

Cranial Polyneuropathy from Multiple Myeloma with CNS Involvement

ROBERT J. QUON, PhD; SASMIT SARANGI, MD; JEFFREY M. ROGG, MD; MAURICIO F. VILLAMAR, MD

KEYWORDS: multiple myeloma, neurooncology, nervous system neoplasms, chemotherapy, radiation therapy

A 77-year-old woman was diagnosed with non-secretory multiple myeloma (MM) of IgA kappa subtype, with 17p (*TP53*) deletion. There was an initial response to radiation and chemotherapy, but she eventually developed extramedullary involvement and multifocal plasmacytomas.

Inpatient neurology consultation was requested for evaluation of 2 months of severe right hemifacial pain, profound right-sided hearing loss, prominent right peripheral-type facial palsy, and blurry vision affecting the right eye (**Image 1**). Brain MRI showed enhancing lesions involving the right internal auditory canal, cerebellopontine angle, anterior clinoid process, optic canal, right middle cranial fossa floor, and tentorial margin (**Image 2**). These lesions were not present on a pre- and post-gadolinium MRI performed for disease staging approximately a week before the onset of the patient's symptoms. The patient received cranial radiotherapy (20 gray over 5 fractions) and salvage chemotherapy (daratumumab, pomalidomide, and dexamethasone), with no substantial improvement. She transitioned to hospice and died 7 months after the onset of cranial neuropathies.

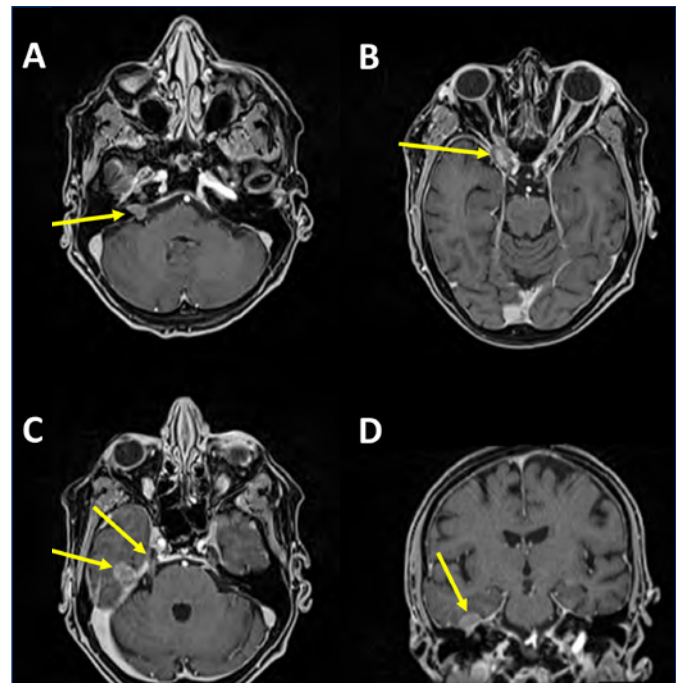
Image 1. Multiple cranial neuropathies due to central nervous system involvement by multiple myeloma

In these images, the patient was asked to smile (**A**) and frown (**B**). She had severe right peripheral-type facial palsy. In addition, she reported severe, stabbing, near-constant right-sided facial pain that could affect the V1, V2, or V3 divisions of the trigeminal nerve, marked right-sided hearing loss, and blurry vision affecting the right eye.



Image 2. T-1 weighted post-contrast brain MRI showing central nervous system involvement from multiple myeloma

(**A**) Contrast-enhancing soft tissue tumor involving the right internal auditory canal and extending into the cerebellopontine angle (arrow), encasing cranial nerves VII and VIII. (**B**) Contrast-enhancing soft tissue tumor infiltrating and expanding the right anterior clinoid process with extension into the right optic canal (arrow). (**C**) and (**D**) Dural-based contrast-enhancing soft tissue tumor at the floor of the right middle cranial fossa (arrows), extending laterally and anteriorly to involve the tentorial margin with encroachment on the adjacent cranial nerve V.



Central nervous system (CNS) involvement occurs in <1% of MM patients.¹ Patients can develop leptomeningeal myelomatosis (70%), direct extension of MM with dural involvement (35%), intraparenchymal metastases (26%), or combinations of these findings.² Both hematogenous and contiguous spread may contribute to CNS involvement.^{3,4} CNS involvement is associated with poor prognosis, with median overall survival <6 months.^{2,3} Negative prognostic factors include multiple lines of chemotherapy and an unfavorable cytogenetic profile.⁵ Radiation therapy and CNS-penetrating immunomodulatory imide drugs (IMiDs)

are frequently used for symptom palliation.^{2,3} Despite the outcome of this case, earlier recognition of CNS MM could potentially be associated with more favorable outcomes.²

References

1. Fassas ABT, Muwalla F, Berryman T, et al. Myeloma of the central nervous system: association with high-risk chromosomal abnormalities, plasmablastic morphology and extramedullary manifestations: Myeloma of the Central Nervous System. *Br J Haematol.* 2002;117(1):103-108. doi:10.1046/j.1365-2141.2002.03401.x
2. Paludo J, Painuly U, Kumar S, et al. Myelomatous Involvement of the Central Nervous System. *Clin Lymphoma Myeloma Leuk.* 2016;16(11):644-654. doi:10.1016/j.clml.2016.08.010
3. Chen CI, Masih-Khan E, Jiang H, et al. Central nervous system involvement with multiple myeloma: long term survival can be achieved with radiation, intrathecal chemotherapy, and immunomodulatory agents. *Br J Haematol.* 2013;162(4):483-488. doi:10.1111/bjh.12414
4. Gozzetti A, Cerase A, Lotti F, et al. Extramedullary intracranial localization of multiple myeloma and treatment with novel agents: A retrospective survey of 50 patients. *Cancer.* 2012;118(6):1574-1584. doi:10.1002/cncr.26447
5. Jurczynski A, Grzasko N, Gozzetti A, et al. Central nervous system involvement by multiple myeloma: A multi-institutional retrospective study of 172 patients in daily clinical practice: Nervous System Involvement by Myeloma. *Am J Hematol.* 2016;91(6):575-580. doi:10.1002/ajh.24351

Authors

Robert J. Quon, PhD, The Warren Alpert Medical School of Brown University, Providence, RI

Sasmit Sarangi, MD, Department of Neurology, Rhode Island Hospital, Department of Neurology, Division of Neuro-Oncology, The Warren Alpert Medical School of Brown University, Providence, RI

Jeffrey M. Rogg, MD, Department of Diagnostic Imaging, Rhode Island Hospital, Providence, RI

Mauricio F. Villamar, MD, Department of Neurology, Neurohospitalist Division, The Warren Alpert Medical School of Brown University, Providence, RI; Department of Medicine, Kent Hospital, Warwick, RI

Acknowledgments

The authors thank the patient and her family for consenting to this publication. In accordance with the Journal's policy, written informed consent was obtained from the patient to publish this report.

Disclosures

Study sponsorship/funding: None

Declaration of conflicting interests: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Correspondence

Mauricio F. Villamar, MD
593 Eddy St, APC 5
Providence, RI 02903
401-921-7508
Fax 401-736-1057
mauricio_villamar@brown.edu