Epidemiology of glial and non-glial brain tumours in Europe

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Abstract To the central nervous system (CNS) belong a heterogeneous group of glial and non glial rare cancers. The aim of the present study was to estimate the burden (incidence, prevalence, survival and proportion of cured) for the principal CNS cancers in Europe (EU27) and in European regions using population-based data from cancer registries participating in the RARECARE project. We analysed 44,947 rare CNS cancers diagnosed from 1995 to 2002 (with follow up at 31st December 2003): 86.0% astrocytic (24% low grade, 63% high grade and 13% glioma NOS), 6.4% oligodendrogial (74% low grade), 3.6% ependymal (85% low grade), 4.1% Embryonal tumours and 0.1% choroid plexus carcinoma. Incidence rates vary widely across European regions especially for astrocytic tumours ranging from 3/100,000 in Eastern Europe to 5/100,000 in United Kingdom and Ireland. Overall, about 27,700 new rare CNS cancers were estimated every year in EU27, for an annual incidence rate of 4.8 per 100,000 for astrocytic, 0.4 for oligodendrogial, 0.2 for ependymal and embryonal tumours and less than 0.1 for choroid plexus carcinoma. More than 154,000 persons with rare CNS were estimated alive (prevalent cases) in the EU at the beginning of 2008.
Central Nervous System (CNS) cancers are a group of different tumour entities anatomically close to each other but diverse in terms of morphology, site, molecular biology and clinical behaviour and presumably aetiology.1

In Europe, the standardised (World) incidence of primary CNS cancers ranges from 4.5 to 11.2 cases per 100,000 men and from 1.6 to 8.5 per 100,000 women.2 The two most common CNS cancers, high-grade glioma and brain metastasis occur more frequently during adulthood and especially among the elderly. In Europe, the peak of incidence is 18.5/100,000 in people aged ≥65 years.2 The relative frequency of CNS tumours is however highest during childhood, when they account for 23% of all the cancers diagnosed.3

In adults the 5-year survival rate for the primary CNS cancers in Europe was 17% for males and 19% for females (1995–2002),4 with differences across European regions.5 Survivorship is higher for young European patients – 63% – than for the elderly ones.6

Statistics on CNS tumours are estimated by grouping all malignancies arising in all the CNS anatomic sites: meninges, brain, spinal cord, cranial nerves and other localisation of CNS (ICD-10 topography codes C70-C72).7

However, rare tumours are more appropriately defined as a combination of topographical and morphological characteristics, as defined by the International Classification of Diseases for Oncology (ICD-O).5 The Surveillance of Rare Cancers in Europe (RARECARE; www.rarecare.eu) project, is a large collaboration of population-based cancer registries (CRs) across Europe which provides a list of rare cancers on the basis of topography and morphology. Clinical factors, such as difficulties in achieving diagnosis, in clinical decision making and in conducting clinical studies and the lack of standardised treatment mainly affected the definition of the type of cancers of the RARECARE list. Under the threshold proposed by the RARECARE project, incidence lower than 6 per 100,000 per year, CNS malignant tumours are included. However, despite the rarity of entities selected by using this cut-off, there are many sub-entities for each group (i.e. different histologies and WHO grade of glial tumours) with different prognosis and different treatment approaches that complicate the picture. This means that management of these rare brain tumour subgroups has to be considered hyper-specialistic, as well as treatment options centralised in large volumes hub centres with recognised expertise in this field. Furthermore, central pathology review in CNS tumours is particularly important. The experience of modern clinical trials (i.e. European Organisation for Research and Treatment of Cancer (EORTC) and Radiation Therapy Oncology Group (RTOG) studies on grade III gliomas) showed very high (up to 50%) inter-observer variability among neuropathologists in the differential diagnosis between astrocytic and oligodendrogial tumours.9

The objective of this study was to establish a picture of incidence, prevalence and survival of rare CNS cancers in Europe based on the new RARECARE list of tumours.

2. Materials and methods/cancer cases

Rare cancers of the CNS presented in this paper are based on the new list of cancer types provided by RARECARE. The list is organised into three tiers. The bottom tier (tier 3) corresponds to the World Health Organisation (WHO) name of individual cancer entities [http://www.iarc.fr/en/publications/pdfs-online/pat-gen/] and their corresponding ICD-O-3 morphology and topography codes. Tier 3 entities were grouped into categories of cancers (tier 2) considered similar from the point of view of clinical management and research. These categories were further grouped into more general categories (tier 1), considered to involve the same clinical expertise and patient referral structure. Accordingly the following cancer entities were identified and will be described in this paper:

Glial tumours (tier 1 entity) which include astrocytic, oligodendrogial, oligoastrocytic and ependymal...