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Case of Uncommon Meningioma's With Rhabdoid Variant

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Abstract: Rhabdoid meningioma is a rare subtype of meningioma's accounting for 1-3% of all intracranial meningioma's and classified as WHO Grade III tumor. It has an aggressive course and needs to be treated by both surgery and radiotherapy. It has been suggested that rhabdoid morphology is associated with a poor prognosis. Rhabdoid morphology is defined as sheets of loosely cohesive cells with eccentric nuclei and hyaline, Para nuclear inclusions. Ultra structurally, the latter consisted of whorls of intermediate filaments often entrapping lysosomes or other organelles. We report a case of 60 year female operated by left temporo parietal craniotomy for meningioma histopathologically reported as meningioma with rhabdoid differentiation (WHO GRADE2). Patient managed by regular follow up with MRI brain scan.

Keywords: Intracranial; Meningioma; Rhabdoid.

INTRODUCTION

Most meningioma's are benign and classified as grade I according to World Health Organization (WHO) standards [1]. However, subtypes such as atypical, clear cell, chordoid ,Rhabdoid, and malignant meningioma's display less favorable clinical outcomes and are classified as grades II and III [2–5]. Atypical meningioma's account for between 4.7 and 7.2% of all meningioma's [1]. Malignant meningioma's are less common, comprising between 1.0 and 2.8% [1]. Malignant and atypical meningiomas are more prone to recurrence and rapid growth [2]. The distinction between benign and atypical or malignant meningioma represents important surgical information, because surgical and treatment planning as well as prognostication will depend on those pathologic types.

Rhabdoid meningiomas were described for the first time in 1998 as an unusual variant with increased proliferative activity. Later, in 2000, these tumors were included in the revised WHO classification of CNS tumors as an aggressive meningioma corresponding to WHO grade III [5].

We reported this case because of its unusual appearance on histopathogical aspect and rare presentation and only rhabdoid differentiation without other histological features of malignancy, hence classified as WHO GRADE 2.

CASE REPORT:

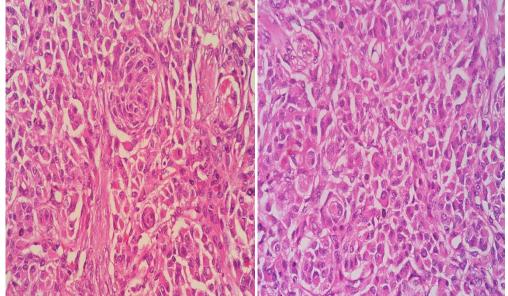
60 year old female patient came with history of left side headache since 1 month, no history of vomiting ,LOC ,seizure , history of right sided mild weakness, on clinical examination patient was conscious oriented to time place person, cranial nerve appears to normal, power of right upper and lower limb 3+/5. Patient evaluated with MRI brain and reported as A welldefined enhancing extra axial lesion 53/44/45 mm seen involving the left temporal region with extension to frontal region with mass effect, adjacent bony sclerosis noted. Enhancing dural tail lesion .features suggestive of left temporal meningioma (? Atypical) with mass effect. Patient underwent left frontal craniotomy with excision of tumor, operative finding showed bone thickened with some area having dural tear +, tumor was firm ,vascular adherent to dura on convexicity and lateral and middle 3 rd of sphenoid wing. Clear demarcation from surrounding brain. Histopathological section studied was consistent with meningothelial meningioma's with focal rhabdoid diffentiation, (grade Behavior of meningioma's with rhabdoid differentiation in the absence of other histological feature of malignancy is not determined hence it was kept in WHO grade 2 ,Post-operative period was uneventful, Patient was followed up regularly with MRI brain to look for recurrence but follow up brain scan did not show any recurrence hence this patient on regular follow up.



PRE OPERATIVE T 1 WEIGHTED CONSTRAST



SAGITAL POST OPERATIVE T 1 WEIGHTED CONSTRAST



HISTOPATHOLOGY SLIDE H&E LOW FIELD HIGH FIELD SHOWING WORL PATTER(RHABDOID)

DISSCUSION

The term rhabdoid morphology in tumor refers to the characteristic resemblance of the cells to a rhabdomyoblasts without true skeletal muscle differentiation [2]. Because this same histology has been demonstrated in a wide variety of malignancies including carcinomas, sarcomas, gliomas, and melanomas, many have argued that neoplasms with rhabdoid morphology occurring at extra renal sites simply represent aggressive, poorly differentiated tumors with a common phenotype [2]. These tumors commonly show mutation of the INI1 gene on 22.q11.2. Tumors with different histogenesis including

carcinomas, sarcomas, gliomas and melanomas (composite extra renal rhabdoid tumor or ERT) also may show rhabdoid phenotype, but they don't show 22q11.2 deletion [4]. But an important feature, common to all rhabdoid tumors is aggressive behavior and poor outcomes irrespective of their histogenesis. In 1998, Kepes *et al.*; and Perry *et al.*; described the first 2 series of meningiomas with rhabdoid transformation [5, 6]. This meningioma's often recur, invade brain and lead to extra cranial metastasis.

Besides the rhabdoid cells, cytoarchitectural features of atypical meningioma like more than or equal

to 4 mitoses per 10 high power fields, high cellularity, sheeting architecture, nuclear atypia, necrosis are often seen in most cases [7]. Clinically these tumors arise in elderly persons with equal incidences in male and female. Most patients present with neurological features like hemiparesis, diplopia, vomiting etc. .

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 meningioma's, following incomplete resection and upto 60 Gy for meningioma's that have atypical or malignant features

Our case had histological feature showing only rhabdoid differentiation and only few mitotic figures noted (<4/10 HPF) and section showed no evidence of pleomorphism, necrosis, atypical mitosis and brain parenchymal invasion hence it is classified as WHO GRADE 2. Patient on regular follow up with MRI brain did not showed any recurrence.

CONCLUSION

Rhabdoid meningioma's are usually encountered in higher grade meningioma's, seen to emerge as a recurrent growth during the process of malignant transition, are seen infrequently in pure form, and have poor outcome. Hence close follow up and aggressive treatment is mandatory in such cases.

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