**Rhabdoid Meningioma in a Eight Year Old Child**

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**CASE REPORT SUMMARY**

**Introduction:** We report a case of Rhabdoid meningioma in a eight year old child which is the fifth case in the world according to our knowledge. Material and method: A 8 – year old girl was seen in our University Clinic of Neurosurgery with one month history of increasing headaches, vomiting and increasing frequency of grand mal seizures (known history of epilepsy). Her past medical history was not significant, except the epilepsy. After opening the dura mater, an extra axial tumor was found. The neoplasm invaded the brain and was highly vascular. The lesion was totally excised and 2cm of adjacent dura mater was excised – Simpson O. Results: The postoperative course was without significant events. The patient was discharged home on the seventh postoperative day. Seizures were controlled with anticonvulsant therapy. Postoperative control head CT scan demonstrated total removal of the lesion. HP analysis: Paraffin-embedded tissue sections stained with H&E revealed meningothelial tumor with rhabdoid morphology characterized by sheets of tumor cells with eccentric nuclei, variably abundant eosinophilic cytoplasm and intracytoplasmic hyaline paranuclear inclusions. The number of mitoses was up to four in ten high-power fields, and the Ki-67 proliferation index was 4,4%. The histopathological diagnosis was rhabdoid meningioma (grade III). Discussion: Radical surgery (Simpson grade 0) has been shown to significantly enhance prognosis in atypical and malignant meningiomas. Conclusion: Rhabdoid meningioma is an anaplastic, very rare subtype of malignant meningioma. The prognosis for rhabdoid meningioma depends on their proliferative activity and the possibility of radical removal.

**Keywords:** childhood brain tumor, extrarenal rhabdoid tumor, malignant meningioma, rhabdoid meningioma

1. **INTRODUCTION**

Meningiomas occur in 1%~4% of primary intracranial tumors in the pediatric group, and is increasing in incidence with age (1,2,3,4,5). There is a marked female predominance in adults, while in children these tumors affect both sexes equally (6,7,8,9). Since Beckwith and Palmer introduced the term ‘rhabdoid tumor’ in 1978 in reference to a subgroup of childhood malignant renal tumors, many tumors with a rhabdoid morphology have been reported in various sites, including the central nervous system. In 1998 Kepes et al. and later Perry et al. both described two series with clinically aggressive tumors that were a histologically distinct subgroup of malignant meningioma. They suggested the term rhabdoid meningioma which was adopted into the WHO classification (grade III) in 2000. Rhabdoid meningioma appears to have a particularly poor prognosis and is classified as a Grade III neoplasm in the 2002 World Health Organization classification. We report a case of Rhabdoid meningioma in a eight year old child which is the fifth case in the world to the best of our knowledge (10,11,12,13).

2. **MATERIAL AND METHOD**

A 8 – year old girl was seen in our University Clinic of Neurosurgery with one month history of increasing headaches, vomiting and increasing frequency of grand mal seizures (known history of epilepsy). Her past medical history was unremarkable – except epilepsy. On examination she was conscious and oriented. The girls' neurological examination showed light hemiparesis on her left side. Muscle power on left limbs was grade 4-/5. No neurocutaneous stigmata were present. Routine laboratory results (hemoglobin, complete blood count, renal and liver function, electrolytes and coagulation tests) were normal. Renal ultrasound and chest radiograph were also normal. The tumor caused severe vasogenic edema with significant midline shift.

**Operation:** On 20 December 2008 patient underwent a right parieto – temporop – basal osteoplastic craniotomy. Opening the dura mater, an extra axial tumor was found. The neoplasm invaded the brain and was highly vascular. The lesion was totally excised and 2cm of adjacent dura mater was excised – Simpson O. The dural defect was repaired with pericranial fascia. Bone flap looked abnormal, the part infiltrated with tumor was resected as well.

3. **RESULTS**

The postoperative course was unremarkable. The patient was discharged home on the seventh postoperative day. Seizures were controlled with anticonvulsant therapy. Postoperative control head CT scan demonstrated total removal of the lesion (Fig. 3 and Fig. 4). Postoperative CT showed no residual tumor.

**Histological studies:** Paraffin-embedded tissue sections stained with H&E revealed meningothelial tumor with rhabdoid morphology characterizing by sheets of tumor cells with eccentric nuclei, variably abundant eosinophilic cytoplasm and intracytoplasmic hyaline paranuclear inclusions. The number of mitoses was up to four in ten high-power fields, and the Ki-67 proliferation index was 4,4%. The histopathological diagnosis was rhabdoid meningioma (grade III). Discussion: Radical surgery (Simpson grade 0) has been shown to significantly enhance prognosis in atypical and malignant meningiomas. Conclusion: Rhabdoid meningioma is an anaplastic, very rare subtype of malignant meningioma. The prognosis for rhabdoid meningioma depends on their proliferative activity and the possibility of radical removal.

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characterized by sheets of tumor cells with eccentric nuclei, variably abundant eosinophilic cytoplasm and intracytoplasmic hyaline paranuclear inclusions. The meningothelial features were evidenced by the focal whorl formation of tumor cells and immunohistochemical coexpression of epithelial membrane antigen and vimentin. The number of mitoses was up to four in ten high-power fields, and the Ki-67 proliferation index was 4.4%. The histopathological diagnosis was rhabdoid meningioma (grade III). Radiation therapy were performed after surgery. There was no recurrence of tumor in a follow-up examination after 1.5 years.

4. DISCUSSION
Rhabdoid meningiomas constitute a recently described and rare subtype of meningioma that behaves aggressively and leads to a high rate of recurrence and mortality (7,13). It has been classi\-fied as grade III according to the WHO criteria of brain tumors. Tumors with rhabdoid morphology were first described by Beckwith and Palmer (1) in 1978 as aggressive renal tumors in very young children. Subsequently, rhabdoid tumors and tumors with rhabdoid features have been described in many other organ systems, and all tend to have a poor outcome no matter what the histogenesis of the original tumor. In 1994, Germano et al. reported their experience with meningiomas of the first two decades of life and reviewed 14 series of patients in this age group, which accounted for a total number of 278 cases (3). These authors described an incidence of pediatric meningiomas of 1–4.2% in relation to adult cases (3). Sixty to 80% of adult meningiomas occur in women, while in younger patients there is characteristically no female predominance. Clinical presentation in children is with focal neurological deficits (33%), seizures (25%), and symptoms of raised intracranial pressure (25%) (3). In the experience of Germano et al., most meningiomas of these juvenile patients were supratentorial (70%), and only 13% were intraventricular. Radical surgery (Simpson grade I) has been shown to significantly enhance prognosis in atypical and malignant meningiomas. Some surgeons advocate preoperative embolization to reduce intraoperative blood loss. However, with this technique there is the risk of embolic stroke and hemorrhage into a necrotic tumor (6). Early conventional radiotherapy is recommended for atypical and anaplastic meningiomas irrespective of completeness of surgical excision with numerous studies showing improved disease-free survival. Median survival exceeding 5 years with combined radiochemotherapy has been reported, the majority of that survival benefit probably being due to radiotherapy as current chemotherapeutic regimens are poorly effective against atypical/anaplastic meningiomas (4,10,12). Other therapeutic modalities are being increasingly explored; Stereotactic radiosurgery (SRS) has been reported to be beneficial, even as a primary alternative to patients unable to undergo surgery (12). However, a recent series showed no benefit for anaplastic tumours and poor control of atypical tumours. It seems reasonable to use SRS as an adjunct immediately postoperatively for any tumour residuum, but not as a replacement for standard radiotherapy. Moreover, the use of both significantly increases the incidence of radiation complications. The prognosis for these tumors seems to depend on their proliferative activity (estimated by the Ki-67 index and the mitotic rate) and on the possibility of a radical removal, which, in turn, is determined by their location (11,12,13). The significance of brain invasion by the neoplasms is disputed.

5. CONCLUSION
We have reported the case of a 8 year old girl with an rhabdoid meningioma. Rhabdoid meningioma is an anaplastic, very rare subtype of malignant meningioma. The prognosis for rhabdoid meningioma depends on their proliferative activity and the possibility of radical removal. Adjuvant therapy is essential in prolonging survival and management should at least consist of radical surgery and postoperative radiotherapy.

REFERENCES