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Descriptive epidemiology of malignant mucosal and uveal melanomas and adnexal skin carcinomas in Europe

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ABSTRACT

This work provides descriptive epidemiological data of malignant mucosal and uveal melanomas and adnexal skin carcinomas in Europe as defined as in the RARECARE project. We analysed 8669 incident cases registered in the period 1995–2002 by 76 population-based cancer registries (CRs), and followed up for vital status to 31st December 2003. Age-standardised incidence to the European standard population was obtained restricting the analysis to 8416 cancer cases collected by 64 not specialised CRs or with information available only for some anatomical sites. Period survival rates at 2000–2002 were estimated on 45 CRs data. Twenty-two CRs which covered the period 1988–2002 were analysed to obtain the 15-year prevalence (1st January 2003 as reference date). Complete prevalence was calculated by using the completeness index method which estimates surviving cases diagnosed prior to 1988 ('unobserved' prevalence). The expected number of new cases per year and of prevalent cases in Europe was then obtained multiplying the crude incidence and complete prevalence rates to the European population at 2008. We estimated 5204 new cases per year (10.5 per million) to occur in Europe, of which 48.7% were melanomas of uvea, 24.8% melanomas of mucosa and 26.5% adnexal carcinomas of the skin. Five-year relative survival was 40.6% and 68.9% for mucosal and uveal melanomas, respectively. Adnexal skin carcinomas showed a good prognosis with a survival of 87.7% 5 years after diagnosis. Northern Europe, United Kingdom (UK) and Ireland showed the highest 5-year

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survival rate for uveal melanomas (72.6% and 73.4%), while Southern Europe showed the lowest rate (63.7%). More than 50,000 persons with a past diagnosis of one of these rare cancers were estimated to be alive at 2008 in Europe, most of them (58.8%, $n = 29,676$) being patients with uveal melanoma. Due to the good prognosis and high incidence of uveal melanomas, these malignancies are highly represented among the long-term survivors of the studied rare cancer types. Therefore, maximising quality of life is particularly important in treatment of uveal melanoma. As regards mucosal melanomas, the centralisation of treatment to a select number of specialist centres as well as the establishment of expert pathology panels should be promoted. The geographical differences in incidence and survival should be further investigated analysing the centre of treatment, the stage at diagnosis and the treatment.

1. Introduction

This work is focused on three tumour types which are rare and understudied, thus, poorly understood: uveal and mucosal melanomas, adnexal skin carcinomas. Uveal melanoma is the most common adult intraocular tumour, arising from melanocytes in the uvea. Mucosal melanoma develops in the mucous membrane that lines the nose, mouth, oesophagus, anus, urinary tract and vagina. Adnexal skin tumours are extremely diverse group of neoplasms, arising from cutaneous appendages particularly the sebaceous, apocrine and eccrine glands. Because of their rarity, even the basic descriptive epidemiology of these three tumour types is sparse, restricted to specific anatomic sites and confined to case reports or clinical series. To estimate the cancer burden, the most appropriate data are provided by population-based cancer registries (CRs) which include all cases diagnosed in a well-defined population. The Surveillance of Rare Cancers in Europe[1] (RARECARE) is a large collaboration project of population-based CRs across Europe funded to deal with the issue of rare cancers. The RARECARE working group produced a new list of both rare and common tumours, and developed a new operational definition of rarity. In this work we describe incidence, prevalence and survival patterns in Europe for malignant melanomas of mucosa and uvea, and adnexal skin carcinomas as defined in the RARECARE list of rare tumours.

2. Materials and method

2.1. Data and tumours definition

The RARECARE project extracted data on patients registered by 89 CRs in the period 1978–2002 and followed up for vital status at least to 31st December 2003, from the EUROCARE-4 database.[2] The mean population covered was about 162,000,000 corresponding to 32% of the European population (EU27). Malignant melanoma of mucosa, of uvea and adnexal carcinomas of the skin were drawn from the RARECARE list of tumours defined on the base of morphology and topography codes, according to the third revision of the International Classification of Diseases for Oncology (ICD-O-3).[3] These cancer entities were conceived to be meaningful for clinical decision-making and clinical studies, and identified as rare cancers on the base of their annual incidence rate of less than 6/100,000 per million person-years (pyr). Incidence, which is not affected by variations in life expectancy was considered as a better criterion than prevalence of the burden posed on organisation of health care by rare cancers.

2.2. Statistical analysis

The study was done on 9844 incident malignant cases diagnosed in the period 1995–2002 and registered in 76 CRs. Crude incidence by age as the number of new cases occurring in 1995–2002 divided by the total pyr in the general population (male and female) was obtained restricting the analysis to 8416 cases collected by 64 CRs since we excluded specialised CRs or other not specialised with information available only for some anatomical sites. The European standard population was used to estimate the age-standardised incidence (ASR), overall, by sex and by the following European regions: Northern Europe, Central Europe, Eastern Europe, Southern Europe, United Kingdom (UK) and Ireland.

Relative survival rates were estimated by the period approach[4] in 2000–2002 as the ratio of absolute survival to the expected survival in the general population of the same age and sex. For this analysis, 45 CRs contributing to the considered period were used.

The counting method was applied to 22 CRs which covered the period 1988–2002, choosing 1st January 2003 as reference date, to obtain the observed prevalence[5] of cases diagnosed within 2, 5 and 15 years of the index date. A completeness index was used to estimate the complete prevalence by adding the estimated surviving cases diagnosed prior to 1988 ('unobserved' prevalence) to those counted in 1988–2002[6] (15-year observed prevalence). The completeness indices were obtained by modelling 1985–1999 incidence data with a logistic exponential or polynomial function on age and 1988–1999 survival data with parametric cure models.[7] The expected number of new cases per year and of prevalent cases in Europe (EU27) was calculated by multiplying the incidence and complete prevalence to the 2008 European population (497.5 million) provided by EUROSTAT.[8]

The incidence, survival and prevalence rates and their corresponding standard errors and 95% confidence intervals have been calculated by using SEER*Stat software.[9] We used the SAS software[10] to model incidence and survival data, and the ComPrev software[11] to calculate the completeness index.