Incidence, survival and prevalence of myeloid malignancies in Europe

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Abstract
Background: The Surveillance of Rare Cancers in Europe (RARECARE) project aims at increasing knowledge of rare cancers in Europe. This manuscript describes the epidemiology of myeloid malignancies (MMs), taking into account the morphological characterisation of these tumours.

Methods: We used data gathered by RARECARE on cancer patients diagnosed from 1995 to 2002 and archived in 64 European population-based cancer registries, followed up to 31st December 2003 or later.

Results: The overall annual crude incidence of MMs was 8.6 per 100,000. Acute myeloid leukaemia (AML) and myeloproliferative neoplasms (MPN) were most common, with incidence rates of 3.7 and 3.1 per 100,000 year respectively, followed by 1.8 for myelodysplastic syndromes (MDS) and myelodysplastic/myeloproliferative neoplasms (MD/MPN) and 0.1 for histiocytic and dendritic cell neoplasms (HDCN). The 5-year relative survival rate ranged from 18% for chronic myelomonocytic leukaemia, 19% for AML, 29% for MDS and 44% for chronic myeloid leukaemia to relatively favourable rates for MPN (62%) and HDCN.
1. Introduction

Haematological malignancies (HMs) are thought to derive from a pluripotential or multipotential stem cell that gives rise to very diverse proliferations, some of them being characterised by specific genetic abnormalities.\(^1\) Changing definitions and classifications of HMs complicate epidemiological studies and comparative analyses of incidence, survival and prevalence of these diseases.

In the past, the generic term “leukaemia” was used to group those types of lymphoid and myeloid proliferations which mainly involved the peripheral blood, subdivided into chronic and acute leukaemia. In the third edition of the International Classification of Diseases for Oncology (ICD-O-3),\(^3\) HMs are divided into two main categories based on cell lineage — myeloid and lymphoid. Within each of these groups, HMs are further sub-divided according to cell of origin, extent of maturation, morphology, immunohistochemistry, genetic characteristics and clinical behaviour.

This disease grouping is more useful for epidemiologic and public health studies on incidence, survival and prevalence and for testing aetiological hypotheses, because these groups are likely to have a distinct physiopathology and prognosis, and are more compatible with clinical classifications than the broad categories used by cancer registries (CRs) in the past.

In 2008, 78,416 cases of leukaemia - including both lymphoid and myeloid leukaemia - were estimated in Europe by the GLOBOCAN project, accounting for approximately 2.4% of all cancers.\(^4\) The overall age standardised (world population) incidence rate of leukaemia was 6.8 per 100,000 (crude rate: 10.7).\(^4\) Figures by morphological subtype were recently provided for the 20 European countries represented in the cancer registry-based project on Haematological malignancies (HAEMACARE), in which the age-standardised incidence rate per 100,000 was 7.5 for myeloid malignancies (MMs) and 24.5 for lymphoid malignancies.\(^5\)

The aim of the present study is to provide estimates of incidence, prevalence and survival for the MMs in Europe according to the grouping proposed by the project Surveillance of Rare Cancers in Europe (RARECARE). In Europe there was no internationally agreed definition of rare cancers thus RARECARE has provided a definition of rare cancers and a list of rare entities.\(^6\) An international group of experts agreed that rare cancers are those that present specific problems in clinical decision making, health care organisation and clinical research because of their low frequency. Thus, the definition of rare cancers was based on their frequency and rare cancers were defined as those with an annual incidence below six per 100,000. According to this definition all MMs are actually rare.

2. Material and methods

The present analyses are based on the list of cancers provided by RARECARE.\(^6\) The grouping of rare cancers was based on the ICD-O-3 classification system.\(^3\) However, the groups of tumours were identified not only by considering their morphology, but also by aggregating single tumours to clinically meaningful groups (i.e. perceived by clinicians as single diseases). Such grouping is more relevant for clinical decision making and research. The RARECARE list classifies cancer entities into a two-tier system (Table 1). Tier 1 is the more general category with cancers considered to involve the same clinical expertise and patient referral structure; the tier 2 subcategories include cancers similar from the point of view of clinical management and research.

In the present study, we investigated cancers belonging to four RARECARE tier 1 categories: “acute myeloid leukaemia (AML) and related precursor neoplasms”, “myeloproliferative neoplasms (MPN)”, “myelodysplastic syndrome (MDS) and myelodysplastic/myeloproliferative neoplasms (MD/MPN)” and “histiocytic and dendritic cell neoplasms (HDCN)” (Table 1). AML and related precursor neoplasms comprise only two tier-2 entities: “acute promyelocytic leukaemia (APL)” and “other AML, including also those not otherwise specified (NOS)”. MPN comprise three tier-2 entities: “chronic myeloid leukaemia (CML)”, “mast cell tumours” and “other myeloproliferative neoplasms”. MDS and MD/MPN comprise 4 tier-2 entities: “MDS with 5q- syndrome”, “other and unspecified MDS”, “chronic myelomonocytic leukaemia (CMML)” and “atypical chronic myeloid leukaemia, BCR/ABL negative (aCML)”. HDCN do not have any tier-2 entities. The corresponding ICD-O-3 morphology codes for all entities are shown in the last column of Table 1.