Multinodular and vacuolating neuronal tumor-like lesion of the spinal cord : two case reports

MVNT-like lesion of the spinal cord: two case reports

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Abstract

We describe two cases of a spinal cord lesion with imaging features closely resembling those described in supratentorial Multinodular and Vacuolating Neuronal Tumor (MVNT) or infratentorial "multinodular and vacuolating posterior fossa lesions of unknown significance" (MV-PLUS). Multiple well-delineated non-enhancing T2-hyperintense intramedullary cystic ovoid nodules were visualized within the white matter of the spinal cord, including immediately abutting the grey matter. No alterations in signal intensity or morphology were detected in a follow-up. Moreover, the lesions did not present any relevant clinical symptoms attributable to this lesion. We describe these lesions as presumed MVNT, and therefore use the term MVNT-like spinal cord lesions.

Keywords & Abbreviations

Cervical spine; Spinal cord; Magnetic resonance imaging (MRI) ; Multinodular and Vacuolating Neuronal Tumor (MVNT); Multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS)

Abstract

We describe two cases of a spinal cord lesion with imaging features closely resembling those described in supratentorial Multinodular and Vacuolating Neuronal Tumor (MVNT) or infratentorial "multinodular and vacuolating posterior fossa lesions of unknown significance" (MV-PLUS). Multiple well-delineated non-enhancing T2-hyperintense intramedullary cystic ovoid nodules were visualized within the white matter of the spinal cord, including immediately abutting the grey matter. No alterations in signal intensity or morphology were detected in a follow-up. Moreover, the lesions did not present any relevant clinical symptoms attributable to this lesion. We describe these lesions as presumed MVNT, and therefore use the term MVNT-like spinal cord lesions.

Introduction

MVNT is a fairly new and distinctive entity in the 5th edition of the WHO classification of central nervous system tumors.[1] It was first described in 2013 by Huse et. al., showing a case series of a benign seizure-associated intra-axial lesions.[2] Imaging features on MRI include clusters of multiple well-delineated T2-hyperintense millimetric ovoid nodules subcortically in the white matter (*"bubbly appearance"*), which can also be found juxtacortically. The grey matter is generally not affected. Classically there is no contrast enhancement, no diffusion restriction and there is no signal loss on FLAIR-sequences. Cerebral MVNT's are described as a 'leave me alone'-lesion. The microscopic appearance of a MVNT consists of immature neuro-epithelial cells organized in nodules with prominent vacuolating matrix [3], which is almost pathognomonic.

Case Series

Case 1:

A 45-year-old female presented with right-sided cervicobrachialgia for several weeks. Except for discrete paresthesias in the right thumb and index finger, no neurological symptoms were present. There was no significant medical history.

Alongside a disc extrusion and degenerative uncovertebral changes on the right, magnetic resonance imaging (MRI) showed a large multilocular intradural intramedullary lesion at the cervicothoracic junction (C5-T2), consisting of multiple well-defined homogeneous T2-hyperintense and T1-hypointense cystic-like changes. These cystic-like changes were located in the white matter of the spinal cord, including immediately abutting the grey matter. There was no contrast enhancement and no diffusion restriction. No perilesional signal changes were detected. Follow-up MRI after 12 months showed no volume and/or signal intensity changes of the lesion. To exclude intracranial pathology, an MRI-examination of the brain was performed, but showed no abnormalities. All lab values, including infectious serology, were normal.

Case 2:

A 37-year-old female with history of BRCA1 carrier status and ulcerative colitis (UC), presented with approximately 9 months of chronic headache. She reported the headaches began following an intrauterine fetal demise in the setting of a UC flare, complicated by retained products of conception requiring dilation and curettage. She reported a dull nonlocalizable headache 2/10 in severity but denied other neurologic symptoms.

For workup of her headaches, she was referred for MR-imaging of the head, which revealed numerous well-demarcated, non-enhancing T2-hyperintense cystic-like lesions in the medulla and visualized upper cervical spinal cord. Subsequent dedicated spine MRI revealed the extent of these intramedullary lesions within the cervical spinal cord down to the C4 level. Follow-up imaging over the next 4 years remained stable, without development of clinical symptoms. Laboratory values and infectious workup were unrevealing. Cerebrospinal fluid sampling revealed mildly elevated protein but was otherwise normal.

Discussion

In our cases, we see lesions that include a cluster of multiple well-delineated T2-hyperintense millimetric ovoid nodules in the spinal cord white matter, some immediately next to the grey matter. All imaging characteristics are exactly as those described in MVNT. Moreover, the lesions did not present any clinical symptoms and did not change on follow-up. We therefore describe these lesions as MVNT-like spinal cord lesions.

We present two cases of cervical multilocular intradural intramedullary lesions. Differential diagnosis includes neoplastic, inflammatory, vascular or infectious lesions. Neoplasms such as glioma, ependymoma, hemangioblastoma, metastasis and diffuse leptomeningeal glioneuronal tumor (DLGNT) have a different morphology and generally show contrast enhancement. Due to its similar T1- and T2- signal characteristics, spinal cord subependymoma should also be considered. Spinal subependymomas tend to present as diffuse T2-hyperintense lesions with cord enlargement and possibly some enhancement. As they arise from subependymal glial cells, these lesions are typically centrally located in the spinal cord, rather than peripheral. The lesions are larger than the typical 'cysts' seen in MVNT and have a lower signal intensity compared to cerebrospinal fluid. In MVNT, and in our cases, cystic portions are isointense on T2- and T1-weighted images. Various neurological symptoms have been described in patients with spinal cord subependymoma but can also be asymptomatic. Infectious diseases such as cryptococcosis and echinococcosis may also be considered, however all lab values were negative and there were no clinical signs of infection and, again, the lesions did not show any enhancement. Inflammatory disorders of the cervical spinal cord with T2-hyperintense lesions mainly consist of multiple sclerosis, neuromyelitis optica and acute disseminated encephalomyelitis (ADEM). Both lesions did not show any change on follow-up examinations and there are no clinical symptoms or laboratory findings attributable to neither of the lesions. Vascular pathology (e.g. vasculitis) may mimic this imaging presentation. However, lesions associated with vasculitis usually show contrast enhancement and may be irregularly delineated. Perivascular spaces, also called "Virchow-Robin spaces" are also included in the differential diagnosis of a cerebral MVNT but dilated perivascular spaces in the spinal cord have only been reported on microscopy and not on MRI.

In literature, MVNT's originally were exclusively described within the cerebrum. Early case series reported that MVNT's occur supratentorially, in all lobes [3] and a recent publication has shown that these benign lesions don't change over long-term follow up. [4] Lesions although originally associated with seizures, are very often

asymptomatic incidental findings. [5–6] Recent publications also demonstrated cystic multinodular lesion infratentorially in the posterior fossa, with imaging characteristics compatible with MVNT. [7–9] These were reported within the cerebellar hemispheres and vermis. None were histologically proven. The term multinodular and vacuolating posterior fossa lesions of unknown significance (MV-PLUS) was proposed. [7]

No case reports were found describing MVNT in the spinal cord. However, the imaging features of these lesions in our patients and the stable findings on follow-up MRI, were MVNT-like lesions. It is reasonable to hypothesize that MVNTs can also occur within the spinal cord, as they may also occur in the posterior fossa. Short-term follow-up is a limitation of this case-report. There's a follow-up period of 12 months in case 1 and a 4-year follow-up in case 2. Also, no histopathological confirmation was obtained, due to the asymptomatic characteristics of the lesions in combination with their location in the spinal cord. However, other publications regarding MV-PLUS, also have no histopathological confirmation.

Figures

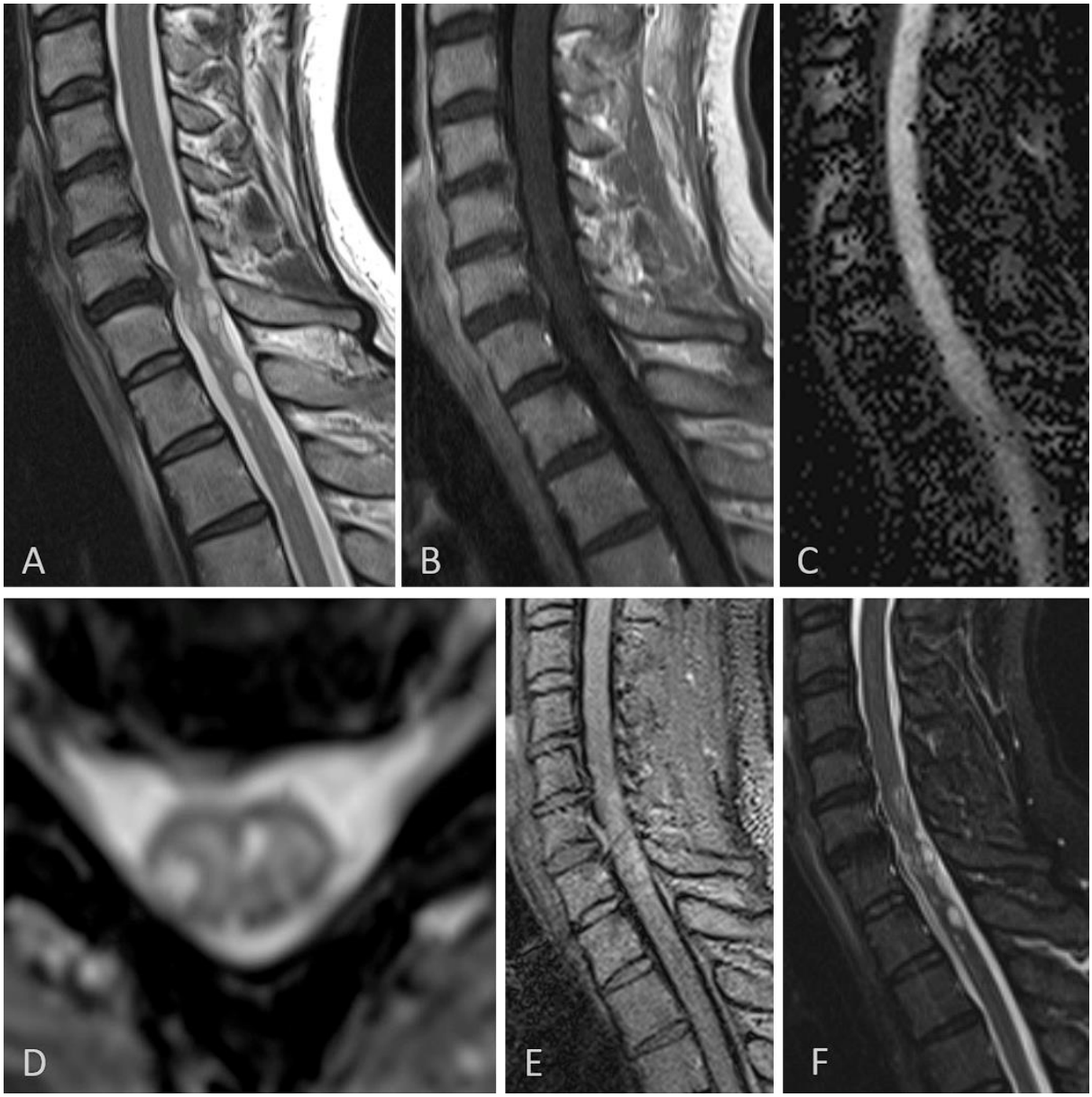

Figure 1: (Case 1) 45-Year-old Female. (A) Sagittal TSE T2-weighted images shows a multilocular intradural intramedullary lesion. There are multiple cystic-like changes within the spinal cord at the cervicothoracic junction at different levels. Note there is no syringomyelia, nor any T2-hypointense changes within the medulla. A cervical disc extrusion at C6-C7 can also be seen. (B) Sagittal TSE T1-weighted images after intravenous administration of gadolinium. No contrast enhancement can be seen. (C) Sagittal EPI Diffusion weighted images show no diffusion restriction within the lesion. (D). Para-axial GRE T2-weighted images depicts the cystic-like intra-axial lesion. Note the sharp delineation of the lesions without perilesional edema. These lesions are located within the white matter of the spinal cord, without affecting the grey matter. (E) Sagittal FLAIR T2-weighted images show the T2-hyperintense lesion. Unlike perivascular spaces ("Virchow-Robin spaces"), there is no signal loss on FLAIR. (F) Sagittal STIR T2-weigted images in a 12-month follow-up. No alterations in signal intensity or morphology were detected. Stable lesion.

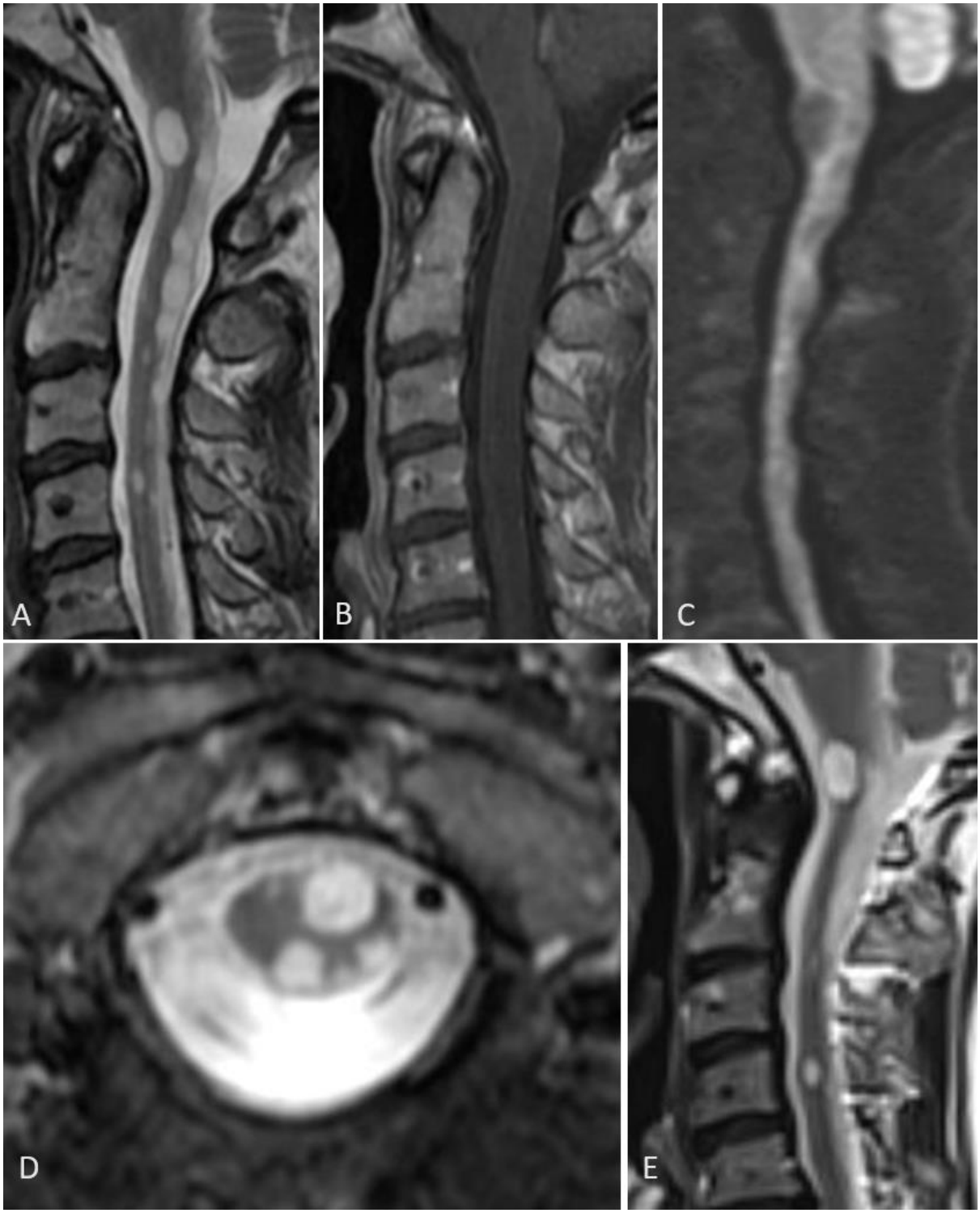

Figure 2: (Case 2) 37-Year-old Female. (A) Sagittal TSE T2-weighted images shows multilocular intradural intramedullary lesions. Similar to the other case, there is no syringomyelia, nor any T2-hypointense changes within the medulla. (B) Sagittal T1-weighted images after intravenous administration of gadolinium. No contrast enhancement can be seen. (C) Sagittal EPI Diffusion weighted images show no diffusion restriction within the lesions. (D) Para-axial T2-weighted images depicts the cystic-like intra-axial lesions. Note the sharp delineation of the lesions. These lesions are located within the white matter of the spinal cord, without affecting the grey matter. (E) Sagittal T2-weighted image in a 4-year follow-up MR-examination. No volume changes and/or signal intensity changes could be detected allowing for differences in field of view.

References

1. 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. doi:10.1093/neuonc/noab106

2. Huse JT, Edgar M, Halliday J, Mikolaenko I, Lavi E, Rosenblum MK. Multinodular and Vacuolating Neuronal Tumors of the Cerebrum: 10 Cases of a Distinctive Seizure-Associated Lesion. *Brain Pathol*. 2013;23(5):515-524. doi:10.1111/bpa.12035

3. 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 Am J Neuroradiol*. 2017;38(10):1899-1904. doi:10.3174/ajnr.A5281

4. Dogra S, Zagzag D, Young M, Golfinos J, Orringer D, Jain R. Long-Term Follow-up of Multinodular and Vacuolating Neuronal Tumors and Implications for Surveillance Imaging. *Am J Neuroradiol*. Published online July 27, 2023. doi:10.3174/ajnr.A7946

5. Arbuiso S, Roster K, Gill A, et al. Multinodular and Vacuolating Neuronal Tumor: Incidental Diagnosis of a Rare Brain Lesion. Cureus. 2021;13(12):e20674. Published 2021 Dec 25. doi:10.7759/cureus.20674

6. Makrakis D, Veneris S, Papadaki E. Multinodular and vacuolating neuronal tumor incidentally discovered in a young man: Conventional and advanced MRI features. Radiol Case Rep. 2018;13(5):960-964. Published 2018 Aug 6. doi:10.1016/j.radcr.2018.07.016

7. Lecler A, Bailleux J, Carsin B, et al. Multinodular and Vacuolating Posterior Fossa Lesions of Unknown Significance. Am J Neuroradiol. 2019;40(10):1689-1694. doi:10.3174/ajnr.A6223

8. Morassi M, Bagatto D. Infratentorial Multinodular and Vacuolating Neuronal Tumor or Multinodular and Vacuolating Posterior Fossa Lesions of Unknown Significance? Clinico-Radiologic Findings from 2 Cases. World Neurosurg. 2020;136:58-61. doi:10.1016/j.wneu.2019.12.166

9. Múnera V, Vargas S, Herrera Jaramillo DA. Lesión multinodular y vacuolar de fosa posterior. Presentación de caso en Colombia. *Rev Colomb Radiol*. 2020;31(2):5339-5341. doi:10.53903/01212095.45