Teaching NeuroImages: Intracranial malignant triton tumor

An uncommon location of a rare tumor

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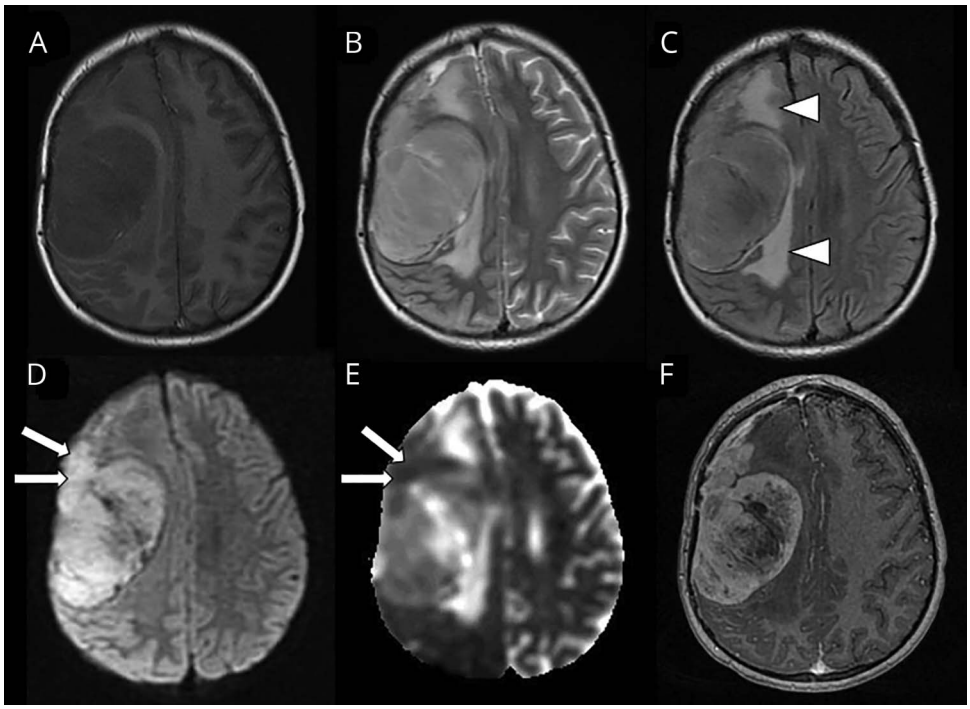
Figure 1 Brain MRI



Axial brain MRI reveals a large extra-axial right frontal mass, hypointense on T1-weighted imaging (A) and hyperintense on T2-weighted imaging (B), associated with moderate surrounding vasogenic edema (arrowheads), better seen on fluid-attenuated inversion recovery (C). The lesion demonstrated areas of diffusion restriction (arrows) on diffusion-weighted imaging (D) and apparent diffusion coefficient (E) and heterogeneous enhancement on postcontrast T1-weighted imaging (F).

A 5-year-old girl with neurofibromatosis type 1 (NF1) presented with a 1-month history of progressive left hemiparesis. Brain MRI revealed a large extra-axial right frontal mass (figure 1). The pathologic findings after complete resection were compatible with malignant triton tumor (MTT) (figure 2). MTT is an aggressive subtype of malignant peripheral nerve sheath tumor, histopathologically characterized by a high-grade spindle cell tumor, with immunohistochemistry demonstrating reaction to skeletal muscle stains, such as desmin and myogenin.[1,2] MTT is associated with NF1 in 38% of cases.[1] Intracranial MTT is exceedingly rare, usually presenting radiologically as a rapidly enlarging, heterogeneous, and enhancing mass.[2]

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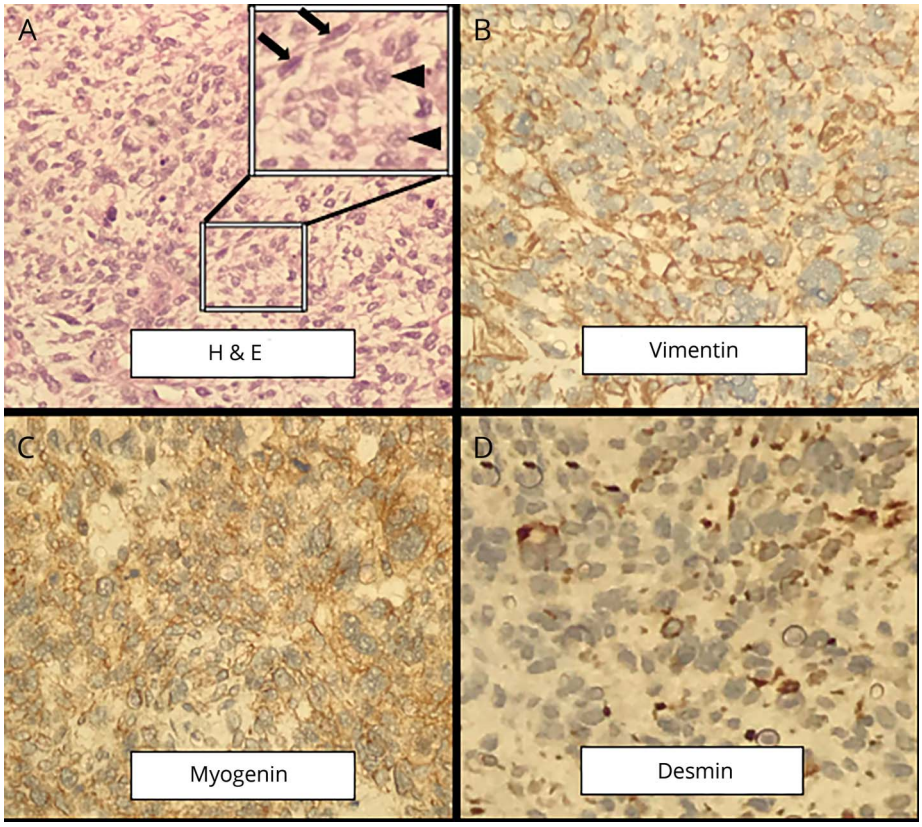
Figure 2 Pathologic findings after excision of the intracranial mass



(A) Histologic evaluation (hematoxylin & eosin, ×200) shows a neoplastic process composed of spindle cells showing a fascicular growth pattern (arrows), extensive pleomorphism, and high mitotic index (arrowheads). Immunohistochemical analysis (×200) demonstrates positive immunoexpression for vimentin (B), myogenin (C), and desmin (D), reflecting areas of rhabdomyoblastic differentiation.

Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

Appendix Authors

Name	Location	Role	Contribution
Adriano Basso Dias, MD	Irmandade Santa Casa de Misericórdia de Porto Alegre, Brazil	Author	Study concept and manuscript writing
Eduardo Cambruzzi, PhD	Irmandade Santa Casa de Misericórdia de Porto Alegre, Brazil	Author	Major role in the acquisition of data
Cláudia Scherber Giugno, MD	Irmandade Santa Casa de Misericórdia de Porto Alegre, Brazil	Author	Major role in the acquisition of data
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Appendix *(continued)*

Name	Location	Role	Contribution
Lázaro Luís Faria do Amaral, MD	Beneficência Portuguesa de São Paulo, Brazil	Author	Critical revision of the manuscript for important intellectual content
Rene Lenhardt, MD	Irmandade Santa Casa de Misericórdia de Porto Alegre, Brazil	Author	Critical revision of the manuscript for important intellectual content, study supervision

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