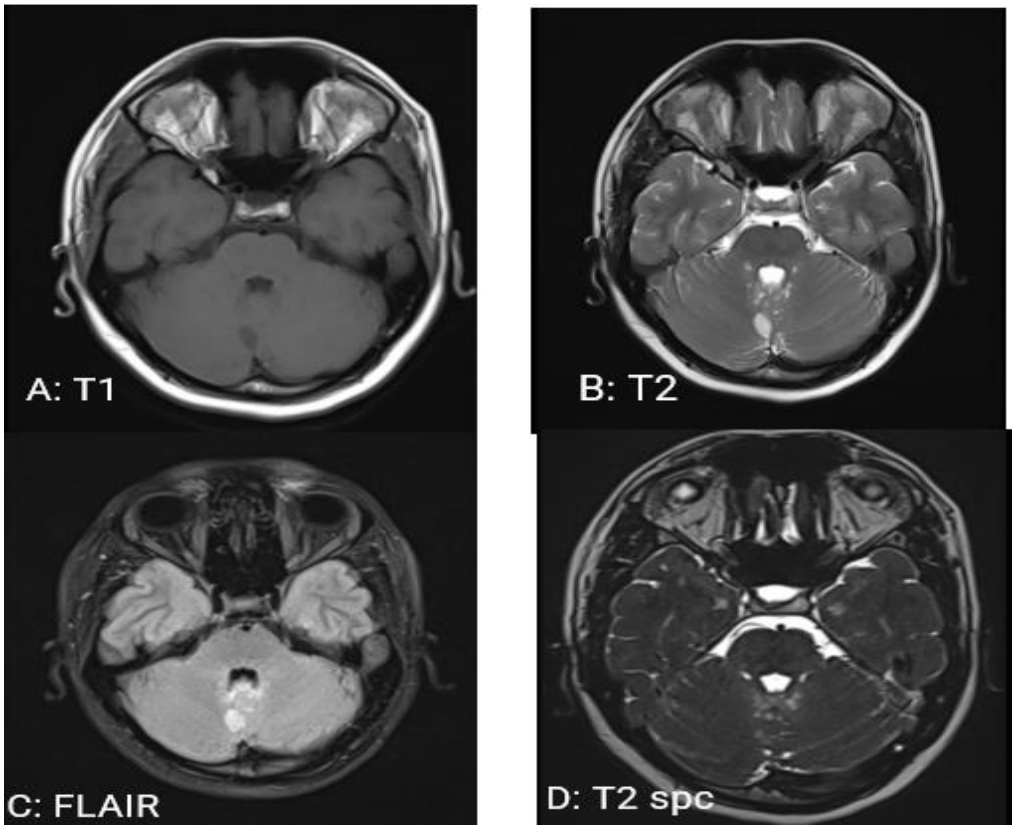

Figure 1: T1 axial(A), T2WI axial(B), FLAIR axial(C) and show an ovoid lesion in the right cerebellar hemisphere which is T2/FLAIR hyperintense and T1 hypointense.T2 space(D) shows multiple clustered hyperintense foci in the paravermian region.

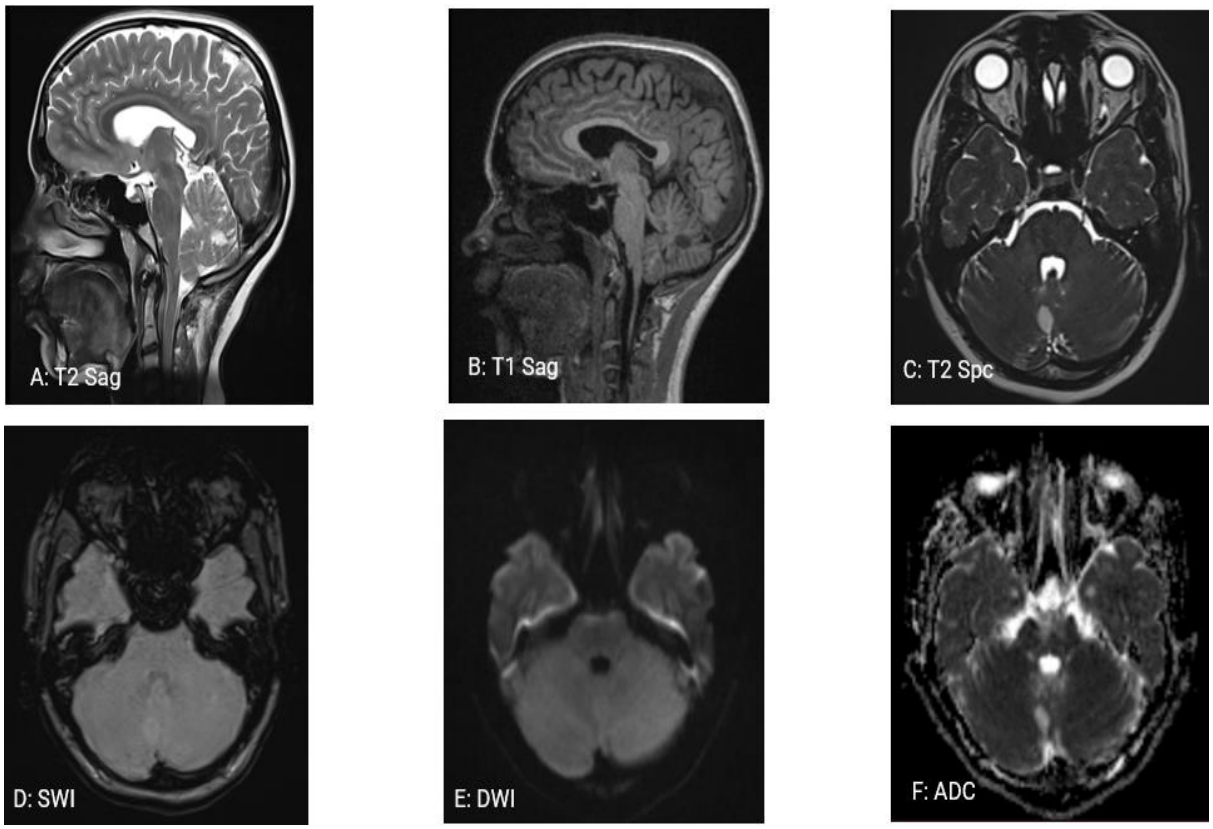

Figure 2: A-F: T2 Sagittal(A), T1 Sagittal(B) and T2 space(C) shows the right cerebellar hemisphere lesion. SWI(D) shows no blooming. DWI(E) with ADC(F) shows no diffusion restriction.

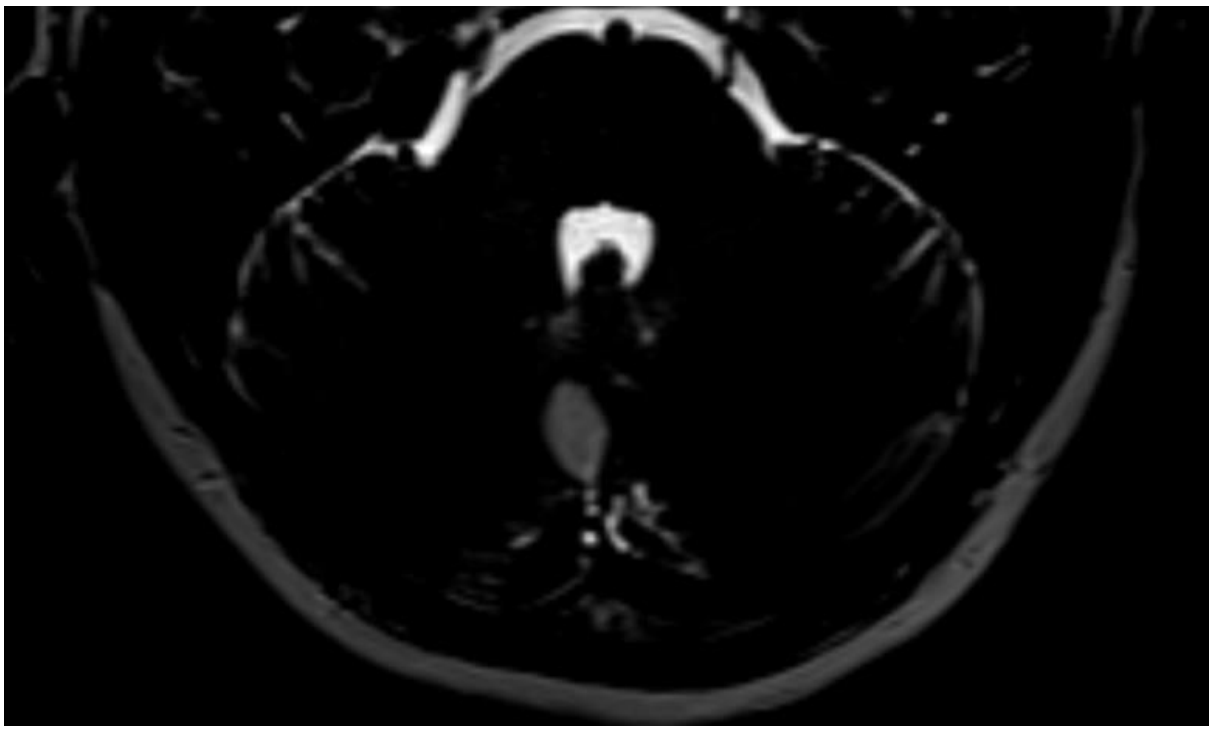

Figure 3: T2 space axial image shows a intralesional tiny hypointense focus suggestive of central dot sign

DISCUSSION

MVNT is a rare supratentorial brain tumor associated with seizures or can be asymptomatic and are discovered incidentally on imaging. Nodules appear isointense or mildly hypointense on T1W and hyperintense to the adjacent white matter on T2W and FLAİR images. They exhibit no diffusion restriction on DWI and ADC sequences or on contrast enhancement. They showed no blooming on susceptibility-weighted imaging (SWI), nor did they show any mass effect.[1,5]

It is unclear whether MVNTs correspond to true neoplasia or hamartomatous or malformative dysplasia. The pathophysiology and frequency of these lesions have yet to be determined.[1]

Multinodular lesions in Infratentorial location resembling MVNT (Multinodular and Vacuolating Neuronal Tumor of Supratentorial location) are extremely rare and have been reported in very few literatures. They are also known as multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS) and are not histologically proven; they consist of the coalescence of small nodules with imaging features strikingly similar to MVNT. To date, only 14 cases of infratentorial MVNT or MV-PLUS have been described, and due to the absence of malignancy criteria and the lack of evolution on follow-up MRIs, it is considered a “leave me-alone” lesion. It is similar to MVNT in that it shows no signs of malignancy.[6,7]

In the current case, MR showed a well-defined T1 hypointense, T2/FLAIR hyperintense, oval-shaped lesion in the right cerebellar hemisphere with no evidence of restricted diffusion and blooming. There were few punctate T2/FLAIR hyperintense lesions also in the bilateral cerebellar periventricular region and vermis. Flattening of the posterior disc with increased peri optic halo was also noted bilaterally.

In asymptomatic patients biopsy and surgical excision is contraindicated. In symptomatic patients (headaches and seizures) conservative treatment with antiepileptics and follow up is recommended. Surgical excision of the tumor is considered when it is associated with repeated episodes of seizures. No tumor recurrence has been reported regardless of the extent of the excision.[5]

LIMITATIONS

The patient did not report for follow up imaging resulting in loss to follow up. Additionally, the patient did not consent for contrast enhanced MRI. A histopathological examination could not be done as excision was contraindicated due to the location of the lesion in posterior fossa.

CONCLUSION

MVNTs and MVNT-PLUS are considered as benign lesions. They are considered malformative, hamartomatous lesions rather than a true neoplasm. It is essential to recognize the unique appearance of MVNTs. Identifying these tumors in imaging studies can prevent unnecessary surgical excision or biopsy.

DIFFERENTIAL DIAGNOSIS

Enlarged Perivascular Spaces- Generally found supratentorial and are asymptomatic.

Tumefactive Demyelinating lesions- show high ADC values as well as variable contrast enhancement with features of demyelination.

LIST OF ABBREVIATIONS

MVNT- Multinodular and vacuolating neuronal tumor

MVNT-PLUS - Multinodular and vacuolating posterior fossa lesions of unknown significance

WHO - World Health Organization

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