Rare Skin Cancer In England

NCIN Data Briefing

KEY MESSAGE:

Rare skin cancers are a mixed group in terms of causation, pathogenesis and outcome. Their rarity makes it challenging to study them. This data briefing describes their incidence and highlights the poor outcome for patients with Merkel cell carcinoma

Introduction

Rare skin cancers are a mixed group of non-melanoma skin cancers (ICD code C44) but can be classified into 2 broad categories, dermal and epidermal (Table 1). (http://www.cancerhelp.org.uk/type/skin-cancer/about/the-skin).

Cutaneous T cell lymphomas are considered as rare skin cancers but are not included in this analysis. Although these cancers all arise in the skin their evolution and biology differs and while the majority are managed by skin cancer MDTs some, such as dermatofibrosarcomas, may also be managed by sarcoma services making analysis of their clinical pathways complex.

Merkel cell carcinoma, the most frequent type of rare skin cancer, is most common on sun-exposed skin of the elderly. Discovery of the Merkel cell polyomavirus and evidence of increased risk in immunosuppression has changed our understanding of the tumour in recent years. In 2008 the age standardised incidence rate for rare skin cancer in England was 0.9 per 100,000 population compared with 16.1 per 100,000 population for malignant melanoma.

Table 1: Rare Skin Cancer Morphology codes, definitions and number of cases in the ten year period (1999-2008)

Epidermal			***Dermal***		
8022/3	*Pleomorphic carcinoma of the skin*	*5*	*8804/3*	*Epithelioid cell sarcoma*	*11*
8093/3	*Fibro-epithelial tumour of Pinkus*	*201*	*8830/3*	*Malignant fibrous histiocytoma*	*113*
8110/3	*Pilomatrix carcinoma*	*53*	*8832/3*	*Dermatofibrosarcoma, NOS*	*1,228*
8200/3	*Adenoid cystic carcinoma*	*179*	*8850/3*	*Liposarcoma, NOS*	*16*
8247/3	*Merkel cell carcinoma*	*1,515*	*8890/3*	*Leiomyosarcoma, NOS*	*289*
8390/3	*Skin appendage carcinoma*	*387*	*8900/3*	*Rhabdomyosarcoma, NOS*	*7*
8400/3	*Sweat gland adenocarcinoma*	*176*	*9120/3*	*Haemangiosarcoma*	*108*
8401/3	*Apocrine adenocarcinoma*	*52*	*9130/3*	*Malignant haemangioendothelioma*	*2*
8410/3	*Sebaceous adenocarcinoma*	*713*	*9133/3*	*Malignant epithelioid haemangioendothelioma*	*1*
8430/3	*Mucoepidermoid carcinoma*	*25*	*9540/3*	*Neurofibrosarcoma*	*2*
8480/3	*Mucinous adenocarcinoma*	*76*			
8940/3	*Mixed tumour, NOS*	*10*			

Method and main data

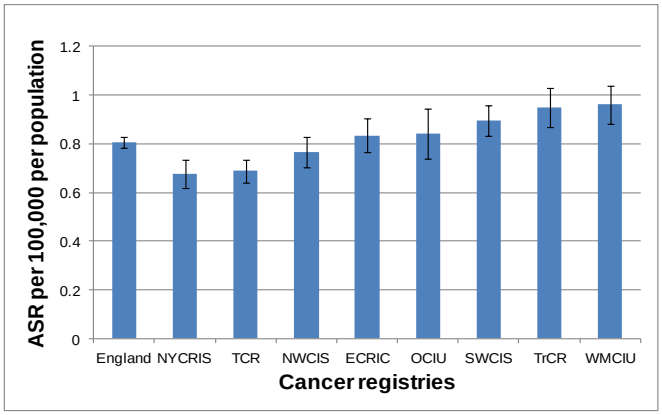
Data were extracted from the National Cancer Data Repository (NCDR) for the years 1999 to 2008. The cases were selected by ICD code C44 (other malignant neoplasm of skin) and morphology codes as listed in Table 1 were used.

5,169 new cases of rare skin cancers were registered from 1999 to 2008 in England. 3,392 were epidermal (including 1,515 cases of Merkel cell carcinoma, 713 cases of sebaceous adenocarcinoma and others) and 1,777 were dermal (including 1,228 cases of dermatofibrosarcoma, 289 cases of leiomyosarcoma and others).

Incidence

The numbers of rare skin cancers are low, (Table 1), so that risk estimates are subject to error, but this study shows evidence for increasing standardised incidence rates (ASR) of Merkel cell carcinoma and dermatofibrosarcoma over the last ten years (0.1 to 0.2 and 0.2 to 0.3 per 100,000 population respectively) in England. The 10 year age standardised incidence rate for all rare skin cancers varied between English cancer registries (Figure 1) ranging from 0.7 per 100,000 population for the Northern Yorkshire Cancer Registry and Information Service to 1 per 100,000 population for the West Midlands Cancer Intelligence Unit and 0.8 per 100,000 population for England overall, possibly reflecting case ascertainment.

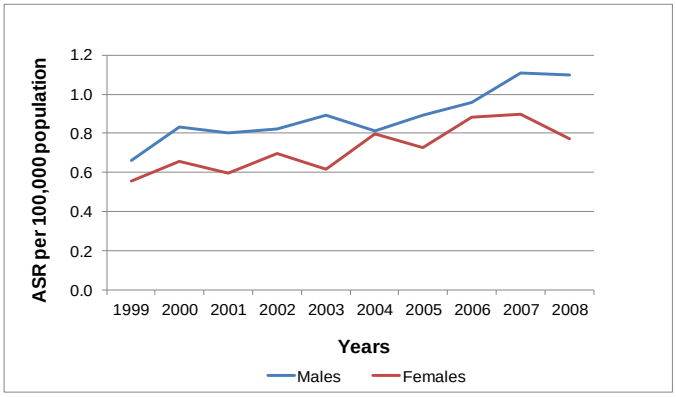
Figure 1: 10 Year Incidence Rate for Rare Skin Cancer across English Cancer Registries (1999-2008)



Trends over the last 10 years of age standardised incidence rates (ASR) for England

Considering the incidence over the last 10 years for England (Figure 2), it is clear that although a slight decrease was observed for females in 2008, the general trend was gradual but upward for both males and females (70% ASR ($p<0.01$) increase for males and 40% ($p<0.01$) for females). The incidence for males was higher than for females. Despite the overall low incidence the trend was similar to malignant melanoma and non melanoma skin cancer.

Figure 2: Incidence of Rare Skin Cancer in England for the last ten years by gender (1999-2008)



Mortality

Outcomes for the rare skin cancers are mixed. Merkel cell carcinoma, which accounts for 29.3% of the rare skin cancers, is known to have a poor prognosis. Using mortality data from NCDR for the 10 year period (1999-2008) 79% of these patients died within 2 years of diagnosis.

Conclusion

Incidence rates appear to be increasing although improvements in diagnosis and/or registration cannot be excluded as a cause. Variation by cancer registries also points to possible variation in case ascertainment. Small numbers require national and international studies to study the epidemiology of these cancers. Some, such as Merkel cell carcinoma have very poor prognosis which is masked in routine analysis when they are grouped with non-melanoma skin cancers. The association between Merkel Cell Cancer and immunosuppression and increased numbers of organ transplants is further justification for improving monitoring of incidence.

FIND OUT MORE:

South West Public Health Observatory is the lead Cancer Registry for Skin Cancer:
http://www.swpho.nhs.uk

Other useful resources within the NCIN partnership:
Cancer Research UK CancerStats – Key facts and detailed statistics for health professionals
http://info.cancerresearchuk.org/cancerstats/

The National Cancer Intelligence Network is a UK-wide initiative, working to drive improvements in standards of cancer care and clinical outcomes by improving and using the information collected about cancer patients for analysis, publication and research. Sitting within the National Cancer Research Institute (NCRI), the NCIN works closely with cancer services in England, Scotland, Wales and Northern Ireland. In England, the NCIN is part of the National Cancer Programme.