Anaplastic meningioma presenting as a scalp mass with heterozygous CDKN2A deletion: a 13-year recurrence-free survival

V. Matthijs MD (1 2), W. Maenhoudt MD (2), J. Van Lerbeirghe MD (2), S. Du Four MD PhD (2),O. Van Damme MD (2), D. Vanhauwaert MD PhD (2 3)

[1] Neurosurgical training programme, Ghent University
[2] Department of Neurosurgery, AZ Delta, Roeselare, Belgium
[3] Academic consultant, Department of Human Structure and Repair,Ghent University

Introduction

Anaplastic meningiomas (WHO grade 3) are rare, highly aggressive tumours. Extracranial or scalp presentation is exceptional and often initially mistaken for primary skin or metastatic carcinoma. We describe a case with atypical clinical presentation and delayed molecular characterization revealing a heterozygous CDKN2A deletion.

Case

A 58-year-old man was referred in 2012 after a biopsy for a rapidly enlarging scalp lesion. Pathology revealed a large cell undifferentiated anaplastic malignancy. Subsequent neuroimaging showed a right frontoparietal extra-axial mass invading the calvarium and subcutaneous tissue, compatible with a meningioma. En bloc resection including dura and bone was performed, followed by adjuvant radiotherapy 56Gy. Histopathology confirmed an anaplastic meningioma (WHO grade 3) with marked atypia and high mitotic index. The postoperative course was complicated by a severe left sided sensory and motoric hemiplegia and CSF fistula which required revision surgery. Postoperative hemiparesis improved after multidisciplinary rehabilitation, with a residual weakness in the left sided plantar flexors and extensors (KPS 80-90). Next-generation sequencing (2024) of archival tissue identified a heterozygous CDKN2A deletion. At last follow-up (164 months) the patient remains free of recurrence, but clinically deteriorated in the last 2 years with predominantly paresis of the left lower limb and associated ataxia (KPS 60-70). This has been attributed to the late term effect of the EBRT.

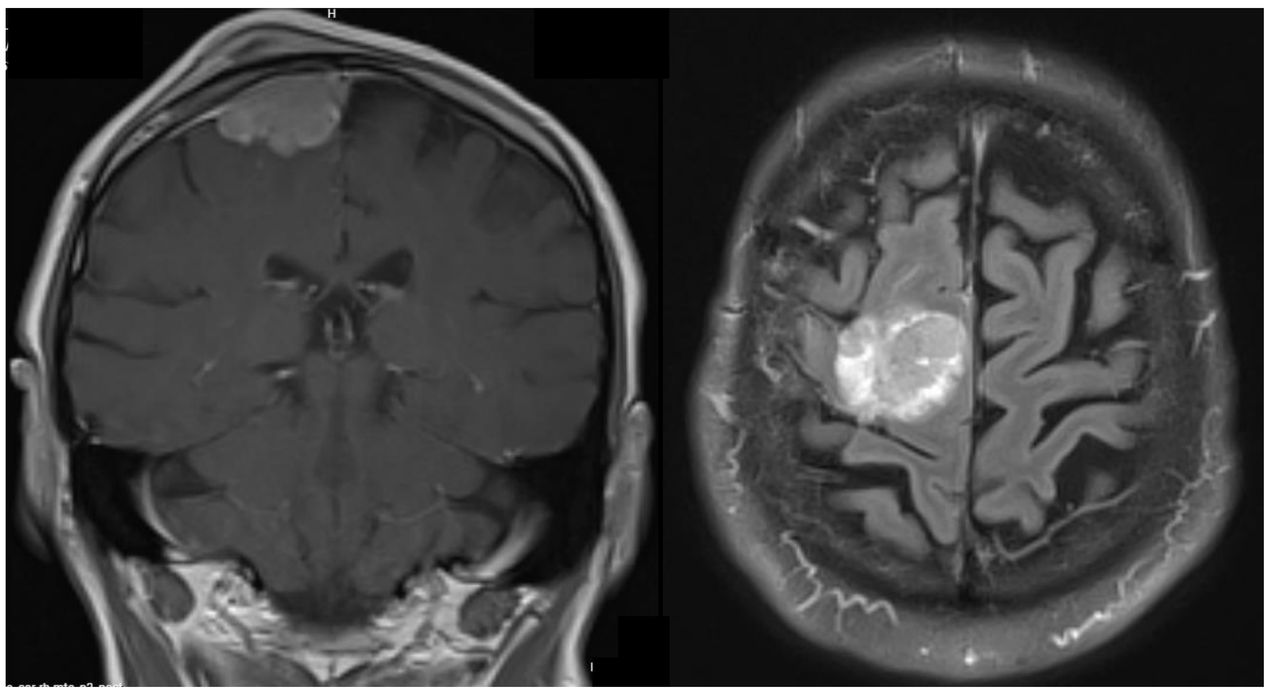

Fig 1. Initial presenting MRI scan revealing a large extra-axial lesion in the right frontal region. There is evident trans-osseous invasion into subcutaneous tissue and overlying skin.

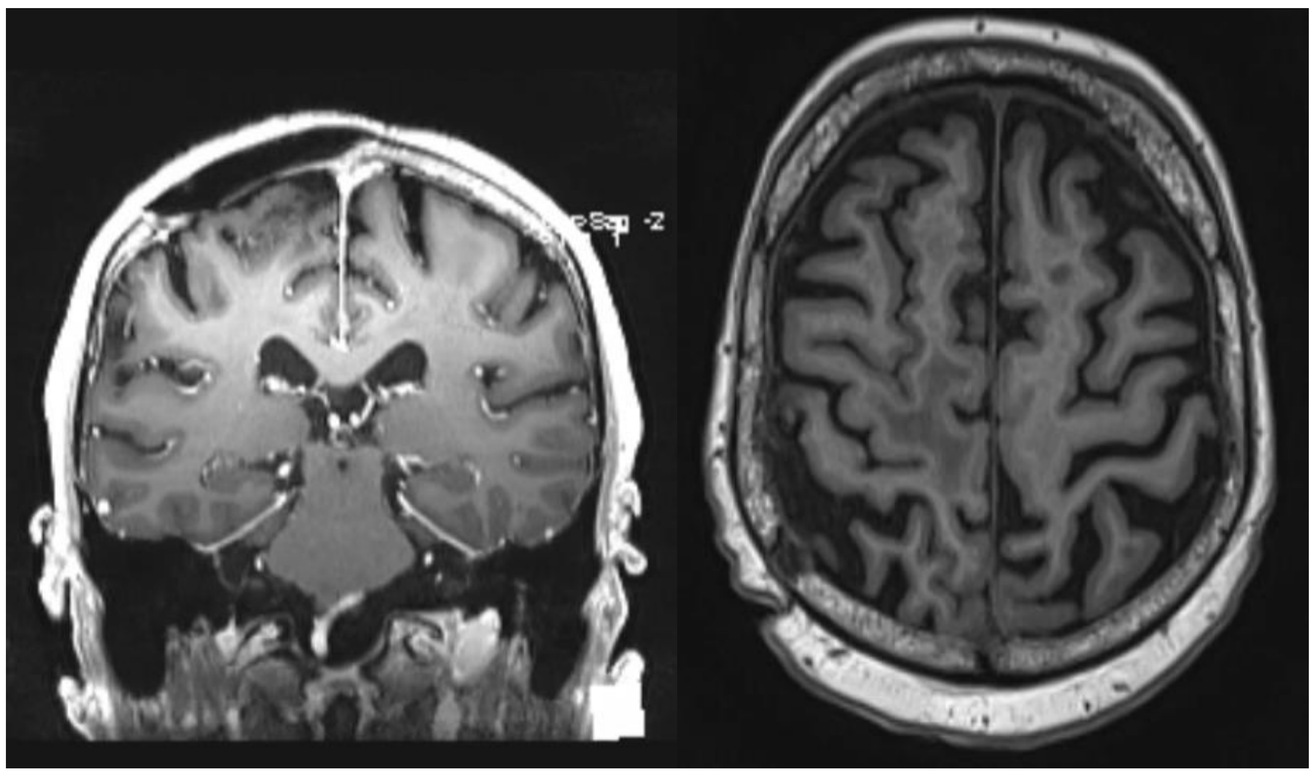

Fig 2. Follow-up MRI 13 years after initial resection.

Conclusion

This case illustrates a rare scalp presentation of an anaplastic meningioma with transosseous extension and long-term disease control after surgery and radiotherapy. The presence of a heterozygous CDKN2A deletion, while not diagnostic for grade 3 meningioma, may indicate increased biological aggressiveness. Late molecular profiling can provide valuable insights into tumour biology and help refine prognostic assessment and follow-up strategies.

References

1. Tosefsky K, Rebchuk AD, Wang JZ, Ellenbogen Y, Drexler R, Ricklefs FL, et al. Grade 3 meningioma survival and recurrence outcomes in an international multicenter cohort. J Neurosurg. 2024 Feb 1;140(2):393–403.
2. Wach J, Basaran AE, Arlt F, Vychopen M, Seidel C, Barrantes-Freer A, et al. CDKN2A/B deletions are strongly associated with meningioma progression: a meta-analysis of individual patient data. Vol. 11, Acta Neuropathologica Communications. BioMed Central Ltd; 2023.
3. Tabor J, O'brien J, Morales Valero SF, Pappajohn A, Declan Mcguone ;, Erson-Omay Z, et al. Heterozygous CDKN2A Loss is Associated with Recurrence and Survival in High, But Not Low Grade Meningiomas. 2024.
4. Goldbrunner R, Stavrinou P, Jenkinson MD, Sahm F, Mawrin C, Weber DC, et al. EANO guideline on the diagnosis and management of meningiomas. Neuro Oncol. 2021 Nov 1;23(11):1821–34.