Neuroimaging Highlight

Editors: Richard Farb, David Pelz

Chordoid Glioma: Imaging Pearls of a Unique Third Ventricular Tumor

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A 46-year-old male presented with unprecedented syncopal episodes and was subsequently worked up for a neurologic etiology. Computed tomography of the head was done which revealed an incidental anterior third ventricular lesion. There was no associated hemorrhage or mass effect. Magnetic resonance imaging (MRI) was subsequently performed and the reporting radiologist interpreted the imaging as suspicious for a chordoid glioma or craniopharyngioma. An open subtotal resection was completed via a frontal transcallosal trans-ventricular approach. The final pathological diagnosis was a chordoid glioma. During the follow-up period, the patient underwent permanent cerebrospinal fluid diversion in the form of a ventriculoperitoneal shunt. The patient also was seen in consultation by a radiation oncologist and it was elected to defer any radiotherapy and follow the residual lesion with frequent imaging.

Chordoid glioma is a relatively recently defined clinico-pathological entity with the first case series described in 1998 by Brat[1]. Since then, over 40 cases have been reported in the literature[1-16]. Patients have ranged in age from 7 - 70 years with a median age of 45; there have been only two documented cases in the pediatric population[6]. There is a female predilection although *in vitro* studies have failed to demonstrate estrogen or progesterone receptor reactivity[17]. Patients may present with symptomatic obstructive hydrocephalus, endocrine disturbance, or symptoms secondary to visual apparatus compression. This tumor has been classified a WHO grade II neoplasm which belies its histologically benign nature. Gross surgical excision is felt to be curative but is difficult to achieve given the location and adherent nature of this tumor[11]. There are no documented cases of delayed malignant transformation and no long-term studies of the natural history of this lesion as of yet.

Radiographically, this lesion demonstrates a number of consistent features. It is found in the anterior third ventricle although a single report has described an occipital horn location[6]. On CT, the lesion has well defined borders, hyperdense to brain and demonstrates uniform contrast enhancement. Magnetic resonance imaging is more informative owing to enhanced anatomic resolution. This lesion is felt to derive from the lamina terminalis and often demonstrates a pedicled origin. It is well circumscribed with an ovoid shape and an axis greatest in the superior to inferior direction. There is typically minimal reaction of the surrounding suprasellar and hypothalamic structures[15] and minimal displacement of surrounding structures. The optic chiasm is classically displaced inferiorly by the tumor mass.

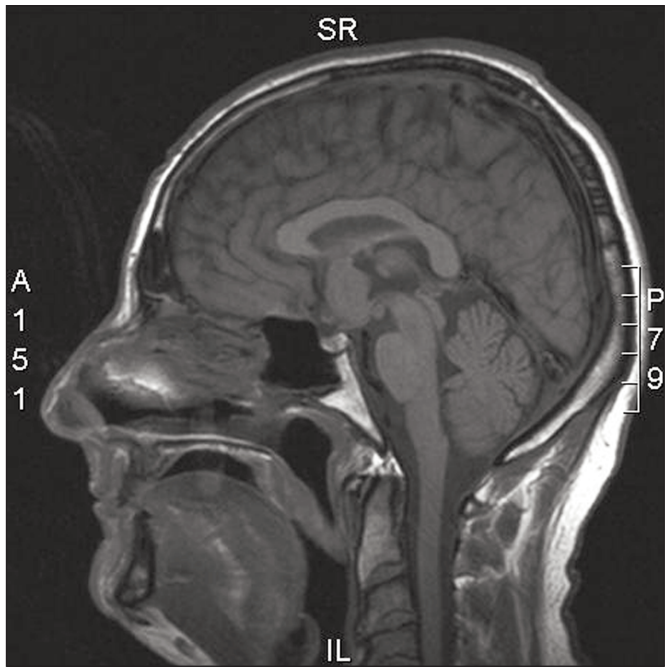

Figure 1: *MRI T1 sagittal section without gadolinium. The optic apparatus is inferiorly displaced by the tumor mass.*

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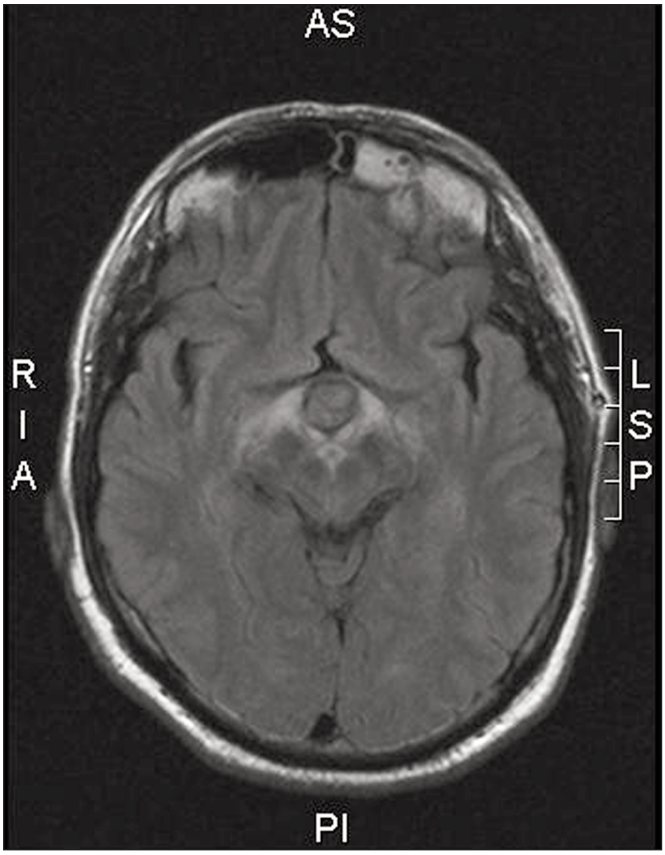

Figure 2: *MRI FLAIR Axial.*

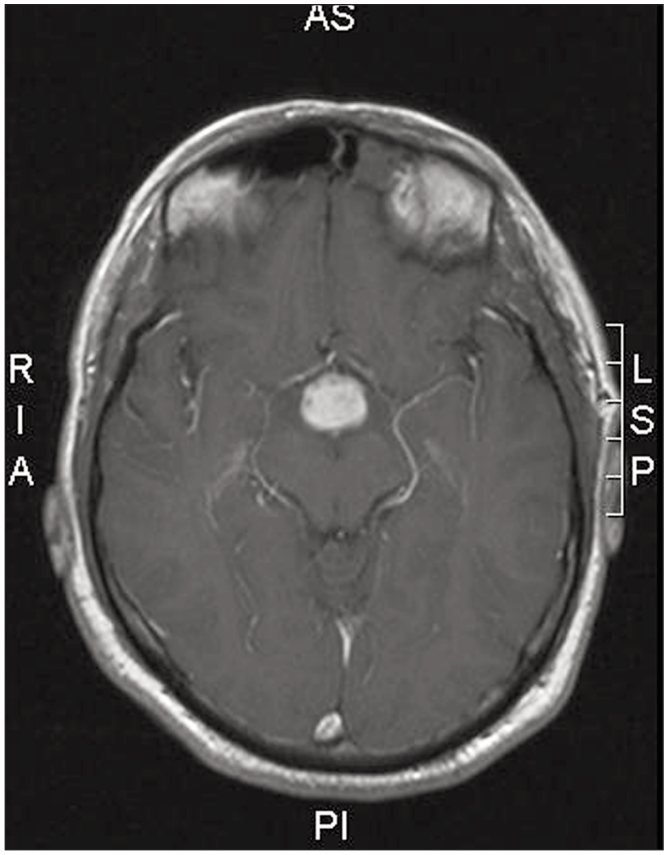

Figure 3: *MRI T1 Axial post gadolinium.*

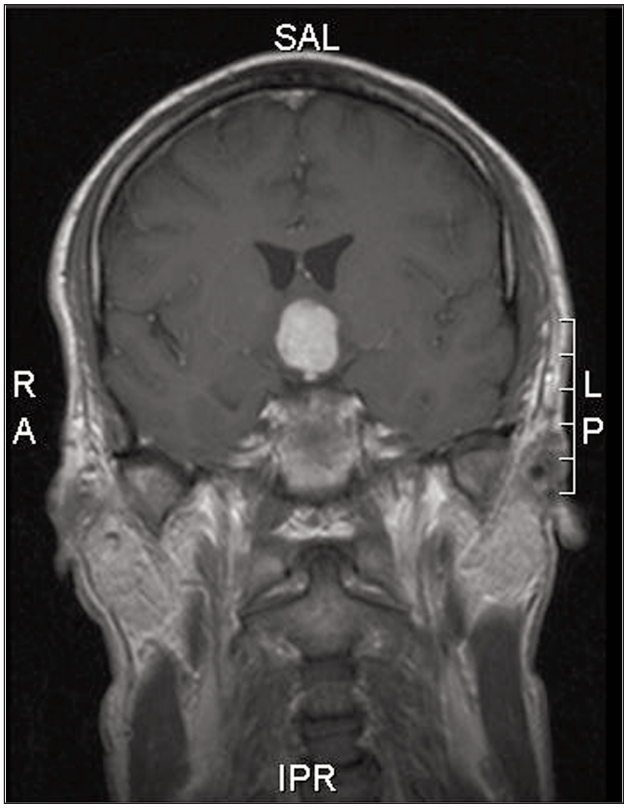

Figure 4: *MRI Coronal T1 post gadolinium. Lesion measures 2.3 x 1.9 x 1.6 cm at greatest dimensions. It is located in the anterior third ventricle adjacent to the hypothalamus. Note uniform enhancement and well-defined borders. Also note that lesion is distinct from and inferiorly displaces pituitary structures. There is also surrounding increased signal on FLAIR imaging in the adjacent lateral hypothalamus.*

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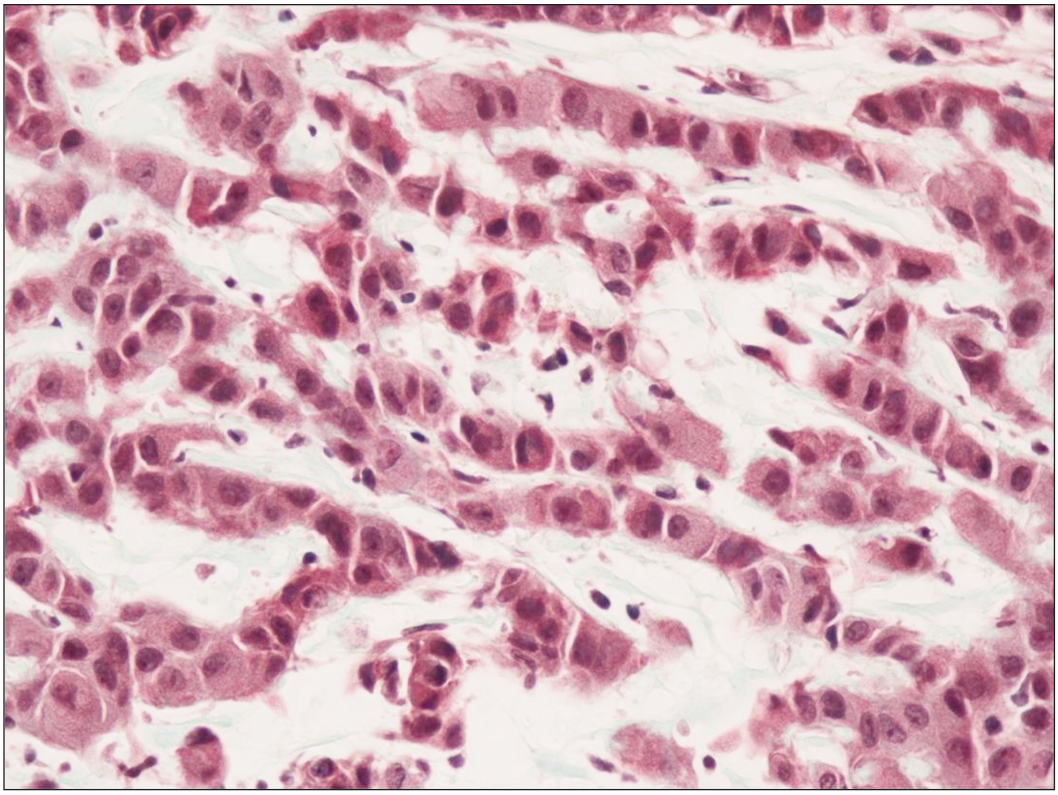

Figure 5: *Gomori trichrome stain showing trabecular arrangement of neoplastic cells mimicking chordoma.*

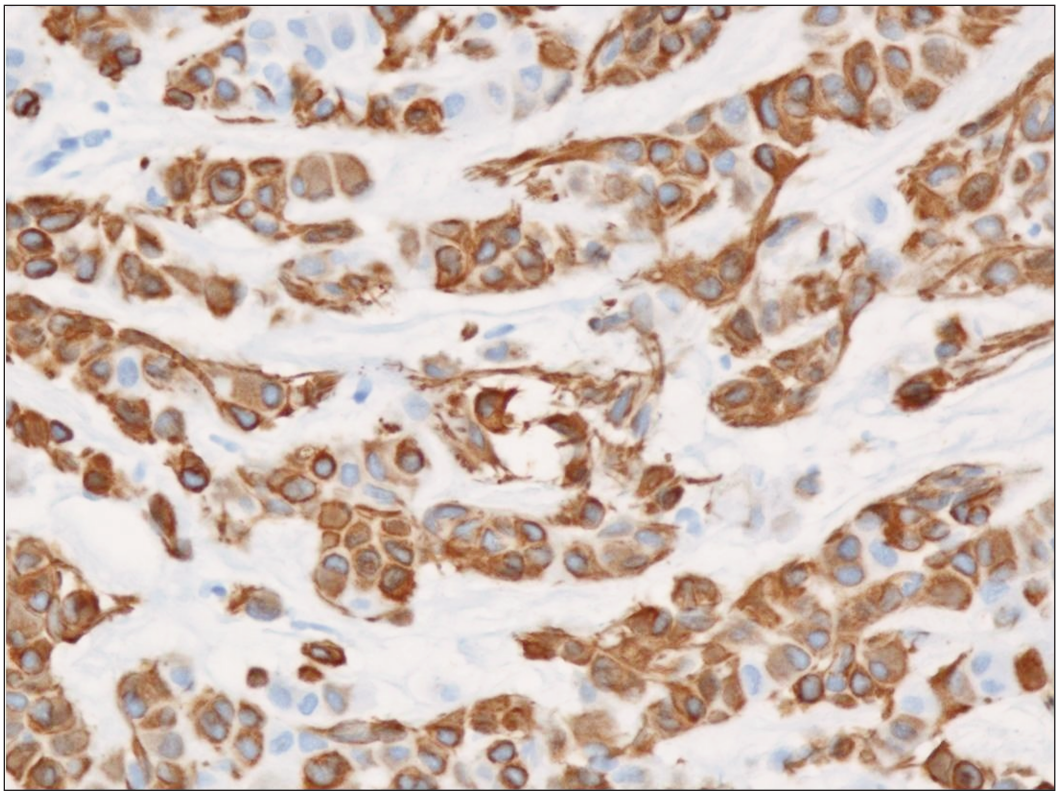

Figure 6: *Immunolabeling for Glial Fibrillary Acidic Protein (GFAP) confirms the glial nature of the neoplastic cell.*

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Furthermore this tumor is isointense on T1 and isointense to mildly hyperintense on T2 weighted imaging[6] although a small central cystic component has been observed in a minority of cases.[1,6,16] A number of different pathological entities occupy the suprasellar / third ventricular space. A comprehensive list can be found in most radiology textbooks. The intimate relationship of this lesion to the lamina terminalis, the displacement of the optic apparatus inferiorly, the predominantly solid enhancing characteristics of this lesion limit the differential diagnosis. The most common other lesions in the differential include a craniopharyngioma, germinoma or hypothalamic glioma.

Chordoid gliomas are uncommon lesions with an oncologically benign nature. While rare, this lesion should be considered in the radiographic differential of tumors of the anterior third ventricle.

References

1. Brat DJ, Scheithauer BW, Staugaitis SM, Cortez SC, Brecher K, Burger PC. Third ventricular chordoid glioma: a distinct clinicopathologic entity. J Neuropathol Exp Neurol. 1998;(57): 283-90.
2. Buccoleiro AM, Caldarella A, Gallina P, Di Lorenzo N, Taddei A, Taddei GL. Chordoid glioma: clinicopathologic profile and differential diagnosis of an uncommon tumor. Arch Pathol Lab Med. 2008;(128):141-5.
3. Castellano-Sanchez AA, Schemankewitz E, Mazewski C, Brat DJ. Pediatric chordoid glioma with chondroid metaplasia. Pediatr Dev Pathol. 2001;4(6):564-7.
4. Cenacchi G, Roncaroli F, Cesoli S, FIcara G, Merli GA, Giangaspero F. Chordoid glioma of the third ventricle: an ultrastructural study of three cases with a histogenetic hypothesis. Am J Surg Pathol. 2001;(25):401-5.
5. Grand S, Pasquir B, Gay E, Kremer S, Remy C, Le Bas JF. Chordoid glioma of the third ventricle: CT and MRI including perfusion data. Neuroradiology. 2002;(44):842-6.
6. Jain D, Sharma MC, Sarkar C, et al. Chordoid glioma: report of two rare examples with unusual features. Acta Neurochir. 2008; (150):295-300.
7. Kurian KM, Summers DM, Statham PF, Smith C, Bell JE, Ironside JW. Third ventricular chordoid glioma: clinicopathological study of two cases with evidence for a poor clinical outcome despite low grade histological features. Neuropathol Appl Neurobiol. 2005;(31):354-61.
8. Nakajima M, Nakasu S, Hatsuda N, Takeichi Y, Watanabe K, Matsuda M. Third ventricular glioma: case report and review of the literature. Surg Neurol May. 2003;59(5):424-8.
9. Pasquier B, Peoc'h M, Morrison AL, et al. Chordoid glioma of the third ventricle: a report of two new cases, with further evidence supporting an ependymal differentiation and review of the literature. Am J Surg Pathol. 2002;(26):1330-42.
10. Pomper MG, Passe TJ, Burger PC, Sheithauer BW, Brat DJ. Chordoid glioma: a neoplasm unique to the hypothalamus and anterior third ventricle. Am J Neuroradiol. 2001;(22):464-9.
11. Raizer JJ, Shetty T, Guntin PH, et al. Chordoid glioma: report of a case with unusual histologial features, ultrastructural study and review of the literature. J Neurooncol. 2003;(63):39-47.
12. Reifenberger G, Weber T, Weber RG, et al. Chordoid glioma of the third ventricle: immunohistochemical and molecular genetic characterization of a novel tumor entity. Brain Pathol. 1999;(9): 617-26.
13. Sato K, Kubota T, Ishida M, Yoshida K, Takeuchi H, Handa Y. Immunohistochemical and ultrastructural study of chordoid glioma of the third ventricle: its tancytic differentiation. Acta Neuropathol. 2003;(106):176-80.
14. Suh YL, Kum NR, Kim JH, Park SH. Suprasellar chordoid glioma combined with Rathke's cleft cyst. Pathol Int. 2003;(52):780-5.
15. Tarazewska A, Boguchi J, Andrychowski J, Koszewski W, Czernicki Z. Clinicopathological and ultrastructural study in two cases of chordoid glioma. Folia Neuropathol. 2003;(41):175-82.
16. Tonami H, Kamehiro M, Oguchi M, et al. Chordoid glioma of the third ventricle: CT and MRI findings. J Comput Assist Tomogr. 2000;(24):336-8.
17. Vajtai I, Varga Z, Sheithauer BW, Bodosi M. Chordoid glioma of the third ventricle: confirmatory report of a new entity. Hum Pathol. 1999;(6):723-6.