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Registration of Cutaneous T-Cell Lymphoma (CTCL) in England

National Cancer Registration and Analysis Services Short Report

Key messages

- the average annual number of registered CTCL cases in England is 332
- men have higher incidence of CTCL than women
- percentage of completed stage was low but appears to be improving

Introduction

Cutaneous T-Cell lymphoma (CTCL) is a rare type of non-Hodgkin lymphoma arising in the skin and represents a heterogeneous group of diseases which can be difficult to classify. It encompasses certain subtypes, the best defined of which are: mycosis fungoides representing around 50% of the CTCL, primary cutaneous CD30+ lymphoproliferative disorder (30%) and Sezary syndrome (3%)[1].

Mycosis fungoides is a disease of slow progression mainly affecting middle aged people but also occurs in younger individuals. The UK incidence rate of CTCL was 0.4 per 100,000 per year in 2003[2]. CTCL cases diagnosed at early stage have a good survival prognosis but diseases such as Sezary syndrome have a poor prognosis. Depending on the stage and diagnosis at presentation, life expectancy can vary from less than five years after diagnosis to full life expectancy[3].

The aims of this short report are to explore the CTCL data available from the National Cancer Registration and Analysis Service (NCRAS) in order to estimate the number of cases diagnosed and to consider the CTCL disease distribution across the strategic clinical networks (SCNs) in England from 2009 to 2013.

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Methodology

New cases of CTCL were identified from the Cancer Analysis System (CAS) using morphology codes (9700/3, 9701/3, 9708/3, 9709/3, 9718/3, 9719/3, 9726/3) based on the World Health Organization-European Organization for the Research and Treatment of Cancer (WHO-EORTC) classification[1]. Data on sex, age, stage and patients' SCN of residence were extracted. Age standardised incidence rates (ASR) were calculated using the 2013 European standard population.

Results

Table 1: Number of Cutaneous T-Cell Lymphoma cases by morphology and ICD10 code in England 2009-2013

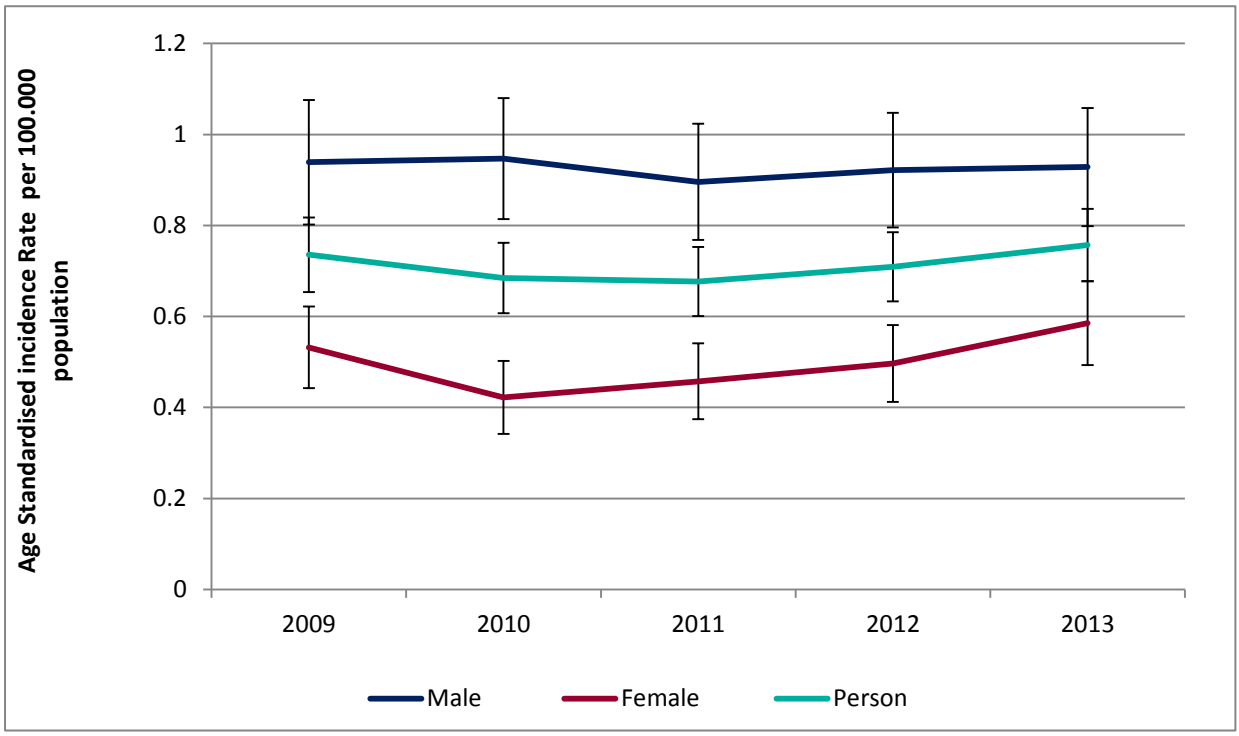
	ICD 10 CODE			
Morphology code and description	**C84**	**C85**	**Unknown**	**Total**
9700/3: Mycosis fungoides	919	1		**920**
9701/3: Sezary syndrome	42			**42**
9708/3: Subcutaneous panniculitis-like T-cell lymphoma	9			**9**
9709/3: Cutaneous T-cell lymphoma (NOS)	465		1	**466**
9718/3: Primary cutaneous CD30+ T-cell lymphoproliferative disorder	160			**160**
9719/3: NK/T-cell lymphoma, nasal and nasal-type	1	59		**60**
9726/3: Primary cutaneous gamma-delta T-cell lymphoma	2			**2**
Total	**1,598**	**60**	**1**	**1,659**

Source: CAS Snapshot 1601

Overall 1,659 CTCL cases were recorded on CAS covering a period of five years (Figure1). The majority of CTCL cases were coded under the ICD 10 Code C84 (peripheral and cutaneous T cell- lymphoma). However, 61 cases were coded under the ICD10 Code C85 (other and unspecified types of non-Hodgkin lymphoma) and one as unknown. 55% (920/1,659) cases were mycosis fungoides, 9.6% (160/1,659) were primary cutaneous CD30+ T-cell lymphoproliferative disorder and 2.5% (42/1,659) were Sezary syndrome. 28% (466/1,659) of cases were recorded as cutaneous T-cell lymphoma (NOS) which might be used as a generic code for CTCL.

It should be noted that morphology codes 9718/3 (primary cutaneous CD30+ T-cell lymphoproliferative disorder) and 9709/3 (cutaneous T-cell lymphoma (NOS)) may include subtypes such as: primary cutaneous anaplastic large cell lymphoma, lymphomatoid papulosis, primary cutaneous CD4 positive small/medium pleomorphic T cell lymphoma and primary cutaneous CD8 positive aggressive epidermotropic cytotoxic T cell lymphoma.

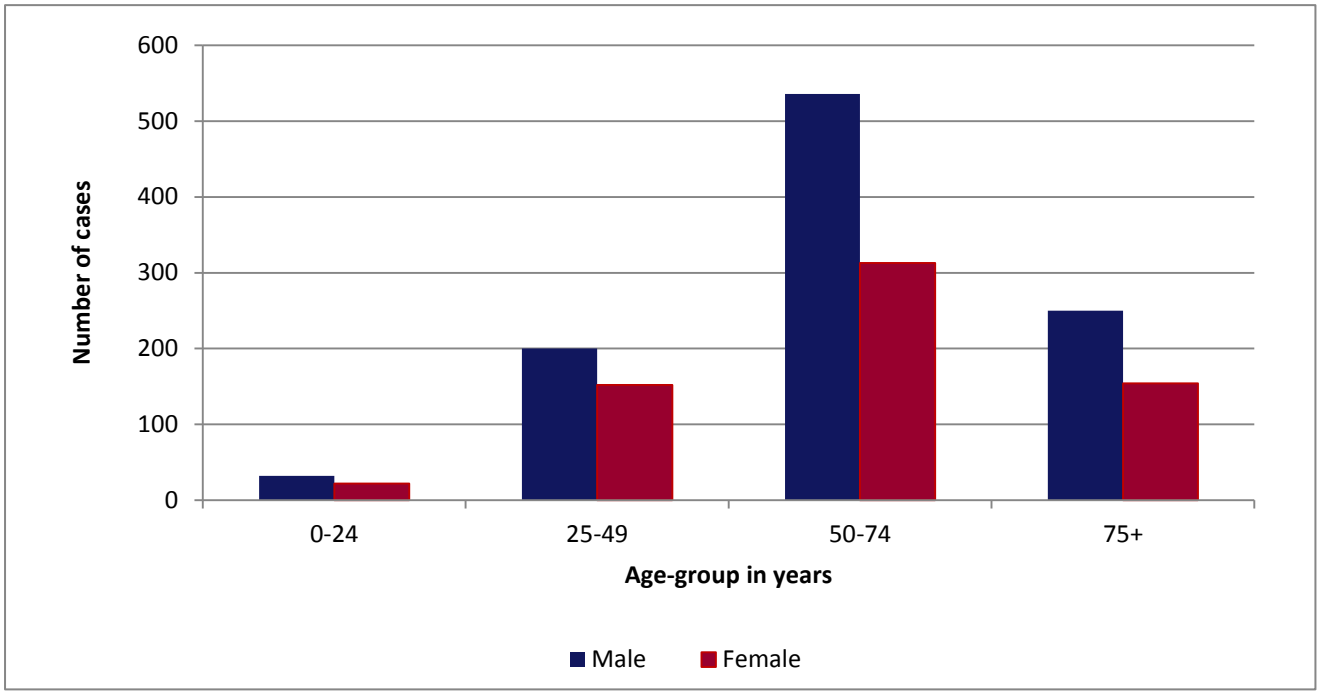
Figure 1: Age-standardised incidence rate per 100,000 population for Cutaneous T Cell lymphoma cases in England 2009- 2013



Source: CAS Snapshot 1601

Overall, for all CTCL cases, the ASR for males (0.93 per 100,000 population in 2013) was significantly higher than for females (0.58 per 100,000 population in 2013). The overall ASR (0.75 per 100,000 in 2013) did not show a significant increase over the five years studied (Figure 1). Considering mycosis fungoides alone, the ASR for males in 2013 was 0.42 per 100,000 population (0.34-0.51) and 0.29 per 100,000 population for females (0.23-0.36).

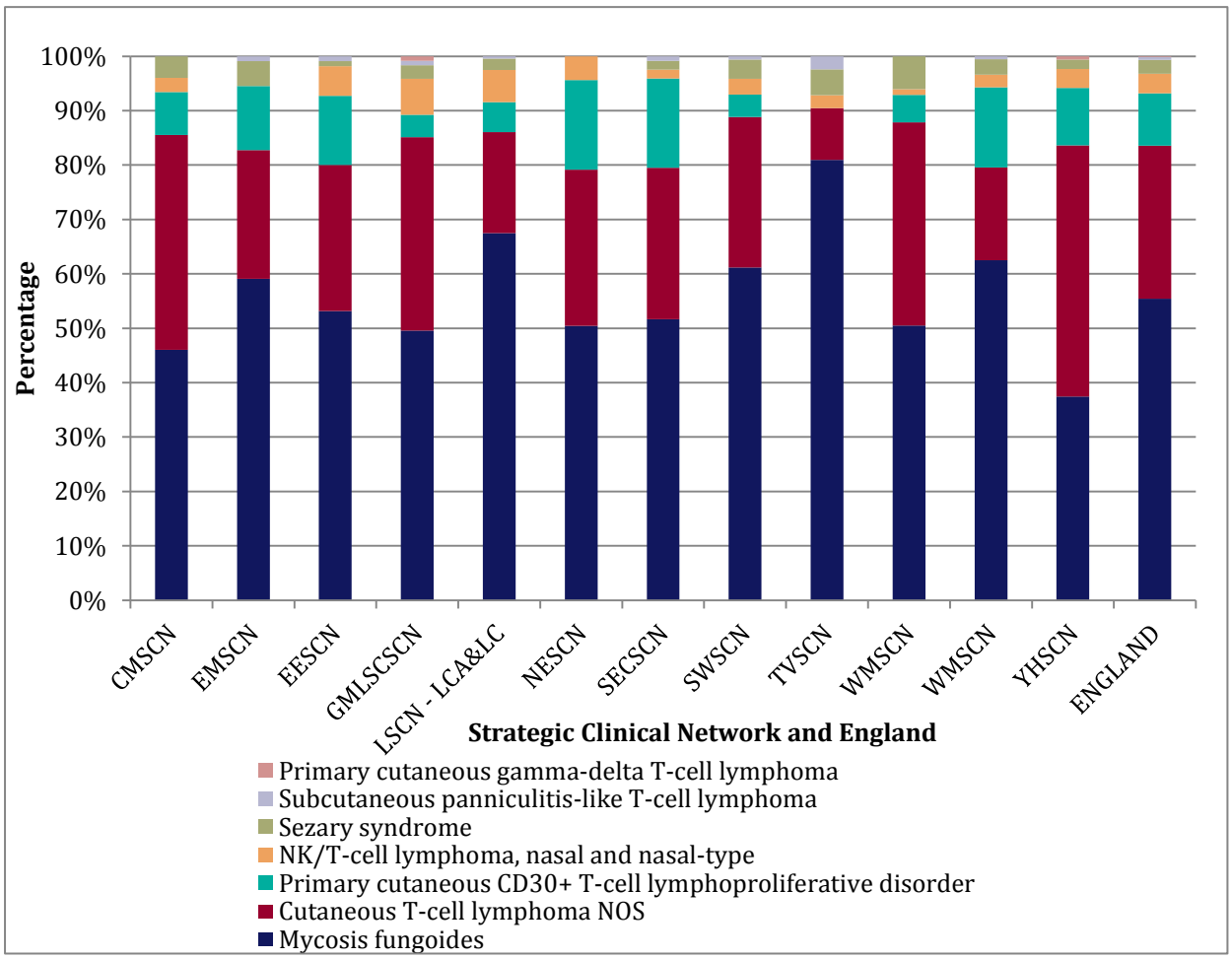
Figure 2: Sex and age-group distribution of the overall cohort of Cutaneous T-Cell lymphoma cases in England 2009- 2013



Source: CAS Snapshot 1601

The majority of CTCL cases were diagnosed in the 50 to 74 age group, with a similar number of cases diagnosed among those aged under 50 and aged 75 and over. The overall male to female ratio was 1.6:1.0. The ratio for mycosis fungoides alone was 1.5:1.0. The age and sex distribution for mycosis fungoides showed a peak in the 50 to 74 age group with a male dominance similar to the overall cohort (Figure 2).

Figure 3: Distribution of Cutaneous T-Cell lymphoma morphology types by English strategic clinical networks and in England: 2009-2013



Source: CAS Snapshot 1601

Notes: Abbreviations for the names of SCNs are as follows:

CMSCN - Cheshire and Mersey, EMSCN - East Midlands, EESCN - East of England, GMLSCSCN - Greater Manchester Lancashire and South Cumbria, LSCN– LCA & LC London Cancer Alliance and London cancer, NESCN- Northern of England, SECSCN - South East Coast, SWSCN - South West, TVSCN - Thames Valley, WMSCN - West Midlands, WSCN – Wessex, YHSCN - Yorkshire and the Humber .

There was a variation in the recorded distribution of the disease sub-types across the SCNs. For example, in the Thames Valley SCN, 81% of cases were mycosis fungoides, in Yorkshire and the Humber SCN it accounted for 37.4% of cases versus a national level of 55% (Figure 3). When considering the number of cases by morphology type over five years there was little variation in the number recorded for each disease, illustrating little local change in disease recording within each SCN, although differing coding practices may be a source of the variation in recorded sub-types.

The percentage of cases with a recorded stage was low over the five year period, but there did appear to be an improvement as the completeness increased from 7% recorded stage in 2009 to 20% in 2013.

Conclusion

The incidence of CTCL in England for the time period examined was higher in males and in the 50 to 70 year old age group of the population, as described in the literature. The ASR did not show a significant increase over the last five years. Whilst genuine regional variation in CTCL sub-type due to geographical and socio-economic factors could not be excluded, it could also reflect variable pathology reporting conventions or different coding practices regarding CTCL morphology codes. The proportion of mycosis fungoides in our data is overall concordant with that reported in prior literature[1,4]. This report provides a foundation for further linkages with other cancer datasets such as the chemotherapy dataset (SACT) to enable investigation of treatment patterns and other predictors of outcomes.

References

1. Willemze R. et al, Blood, 15 May 2005, Vol 105, 10, 3768-3785
2. Whittaker S.J.et al, J British Journal of Dermatology 2003, 149: 1095-1107
3. NICE. Guidance on Cancer Services, Feb 2006, 114-119
4. Kempf W. et al, Histopathology 2010, 56, 57-70

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http://www.ncin.org.uk/publications/reports/

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