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

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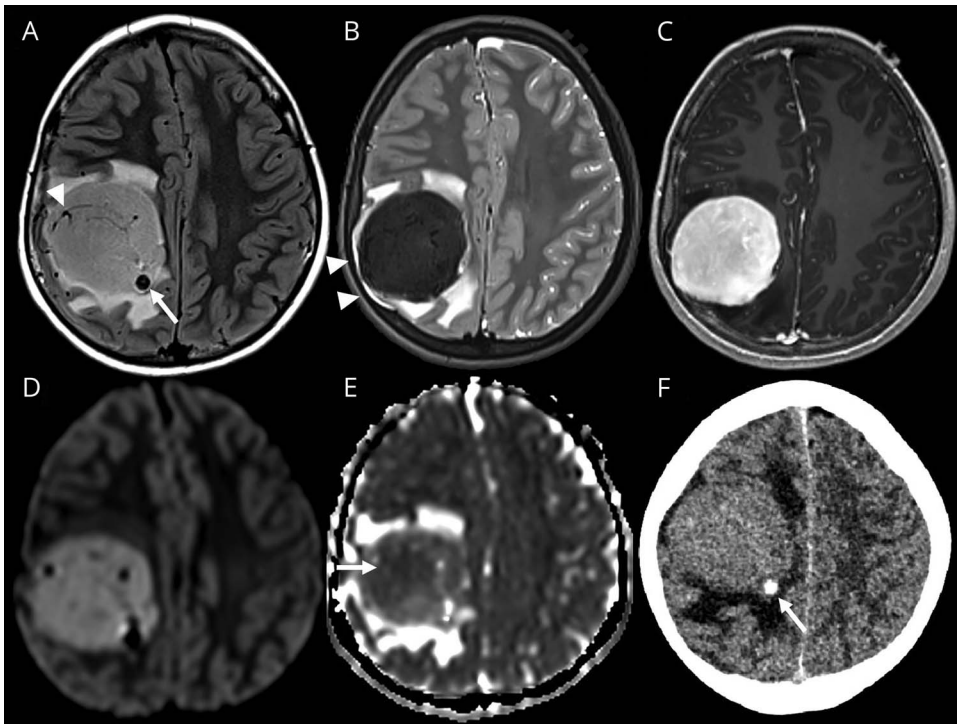
Figure 1 MRI brain



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]

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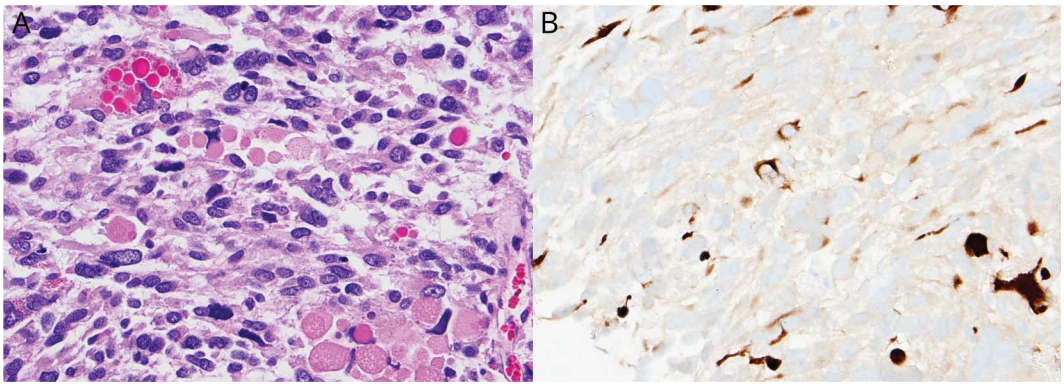
Figure 2 Histopathologic features of *DICER1*-associated sarcoma



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

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Disclosure

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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
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

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