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Anaplastic ganglioglioma is a rare tumor and diagnosis has been based on histological criteria. The 5th edition of the World Health Organization Classification of Tumours of the Central Nervous System (CNS WHO) does not list anaplastic ganglioglioma as a distinct diagnosis due to a lack of molecular data in previous publications AIM:

Reinhardt et al. retrospectively compiled a cohort of 54 histologically diagnosed anaplastic gangliogliomas to explore whether the molecular profiles of these tumors represent a separate type or resolve into other entities METHODS: Samples were subjected to histological review, DNA methylation profiling, and next-generation sequencing. Morphologic and molecular data were summarised to an integrated diagnosis RESULTS: The majority of histologically diagnosed anaplastic gangliogliomas resolved into CNS WHO diagnoses of glial tumors, most commonly pleomorphic xanthoastrocytoma (16/54), glioblastoma, IDH wildtype, and diffuse pediatric-type high-grade glioma, H3 wildtype and IDH wildtype (11 and 2/54) followed by low-grade glial or glioneuronal tumors including pilocytic astrocytoma, dysembryoplastic neuroepithelial tumour and diffuse leptomeningeal glioneuronal tumor (5/54), IDH mutant astrocytoma (4/54) and others (6/54). A subset of tumors (10/54) was not assignable to a CNS WHO diagnosis and common molecular profiles pointing to a separate entity were not evident.

In summary, they showed that tumors histologically diagnosed as anaplastic ganglioglioma comprise a wide spectrum of CNS WHO tumor types with different prognostic and therapeutic implications. They, therefore, suggest assigning this designation with caution and recommend a comprehensive molecular workup ¹⁾.

Anaplastic ganglioglioma (AGG) is a very rare type of brain tumor that is a type of ganglioglioma. In general, gangliogliomas are classified as grade I or low-grade tumors, meaning that they grow slowly and are considered benign. Anaplastic gangliogliomas, however, are considered grade III or high-grade tumors, which means that they are usually aggressive, malignant tumors. The main treatment is the removal of the entire tumor during surgery. If the entire tumor is not removed, it has the potential to recur and may require additional surgery or treatments, such as radiation therapy or chemotherapy. Unfortunately, because gangliogliomas are quite rare, there is limited information to show that radiation therapy or chemotherapy are effective treatments for this condition ²⁾.

1)

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2)

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