Original Article

Meningioma With Rhabdoid Differentiation: A Rare Case Report And Review Of Literature

***Dr. Yesha Parimalbhai Lad[1], Dr. Banyameen Iqbal[2], Dr. Nirali Patel[3], Dr. (col.) Harsh Kumar[4], Dr. Charusheela Gore[5], Dr. Shirish Sahebrao Chandanwale[6]**

1. Resident Doctor, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.
2. Professor, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.
3. Resident Doctor, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.
4. Professor, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.
5. Professor and HOD, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.
6. Professor, Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India.

*****Corresponding author:
Dr. Yesha Parimalbhai Lad, Resident Doctor, Department of Pathology, Dr D.Y. Patil Medical College, Hospital and Research Center, Dr. D.Y. Patil Vidyapeeth, Pimpri, Pune – 411018, India
Email: yeshalad@gmail.com
DOI: 10.47750/pnr.2022.13.S09.1223

Abstract

Meningiomas are tumors of meningothelial origin that develop slowly and have a variety of histological characteristics. As per WHO classification, some subtypes are recognized which display atypical cells such as papillary, rhabdoid and anaplastic variants and are classified as WHO grade III tumors. In this case report we are presenting a rare case of 34-year-old male presented with headache and seizures and was diagnosed with this unusual type of meningioma.

Keywords: Atypical meningioma, Meningioma, Rhabdoid

Introduction

Rhabdoid meningiomas were first recognized in 1998 as a rare kind of meningioma with enhanced proliferative activity. In the year 2000, these tumors were classified as an aggressive meningioma, corresponding to WHO grade III, in the updated WHO classification of CNS tumors. Meningiomas are tumors of meningothelial origin that develop slowly and have a variety of histological appearances.[1] However, certain subtypes, such as atypical, clear cell, chordoid meningiomas (WHO grade II) and papillary, rhabdoid, and anaplastic variants (WHO grade III) demonstrate aggressive behavior, according to WHO 2021 classification. Anaplastic meningiomas account for around 2% of all meningiomas.[1] These tumors on microscopy include loosely cohesive sheets of plump cells with nuclei pushed to one side and glassy, eosinophilic inclusion-like cytoplasm. The tumor cells resemble rhabdoid cells, which have been reported in tumors from different locations, including the kidney and in atypical teratoid rhabdoid tumors of the brain.[2] These aggressive tumors are characterized by rapid growth, brain invasion, and frequent recurrences.[1]

Case Report

A 36-year-old male was admitted with severe headache for 5 days. Patient had one episode of seizures 8 months back, for which he was admitted to hospital and treated for the same. He was asymptomatic in later months. He last had seizures 8 days before the admission, for which an MRI was done. There was no history of trauma, fever, blurred vision, diplopia, hearing loss or dysphagia. The patient was neither diabetic, hypertensive, or suffering from TB or asthma.

On examination patient was conscious and oriented and all the vitals were normal. Higher motor functions were normal and sensory functions were intact. Bilateral upper and lower limb tone and power was also normal (5/5).

MRI showed a well-defined extra axial heterogeneously enhancing lesion in the left fronto-temporal region measuring 3.5 x 3.2 x 3.0 cm suggestive of meningioma. The lesion appeared hyperintense on T2WI, showed minimal blooming on GRE, minimal restricted diffusion, isointense on T1 and showed heterogenous post contrast enhancement. Thickening and enhancement of the overlying dura was noted (Dural Tail sign). The lesion was causing mass effect on the insular cortex, perisylvian region and adjacent fronto-temporal lobe. There was no shift of midline structures. The thalami, brain stem, cerebellum and other regions did not show any abnormality. **[Figure 1]**

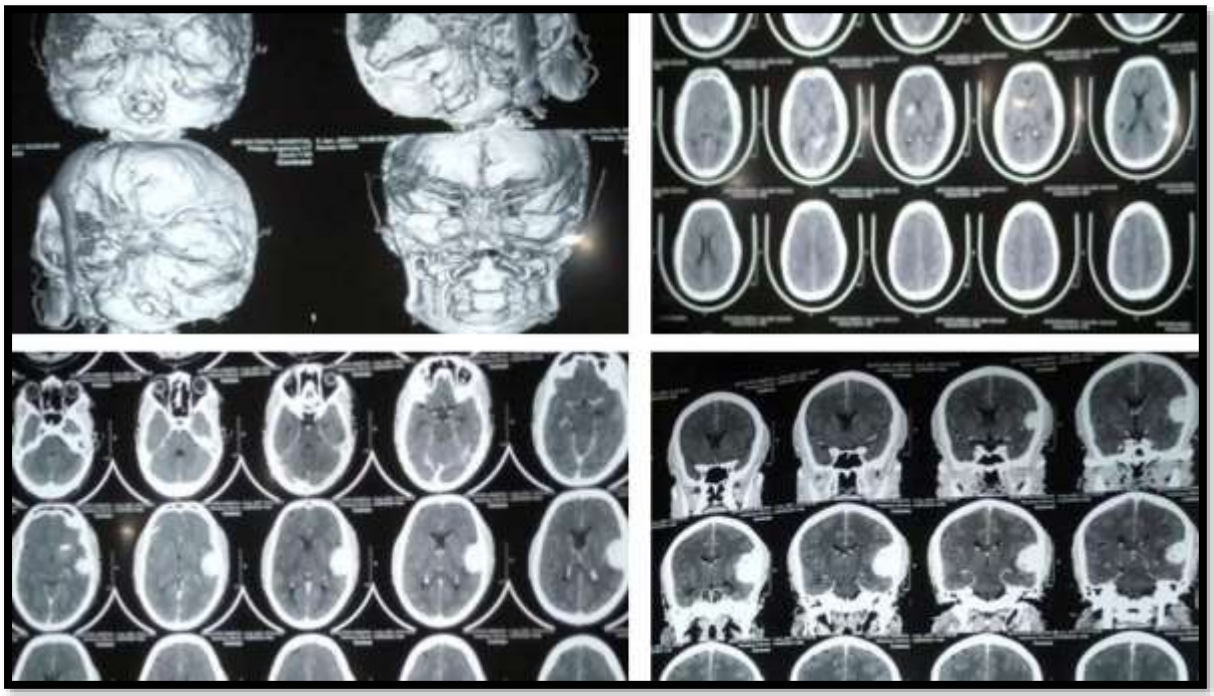

Figure 1: MRI showed a well-defined extra axial heterogeneously enhancing lesion in the left fronto-temporal region.

Left fronto-temporal craniotomy with tumor excision was done. Craniotomy was done after making burr holes and durotomy was done. The tumor was 3x3x3 cm in size. Tumor was greyish white in colour with moderate vascularity. Haemostasis was achieved and incision was closed in layers.

Grossly, multiple greyish white tissue pieces with few brownish areas were received. Largest piece was measuring 3 x 3 x 0.8 cm and smallest piece was measuring 2 x 1.5 x 0.4 cm. Few congested dilated blood vessels were seen. It was soft to firm in consistency.

Histopathology was done which showed large to medium sized nests of cells arranged in a syncytial pattern. **[Figure 2A]** Many cells show round eccentric nuclei with occasional nucleoli. In some foci these cells showed an abundant granular or clear cytoplasm while in other areas there is a fibroblastic appearance. **[Figure 2B]** Abundant thin-walled blood vessels were seen. **[Figure 2C]** A few scattered mitotic figures were seen. No necrotic areas were seen.

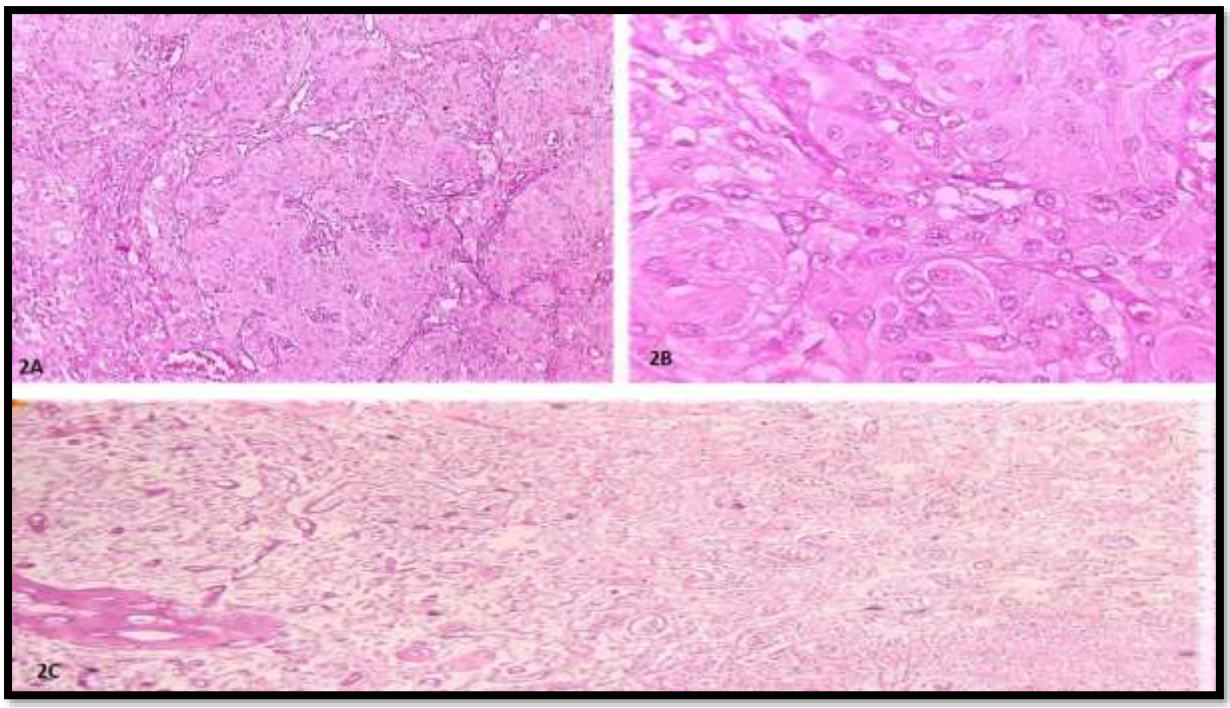

Figure 2: (A) Photomicrograph showing large to medium sized nests of cells arranged in a syncytial pattern [H&E,x10]. **(B)** Cells with round eccentric nuclei with occasional nucleoli. [H&E,x40] **(C)** Photomicrograph shows the presence of abundant thin-walled blood vessels. [H&E,x40]

Immunohistochemistry was performed which showed positivity for Epithelial membrane antigen (EMA) and Vimentin while as negativity for Glial fibrillary acidic protein (GFAP) and Desmin. **[Figure 3A, B, C, D]**

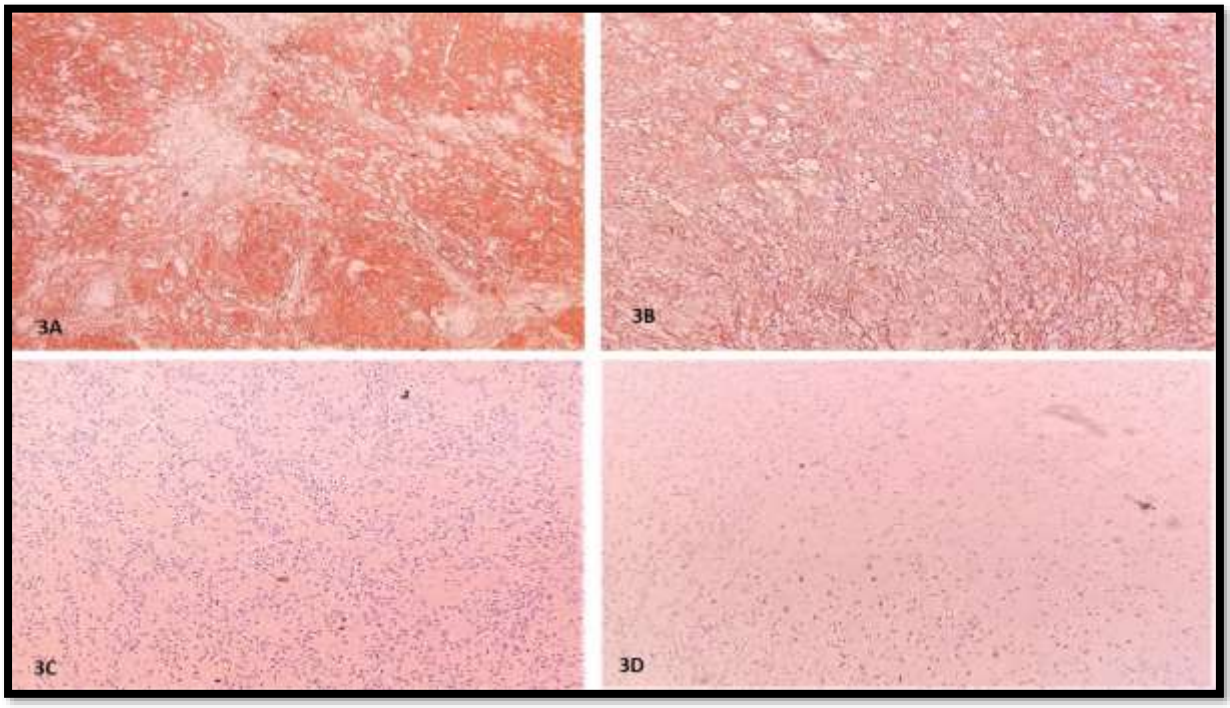

Figure 3: Immunohistochemistry (A) Photomicrograph showing Positivity for Epithelial membrane antigen [EMA] (B) Positive for Vimentin. (C) Negative for GFAP. (D) Negative for Desmin

The above microscopic features favoured diagnosis of Meningioma – mixed type with Rhabdoid differentiation (WHO Grade III)

Discussion

The term rhabdoid morphology in tumor refers to the cells' resemblance to rhabdomyoblasts without true skeletal muscle differentiation. The term malignant rhabdoid tumor (MRT) was used to characterize a specific type of paediatric renal tumor.[1] Following then, the name was used to other types of extra renal paediatric tumor (extra renal MRT), such as atypical teratoid / rhabdoid tumor (AT/RT) of the CNS. The INI1 gene on 22.q11.2 is more typically mutated in these cancers.[3,4] Tumors with distinct histogenesis, such as carcinomas, sarcomas, gliomas, and melanomas (composite extrarenal rhabdoid tumor or ERT), may have a rhabdoid phenotype but may not have a 22q11.2 deletion. A conventional meningioma component may be seen in most instances of rhabdoid meningioma, at least focally. Completely rhabdoid meningiomas with no conventional meningothelial characteristics are uncommon. In such circumstances, immunohistochemical and/or ultrastructural evidence of meningothelial differentiation must be established.[5] Aggressive activity and poor outcomes are typical features of all rhabdoid tumours, regardless of histogenesis. In 1998, Kepes et al and Perry et al reported first two series of rhabdoid meningiomas. These meningiomas frequently recur and enter the brain, resulting in extracranial metastases.[6-8] Aside from rhabdoid cells, most atypical meningiomas include cytoarchitectural characteristics such as greater than or equal to 4 mitoses per 10 high power fields, high cellularity, sheets of cell, nuclear atypia, and necrosis.[9] Clinically these tumors arise equally in elderly individuals. The majority of patients present with neurological symptoms such as hemiparesis, diplopia, vomiting and other neurological symptoms.[10] Metastatic carcinoma, melanoma, glioma, sarcoma, and AT/RT are among the possible diagnoses. The presence of evidences of meningothelial differentiation, such as whorls, nuclear grooving, intranuclear pseudo inclusions, immunohistochemistry findings of expression of epithelial membrane antigens, vimentin, progesterone receptor positivity, or electron microscopic findings of interdigitating cytoplasmic membrane, intercellular junction, and so on, is required for the diagnosis of this tumor. Malignant meningiomas are characterized by abundant epithelial membrane antigen (EMA) and poor cytokeratin. Ki - 67 (MIB-1) is an essential marker for meningioma malignancy. The mean labelling index for these tumors is 14.7% ± 9.8%.[1]

Recent research suggests that categorising these tumors should not be based solely on rhabdoid or papillary architecture. SMARCE1 (clear cell subtype), BAP1 (rhabdoid and papillary subtypes), and KLF4/TRAF7 (secretory subtype) mutations, TERT promoter mutation and/or homozygous deletion of CDKN2A/B (CNS WHO grade 3), H3K27me3 loss of nuclear expression (potentially worse prognosis) and methylome profiling are some molecular biomarkers related with classification and grading (prognostic subtyping).[

Conclusion

Rhabdoid meningioma is an uncommon type of meningioma that typically affects the elderly. It can, however, be infrequently seen in middle-aged/young patients. These anaplastic meningiomas account for around 2-3% of all meningiomas. Hence, while reporting for meningiomas a thorough screening is a must to rule out these atypical features like rhabdoid differentiation, papillary formations and the like, which display an aggressive behavior and are graded as WHO Grade III tumors.

References

1. Kar A, Rath G, Mohanty L, Behera P, Mohapatro S, Lenka A et al. Rhabdoid Meningioma-An Uncommon and Aggressive Variant. J Cancer Sci Ther.2013;5(10):343-5.
2. Vaubel RA, Chen SG, Raleigh DR, Link MJ, Chicoine MR, Barani I et al. Meningiomas With Rhabdoid Features Lacking Other Histologic Features of Malignancy: A Study of 44 Cases and Review of the Literature. J Neuropathol Exp Neurol. 2016 ;75(1):44-52.
3. Rorke LB, Packer RJ, Biegel JA.Central nervous system atypical teratoid/rhabdoid tumors of infancy and childhood: definition of an entity. J Neurosurg 1996;85:56-65.
4. Mardi K, Thakur RC, Biswas B. Rhabdoid meningioma lacking malignant features: Report of a rare case with review of literature. Asian J Neurosurg. 2015;10(2):172-4.
5. E Matyja, W Grajkowska, P Nauman, W Bonicki, P Bojarski, A Marchel. Necrotic rhabdoid meningiomas with aggressive clinical behavior. ClinNeuropathol. 2010;29(5):307–16.
6. Perry A, Scheithauer BW, Stafford SL, Abell-Aleff PC, Meyer FB."Rhabdoid" meningioma: an aggressive variant. Am J Surg Pathol 1998;22(12):1482-90.
7. Kepes JJ, Moral LA, Wilkinson SB, Abdullah A, Llena JF. Rhabdoid transformation of tumor cells in meningiomas: a histologic indication of increased proliferative activity: report of four cases. Am J Surg Pathol. 1998;22(2):231-8.
8. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D et al. The 2021 WHO Classification of Tumors of the Central Nervous System: a summary. Neuro Oncol. 2021;23(8):1231-1251.

9. Backer-Grondahl T, Moen BH, Torp SH. The histopathological spectrum of human meningiomas. Int J Clin Exp Pathol. 2012;5(3):231-42.
10. Haas JE, Palmer NF, Weinberg AG, Beckwith JB. Ultrastructure of malignant rhabdoid tumor of the kidney: A distinctive renal tumor of children. Hum Pathol.1981;12(7): 646-57.