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Image Report

Cervical neuromyelitis optica with thoracic ependymoma

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ABSTRACT

Background: The occurrence of cervical neuromyelitis optica (NMO) in a patient with a thoracic ependymoma is uncommon. Here, we present a patient with a spinal ependymoma who developed the new onset of NMO 2 months later.

Case Description: A 66-year-old male presented with right lower limb weakness. The magnetic resonance (MR) revealed an intramedullary spinal cord tumor at the T2-T4 level. It was surgically excised and proved pathologically to be an ependymoma. 2 months later, the patient presented with an acute partial quadriparesis and a high signal intensity cord lesion at the C2-C3 level attributed to seropositive NMO (i.e. additional diagnostic studies confirmed this diagnosis).

Conclusion: Patients with intramedullary thoracic ependymomas may also develop NMO resulting in recurrent/new neurological deficits. Critical studies utilized to diagnose NMO include brain and spine MRs showing unique intramedullary brain/cord lesions, aquaporin-4 positive serology, and classical abnormal visual studies. If the diagnosis of NMO is established, multiple additional medical therapies are warranted.

Keywords: Aquaporin-4 antibodies, Ependymoma, Neuromyelitis optica, Rehabilitation, Spine

INTRODUCTION

Ependymoma is a frequent condition seen in the neurosurgical unit. However, the occurrence of cervical neuromyelitis optica (NMO) with a thoracic ependymoma in a single patient is not commonly encountered in the clinical setting. Here, we present a patient with a spinal ependymoma who developed the new onset of NMO 2 months later.

CASE DESCRIPTION

A 66-year-old male presented with right lower limb paresis. The urgent magnetic resonance (MR) revealed a T2 to T4 intramedullary lesion that enhanced with contrast consistent with the diagnosis of an ependymoma. Following a T3 to T4 laminectomy, this diagnosis ependymoma was confirmed [Figure 1]. Postoperatively, the patient regained full neurological function within 6 weeks.

However, 2 months later, he was readmitted with the acute onset of left palm numbness, left lower limb paresis, and urinary frequency (i.e. partial quadriparesis). The repeat neuroaxis MR imaging (MRI) studies documented a new C2-C3 intramedullary his signal lesion [Figure 2]. However, at this point, the patient was diagnosed with NMO based upon sero-positive aquaporin-4 antibodies (AQP4), and brain MR findings of hyperintense patches in the subcortical and deep white

matter on T2-weighted images. The NMO was treated with a combination of intravenous methylprednisolone, plasma exchange, oral steroids, and mycophenolate, resulting in partial resolution in his neurological deficits (i.e., at discharge, he ambulated with a cane, and could perform daily activities). MRI scans three months later revealed partial resolution of the C2-3 intramedullary NMO lesion. Nevertheless, 18 months after that, he again presented with another NMO lesion (cervical MR with extensive patchy enhancement of the cord from C2 to C7 with diffuse cord swelling) now contributing to an acute quadriparesis [Figure 3].

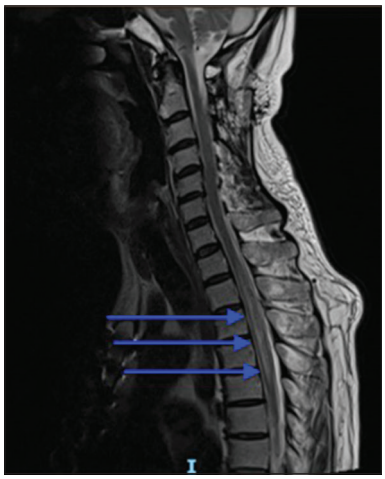

Figure 1: Magnetic resonance imaging sagittal T2 image of the cervical and thoracic spine in June 2019 revealing intramedullary enhancement over the thoracic T2 to T4 regions. See blue arrows. Histology subsequently confirmed ependymoma. No intramedullary lesions were noted in the cervical spine.

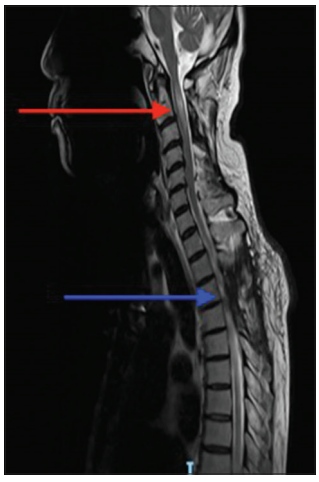

Figure 2: Magnetic resonance imaging sagittal T2 image of the cervical and thoracic spine in August 2019 revealing new intramedullary enhancement over the cervical C2-3 region before treatment with steroids and immunosuppressive agents. See red arrow. The blue arrow indicates the post-surgical excision site for the thoracic ependymoma.

Despite immediate medical treatment with steroid and immunosuppression, the motor and sensory recovery remained incomplete [Figure 4].

DISCUSSION

Here, we presented a patient who originally presented with a paraparesis attributed to a T2-T4 thoracic ependymoma, who 2 and 18 months later developed acute quadriparesis due to NMO (C2-C3, and C2- 7, respectively). The diagnosis of NMO utilizing brain/holo spinal MR and positive AQP4 serology was clearly established, thus avoiding the need for an intramedullary cervical cord biopsy (i.e., to rule out metastatic ependymoma)and its inherent risks.[1,4]

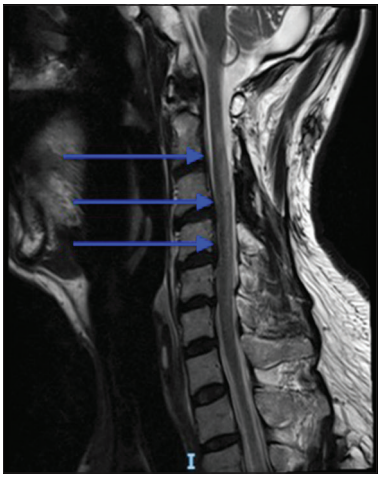

Figure 3: Magnetic resonance imaging sagittal T2 image of the cervical spine in May 2021 during the neuromyelitis optica flare before medical treatment revealing significant cord swelling from C2 to C7 as illustrated by the blue arrows.

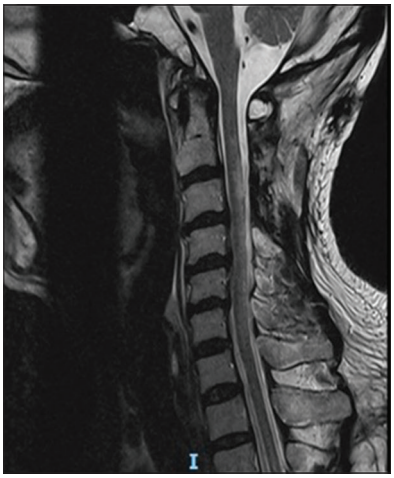

Figure 4: Magnetic resonance imaging sagittal T2 image of the cervical spine in May 2021 after medical treatment revealing significant cord swelling reduction from C2 to C7.

Classical laboratory and MR findings NMO

Classical NMO-immunoglobulin G (IgG) tests and c MR findings of NMO helped confirm this diagnosis in this patient, ruling out metastatic ependymoma. NMO-IgG acts against the AQP-4 water channels which are highly populated in the astrocytes for water transportation across the cell membrane, and NMO on brain/spinal MRI imaging, are typically hyper-intense on T2-weighted studies with lesions distributed to the peri-ependymal regions.[2,3]

CONCLUSION

It is unusual for a patient to present with paraparesis due to an intramedullary thoracic ependymoma (T2-T4) followed by the diagnosis of NMO. As these two entities may mimic each other, it is important to differentiate between the two utilizing MRI brain/spine imaging, AQP4 serology, and visual tests.

Declaration of patient consent

Patient's consent is not required as patients identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Fanous AA, Olszewski NP, Lipinski LJ, Qiu J, Fabiano AJ. Idiopathic transverse myelitis mimicking an intramedullary spinal cord tumor. Case Rep Pathol 2016;2016:8706062.
2. Mpateni SS, Sihlali NC, Gardiner EC, Gigi N. Magnetic resonance imaging findings in a patient with seropositive neuromyelitis optica. SA J Radiol 2018;22:1306.
3. Oh SH, Yoon KW, Kim YJ, Lee SK. Neuromyelitis optica mimicking intramedullary tumor. J Korean Neurosurg Soc 2013;53:316-9.
4. Woo PY, Chiu JH, Leung KM, Chan KY. Seropositive neuromyelitis optica imitating an intramedullary cervical spinal cord tumor: Case report and brief review of the literature. Asian Spine J 2014;8:684-8.

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