DOI: 10.7860/JCDR/2015/11163.5571

Case Report

Surgery Section

Rhabdoid Meningioma: Report of Two Cases

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ABSTRACT

Rhabdoid meningioma is an uncommon anaplastic variant of meningioma. We describe the clinicoradiological and histomorphological features of two such cases of meningioma, with a brief review of literature. One case in a 9-year-old girl, presented initially as an atypical meningioma in the right fronto-parietal region, which on recurrence 18 months later, evolved into a rhabdoid meningioma. The second case in a 33-year-old male was located in the right parieto-occipital region.

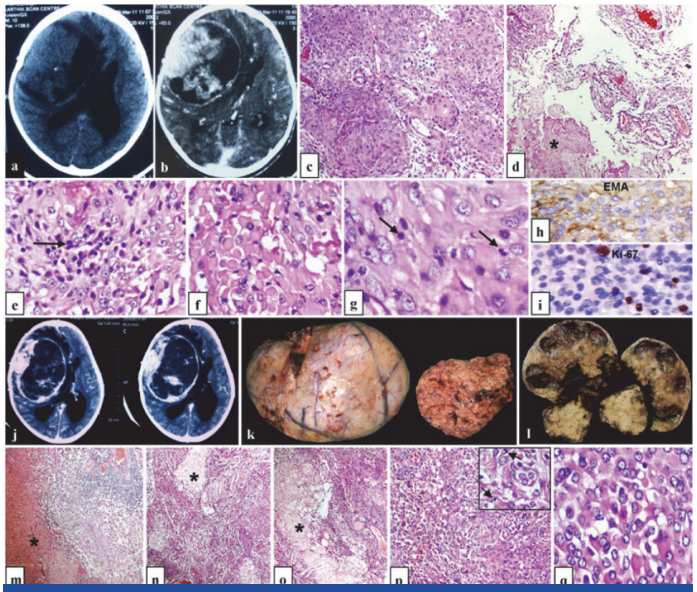
Keywords: Atypical, Anaplastic, Meningioma, Rhabdoid

CASE REPORT

Case 1: A female child presented at the age of 6 years 6 months, with twitching of eyelids and generalized tonic-clonic seizures of four months duration. There was past history of headache and vomiting of 15 days duration at the age of four, for which she has been treated conservatively with medication elsewhere (records of the same are not available) and the symptoms subsided then. Computed tomogram (CT) of brain showed large, well-defined heterogeneous lesion measuring 8 x 6.6 cms in the right fronto-parietal region with effacement of sulci and perilesional oedema [Table/Fig-1a], and heterogeneously contrast enhancing solid elements within [Table/Fig-1b]. Per-operatively the tumor was necrotic and highly vascular. Decompression of the tumor was done. Microscopy revealed polygonal to spindle-shaped neoplastic meningothelial cells arranged in sheets, whorls and fascicles [Table/Fig-1c] and interspersed necrosis [Table/Fig-1d]. Tumor cells with clear nuclei [Table/Fig-1c,e], moderate degree of nuclear pleomorphism, small cells [Table/Fig-1e], focus of rhabdoid cells [Table/Fig-1f] and 6-7 mitotic figures/10 high power field [Table/Fig-1g] were noted. The tumor cells were negative for glial fibrillary acid protein (GFAP: ready to use - BioGenex) and vimentin (ready to use - BioGenex) immunostaining, but showed strong cytoplasmic membrane positivity for Epithelial membrane antigen (EMA: ready to use – Bio Genex) [Table/Fig-1h]. Ki-67 (ready to use - BioGenex) labelling index was 8% [Table/Fig-1i]. Histolopathological diagnosis was atypical meningioma – WHO grade II.

The patient was lost to follow-up and presented 18 months later, at the age of 9 years, with left hemiparesis and tonic-clonic seizures of one month duration. The CT scan brain showed similar radiological picture of heterogeneously enhancing lesion with effacement of sulci and perilesional oedema [Table/Fig-1j]. Right fronto-parietalcraniotomy with gross total excision of the tumor was done. Grossly the recurrent tumor exhibited marked hemorrhage, cystic change and necrosis [Table/Fig-1k,l]. Histological examination revealed hemorrhage, [Table/Fig-1m], cystic change with foamy histiocytes [Table/Fig-1n], necrosis [Table/Fig-1o], predominantly sheets of rhabdoid cells [Table/Fig-1p,q], moderate nuclear atypia and 16-18 mitotic figures/10 high power fields [Table/Fig-1p inset]. Histopathological diagnosis was Rhabdoid meningioma (RM) – WHO (World Health Organization) grade III. The child recovered from hemiparesis and there were no seizures till three months period of follow-up and is undergoing 54 Gy fractionated radiotherapy.

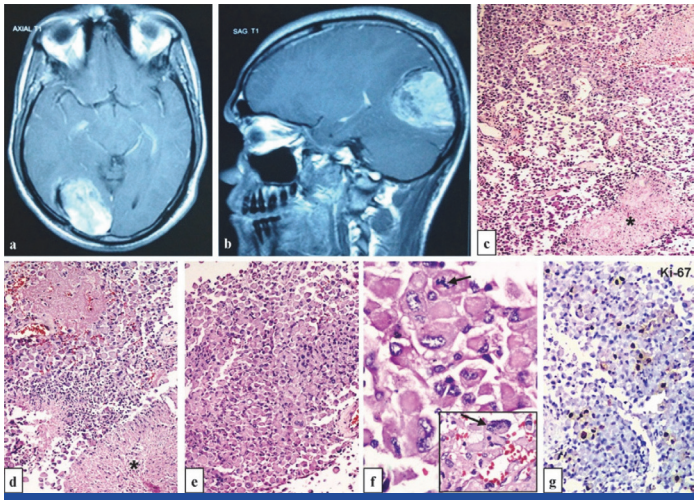
Case 2: A 33-year-old male presented with left sided hemiparesis since six months, vomiting and giddiness since two months, and left-sided upper motor neuron type facial weakness since one month. Axial and sagittal T1 weighted, post-gadolinium images showed a predominantly homogeneously enhancing lesion in the right parieto-occipital region measuring 5 x 4 x 5 cms. The lesion is buckling and effacing the adjacent sulci and attached to the adjacent falx cerebri and adjacent cerebrospinal fluid (CSF) cleft confirming the extra-axial location [Table/Fig-2a,b]. Right parietal craniotomy and complete excision of lesion was done. Histopathology revealed a tumor composed entirely of rhabdoid cells in dyscohesive sheets [Table/Fig-2c] with multiple, interspersed foci of necrosis [Table/Fig-2d]. The rhabdoid cells had eccentric round to oval nuclei, fine to coarse chromatin and prominent nucleoli, abundant eosinophilic cytoplasm [Table/Fig-2e]. Moderate nuclear pleomorphism [Table/Fig-2f], bi- and multinucleation [Table/Fig-2g], atypical mitosis [Table/Fig-2f] and 8-10 mitotic figures/10 high power fields were noted. The Ki-67 labelling index was 10-12%. Histopathological diagnosis was RM – WHO grade III. Postoperatively there was improvement in the neurological deficit and subsequently the patient was discharged, but later lost to follow-up.



[Table/Fig-1]: Axial CT: right fronto-parietal, solid-cystic lesion (a), contrast enhancing (b), perilesional oedema and midline shift. Tumor cells in sheet and whorls (cx100) with necrosis (d x40;asterix), small cells (e x400; arrow), focus of rhabdoid cells (fx400), mitosis (gx400; arrow). EMA positivity (hx400). Ki-67 index: 8% (ix400). Axial contrast CT: recurrent tumor (j). Solid tumor with hemorrhage, necrosis and cystic change (k,l). Foci of hemorrhage (m), foamy histiocytes (n) and necrosis (o) [x100; asterix], sheets of rhabdoid cells (px100,qx400), and mitosis (px400; inset arrow)

DISCUSSION

Meningioma arises from the arachnoidal cap cells of arachnoid villi in meninges. They are classified / graded as benign, atypical and anaplastic meningiomas. Kepes et al., [1] and Perry et al., [2] first described RM in the same year for a series of aggressive meningiomas with rhabdoid morphology. It is a named variant of anaplastic meningioma and belongs to WHO grade III category [3].



[Table/Fig-2]: Contrast MRI - right parieto-occipital, heterogeneously contrast enhancing lesion (a axial T1W; b sagittal T1W). Tumor has foci of necrosis (cx40,dx100;asterix), sheets of rhabdoid cells (e x100), nuclear pleomorphism, atypical mitosis (fx100; arrow) and multinucleation (fx400; inset). Ki-67 index of 12%(g x100)

Meningiomas account for up to 24-30% of all primary intracranial neoplasms and majority have a benign course. Incidence of anaplastic meningioma is 1–2.8%, and is associated with increased recurrence and shorter median survival [3]. RM is an uncommon, aggressive meningioma classified under anaplastic – grade III tumor [3]. Review of literature shows RM accounts for 0.004% of all meningiomas, with 102 published cases [4]. These tumors have an increased propensity for local recurrence, remote metastasis [5] and CSF dissemination [5,6]. A minority of meningiomas show rhabdoid features focally and other histological features of malignancy [5]. Rhabdoid cells may become increasingly evident with tumor recurrences [3] and majority of RMs are a result of secondary rhabdoid transformation in recurrent meningiomas [5], as seen in case 1.

The tumors are predominantly isointense on T1-weighted images and hyperintense on T2-weighted images. Homogeneous enhancement on contrast is seen in majority of lesions. Cystic components and heterogeneous enhancement is also encountered. Peritumoral oedema with marked mass effect (as in case 1), hyperostosis and bony destruction are frequent features [7].

In majority of the tumors necrosis is seen with clustering of neoplastic cells around blood vessels [5]. Vesicular, eccentrically located nuclei with distinct nucleoli and abundant cytoplasm containing eosinophilic hyaline inclusions, is typical of rhabdoid morphology. Rhabdoid cells stain diffusely and intensely positive for vimentin and EMA, focally positive for S-100 and pancytokeratin, and negative for GFAP, smooth muscle actin, desmin, carcinoembryogenic antigen, thyroid transcription factor-1, thyroglobulin, calcitonin, synaptophysin, chromogranin, estrogen and progesterone receptors [8]. The rhabdoid areas of the tumors exhibit high mitotic activity with a variable Ki-67 labelling index of 30% [8] to 80 - 90% [5]. In recurrent tumor of case 1 Ki-67 labelling study was not done, owing to histological features of high grade neoplasia and obvious rhabdoid morphology. Ki-67 labelling of 10-12% in case 2, still connotes this tumor as RM, because of characteristic microscopic features. Rhabdoid meningiomas have been reported to coexist with papillary and clear cell meningioma [6,8].

The differential diagnoses of RM are atypical teratoid/rhabdoid tumor (AT/RT) and metastatic carcinoma [5]. Metastatic carcinoma can be ruled out by proper history elucidation, extensive radiological investigation and panel of immunohistochemical markers. Intra-axial location of tumor and positivity for glial, mesenchymal, epithelial and neuronal immunohistochemical markers and absence of SNF5 (INI1) protein expression, are diagnostic of AT/RT [3].

Prognosis of meningioma depends on histological grade and extent of resection. Patients harboring either WHO grade II or III meningiomas have higher recurrence rates, varying between 29-52 and 50-94%, respectively [9]. Simpson grade IV resection is associated with a significantly shorter recurrence free survival (RFS) than Simpson grade I, II, or III resection ($p<0.001$). Statistically no significant difference was noted in RFS between Simpson grades I, II, and III resections [10]. Accordingly adjuvant radiotherapy is recommended for subtotally resected WHO grade II and all grade III meningiomas [9].

CONCLUSION

Rhabdoid meningioma is an aggressive tumor, with propensity of remote metastasis and CSF dissemination, and mimics metastatic carcinoma and AT/RT, microscopically. Complete excision along with dural attachment, appropriate histological diagnosis and grading, and adjuvant radiotherapy are imperative for proper management.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Sep 08, 2014**
Date of Peer Review: **Nov 28, 2014**
Date of Acceptance: **Dec 30, 2014**
Date of Publishing: **Feb 01, 2015**