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Pathology

Chordoid Meningioma: A Rare Entity

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Abstract **Case Report**

Meningiomas are fairly common brain tumors that develop from the meninges. There are several histological variants of different grades. Chordoid meningioma (CM) is a rare grade 2 variant according to the WHO. The diagnosis is essentially anatomopathological which is based on a morphological study followed by an immunohistochemical complement. The reference treatment is surgery sometimes followed by radiotherapy.

Keywords: Meningioma, Histology, Immunohistochemistry, Brain, Recurrence, Surgery.

INTRODUCTION

Meningiomas are fairly common brain tumors with a clear female predominance. There are several histological variants. The chordoid variant is rare. It accounts for <0.5% of all meningioma types [1].

The differential diagnosis includes chordomas, myxoid chondrosarcoma and chordoid glioma [2].

Chordoid meningioma is classified as grade 2 according to the WHO classification [3]. The diagnosis is anatomopathological and is based on a morphological study, most often supplemented by an immunohistochemical study. The standard treatment is surgery, which is based on complete excision of the tumor whenever possible. Recurrences are frequent.

OBSERVATION

This is a 56-year-old patient who has had a sudden onset headache for 2 months with a notion of right hemiparesis. A magnetic resonance imaging (MRI) with Gadolinium injection showing an extra-axial lesion with a broad base of implantation, exhibiting heterogeneous signal enhancement, strongly enhanced after injection with dural enhancement (dural tail), and exerting a mass effect on midline structures (Figure 1).

The patient was admitted to the operating room for tumor resection. The specimen was sent to the pathology laboratory for anatomopathological study.

Morphological analysis shows tumor proliferation made up of cords and cellular trabeculae within a basophilic extracellular matrix. The tumor cells are medium-sized with abundant eosinophilic cytoplasm and nuclei with moderate atypia. The tumor proliferation strongly expresses EMA antibodies and progesterone receptors (figure 2).

This pathological examination led to the diagnosis of grade II chordoid meningioma according to the WHO.

Patient benefited from stereotactic radiotherapy.

The evolution was stationary without any notion of recurrence for 6 months after surgery.

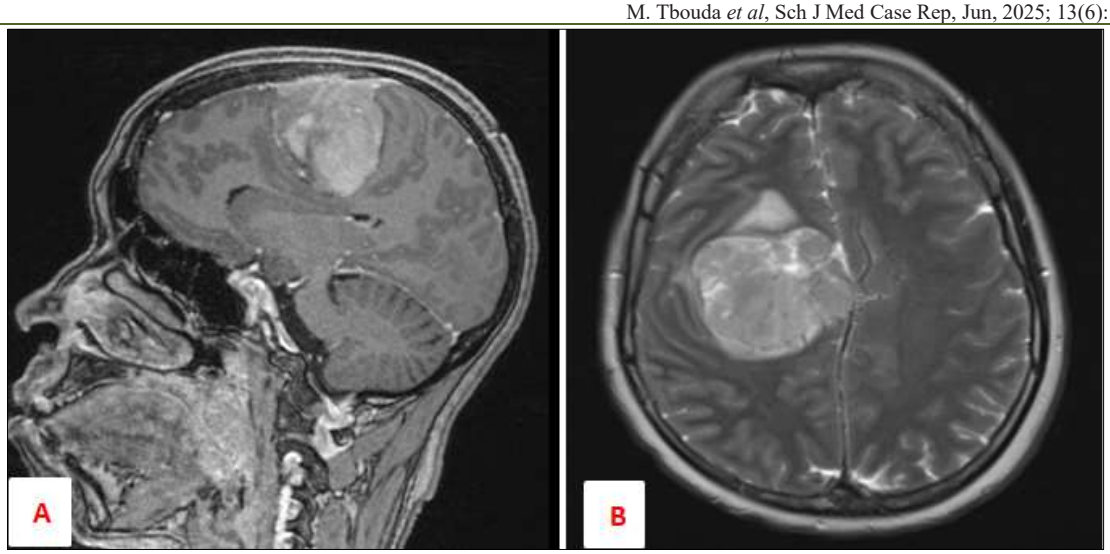

Figure 1: Sagittal T1 (A) fat-saturated MRI scans with Gadolinium injection and axial T2 (B) showing an extra-axial lesion with a broad base of implantation, exhibiting heterogeneous signal enhancement, strongly enhanced after injection with dural enhancement (dural tail), and exerting a mass effect on midline structures.

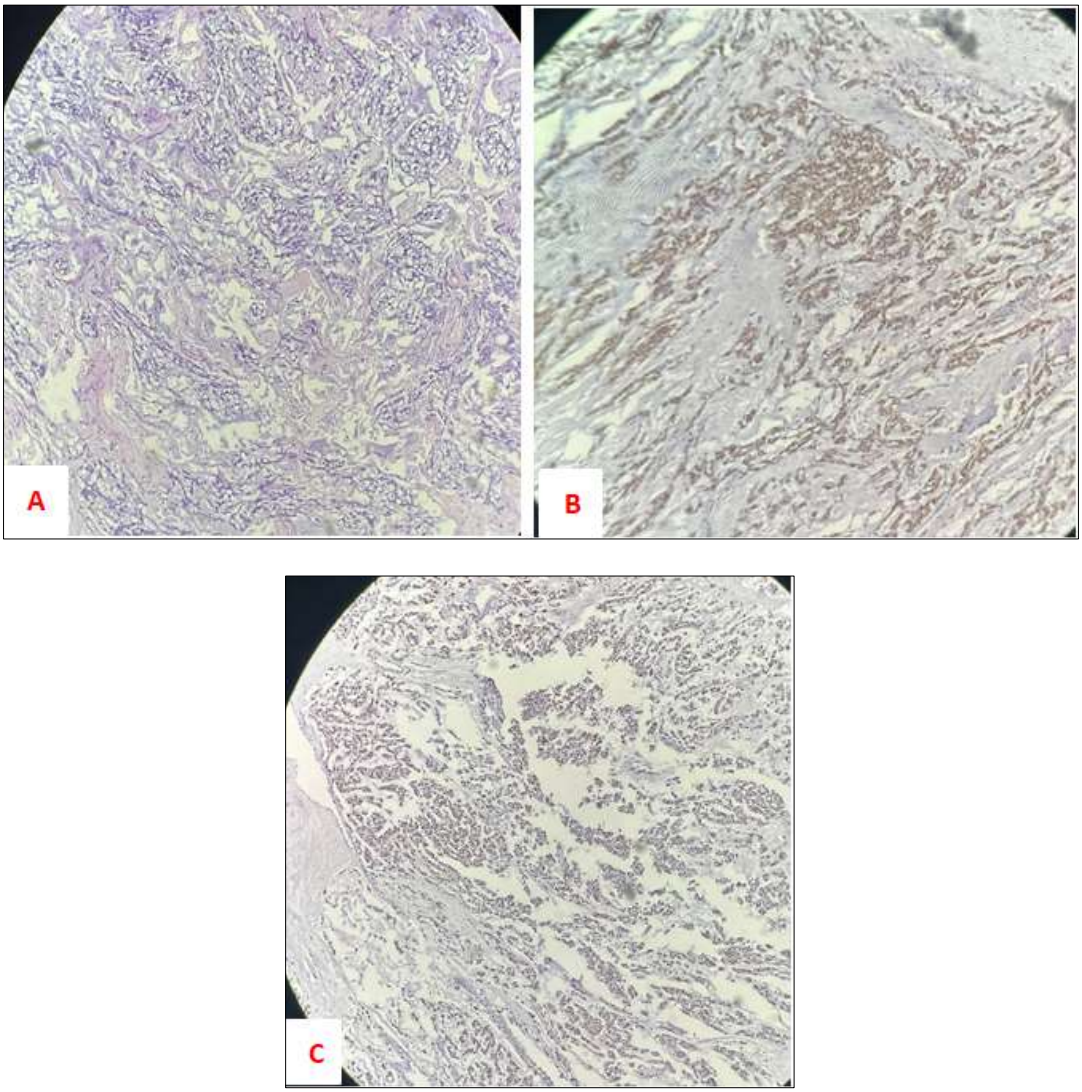

Figure 2: Chordoid meningioma: tumoral proliferation of cellular trabeculae within abundant myxoid stroma (A) strongly expressing EMA (B) and Progesterone receptor antibodies (C)

DISCUSSION

A meningioma is a type of tumor that develops in the meninges, the protective layers surrounding the brain and spinal cord. Most meningiomas are benign and slow-growing, but some can be atypical or malignant.

Chordoid meningioma is a rare WHO Grade II meningioma, meaning it has a higher risk of recurrence compared to typical (Grade I) meningiomas. It has a distinct histological appearance, resembling chordoma (a different type of tumor), with cords or strands of tumor cells in a mucoid (gel-like) background.

It has been previously reported that following the subtotal resection of CM, there is a high tendency for recurrence [4].

Chordoid meningiomas occur very rarely, corresponding to up to 1% of surgically removed meningiomas, even in large cohorts [5]. The diagnosis is anatomopathological which is based on a morphological study which shows a chordoid-like tumor proliferation.

However, sometimes the histological diagnosis is difficult because it is comparable morphology in other intracranial tumors, such as chordoid glioma, chordoma, extraskeletal chondrosarcoma, myxopapillary ependymoma and metastatic tumors [6].

Immunohistopathological studies have shown that this tumor shows positivity for vimentin, EMA and D2–40, and occasional positivity for S-100 protein, on the other hand demonstrating negativity for GFAP and cytokeratin [7].

The differential diagnosis is essentially with tumors showing myxoid differentiation or a so-called chordoid pattern, in particular certain chordoid gliomas, chordomas, myxoid chondrosarcomas, myxoid ependymomas and not forgetting certain secondary processes. In these cases, immunohistochemistry is of great help.

Treatment is essentially surgical. Ideally, the entire tumour should be resected with a healthy margin. This last option is sometimes difficult, especially in the case of large and/or infiltrating tumors. Stereotactic radiotherapy remains a therapeutic option, either alone or as an adjunct to incomplete surgery.

CONCLUSION

Chordoid meningioma is a rare histological variant, WHO grade 2. Diagnosis of certainty is anatomopathological, often requiring immunohistochemistry to rule out other brain tumors showing a myxoid pattern. Standard treatment is complete surgery.

Recurrences are frequent.

Conflicts of Interest: None.

REFERENCES

1. Couce, M. E., Aker, F. V., & Scheithauer, B. W. (2000). Chordoid meningioma: a clinicopathologic study of 42 cases. *The American journal of surgical pathology*, *24*(7), 899-905.
2. Jee, T. K., Jo, K. I., Seol, H. J., Kong, D. S., Lee, J. I., & Shin, H. J. (2014). Clinical features and treatment outcome of chordoid meningiomas in a single institute. *Journal of Korean Neurosurgical Society*, *56*(3), 194-199.
3. Presta, I., Guadagno, E., Di Vito, A., Malara, N., Mignogna, C., Maisano, D., ... & Donato, G. (2017). Innate immunity may play a role in growth and relapse of chordoid meningioma. *International Journal of Immunopathology and Pharmacology*, *30*(4), 429-433.
4. Louis, D. N., Perry, A., Reifenberger, G., Von Deimling, A., Figarella-Branger, D., Cavenee, W. K., ... & Ellison, D. W. (2016). The 2016 World Health Organization classification of tumors of the central nervous system: a summary. *Acta neuropathologica*, *131*, 803-820.
5. Junior, C. R. M., de Morais, M. V., Figueiredo, R. P., Morales, A. G., & de Melo, P. M. P. (2020). Chordoid Meningioma: Literature Review. *Arquivos Brasileiros de Neurocirurgia: Brazilian Neurosurgery*, *39*(01), 012-015.
6. Siraj, F., Ansari, M. K., Sharma, K. C., & Singh, A. (2015). Chordoid meningioma: A diagnostic dilemma. *Journal of Cancer Research and Therapeutics*, *11*(3), 663.
7. Sugur, H., Shastry, A. H., Sadashiva, N., Srinivas, D., Santosh, V., & Somanna, S. (2018). Chromosomal aberrations in chordoid meningioma–An analysis. *Neurology India*, *66*(1), 156-160.