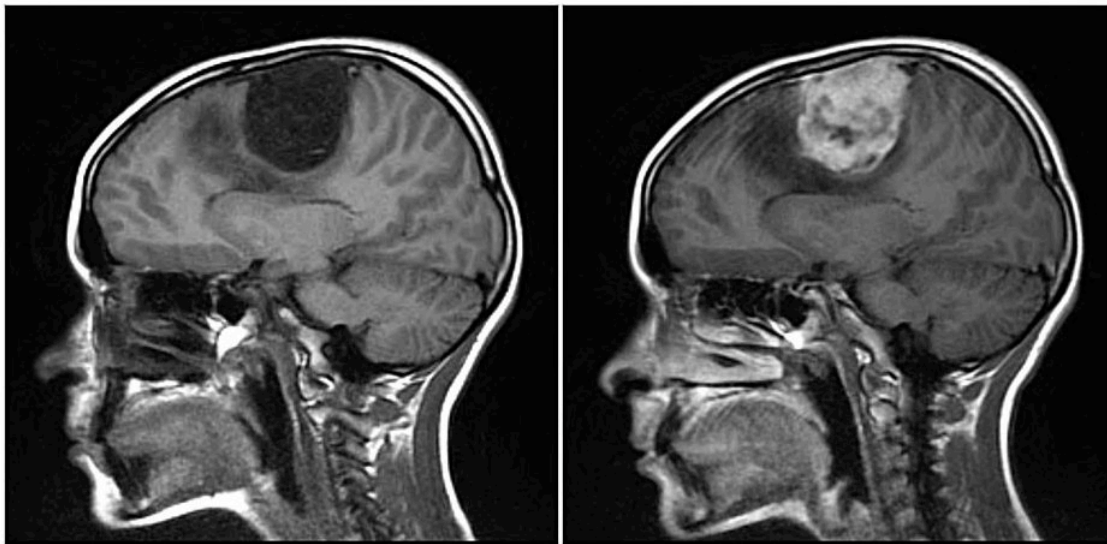


50th ANNUAL DIAGNOSTIC SLIDE SESSION, 2009
DIAGNOSES AND REFERENCES
MODERATOR: Anthony T. Yachnis, M.D.
EDITOR: Leroy R. Sharer, M.D.

Case 2009-4

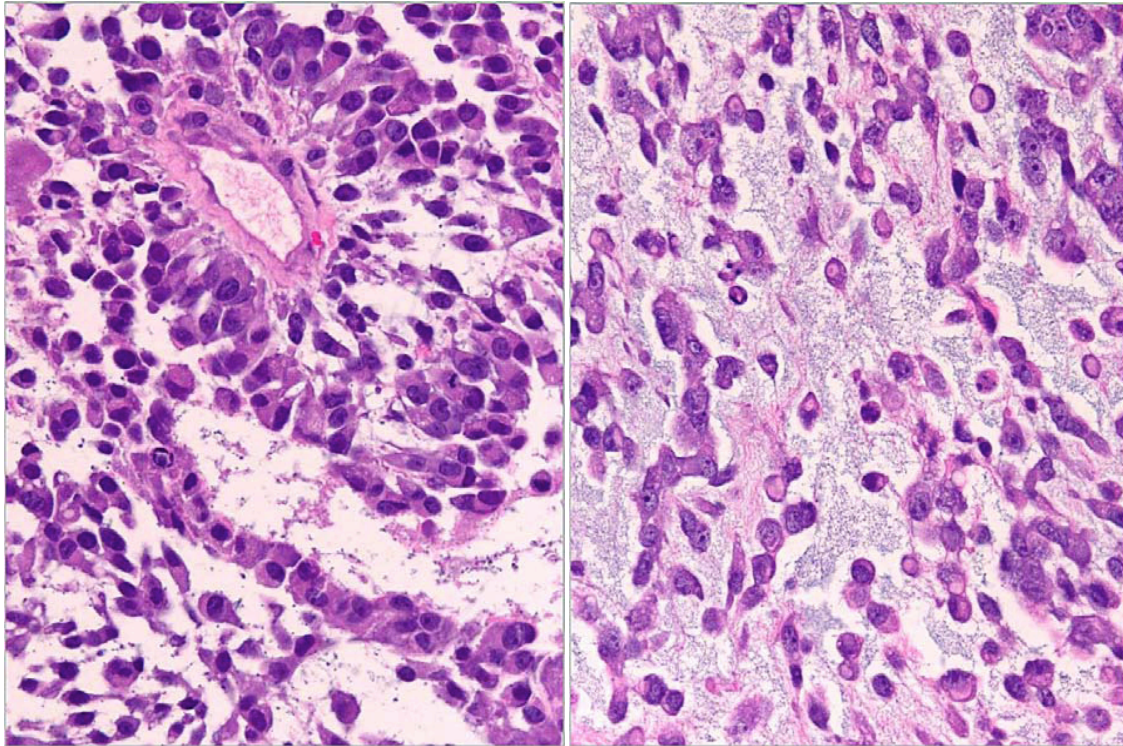
Submitted by: Mark Jentoft, M.D., Caterina Giannini, M.D., Ph.D., and Bernd W Scheithauer, M.D., Mayo Clinic, Rochester MN

A previously healthy 11 year old boy presented to an outside facility with a 4 day history of left sided weakness, bifrontal headache, facial numbness, nausea, and vomiting. A ten pound weight loss had occurred recently. An MRI of the brain showed a 5.3 x 4.7 x 4.3 cm extra-axial mass over the right cerebral convexity at the fronto-parietal junction. The mass had a broad base over the surface of the dura with a small dural tail and the tumor extended into the underlying brain parenchyma. Moderate edema associated with a 7 mm midline shift was noted. A surgical resection of the tumor was performed.



T1

T1 w C



Diagnosis: High grade myxoid rhabdoid neoplasm most consistent with myoepithelial carcinoma; differential diagnosis includes atypical teratoid/rhabdoid tumor

Comment: Various diagnoses were offered by members of the audience, including rhabdoid meningioma, atypical teratoid/rhabdoid tumor (AT/RT), meningioangiomatosis, and sarcoma, with the favored diagnosis being AT/RT. The tumor was immunopositive for cytokeratin Cam 5.2 and S-100 protein, while it was negative for EMA, GFAP and INI1. One INI1 allele was seen on FISH, with a probable point mutation in the other. Cytogenetic study revealed a translocation 3-22, not involving the *INI1* gene. One consultant diagnosed myoepithelial carcinoma, while another favored AT/RT. The lesion was resected in September 2008, and the child later developed pulmonary nodules, which on biopsy were similar to the lesion seen in the brain. He died of disease late in December 2008.

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