World Neurosurg. 2024 Mar 13:S1878-8750(24)00403-0. doi: 10.1016/j.wneu.2024.03.028.
Online ahead of print.

Genetically Distinct Oligosarcoma Arising from Oligodendroglioma: Systematic Review & Illustrative Case Example

Alexander Evans [1], Kiana Y Prather [1], James Battiste [1], Kar-Ming Fung [2], Ian F Dunn [1], Christopher S Graffeo [3]

Affiliations

PMID: 38490447 DOI: 10.1016/j.wneu.2024.03.028

Abstract

Background: Oligosarcoma is a rare central nervous system (CNS) neoplasm that may arise following oligodendroglioma resection, which demonstrates a unique genetic profile and aggressive clinical phenotype. We present a systematic review and illustrative case example emphasizing the clinical and prognostic features of this unusual and unfavorable neuro-oncologic disease.

Methods: Systematic literature review and illustrative case report.

Results: A 41-year-old man who had undergone two neurosurgical resections for a WHO grade II oligodendroglioma (Ki-67=5-10%, 1p/19q co-deleted, IDH2 mutated), without adjuvant chemoradiation, presented with seizures seven years after resection. An extra-axial mass was identified adjacent to the resection cavity, in which gross total resection was achieved. Pathology confirmed WHO grade IV oligosarcoma (Ki-67=20%). Adjuvant chemoradiation was initiated, with disease control observed over 6 months of follow-up. 7 publications met inclusion criteria. Oligosarcoma has been confirmed in 36 lesions, arising in 35 patients; 5 were primary oligosarcoma, while 31 occurred in the setting of prior resected oligodendroglioma or oligoastrocytoma. Features shared by these lesions include re-gain of H3K27me3 expression, 1p/19q codeletion, homozygous deletion of CDKN2A/B, loss of 6q, loss of NF1 and YAP1, and attenuation of CpG island methylator (G-CIMP). Median survival after oligosarcoma diagnosis was 1.3 years (range, 0-5.2; n=35).

Conclusion: Oligosarcoma is a prognostically unfavorable CNS neoplasm with characteristic imaging and pathologic features, and a strong association with previously resected oligodendroglioma. Aggressive treatment is recommended, including gross total resection and adjuvant chemoradiation. Further study is required to define optimal treatment protocol for this CNS malignancy.

Keywords: genetics; oligodendroglioma; oligosarcoma; recurrence.