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Primary brain tumours in adults

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Abstract

The most frequent adult-type primary CNS tumours are diffuse gliomas, but a large variety of rarer CNS tumour types exists. The classification of these tumours is increasingly based on molecular diagnostics, which is reflected in the extensive molecular foundation of the recent WHO 2021 classification of CNS tumours. Resection as extensive as is safely possible is the cornerstone of treatment in most gliomas, and is now also recommended early in the treatment of patients with radiological evidence of histologically low-grade tumours. For the adult-type diffuse glioma, standard of care is a combination of radiotherapy and chemotherapy. Although treatment with curative intent is not available, combined modality treatment has resulted in long-term survival (>10-20 years) for some patients with isocitrate dehydrogenase (IDH) mutant tumours. Other rarer tumours require tailored approaches, best delivered in specialised centres. Targeted treatments based on molecular alterations still only play a minor role in the treatment landscape of adult-type diffuse glioma, and today are mainly limited to patients with tumours with $BRAF^{V600E}$ (ie, Val600Glu) mutations. Immunotherapy for CNS tumours is still in its infancy, and so far, trials with checkpoint inhibitors and vaccination studies have not shown improvement in patient outcomes in glioblastoma. Current research is focused on improving our understanding of the immunosuppressive tumour environment, the molecular heterogeneity of tumours, and the role of tumour microtube network connections between cells in the tumour microenvironment. These factors all appear to play a role in treatment resistance, and indicate that novel approaches are needed to further improve outcomes of patients with CNS tumours.