

# Rhabdoid Meningioma In A Background Of Atypical Meningioma With Lipomatous Metaplasia: Case Report And Review Of Literature

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## Abstract

Meningiomas are heterogenous in their histology, and most of histologic subtypes have no prognostic significance, and are classified as WHO grade 1. Those with clinically increased risk of recurrence are classified as WHO grade II or III. A relatively recent addition to the meningioma family is a rare variant called rhabdoid meningioma, which is associated with increased risk of recurrence and aggressive clinical behavior and therefore classified as WHO grade III meningioma. We report a case of rhabdoid meningioma in a 43 year old man, which exhibited an unusual combination of rhabdoid and lipomatous components, in a background of atypical meningioma. The clinical features, neuroimaging, surgical treatment results and pathological features of this case are described with a limited review of literature regarding prognosis of patients and treatment options.

## INTRODUCTION

The term rhabdoid meningioma (RM) is used to describe tumors with clinical and radiological features of meningioma, but histologically formed of rhabdoid cells (1), or more commonly a conventional meningioma which may show rhabdoid transformation on successive recurrences (2). This new variant was first described in 1991 (3), it has increased risk of recurrence with relatively poor prognosis, and is classified as WHO grade III meningioma (4, 5). We report a case of a 43 year old man with a left frontal meningioma showing prominent rhabdoid cellular features and lipomatous changes in a background of atypical meningioma. This case is presented with emphasis on the importance of identification of rhabdoid component and a limited review of the literature regarding prognosis of patients and treatment options.

## CASE REPORT

**History:** A 43 years old man with history of hypertension, presented with complaints of dizziness, loss of balance and headaches for the last 7 or 8 months. He also reported short-term memory and word finding difficulties and spatial impairment with math tasks.

**Radiology:** MRI showed a left-sided dural-based convexity lesion with homogeneous enhancement located at the region

of motor and premotor cortices (Figure 1A).

**Examination:** Physical examination demonstrated full strength upper and lower extremities with normal muscle bulk and tone. No cranial nerve deficit was present. Neuropsychological assessment revealed high-average superior cognitive function. However, his performance on verbal tasks was impaired clinically, and lower than his performance on non-verbal tasks.

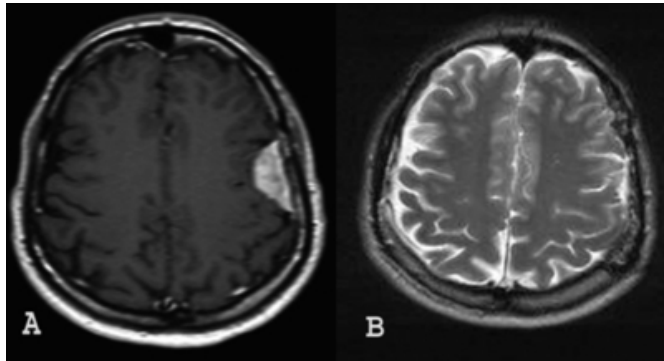
**Operation:** The tumor was removed in toto. No gross evidence of brain invasion was identified. The bone flap looked normal, however, it was decided to shave it with 5mm cutting burr over the entire tumor area for about 3-5mm in depth.

**Post-operative course:** The patient was discharged on the third day of surgery. During a period of two weeks following surgery he suffered from 9 to 10 attacks of numbness of right hand associated with numbness of right perioral area. He had two episodes of speech arrest with no loss of consciousness that lasted for about 15 minutes each. These seizures were controlled by an anticonvulsant. MRI scan eight months post surgery showed no residual tumor (Figure 1B). On last follow-up (eleven months status post surgery), he was doing well with full strength of the upper and lower extremities with normal cranial examination and mentation. No post-

operative radiation or chemotherapy was instituted.

**Figure 1**

Figure 1: A. MRI showed a left-sided dural-based convexity lesion with homogeneous enhancement located at the region of motor and premotor cortices. B. MRI scan eight months post surgery showed no residual tumor.



**PATHOLOGICAL FINDINGS**

Gross: The tumor was received as a 3.5 x 3 x 2.2 cm soft tan rounded mass with dura attached along one aspect of the tumor. The remaining surface was grey-tan granular and uneven. Serial sectioning revealed solid grey-tan cut surface.

Microscopically, the tumor showed atypical meningioma with prominent rhabdoid areas, admixed with multi-focal lipomatous metaplasia (Figure 2A). The rhabdoid areas were formed of nodular to sheet-like aggregates of cells with abundant eosinophilic cytoplasm and eccentrically placed nuclei (Figure 2B). Nucleoli were inconspicuous and nuclear pleomorphism was mild. Occasional, mitotic figures were present, however, necrosis was absent. Tumor cells with rhabdoid features constituted approximately 80% of the neoplasm. Vimentin immunostain highlighted cytoplasm containing whorled intermediate filaments which pushed the nucleus to one side (Figure 2C).

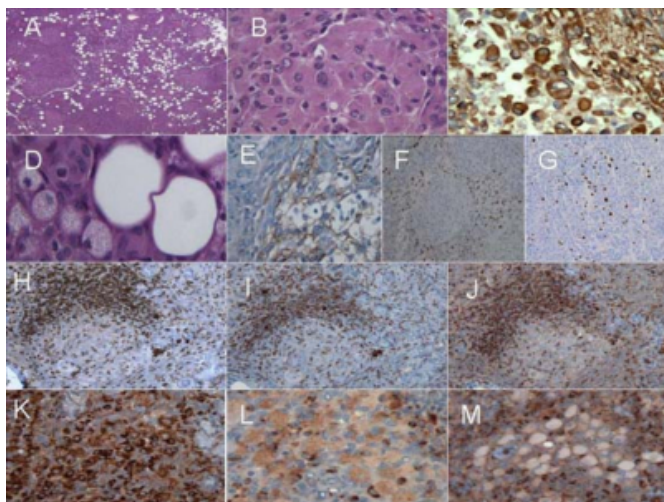
The component of atypical meningioma exhibited areas with high cellular density, sheeting, spindled and small cells with high nuclear cytoplasmic ratio, moderate nuclear pleomorphism and prominent nucleoli. Mitotic figures were up to 4/10 HPF, however, no necrosis was seen.

The lipomatous component (Figure 2D) was composed of clusters, as well as individually scattered cells with finely vacuolated cytoplasm around a central nucleus or cells with nucleus pushed to the periphery by a large fat vacuole, or fatty cysts (likely formed by rupture of two or more lipidized cells). Attached fragments of dura showed involvement of dural sinuses by meningioma. No brain invasion was seen.

Immunohistochemical staining demonstrated focal EMA positivity in all tumor components, including the lipidized cells (Figure 2E). The cells in the rhabdoid component were mostly negative for progesterone receptors compared to approximately 30% of tumor cells staining in the non-rhabdoid (atypical meningioma) component (Figure 2F). Proliferation marker Ki-67 revealed staining of up to 15% nuclei in focal areas of both the rhabdoid as well as the atypical meningioma components (Figure 2G). All tumor components including rhabdoid, atypical and lipomatous exhibited positivity for CD 68, and two lysosomal proteases cathepsin B (CB) and cathepsin H (CH). For all three (CD68, CB, CL) positivity was more pronounced in the atypical component (Figure 2 H, I, J; respectively), as compared to the rhabdoid areas (nodular area in the center).

**Figure 2**

Figure 2: A. Nodular rhabdoid areas are seen in a background of atypical meningioma with lipomatous metaplasia; B. Rhabdoid cells showing abundant eosinophilic cytoplasm with eccentric nuclei, inconspicuous nucleoli and mild nuclear pleomorphism; C. Vimentin immunostain highlighting the cytoplasm containing whorled intermediate filaments; D. Lipidized cells with finely vacuolated cytoplasm and fatty cysts; E. Lipidized cells with EMA positivity suggesting a meningotheial origin of these cells. F. The cells in the rhabdoid component (nodular area shown in the center) were mostly negative for progesterone receptors compared to approximately 30% of tumor cells staining in the non-rhabdoid (atypical meningioma) component. G. Proliferation marker Ki-67 revealed staining of up to 15% nuclei in focal areas of both the rhabdoid as well as the atypical meningioma components. H, I, J. Positivity for CD68 (H); cathepsin B (I) and cathepsin H (J) was more pronounced in the atypical component (top left), as compared to the rhabdoid areas (nodular area in the center of picture). K, L, M. The lipidized (foam) cells exhibited strong cytoplasmic staining for CD68 (K); moderate for cathepsin B (L); and weak for cathepsin H (M).



Lipidized cells were strongly positive for CD68, moderately for CB and weakly for CH (Figure 2 K, L, M; respectively). All tumor components (rhabdoid, non-rhabdoid and lipomatous) were negative for GFAP, cytokeratin and desmin.

**DISCUSSION**

Conventional meningiomas with lipomatous metaplasia although uncommon have been described in literature, and are included under metaplastic meningiomas in the WHO classification (5). Our case represents a very unusual combination of a rhabdoid meningioma occurring in a background of atypical meningioma with lipomatous metaplasia, which has not been reported in English literature,

to the best of our knowledge.

The focal EMA positivity in the areas of lipomatous metaplasia suggested meningotheial origin of these lipid laden cells. Although CD68 was positive in the foam cells, these cells could not be interpreted as histiocytic cells, since most of the meningioma cells, especially those constituting the atypical component also expressed strong CD68 positivity. Lysosomal proteases, cathepsins B and H were also expressed in the neoplastic meningotheial cells, their expression was relatively more abundant in the atypical meningioma component as compared to the rhabdoid component. This differential expression of CD68, CB and CH, in the rhabdoid and atypical meningioma components may suggest that, although both components are aggressive variants of meningioma, they likely acquire divergent pathways to achieve this goal. The marked loss of expression of progesterone receptors in the rhabdoid component compared to the atypical meningioma component further suggests that these two aggressive variants acquire divergent molecular pathways.

Review of data from 35 previously reported cases of RM revealed that overall recurrences occurred in 19 out of 35 cases (6). The presence of rhabdoid morphology in diverse, histogenetically unrelated tumors including pediatric tumors (7) suggested that the rhabdoid pattern is a phenotypic expression associated with an aggressive behavior (8). Kepes et al. in 1998 (4) suggested that the rhabdoid phenotype represents a marker of malignant transformation in meningiomas. The subsequent study of Perry et al. (1) on 15 cases of RM revealed that in 7 of 15 patients that died of disease, the median time of death from initial surgery was 5.8 years, and from the first appearance of rhabdoid morphology was 3.1 years. Unfortunately, in cases with recurrence they did not describe the histologic subtype and grade of meningioma at the time of initial surgery for the meningioma. They only describe the histologic type and grade of meningioma when rhabdoid morphology was noted. Nevertheless, the above study like that of Kepes et al., (4) also concluded that rhabdoid phenotype was a marker of aggressive behavior/ malignant transformation. Molecular changes including loss of heterozygosity of chromosome 22 and NF-2 gene mutation are considered to be among the events that lead to initiation of meningiomas. Both of these markers are present in 50% of sporadic meningiomas, while the latter is present in 100% of NF-2 meningiomas. However, molecular changes specific to aggressive variants

of meningiomas including rhabdoid phenotype have not been well-characterized.

The prognosis of meningiomas is heavily weighed upon extent of surgical excision in addition to the histologic subtype/ grade of tumor. The resection of dural base along with complete removal of tumor has been shown to reduce recurrence rate by half (9,10). In our case, the tumor was removed in toto along with its dural base. Although, the bone flap appeared normal, however, it was decided to shave it with 5 mm cutting burr over the entire tumor area for about 3-5 mm in depth. No radiation or chemotherapy was instituted post-surgery. Eleven months post-surgery, the patient was free of recurrence. In cases where post-operative imaging reveals residual tumor, perhaps due to critical location of tumor, management may include observation, irradiation or re-operation. Post-operative adjuvant radiation therapy should be considered if the residual meningioma is WHO grade 2 or 3 (atypical, rhabdoid, anaplastic etc). Meningiomas that recur, warrant surgical re-excision and/or radiotherapy. If surgery is not feasible, radiation should be given, especially to WHO grade 2 and 3 meningiomas. Since rhabdoid meningiomas are WHO grade III, they should be managed accordingly.

Some cases of meningioma have only focal rhabdoid changes and lack obvious features of atypical or malignant meningioma (11). In addition, RM in pediatric patients seem to be less aggressive than that of their adult counterparts (6). The behavior of such category of meningiomas remains to be determined with follow up studies on a larger number of cases.

## **SUMMARY**

In summary, we present a rhabdoid meningioma with an unusual combination of lipomatous metaplasia in a background of atypical meningioma, from a the left temporal lobe of a 43 year-old man. The rhabdoid and atypical meningioma components showed distinct differences in the expression of CD68, progesterone receptors and two lysosomal proteases CB and CH, suggesting acquisition of

different pathways, each leading to an aggressive behavior. The presence of rhabdoid features, even focal, in a classic meningioma should be reported and quantified in the diagnosis. This is of importance in view of most current literature that suggests that meningiomas harboring a rhabdoid cellular component may have increased risk of recurrence and progression to aggressive behavior with successive recurrences.

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