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Case Report Open Access

Meningioma with Remarkable Multiple Rosette Formation: A Diagnostically Difficult Case

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Abstract

Background: Rosette formation is a rare morphological feature in meningiomas. It can be seen in angiomatous, meningothelial, transitional, secretory, papillary or rhabdoid subtype; however there are very few reports of meningiomas with rosettes, and the feature itself is not criterion for the WHO grade, so this feature sometimes makes the diagnosis difficult.

Case presentation: A 62 y old Japanese woman stumbled and was admitted to a hospital. Magnetic resonance imaging showed a 50 mm extra-axial tumor on the right cerebral falx. Histologically, the tumor contained multiple rosettes that had a central nuclear-free zone without a vessel core. It also showed a sheet-like or whorled growth pattern and pseudopapillary structure. Some round tumor cells had abundantly dense eosinophilic cytoplasm, displaying rhabdoid features. High cellularity, small cells with a high nuclear-to-cytoplasmic ratio and prominent nucleoli were recognized, but the mitotic activity was less than 1/10 high-power fields. The tumor was immunohistochemically positive for EMA and synaptophysin (weakly) but negative for GFAP, progesterone, STAT6, S-100, NeuN and melan A. The Ki-67 labeling index was merely 0.5%. Although ependymoma was considered as the first differential diagnosis, it was ruled out, as perivascular pseudorosettes and ependymal rosettes were not seen. We ultimately made a diagnosis of atypical meningioma with a rosette-like and pseudopapillary pattern, WHO grade II.

Discussion and conclusion: The meningiomas in this case showed remarkable rosette formation, but as this is not criterion for deciding WHO grade, grading was diagnostically difficult. Papillary meningioma was also considered as a critical differential diagnosis; however, our final diagnosis was made after considering the very low mitotic activity and Ki-67 labeling index, which were not consistent with WHO grade III. This case was very unique and diagnostically difficult. This report may be an aid for the diagnosis of pathologically unusual entity.

Keywords: Meningioma; Rosette formation; Atypical meningioma; Pseudopapillary

List of Abbreviations CT: Computed Tomography; MRI: Magnetic Resonance Imaging; WHO: World Health Organization; EMA: Epithelial Membrane Antigen; GFAP: Glial Fibrillary Acidic Protein; STAT6: Signal Transducer and Activator of Transcription 6.

Introduction

Meningiomas are common intracranial neoplasm and categorized as WHO grades I to III. Most meningiomas correspond to WHO grade I. WHO grade II meningiomas consist of chordoid, clear cell or atypical meningioma, and those of WHO grade III contain papillary, rhabdoid or anaplastic meningioma. It has been described that recurrence rates of benign meningioma are about 7-25%, whereas that of atypical meningioma is 29-52%, and anaplastic meningioma recur in 50-94% [1]. A study reported recurrence rate of papillary meningioma up to 59% [2]. Clinically, it is thus very critical to distinguish each grade meningioma only by microscopic findings.

Meningiomas show much variety of morphology and rosette formation is a rare morphological feature of the tumor. It could be seen in angiomatous, meningothelial, transitional, secretory, papillary or rhabdoid subtype among them; however there are very few reports of the tumor with rosettes [3-5]. In addition, the feature itself is not criterion for deciding WHO grade. Therefore the feature sometimes makes diagnosis difficult.

Case Presentation

A 62 y old Japanese woman stumbled and was admitted to a hospital. Computed tomography (CT) and magnetic resonance imaging (MRI) revealed 50 mm extra-axial tumor on the cerebral falx in the front region of right hemisphere (Figure 1A). The tumor showed calcification on CT. The mass showed iso-signal intensity on T1-weighted MRI and slight high-signal intensity on T2-weighted MRI. She was clinically diagnosed with meningioma. The tumor connected to the deeper edge of the cerebral falx was surgically removed.

Histologically, the tumor contained multiple rosettes that had a central nuclear-free zone filled with fibrillar cytoplasmic processes without vessel core (Figures 1B and 1C). It showed a sheet-like or

whorled growth pattern and pseudopapillary structure (Figure 1D). Psammoma bodies, intranuclear pseudoinclusions, abundant hyalinized vessels and brownish-granule deposition were also seen. The tumor was consisted of spindle cells with large, oval-shaped nuclei and eosinophilic cytoplasm. Some round tumor cells had abundantly dense eosinophilic cytoplasm, displaying rhabdoid features (Figure 1E). High cellularity, small cells with a high nuclear-to-cytoplasmic ratio and prominent nucleoli were recognized (Figure 1F), but the mitotic activity was less than 1/10 high-power fields. No obvious necrosis was seen. Immunohistochemistry revealed that the tumor was positive for EMA (Figure 1G), weakly positive for synaptophysin (Figure 1H) and negative for GFAP, progesterone, STAT6, S-100, NeuN and melan A. Ki-67 labeling index of the tumor was merely 0.5%. We ultimately made a diagnosis of atypical meningioma with a rosette-like and pseudopapillary pattern, WHO grade II. No recurrence has been reported for a year since the operation.

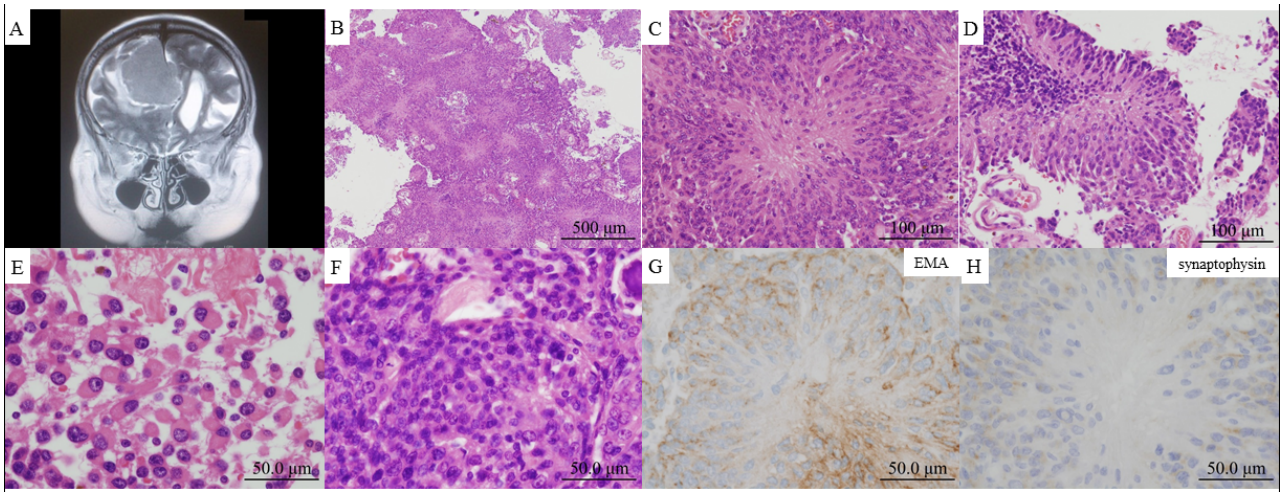

Figure 1: An MRI scan, microscopic findings and immunohistochemical findings of specimen. (A) T2-waighted MRI showed a 50 mm extra-axial tumor on the right cerebral falx. (B) The tumor had multiple rosettes. Bar=500 µm (H&E staining) (original magnification: X40).(C) The rosettes had a central nuclear-free zone without a vessel core. The central area was filled with fibrillar cytoplasmic processes. Bar=100 µm (H&E staining) (original magnification: X200). (D) The tumor also had a pseudopapillary structure. Bar=100 µm (H&E staining) (original magnification: X200). (E) Rhabdoid features were seen. Bar 50 µm (H&E staining) (original magnification: X400). (F) High cellularity, small cells with a high nuclear-to-cytoplasmic ratio and prominent nucleoli were seen. Bar=50 µm (H&E staining) (original magnification: X400). (G and H) Immunohistochemistry revealed that the tumor cells were positive for EMA (G) and synaptophysin (weakly) (H). Ependymal rosettes were not observed on immunostaining for EMA (G). Bar=50 µm (original magnification: X400).

Discussion and Conclusion

The remarkable point of this case was that this tumor showed diffuse and multiple rosette formation. This is a very unique characteristic feature. Rosette formation is a morphological variant of meningioma and a rare feature. Ependymoma was considered as the initial differential diagnosis, since rosette formation is a known sign of ependymal differentiation. Perivascular pseudorosettes and ependymal rosettes are key features for the diagnosis of ependymoma. Perivascular pseudorosettes have a blood vessel core and perivascular anucleate zone. Ependymal rosettes are formed by columnar tumor cells arranged around a central lumen, and EMA immunoreactivity along the luminal surface of ependymal rosettes can be observed. Immunoreactivity for GFAP is usually observed in ependymoma. However rosettes of ependymoma were not seen in our case and the cells were negative for GFAP. Therefore, based on these findings, ependymoma was ruled out.

Papillary glioneuronal tumor, meningeal melanocytoma and solitary fibrous tumor were also considered as differential diagnosis. Papillary glioneuronal tumor consists of glial cells forming a pseudopapillary structure with a hyalinized vessel core and neurocytes filling the interpapillary space and is usually positive for GFAP, S-100, NeuN and synaptophysin. In our case, although the tumor cells were weakly positive for synaptophysin, they were negative for GFAP, S-100 and NeuN. A pseudopapillary structure with glial cells and neurocytes was not observed. Meningeal melanocytoma and solitary fibrous tumor sometimes form a sheet-like and whorled growth pattern that resembles the morphology of meningiomas; the former is positive for melan A, while the latter is positive for STAT6. The tumor cells were also negative for these markers in the present case.

The tumor in our patient showed a whorled growth pattern and was positive for EMA. These findings were consistent with meningioma. While multiple rosette formation was undoubtedly the most remarkable feature, this is not a criterion for deciding on the WHO grade of meningioma, so grading was diagnostically difficult in this case. Papillary meningioma, which is a rare variant defined by the perivascular pseudopapillary pattern that corresponds to WHO grade III, was also considered as a critical differential diagnosis because a pseudopapillary pattern was observed; however the tumor was not consistent with WHO grade III, with its extremely low mitotic activity and Ki-67 labeling index. Considering the increased cellularity, small cells with a high nuclear-to-cytoplasmic ratio and prominent nucleoli, the final diagnosis was atypical meningioma with a rosette-like and pseudopapillary pattern, WHO grade II.

We encountered a very rare and diagnostically difficult case of meningioma with remarkable multiple rosette formation. This unique

case is worth reporting and may be an aid for the diagnosis of pathologically unusual entity.

Declarations

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Conflicts of interests

The authors declare that they have no conflicts of interests.

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