



Central nervous system gliomas



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ABSTRACT

Evidence-based practical guidelines on diagnosis, prognosis, and treatment on the most frequent adult brain tumours are delineated. In Europe, 27,000 new cases of malignant glial tumours and 1000 new cases of malignant ependymal tumours are diagnosed every year. The most common glial tumours are glioblastoma multiforme and anaplastic glioma, comprising more than 50% and 10%, respectively, of the total gliomas.

Prognosis of gliomas is generally poor. Environmental and genetic factors have been correlated with an increased risk of developing brain tumours.

Surgical resection represents the first treatment option for all histotypes.

Role and timing of radiotherapy and chemotherapy as well as treatment for recurrent/progressive disease should be based on age, performance status, histopathological diagnosis, molecular markers, and previous therapy. Impaired neurocognitive and neuropsychological function is common in long-term survivors, regardless of the histology and grade of the tumour and should be taken into account in treatment planning.

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1. General information

1.1. Astrocytic tumours

1.1.1. Incidence

In Europe, about 27,000 new cases of malignant astrocytic tumours are diagnosed per year with a crude annual incidence rate, for the period 2000–2007, of 5 per 100,000 (RARECAREnet, 2017). Incidence is significantly higher in males than in females, the adjusted rates being 5.4 and 3.6, respectively. Incidence increases with age, from 0.9 in children to 12.1 in the elderly (aged 65+ years). There are geographical differences in incidence, with the highest rates in the North of Europe, UK and Ireland (5.7) and the lowest in the South and East of Europe (3.9 and 3.7, respectively). During the period 1995–2007, incidence rates (age-adjusted) slightly and significantly increased from 4.3 (1995–1999) to 4.5 (2003–2007). Geographical differences, at least for southern Europe, unlikely are explained by different access to more sophisticated diagnostic tools and may be attributed to different genetic or environmental ethiology. With regard to time trends, more recent data show stable (or even decreasing) incidence except for the age group 60+ years which shows an increasing trend (Crocetti et al., 2012a), maybe related to improved ability to diagnose brain tumours by stereotactic biopsy, especially in elderly. The most common astrocytic tumours are glioblastoma multiforme and anaplastic glioma, comprising slightly more than 50% and 10%, respectively, of the total astrocytomas (Sant et al., 2012). Glioblastoma multiforme is approximately 5 times higher in elderly (65+ years of age) than in patients 20–64 years of age (Chakrabarti et al., 2005). For anaplastic glioma, the lowest incidence is found among children and adolescents; the highest in the elderly. In the US data, incidence peaked at age 65–79 years (Chakrabarti et al., 2005; Surawicz et al., 1999).

1.1.2. Prevalence

Prevalence is an important indicator of the burden for the health care system. As the prevalence of these tumours was below the threshold of the European Community definition for rare diseases (50 per 100,000) these tumours are classified as rare diseases and may profit from the EU Directive on orphan drugs (European Parliament Council of the European Communities, 2017).

In Europe, over 100,000 people were living with a diagnosis of astrocytic tumours (Crocetti et al., 2012b) at the beginning of 2008. Thirty-one percent of them survived 5 years or less after diagnosis, while about 50% survived 15 years or more after their diagnosis. Five-year relative survival was 14.5% for astrocytic tumours, 54.5% for oligodendroglial tumours.

1.1.3. Aetiology and risk factors

1.1.3.1. Environmental and genetic factors.

Therapeutic ionising radiation is a relevant risk factor for brain tumours (Wrensch et al., 2002). Prior therapeutic irradiation is correlated with an increased risk of developing brain tumours (Hodges et al., 1992). However, other epidemiological studies did not find any significantly increased risk of brain tumours for medical ionising radiation, including radiotherapy and exposure to low doses of ionizing radiation, such as X-rays, scans, and scintigraphy (Blettner et al., 2007). A more recent and large cohort study on about 700,000 people exposed to computed tomography scans in childhood and adolescence, reported a doubling of risk for brain tumours, with a dose-response relationship and greater risk after exposure at younger ages. The average effective radiation dose per scan was estimated as 4.5 mSv (Mathews et al., 2013).

Several occupational studies have shown a high risk of astrocytomas among people with electrical or electronics jobs, or workers exposed to organic chemicals in petroleum refining and chemical

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