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PrimeView
Glioma

Gliomas are primary brain tumours that can occur in children and adults. Prognosis varies widely depending on the molecular characteristics of the tumour. Gliomas are classified according to the 2021 WHO classification of central nervous system (CNS) tumours.

Epidemiology

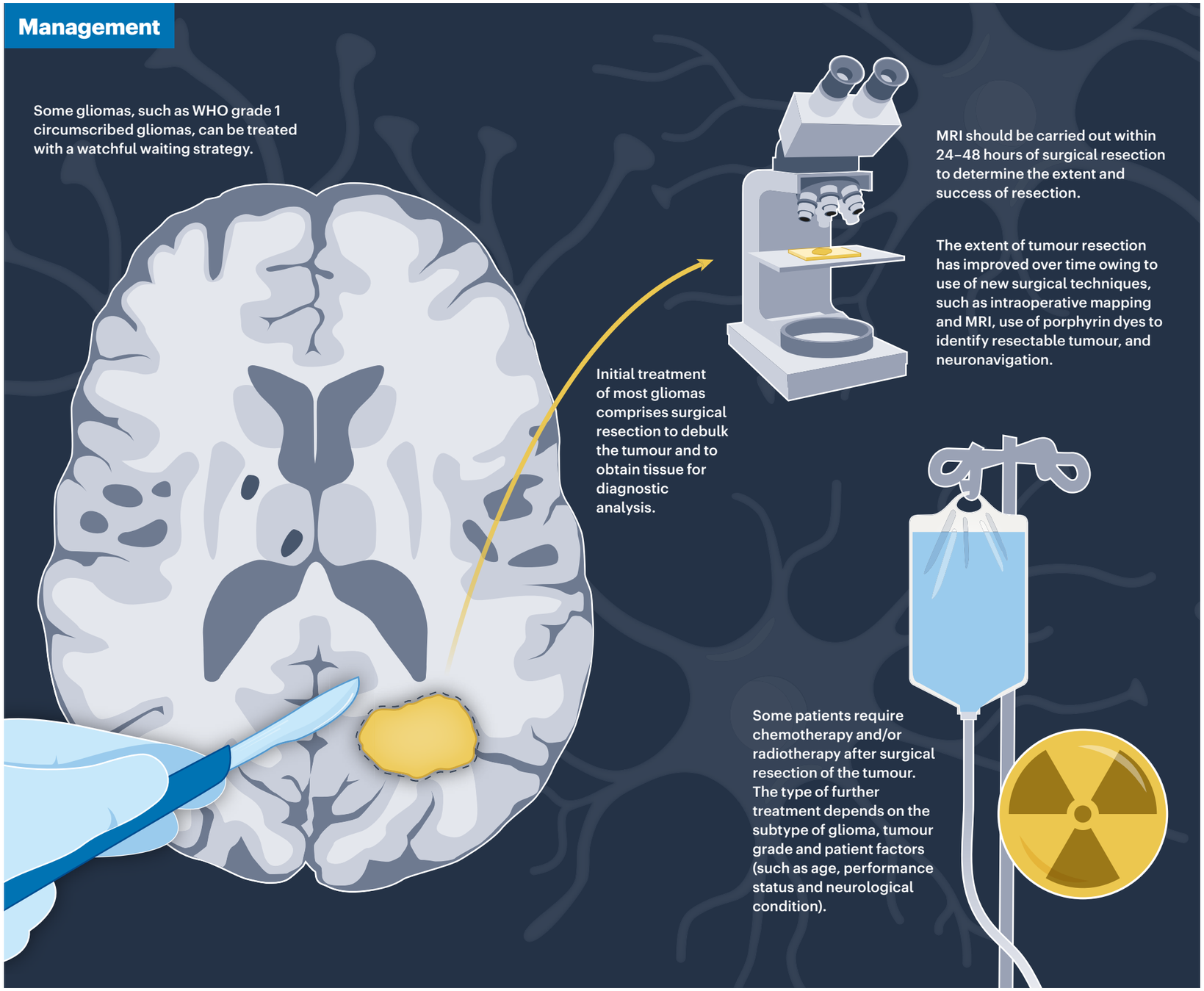
Robust data on the incidence and prevalence of gliomas classified according to the 2021 WHO classification are unavailable. Instead, most epidemiological data are based on gliomas classified according to the 2016 WHO criteria. The average annual age-adjusted incidence of CNS tumours in the USA is 24.8 per 100,000 population. Of these, the most common malignant CNS tumour is glioblastoma, which comprises 14.2% of all tumours and 50.9% of all malignant brain tumours in the Central Brain Tumor Registry of the United States.

- Risk factors for glioma include advanced age and exposure to ionizing radiation, for example, in those with childhood cancers who received radiotherapy. Cell phone use is not associated with increased risk of glioma. Some inherited cancer syndromes convey increased risk of glioma, such as neurofibromatosis type 1 and tuberous sclerosis complex.

Diagnosis

Manifestations of glioma vary by tumour location and include focal neurological deficits such as muscle weakness or seizures. Brain imaging — most commonly MRI — is used to detect gliomas. More advanced imaging techniques can be used to distinguish between gliomas and non-malignant brain tumours and, in some cases, identify specific subtypes of glioma (such as IDH-mutant gliomas by increased 2-hydroxyglutarate levels detected by MR spectroscopy). Histological and molecular evaluation of tumour samples is essential to confirm diagnosis. Tissue samples are evaluated for the presence of several molecular markers of glioma subtypes, such as IDH and *ATRX* mutation status, and to determine tumour grade. Techniques for genetic analysis of these tumours include next-generation sequencing and DNA methylation profiling, in addition to single-gene analysis.

Management



Mechanisms

The genetics of gliomas is very complex. In the 2021 WHO classification, five major groups of gliomas are distinguished based on their molecular characteristics: adult-type diffuse gliomas, paediatric-type diffuse high-grade gliomas, paediatric-type diffuse low-grade gliomas, circumscribed astrocytic gliomas and ependymal tumours. Each of these groups can be further classified into subtypes. The most common group is adult-type diffuse gliomas, which account for >90% of gliomas. These tumours are characterized by the presence or absence of IDH mutations and 1p/19q codeletion. Characteristic mutations and genetic alterations for other groups and subtypes of gliomas vary widely and include mutations in histone genes, gene fusions and hypermethylation. The microenvironment of diffuse gliomas comprises immune and CNS cells. Interactions between the tumour and the microenvironment promote tumour initiation, maintenance, growth, proliferation and invasion.

- Gliomas affect health-related quality of life directly and indirectly, the latter owing to treatment-related adverse effects.

Outlook

The use of high-throughput molecular analysis (next-generation sequencing and DNA methylation profiling) has improved diagnosis of gliomas, but the availability of these analyses varies between regions. Improving the availability and affordability of such techniques is essential for their routine use. Other improvements to diagnosis are also required, such as standardization of techniques used for molecular analysis and neuroimaging protocols.

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