

Epidemiology of primary cutaneous lymphomas in Greece: a twelve-year retrospective study



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Introduction

Primary cutaneous lymphomas (PCL) encompass an heterogeneous group of cutaneous T cell (CTCL) and B cell (CBCL) lymphomas. The scarcity of these clinical entities makes the epidemiological study quite challenging. To date, a few studies of PCL have been conducted in the USA, Europe and Asia. We present epidemiological data of PCL in Greece based on a twelve retrospective study.

Material and Methods

Our study is based on histology reports from the major haemopathology referral centre in Athens, Greece from 2009 to 2020. Clinical data including the type of lymphoma, date of first diagnosis, age at diagnosis and gender were collected and processed. The patients’ classification is according to the 2018 update of WHO-EORTC classification for PCL.

Results

In total, 1171 new cases with PCL have been registered from the 1st of January 2009 to the 31st of December 2020, consisting of 715 males and 456 females, with a male/female ratio of 1,6. The vast majority was CTCL accounting for 83,2% (n=975) of PCL, followed by CBCL with 196 new cases comprising 16,7% of PCL. The male:female ratio was 1,8 for CTCL and 0,9 for CBCL. Mycosis fungoides (MF) was the most common cutaneous lymphoma accounting for 82,3% of CTCL and 68,6% of all PCL (n=804). Classic MF (n:553, 69%) was the most frequent subtype among MF patients followed by folliculotropic MF (n:138, 17,2%). One hundred and twelve patients with MF were presented with atypical forms of the disease. A male predominance was observed among MF patients with 526 newly diagnosed MF in men and 277 in women and a ratio male/female of 1,9. In contrast to the literature, a decreased number of primary cutaneous CD30+ lymphoproliferative disorders was noticed comprising 8,4% of PCL and 10% of CTCL. The most frequent CBCL was the primary cutaneous follicle centre lymphoma (pcFCBCL). Ninety five new pcFCBCL cases were recorded accounting for 48,4% of CBCL and 8,1% of PCL. The marginal zone lymphoma (pcMZBCL) was the second most common CBCL with 86 new cases representing 43,8% and 7,3% of CBCL and PCL, respectively.

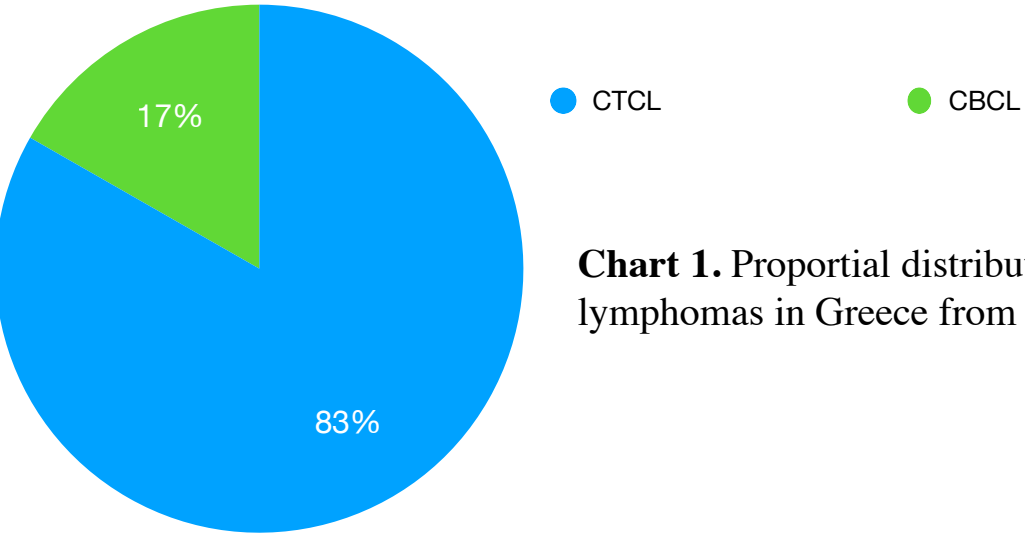


Chart 1. Proportial distribution of new cases of primary cutaneous lymphomas in Greece from 2009 to 2020

Results

PCL type	N	Frequency (%)	Mean Age at diagnosis (SD)
CTCL	975	83,2	
MF, variants included	804	68,7	60,88 (17,27)
SS	16	1,4	66,07 (8,5)
C-ALCL	43	3,7	60,42 (17,63)
LyP	55	4,7	52,60 (16,44)
Adult T cell leukemia/lymphoma	0	0	
Subcutaneous panniculitis-like T cell lymphoma	1	0,1	72
Extranodal NK/T cell lymphoma, nasal type	0	0	
Chronic active EBV infection	0	0	
Primary cutaneous gamma-delta T-cell lymphoma	10	0,9	
CD8+ AECTCL	7	0,6	49,38 (18,51)
Primary cutaneous CD4+ small/medium T-cell LPD	28	2,4	66,45 (15,56)
Primary cutaneous acral CD8+ T-cell	0	0	
Primary cutaneous peripheral T-cell lymphoma, NOS	11	0,9	59,67 (16,24)
CBCL	196	16,7	
PCMZBCL	86	7,3	57,37(18,73)
PCFCBCL	95	8,1	57,45 (15,48)
PCDLBCL, LT	13	1,1	78,23 (9,6)
EBV+ mucocutaneous ulcer	2	0,2	75
Total no.	1171	100	

Table 1. Demographic data of 1171 new PCL cases (SS, Sezary syndrome; C-ALCL, Cutaneous anaplastic large cell lymphoma; LyP, Lymphomatoid papulosis; CD8+ AETCTCL, CD8+ Aggressive epidermotropic cytotoxic T cell lymphoma)

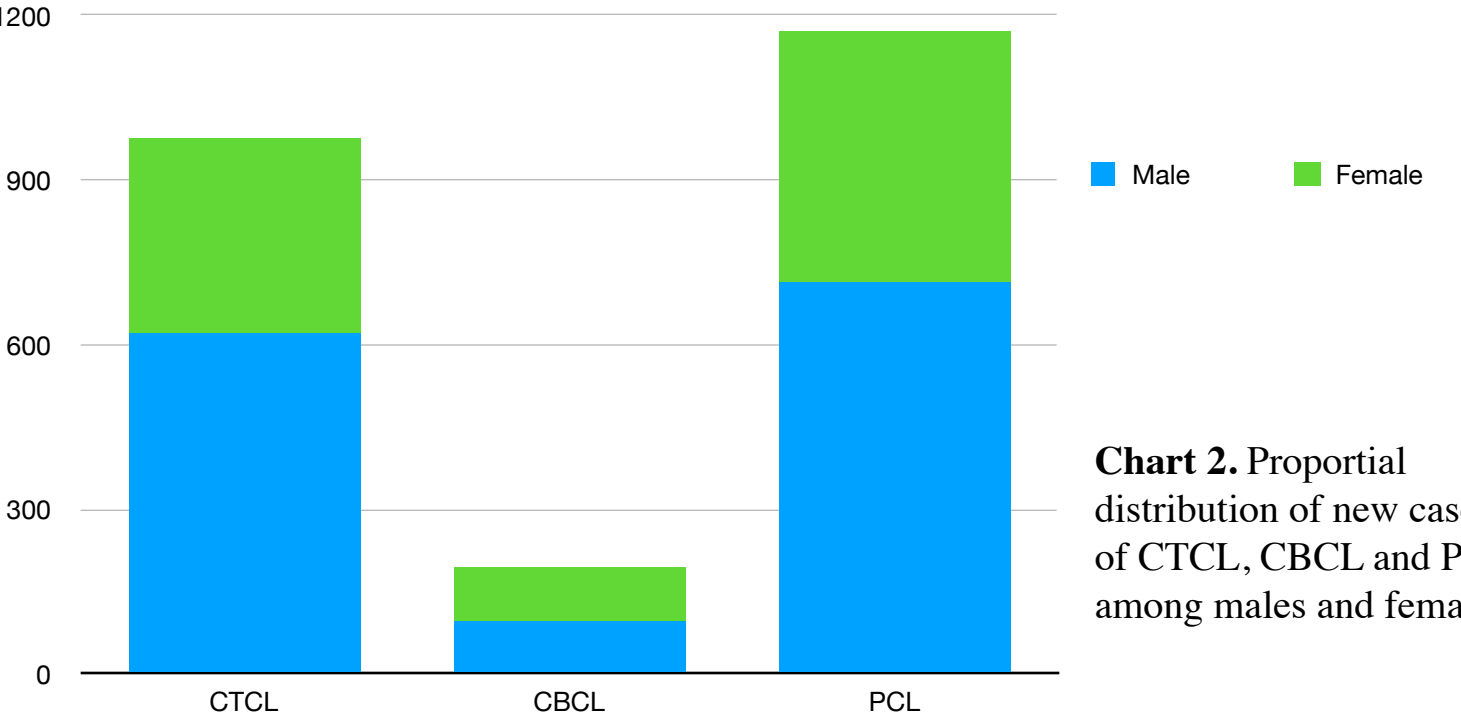


Chart 2. Proportial distribution of new cases of CTCL, CBCL and PCL among males and females

Results

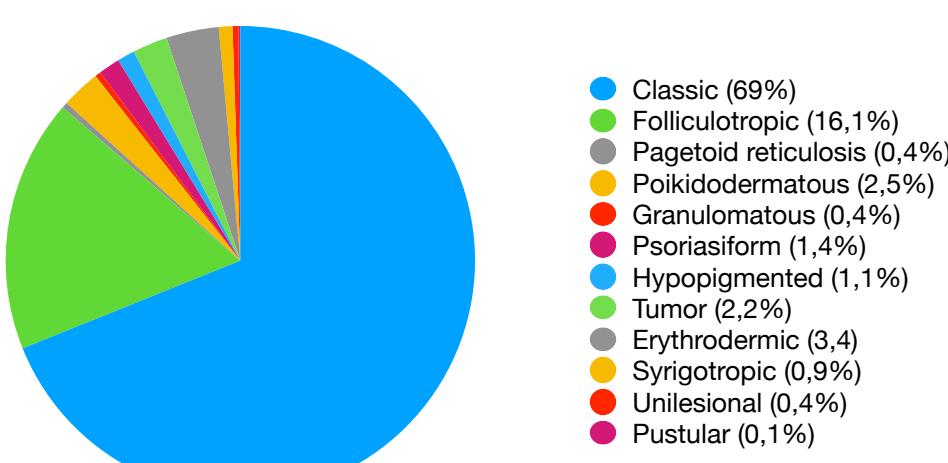


Chart 3. Proportial distribution of MF variants.

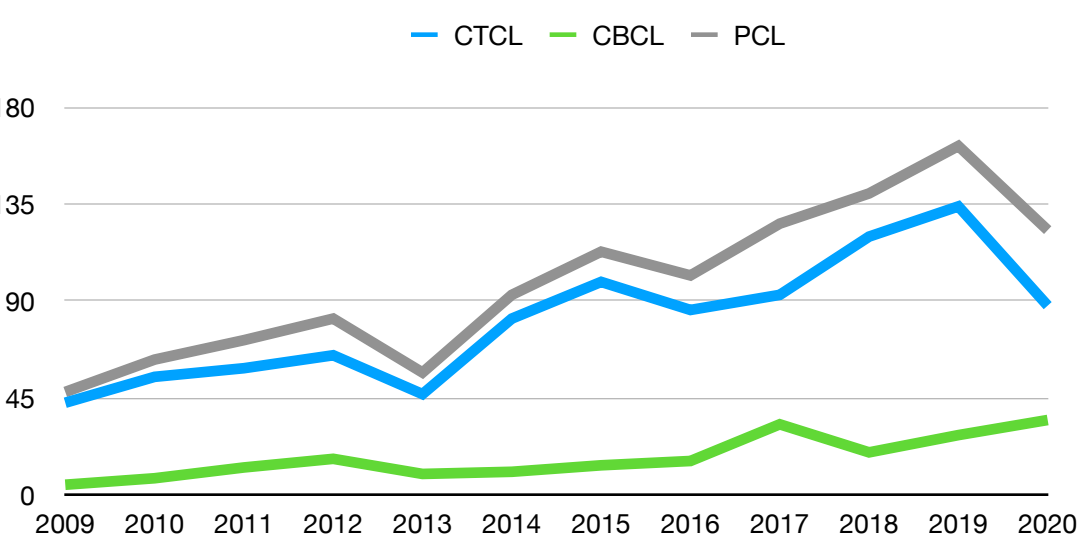


Figure 1. Temporal trends of primary cutaneous lymphomas in Greece from 01st January 2009 to 31st December 2020.

Discussion

In the present study, an increasing trend for PCL was observed through the years. This increase may reflect the improvements in the detection and classification of these lymphomas in conjunction with an increase of the underlying causative agents. It is noteworthy that in 2020, during the COVID-19 era, there was a downward trend in new PCL diagnoses, mainly those of MF, whilst the diagnoses of CBCL were increased. While the COVID-19 pandemic may be held responsible for this, other reasons may be implicated and it needs further monitoring over the next few years. This report will empower the comparison with epidemiological data of PCL from other countries and the observation of the diagnostic trend contributing to the investigation of pathogenesis of the disease and novel therapies in the near future.

REFERENCES

- Willenze R, Cerroni L, Kempf W, et al. The 2018 update of the WHO-EORTC classificaAon for primary cutaneous lymphomas. Blood. 2019 Apr 18;133(16):1703-1714.
- Bradford PT, Devesa SS, Anderson WF, Toro JR. Cutaneous lymphoma incidence paierns in the United States: a populaAon-based study of 3884 cases. Blood. 2009 May 21;113(21):5064-73.
- Richards, Mike et al. "The impact of the COVID-19 pandemic on cancer care." Nature cancer, 1-3. 20 May.