

Anaplastic astrocytoma case reports

A case of [anaplastic astrocytoma](#) (WHO grade III) in an 18-month-old child with [Coffin-Siris syndrome](#) (CSS) due to a de novo germline missense [SMARCE1](#) mutation. Additional molecular features of the tumor are described as well. The role of missense SMARCE1 mutations in tumor predisposition in children with CSS should be further investigated to better inform genetic counselling ¹⁾.

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[Temozolomide](#) (TMZ) for [malignant gliomas](#) is traditionally dosed in 5 out of a 28-day cycle, however alternative regimens exist, including dose-dense. Continuous daily dosing is available, but the acceptable dose and duration of therapy is unknown.

Zhou et al. document a 40-year-old male with recurrent [anaplastic astrocytoma](#), [IDH mutant](#) and MGMT promotor methylation negative, who has well-tolerated continuous daily TMZ for 20 months at 100 mg per day for nearly the length of this period. A trial at 80 mg per day demonstrated disease progression with response upon return to 100 mg per day. Prior to the daily TMZ, the patient underwent three surgical resections, radiation therapy with concurrent TMZ according to the [EORTC NCIC protocol](#), and subsequently bevacizumab in combination with use of the Optune device. Long-term survival of patients with recurrent malignant gliomas is uncommon, and currently no standard treatment strategies exist for these patients. We present this case to demonstrate the tolerability and dose dependency of prolonged daily TMZ dosing as a therapeutic option for recurrent anaplastic astrocytomas ²⁾.

¹⁾

Lin B, Kesserwan C, Quinn EA, Einhaus SL, Wright KD, Azzato EM, Orr BA, Upadhyaya SA. Anaplastic Astrocytoma in a Child With Coffin-Siris Syndrome and a Germline SMARCE1 Mutation: A Case Report. J Pediatr Hematol Oncol. 2018 Nov 28. doi: 10.1097/MPH.0000000000001361. [Epub ahead of print] PubMed PMID: 30499906.

²⁾

Zhou Z, Howard TA, Villano JL. Long-term daily temozolomide with dose-dependent efficacy in MGMT promotor methylation negative recurrent high-grade astrocytoma. Cancer Chemother Pharmacol. 2017 Aug 8. doi: 10.1007/s00280-017-3415-5. [Epub ahead of print] PubMed PMID: 28791452.

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