50th ANNUAL DIAGNOSTIC SLIDE SESSION, 2009
DIAGNOSES AND REFERENCES
MODERATOR: Anthony T. Yachnis, M.D.
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Case 2009-3

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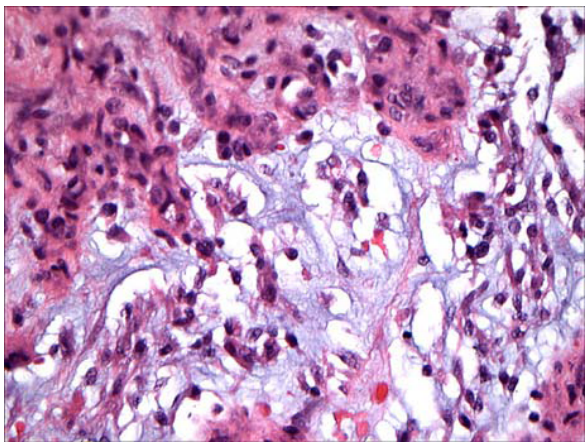
A 61 year-old man presented to his primary care physician complaining of episodes of lightheadness and hot flashes. He also reported subtle word finding difficulties, progressive loss of right leg strength, forgetfulness and difficulty with attention at work. His past medical history included hypercholesterolemia, gastroesophageal reflux, depression, anxiety, sleep apnea, asthma and benign prostatic hypertrophy. MRI showed a heterogenously enhancing lesion with an ovoid shape located in the medial left frontal lobe and cingulate gyrus. The lesion was T2 hyperintense with areas of nodular and peripheral rim enhancement. The mass appeared to extend into the falx with linear enhancement of the anterior and posterior portions of the falx around the lesion. There was a suggestion of cortex displaced posteriorly by this lesion, favoring an extra-axial location. The patient underwent a bifrontal craniotomy for tumor resection.



Diagnosis: Glioblastoma with chordoid features

Comment: Audience members gave various suggestions for diagnosis, most of which are included below in the Comment from the Presenter, with the addition of chondromyxoid fibroma and PXA. Grossly, the resected material was soft, white and mucoid. A smear preparation at the time of surgery had myxoid material as well as cells with pink processes. Necrosis was evident, on histological examination of the fixed specimen.

From the Presenter: Mitoses and microvascular proliferation were present, with a proliferation index >20%. The tumor cells were immunoreactive for GFAP, keratin (presumed cross-reactivity) and S-100 but negative for EMA and brachyury. Rare tumor cells expressed p53. The tumor appeared to be separate from the brain, as

neurofilament did not revealed entrapped axons. FISH for EGFR showed no amplification. The differential diagnosis for lesions with a chordoid or mucinous/myxoid background includes chordoid meningioma, chordoid glioma of the third ventricle, pilomyxoid astrocytoma, chordoma and metastasis.

References:

Horbinski C, Dacic S, McLendon RE, et al: Chordoid glioma: a case report and molecular characterization of five cases. Brain Pathol 2009; 19:439-448.

Sangoi AR, Dulai MS, Beck AH, Brat DJ, Vogel H: Distinguishing chordoid meningiomas from their histologic mimics: an immunohistochemical evaluation. Am J Surg Pathol 2009; 33: 669-681.

Tirabosco R, Mangham DC, Rosenberg AE, et al: Brachyury expression in extra-axial skeletal and soft tissue chordomas: a marker that distinguishes chordoma from mixed tumor/myoepithelioma/parachordoma in soft tissue. Am J Surg Pathol 2008; 32:572-580.