Rare thoracic cancers, including peritoneum mesothelioma

Sabine Siesling a,b,*, Jan Maarten van der Zwan a, Isabel Izarzugaza c, Jana Jaal d, Tom Treasure e, Roberto Foschi f, Umberto Ricardi g, Harry Groen h, Andrea Tavilla i, Eva Ardanaz j,k, on behalf of the RARECARE working group 1

a Department of Registry and Research, Comprehensive Cancer Centre The Netherlands, P.O. Box 330, 9700 AH Groningen, The Netherlands
b Twente University, Health Technology and Services Research, P.O. Box 217, 7500 AE Enschede, The Netherlands
c Basque Country Cancer Registry, Información Sanitaria, Dpt. de Sanidad del Gobierno Vasco, 01010 Vitoria-Gasteiz, Spain
d Department of Radiotherapy and Oncological Therapy, Haematology and Oncology Clinic, Tartu University Hospital, Vallikraavi str. 10, 51003 Tartu, Estonia
e Clinical Operational Research Unit, UCL (Department of Mathematics), 4 Taciton Street, London WC1H 0BT, UK
f Department of Cancer Medicine, Fondazione IHCSS, Istituto Nazionale dei Tumori, Via Venezian 1, 20133 Milan, Italy
g Radiation Oncology, University of Turin, Azienda Ospedaliero-Universitaria San Giovanni Battista di Torino, Via Giuseppe Verdi 8, 10124 Torino, Italy
h University Medical Centre Groningen, Dept. Pulmonology, Hanzeplein 1, P.O. Box 30.001, 9700 RB Groningen, The Netherlands
i Department of Cancer Epidemiology, Istituto Superiore di Sanità, Viale Regina Elena 299, Rome, Italy
j Registro de Cáncer de Navarra, Instituto de Salud Pública de Navarra, Leyre 15, 31003 Pamplona, Navarra, Spain
k CIBER Epidemiology and Public Health CIBERESP, Spain

Available online 9 March 2012

Abstract Rare thoracic cancers include those of the trachea, thymus and mesothelioma (including peritoneum mesothelioma). The aim of this study was to describe the incidence, prevalence and survival of rare thoracic tumours using a large database, which includes cancer patients diagnosed from 1978 to 2002, registered in 89 population-based cancer registries (CRs) and followed-up to 31st December 2003.

Over 17,688 cases of rare thoracic cancers were selected based on the list of the RACECARE project.

Mesothelioma was the most common tumour (19 per million per year) followed by epithelial tumours of the trachea and thymus (1.3 and 1.7, respectively). The age standardised incidence rates of epithelial tumours of the trachea was double in Eastern and Southern Europe versus the other European regions: 2 per million per year. Epithelial tumours of the thymus had the lowest incidence in Northern and Eastern Europe and UK and Ireland and somewhat higher
1. Introduction

Rare thoracic cancers are located in the chest and include those of the trachea, of the thymus and mesothelioma. Apart from mesothelioma, little information is available on their patterns of incidence and survival. This is largely because in the routine statistics and publications these tumours are grouped together with other sites. Tumours of the trachea are grouped with lung and bronchus and tumours of the thymus are often grouped together with those of heart, mediastinum and pleura and called ‘Other thoracic organs’.1

Moreover, the three tumour types have a different aetiology. As with lung cancer, cancer of the trachea is associated with active and passive smoking (environmental exposure). Survival is comparable with the survival of lung cancer, thus very low. The causation of mesothelioma by asbestos has been established for more than 50 years.2 The use of this dangerous carcinogen peaked between 1970 and 1990. Still the worldwide production has not declined significantly, resulting in an ongoing rise in incidence and mortality. In most industrialised countries more than 90% of all (pleural) mesotheliomas are related to asbestos exposure. Tumours of the thymus have a largely unknown aetiology with a complex biology. The most frequent tumours of the thymus are the thymomas. Survival of thyromas is mainly related to the stage at diagnosis, histological type and completeness of resection.3,4

In the present study, population-based data from different European cancer registries (CRs) participating in the RARECARE project, were used to estimate the burden of rare thoracic cancers. This database gives us the unique opportunity to study these rarities. The RARECARE project produced a list of tumours based on both cancer morphologies and topographies according to the third revision of the International Classification of Diseases for Oncology (ICD-O-3),5 using an incidence rate less than 6/100,000 as a threshold for rarity.

The aim of this study was to describe the incidence, prevalence and survival of the epithelial cancers of the trachea, thymus, and mesothelioma. Malignant mesothelioma most commonly arises in the pleura but can also arise in the peritoneum. To give a complete overview of the burden of mesothelioma we included the mesothelioma located on the peritoneum as well in our study. Furthermore, for the first time ever complete prevalence estimates will be reported for these specific types of rare tumours.

2. Patients and methods

2.1. Tumour grouping

The rare thoracic cancers described in this article include the epithelial tumours of the trachea, epithelial tumours of the thymus and malignant mesothelioma, including both mesotheliomas in the pleura and in the peritoneum. The present analyses are based on the list of cancers provided by RARECARE. The list is based on the ICD-O-3 and is organised in two hierarchical tiers (Table 1). Tier 2 includes cancer entities considered similar from the point of view of clinical management and research. Tier 2 cancer entities were grouped into general categories (tier 1 of the list) considered to involve the same clinical expertise and patient referral structure. For rare epithelial thoracic cancers described in this paper, there are three ‘tier 1’: epithelial tumours of the trachea (C33), thymus (C37) and mesothelioma (ICD-O-3 morphology codes 9050–9053).

For epithelial cancer of the trachea three ‘tier 2’ entities were identified: squamous cell carcinoma (ICD-O morphology codes 8004, 8020–8022, 8031–8032, 8050–8076, 8078, 8082–8084, 8560, 8980); adenocarcinoma (8140–8141, 8143–8144, 8147, 8190, 8201, 8210–8211, 8221, 8230–8231, 8255, 8260–8263, 8290, 8310, 8315, 8320, 8323, 8333, 8380–8384, 8440–8441, 8470, 8480–8482, 8490, 8504, 8510, 8512, 8514, 8525, 8542, 8545–8551, 8562–8576); and salivary gland type tumours (8200, 8430, 8982; thus including adenoid cystic carcinoma, mucoepidermoid carcinoma and myoepithelial carcinoma).

For epithelial cancer of thymus five ‘tier 2’ entities were identified: malignant thymoma (8580–8586; thus including not otherwise specified (NOS, 8580), type AB (8582), type A (8581), type B (8583, 8584, 8585), type C (8586)); squamous cell carcinoma (8051–8076, 8078, 8083–8084); undifferentiated carcinoma (8020–8022); lympho-epithelial carcinoma (8082) and adenocarcinoma (the same as for trachea).

For mesothelioma, two ‘tier 2’ entities were recognised: mesothelioma of pleura and pericardium (C38)