Rhabdoid Meningioma of Brain - A Rare Aggressive Tumor

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Abstract

Rhabdoid meningioma is a rare aggressive variant of meningioma, regarded as WHO Grade III type. Histologically and cytologically, it is distinctive type having abundant eosinophilic cytoplasm, cytoplasmic inclusion with eccentrically placed vesicular nuclei and prominent nucleoli. High recurrence rate and poor outcome are important features. Here, we are presenting a rare case of rhabdoid meningioma found in a recurrent meningioma of the posterior fossa in a middle-aged female. We emphasized the squash cytology and histology finding of the rare neoplasm.

Keywords: *Aggressive, rhabdoid meningioma, squash cytology, WHO Grade III*

Introduction

Meningiomas are slow-growing central nervous system (CNS) neoplasm of meningothelial origin. However, some of the meningiomas have aggressive behavior (WHO Grade II: atypical, clear cell and choroid and WHO Grade III-papillary and anaplastic variant).[1] Rhabdoid meningiomas are rare aggressive variant, adopted in WHO classification of CNS neoplasm (2000) and belonged to WHO Grade III subtype.[1,2] This tumor is associated with rapid growth, high incidence of recurrence, and poor survival than others.[1,2] Here, we are reporting a rare case of rhabdoid meningioma in a middle-aged male patient with an emphasis on squash cytology and histology.

Case Report

A 45-year-old female presented with headache, vertigo, and vomiting for the last 2 days and altered sensorium for the last 1 h. She had a past history of posterior fossa tumor and was operated 1 year back. Histologically, it was diagnosed as meningothelial meningioma of posterior fossa (WHO Grade I). Her hematological and biochemical tests were within normal limits except mild anemia. Electrolyte assay showed mild hyponatremia (Na-129 mmol/l). Computed tomography scan revealed a brightly contrast-enhancing dural-based mass at previously operated location [Figure 1]. Clinical and radiological diagnosis was recurrent meningioma. Her visual acuity was normal, but both the fundus showed papilledema. She had undergone repeat surgery, and intraoperative squash cytology was taken. The squash cytology revealed monotonous meningothelial cells arranged in lobules [Figure 2a]. There were some meningothelial cells with characteristic abundant cytoplasm and round nuclei pushing to periphery (rhabdoid cells) [Figure 2b and c]. In gross examination, the resected tumor was a grayish mass 5.5 cm × 4 cm × 3 cm. In histopathology, microscopy revealed a solid mass composed of meningothelial cells in sheets and in whorled pattern [Figure 3a]. There are areas of monomorphous sheets of cells with abundant eosinophilic cytoplasm with eccentrically placed vesicular nuclei and prominent nucleoli [Figure 3b]. The cells contained spherical masses of eosinophilic inclusions pushing the nuclei to the periphery. Areas of necrosis and psammoma bodies are also found. Mitotic count was 4–8/10 high power field. Final histopathological diagnosis was rhabdoid meningioma.

Discussion

Rhabdoid meningioma is a very rare aggressive variant of meningioma.[2] Most of the meningiomas are among Grade I (WHO).[2,3] Only 5%–7% meningioma are atypical (Grade II) and 3% are anaplastic type (WHO Grade III).[2] Rhabdoid transformation of the meningothelial tumor was first described by Perry *et al*. in 1998.[3,4]

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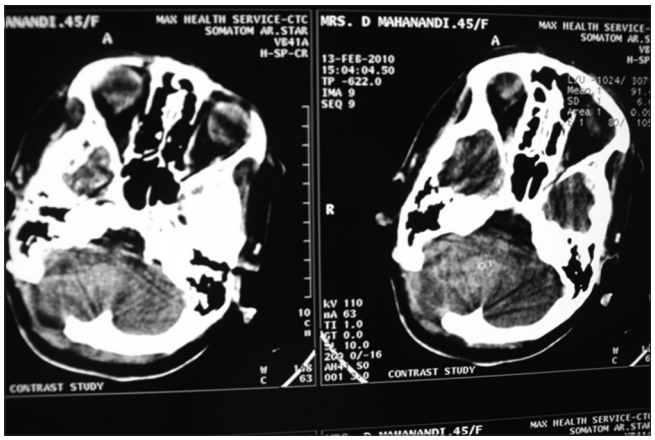

Figure 1: Computed tomography scan image of the recurrence of posterior fossa tumor at previously operated site

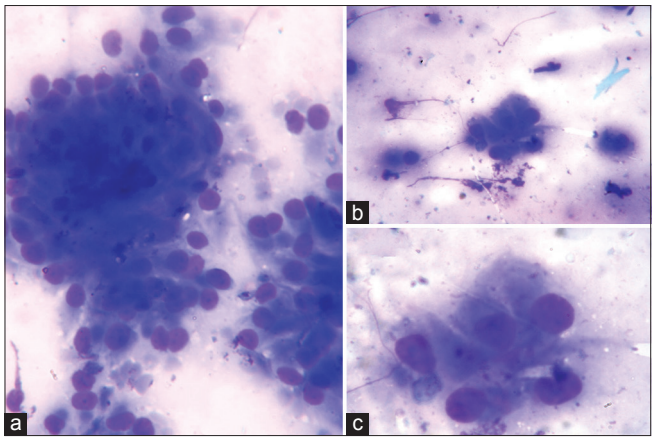

Figure 2: (a) Photomicrography of squash cytology of meningioma showing whorl clusters of meningothelial cells (Leishman and Giemsa, ×40). (b and c) Squash cytology of meningothelial cells with rhabdoid morphology (abundant cytoplasm, large eccentric nuclei) (Leishman and Giemsa, ×40)

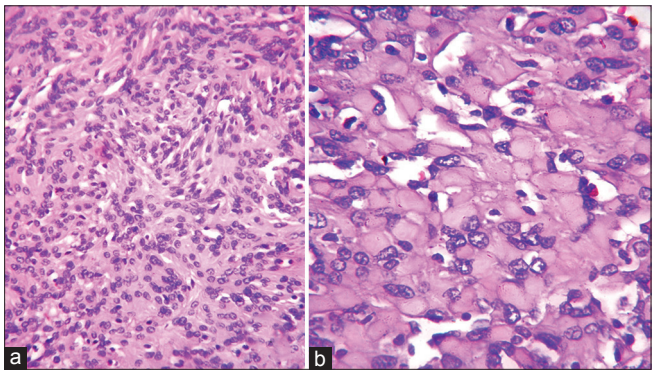

Figure 3: (a) Photomicrograph of histology of the area showing whorled pattern of typical meningothelial cells without significant anaplasia (H and E, ×40). (b) Photomicrograph of histology of the area of monomorphous sheets of cells with abundant eosinophilic cytoplasm, intracytoplasmic inclusions, eccentrically placed vesicular nuclei and prominent nucleoli; typical of rhabdoid meningioma (H and E, ×40)

It was included in WHO classification 2000 as a subtype of meningioma with high risk of recurrence.[3] Although cases have been reported of different age, most of the cases occur in young and middle age with an equal incidence in male and female.[1,3,5] The term rhabdoid refers to the resemblance of neoplastic cells to rhabdomyoblast without true skeletal muscle differentiation.[1] Ultrastructurally, it represents whorls of intermediate filaments expressing vimentin and occasionally cytokeratin.[6] Tumor with rhabdoid morphology was initially introduced in renal neoplasm with aggressive behavior.[6] Rhabdoid morphology has been identified in different tumors such as carcinomas, sarcomas, gliomas, and melanoma.[1]

Histologically, the rhabdoid morphology cells have large round to oval with abundant eosinophilic cytoplasm, eccentrically placed nuclei with prominent nuclei.[3] Paranuclear cytoplasmic eosinophilic inclusions are frequently found.[3] Most of the cases exhibit rhabdoid morphology with histological evidence of meningothelial differentiation.[3] In our case also, the tumor exhibited typical meningothelial differentiation with areas of rhabdoid morphology. Infiltrating growth and focal necrosis are also associated features and were evident in our case. Differential diagnoses of rhabdoid meningioma include atypical teratoid/rhabdoid tumor, metastatic carcinoma, melanoma, sarcoma, and mega cell medulloblastoma.[1,3,5] Histological diagnosis depends on the evidence of meningothelial differentiation (whorls, nuclear characters, eosinophilic inclusion bodies) and immunohistochemistry fi ndings (epithelial membrane antigen, vimentin, and progesterone receptor positive).[1]

Recurrence rate is very high approaching about 87%.[5] In our case, it was a recurrent tumor after 18 months of surgery. Radical surgery and postoperative radiotherapy of 60 GY (conventional fractional) are the therapeutic modality of choice.[1] However, stereotactic radiosurgery, preoperative chemoembolization, and combined radio-chemotherapy are the other optional modalities.[2] Median survival is <3 years after surgical resection with adjuvant therapy.[2]

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Conflicts of interest

There are no conflicts of interest.

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