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CASE REPORT

Rhabdoid meningioma lacking malignant features: Report of a rare case with review of literature

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ABSTRACT

We reported a case of meningioma with rhabdoid morphology but lacking histological features of malignancy in arising from the spinal cord in a 28-year-old male. The tumor showed light microscopic, immunohistochemical evidence of meningothelial differentiation together with diffuse areas exhibiting rhabdoid morphology. The rhabdoid areas were characterized by cells with large cytoplasmic eosinophilic inclusions and eccentric nuclei. Unlike most cases reported in the literature, this case lacked significant mitotic activity and other atypical features. The diagnostic and prognostic significance of this tumor entity is discussed along with a review of the literature.

Key words: Meningioma, rhabdoid, spinal cord

Introduction

The WHO classification of central nervous system (CNS) tumors published in 2000, includes a number of meningioma subtypes, many of which are descriptive and of no prognostic significance.[1] This classification treats rhabdoid meningiomas as a distinct subtype due to their aggressive behavior and high rates of recurrence. One interesting feature of rhabdoid meningiomas is that in a significant number of cases, the rhabdoid cells appear after initial recurrence. We report a case of spinal meningoma in a 28-year-old male that showed "rhabdoid" morphology at the initial presentation.

Case Report

A 28-year-old male presented with a history of progressive weakness and decreased sensation in both lower limbs. He also complained of weakness and numbness up to the nipple. There was no bladder involvement. On examination, there was Grade I to II weakness in both lower limbs and sensory loss (touch, pain, temp) below T3 and T4. Magnetic resonance imaging revealed the intradural extramedullary tumor at C7-T1 level [Figure 1]. Laminectomy C7-T1 was done with complete excision of intradural, extramedullary well circumscribed, firm, vascular tumor arising from dura C7-T1 and compressing the cord significantly. On microscopic examination, of the resected tumor, most of the tumor cells exhibited typical rhabdoid morphology with large, vesicular, often eccentrically located nuclei with distinct nucleoli and abundant cytoplasm containing eosinophilic hyaline inclusions [Figure 2]. Numerous psammoma bodies were also seen. Classical meningothelial features with focal whorl formation were scarce and seen only in a few foci. Immunohistochemically, these rhabdoid cells showed epithelial membrane antigen (EMA) as well as vimentin positivity. By 7th postoperative day, significant improvement was observed, and patient got Grade III power. Sensory loss also started improving. The postoperative period of this patient was uneventful and after surgery was planned for radiotherapy and chemotherapy. Patient has not so far presented with recurrences, 6 months after radiotherapy and chemotherapy.

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Discussion

Although meningiomas are well-known entities with frequent histological variants, Rhabdoid meningioma is a rare subtype of meningiomas accounting for 1–3% of all intracranial meningiomas.[2] Rhabdoid meningiomas were described for the first time in 1998 as an unusual variant with increased proliferative activity.[1,3,4] In 2000, they included in the revised WHO classification of tumors of the CNS as a subtype of meningiomas with increased risk of recurrence and more aggressive growth, corresponding to WHO Grade III. Meningiomas with focal rhabdoid differentiation are considered WHO Grade I. Rhabdoid features should constitute

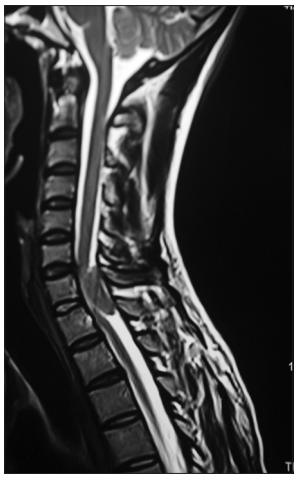

Figure 1: Magnetic resonance imaging revealing intradural extrarmedullary tumor at C7-T1 level

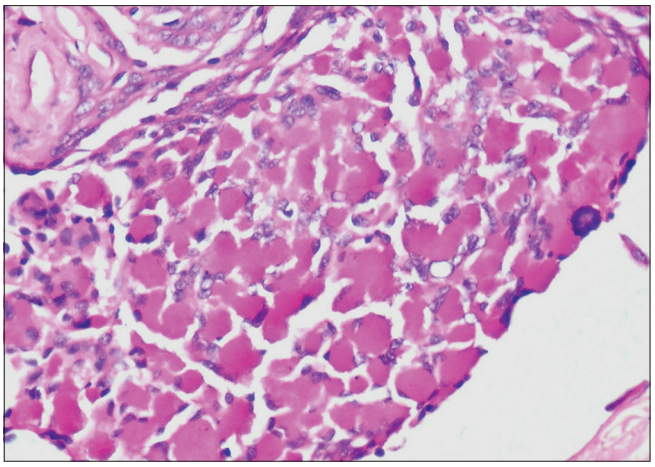

Figure 2: Photomicrograph showing cells with "rhabdoid" morphology. A psammoma body is also seen at the periphery (H and E, ×40)

over 50% of tumor to call a meningiomas as the rhabdoid meningioma (WHO Grade III).[1]

Rhabdoid morphology in tumors refers to the resemblance of the cells to rhabdomyoblasts, without true skeletal muscle differentiation. The cytologic features include abundant eosinophilic cytoplasm, eccentric nuclei, and intracytoplasmic hyaline inclusions. Ultrastructurally, the latter represents whorls of intermediate filaments expressing vimentin and sometimes cytokeratin. The term malignant rhabdoid tumor (MRT) was first used to describe a distinctive pediatric renal tumor.[5] The term was then extended to similar extrarenal pediatric tumors (extrarenal MRT), including atypical teratoid/rhabdoid tumor of the CNS.[6] A rhabdoid phenotype has also been described in a variety of tumors with a different histogenesis, including carcinomas, sarcomas, gliomas, and melanomas.[7]

In 1998, Kepes *et al*.[8] and Perry *et al*.[9] described the first two series of meningiomas with rhabdoid transformation. These meningiomas often recur, and the rhabdoid features become more apparent in subsequent biopsies. Brain invasion, anaplasia, and extracranial metastasis have been reported in rhabdoid meningiomas. Besides the rhabdoid morphology, cytoarchitectural features of atypical meningioma (four or more mitoses per 10 high-power fields, high cellularity, sheeting architecture, nuclear atypia, and necrosis) were seen in most cases. Expression of cytokeratin is common. The wide histological differential diagnosis includes metastatic carcinoma, metastatic melanoma, glioma, and sarcoma. Diagnosis depends on finding evidence of meningothelial differentiation either by light microscopy (whorls, intranuclear pseudoinclusions), immunohistochemistry (expression of vimentin, EMA, and progesterone–receptor positivity), or by electron microscopy (interdigitating cytoplasmic membranes, intercellular junctions).

Perry *et al*.[9] studied a series of 15 meningiomas with rhabdoid features. Histological features of malignancy (brain invasion or anaplasia) were observed in nine cases, and another two tumors being considered malignant on the basis of extracranial metastasis. In the majority, increased cell proliferation was evidenced by a high mitotic rate or MIB-1 labeling index. On follow-up, 13 patients (87%) experienced at least one recurrence and 8 (53%) died of disease. Median time to death was 5.8 years after initial surgery and 3.1 years after the first appearance of rhabdoid morphology.

Kepes *et al*.[8] studied a small series of rhabdoid meningiomas and concluded that rhabdoid transformation of tumor cells in meningiomasis is a histologic indication of increased proliferative activity. They further suggested that rhabdoid meningiomas are highly aggressive tumors, and that the rhabdoid phenotype represents a marker of malignant transformation in meningiomas. However Cooper *et al*.[10] have reported 2 cases of meningiomas with rhabdoid morphology but lacking histological features of malignancy. The tumors showed light microscopic, immunohistochemical and ultrastructural evidence of meningothelial differentiation together with diffuse or focal areas exhibiting rhabdoid morphology. Unlike most cases reported in the literature, these "rhabdoid meningiomas" lacked significant mitotic activity or other atypical features. These cases are similar to our case as there was no evidence of atypia/malignancy in our case. Ho *et al*.[11]suggested that any histological rhabdoid features in recurrent meningiomas or even in primary cases seem to indicate the malignant clinical course and the need for aggressive treatments, because transformation from a benign or atypical one to a malignant one seems to occur at last.

However, in the recent literatures, reports of rhabdoid meningiomas have included cases having the rhabdoid cells in the primary meningioma and cases with a range of the histological atypia from apparently benign to frankly

malignant.[4,8,9] However, until date there are still a lot of arguments about their developmental patterns and the treatment strategy especially for rhabdoid meningiomas lacking other histological features of malignancy.[11]

Very few cases of the rhabdoid meningioma in the spinal location have been reported. Endo *et al*.[12] have described a case of rhabdoid transformation of recurrent meningioma in the cervical cord. At each recurrence of the tumor, they found that population of the rhabdoid cells had increased, and the ability to grow had also increased as was confirmed by the MIB-1 labeling index. They concluded that phenotypic change of the cells with "rhabdoid" morphology may affect meningiomas and that such changes are associated with aggressive biological and clinical behavior. Recently Jeong *et al*.[13] have reported a case of primary intraspinal rhabdoid papillary meningioma with primary manifestation in the spinal cord of a 72-year-old female. They concluded that search for eosinophilic hyaline cytoplasm, rather than a fibrillary one, is critical for distinguishing it from other commonly encountered spinal cord tumors in the total absence of meningothelial whorls.

Rhabdoid meningiomas are associated with rapid growth, brain invasion and frequent recurrences Surgery is the sole modality of treatment followed by the conventional fractionated radiation therapy, usually to doses ranging from 50 to 60 Gy of radiation via conventional fractionation and delivery techniques. Currently, most investigators would recommend oppressively 54 Gy for benign meningiomas, following incomplete resection and upto 60 Gy for meningiomas that have atypical or malignant features.[14]

Conclusion

It is important to recognize rhabdoid morphology in meningiomas because this subtype is associated with recurrences, metastatic potential, sometimes frank malignancy, and poor outcomes. Close follow-up and aggressive treatment is mandatory in such cases.

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