Teaching NeuroImages: Intracranial *DICER1*-associated spindle cell sarcoma in a child

Ali Rashidi, MD, Licia P. Luna, MD, PhD, Fausto Rodriguez, MD, and Aylin Tekes, MD

Neurology® 2020;95:e2176-e2177. doi:10.1212/WNL.0000000000010374

Correspondence

Dr. Luna
lluna6@jhmi.edu

Figure 1 MRI brain

Brain MRI reveals a right frontoparietal intraaxial mass, hyperintense on fluid-attenuated inversion recovery (A), hypointense on T2-weighted imaging (B), with marked enhancement on postcontrast T1-weighted imaging (C), areas of restricted diffusion on diffusion-weighted imaging (D) and apparent diffusion coefficient map (E, arrow), vascular structures (A, arrowhead), and remodeling of the inner table (B, arrowheads). A hyperdense focus on CT (arrow in F) with a signal loss on MRI (arrow in A) was a large vein with mineralization on surgery. Differential considerations include embryonal tumors.

An 8-year-old boy presented with a 2-week history of worsening right-sided headache and left-sided jerking movements. Symptoms were associated with vomiting, photophobia, phonophobia, and diplopia but no loss of consciousness. cEEG demonstrated increased voltage on the right but no evidence of seizures. Brain MRI demonstrated a large right frontoparietal mass (figure 1). The pathologic findings were compatible with *DICER1*-associated pleomorphic spindle cell sarcoma (figure 2). Recently described, *DICER1* mutation predisposes individuals to development of a subgroup of tumors termed “spindle cell sarcoma with rhabdomyosarcoma-like features, *DICER1* mutant” although it may present as “primary intracranial sarcoma, *DICER1*-mutant.”^{1,2}

MORE ONLINE

→Teaching slides

links.lww.com/WNL/B175

From the Russell H. Morgan Department of Radiology and Radiological Science (A.R., L.P.L., A.T.) and Pathology Department (F.R.), Johns Hopkins University School of Medicine, Baltimore, MD.

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Figure 2 Histopathologic features of *DICER1*-associated sarcoma

(A) Pleomorphic mesenchymal-appearing cells, some containing bright eosinophilic globules (×600). (B) Partial desmin immunoreactivity is typical (×600).

Study funding

No targeted funding reported.

Disclosure

The authors report no disclosures relevant to the manuscript. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

Appendix Authors

Name	Location	Contribution
Ali Rashidi, MD	Baltimore, MD	Study concept and manuscript writing
Licia P. Luna, MD, PhD	Baltimore, MD	Study concept, major role in the acquisition of data

Appendix (continued)

Name	Location	Contribution
Fausto Rodriguez, MD	Baltimore, MD	Major role in the acquisition of data
Aylin Tekes, MD	Baltimore, MD	Critical revision of the manuscript for important intellectual content, study supervision

References

1. Lee JC, Villanueva-Meyer JE, Ferris SP, et al. Primary intracranial sarcomas with *DICER1* mutation often contain prominent eosinophilic cytoplasmic globules and can occur in the setting of neurofibromatosis type 1. *Acta Neuropathol* 2019;137:521–525.
2. Sakaguchi M, Nakano Y, Honda-Kitahara M, et al. Two cases of primary supratentorial intracranial rhabdomyosarcoma with *DICER1* mutation which may belong to a “spindle cell sarcoma with rhabdomyosarcoma-like feature, *DICER1* mutant”. *Brain Tumor Pathol* 2019;36:174–182.

Neurology®

Teaching NeuroImages: Intracranial *DICER1*-associated spindle cell sarcoma in a child

Ali Rashidi, Licia P. Luna, Fausto Rodriguez, et al.

Neurology 2020;95:e2176-e2177 Published Online before print July 17, 2020

DOI 10.1212/WNL.0000000000010374

This information is current as of July 17, 2020

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/95/15/e2176.full
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/95/15/e2176.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Pediatric http://n.neurology.org/cgi/collection/all_pediatric CT http://n.neurology.org/cgi/collection/ct MRI http://n.neurology.org/cgi/collection/mri Primary brain tumor http://n.neurology.org/cgi/collection/primary_brain_tumor
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2020 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.