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Clinical, Histopathologic and Genetic Characteristics of Rhabdoid Meningiomas

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Abstract

Rhabdoid meningiomas have variable histological findings, and a wide range of chromosomal copy number alterations are linked to an unpredictable disease course. We analysed 305 RM samples from patients previously reported in the literature, as well as 33 samples from 23 patients studied in our laboratory, in this study. The most common chromosomal alteration was monosomy 22, which involved the minimal but most common recurrent region loss of the 22q11.23 chromosomal region, followed by losses of chromosomes 14,1,6, and 19, polysomies of chromosomes 17,1q, and 20, and gains of 13q14.2, 10p13, and 21q21.2 chromosomal regions. Based on their CNA profile, RM could be divided into two genetic subgroups with distinct clinicopathologic features defined by the presence of chromosomal losses only or combined losses and chromosomal instability.

Keywords: Adhesionrhabdoid meningioma • Diagnosis • Prognosis • Histopathology

Introduction

Meningiomas are the most common subtype of central nervous system tumours, with diverse histopathologic and genetic/molecular features that are usually associated with a benign clinical outcome. Thus, most meningiomas have World Health Organization benign grade 1 histopathologic features, whereas WHO grade 2 and 3 meningiomas are less common. Rhabdoid meningiomas are a rare tumour subtype with distinct histopathologic features. Thus, in the WHO 2021 classification of CNS tumours, RM are defined as "plump cells with eccentric nuclei, open chromatin, macronucleoli, and prominent eosinophilic paranuclear inclusions, appearing either as discernible whorled fibrils or compact and waxy spheres," and RM are classified as WHO grade 3 tumours.

The majority of RM cases, however, exhibit a combined rhabdoid cytology with variable percentages of rhabdoid cells associated with tissue areas that exhibit histopathologic features that are typical of other histological variants of meningiomas. Rhabdoid cells are the hallmark of this rare meningioma subtype, accounting for 50% of total tumour cellularity. In turn, RM are characterised by a high mitotic rate, overt anaplasia, homozygous deletion of the CDKN2A and/or CDKN2B genes, and/or mutations in the TERT promoter. Several authors have recently proposed that rhabdoid cell morphology/histopathology may represent a phenotypic variant rather than a specific subtype of meningioma, citing the fact that some RM cases that were initially diagnosed as WHO grade 3 tumours were reclassified after re-evaluation [1].

Description

Similarly, inconsistent and even contentious findings have been reported regarding the clinical behaviours and outcomes of RM patients. In this regard, it should be noted that the rhabdoid component may already be present at the time of the initial diagnosis or recurrence in men and women of all ages, including adults and children. Furthermore, the number of rhabdoid cells in some patients' recurrent tumours may increase. Not every RM has an aggressive clinical course, according to prognosis. Thus, despite a proclivity for local recurrence, and despite the fact that distant metastasis in RM has previously been reported, the specific mechanisms underlying tumour relapse remain unknown.

Significant progress has been made in the characterization of meningioma genetic/molecular alterations in recent decades. Despite this, there is little information available about the specific genetic features of RM. Thus, preliminary studies have reported chromosome 22 loss, BRCA1 associated protein 1 gene mutations and/or deletion, and high expression of the matrix metallopeptidase 9 gene, all of which are typical of RM. Furthermore, mutations in the PBRM1 gene, which are commonly found in papillary meningiomas, have been found in single RM cases. Similarly, BAP1 gene mutations and/or deletions, which are common in RM, have been reported in papillary meningiomas in conjunction with PBRM1 gene alterations [2].

Despite these findings, the presence and frequency of chromosome copy number alterations (can) other than chromosome 22 losses remain unknown in RM. We report on the histopathologic, clinical, and prognostic features of RM in this study based on a large retrospective series of 305 cases of patients with 12 different series of RM, several RM case reports identified after an extensive review of previous reports, and a group of cases collected retrospectively from 13 centres in Spain. For the first time, we investigated the genetic profile of RM as analysed by whole genome copy number microarrays, as well as its relationship with other features of the disease and patient outcomes, using cases from Spain [3,4].

Out of 305 cases collected, 233 tumours with available histopathologic and/or clinical features consistent with RM were analysed. These included the majority of adult cases (88%) and 12% of childhood RM cases, with median ages of 52 16 years and 12 4 years, respectively. Overall, women outnumbered men in adult tumours but not in childhood tumours. According to the WHO 2021 classification, 32% of the cases were classified as WHO grade 3 meningiomas, 41% as WHO grade 2 tumours, and 27% as WHO grade 1 tumour. The vast majority of tumours had a mixed histological pattern. This was linked to a low mitotic rate of less than 4 mitoses/10 HPF in more than half of the patients, as well as a predominance of low-proliferative tumours in WHO grade 1 (100%) and grade 2 (43%) RM vs. grade 3 (15%) RM. As might be expected, increasing frequencies of cases with a higher mitotic rate and a higher proliferative index were observed in WHO grade

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2 (9%) and grade 3 (48%) RM, but not in grade 1 RM. This resulted in an increased frequency of cases undergoing tumour recurrence (12% vs. 31% and 35%) and deaths [5].

Conclusion

In conclusion, the current study demonstrates the presence of a rhabdoid meningioma cell component in variable proportions and in combination with components of other distinct histopathologic meningioma subtypes across all WHO tumour grades, making cytomorphological diagnosis of RM difficult in the absence of additional criteria. From a genetic standpoint, nearly all RM had monosomy 22/22q deletions, which is usually associated with deletions of chromosomes 1p, 6q, 14, and 19p alone or in combination with gains of chromosomes 17, 1q, and 20 and more complex karyotypes. For the first time, minor common regions of CNA were identified in RM in this study, which involved 22q11.23 losses and gains at the 13q14.2, 10p13, and 21q21.2 chromosomal regions. The two different CNA profiles found among RM were associated with different distributions of WHO tumor grades as well as a distinct patient outcome, which highlights their potential utility for a more robust clinical subclassification of RM.

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Conflict of Interest

There are no conflicts of interest by author.

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