AGMT_PTCL REGISTRY

Austrian Registry and Biobank of Peripheral T-cell Lymphomas

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Protocol Synopsis

Indication: Peripheral T-cell lymphomas (PTCL)

Design: Prospective as well as retrospective, observational, multicentre research initiative

Objectives:
- Epidemiology of this rare disease in Austria
- Information on type of therapy
- Response
- Identification of potential prognostic and predictive factors

Rationale

Peripheral T-cell lymphomas (PTCL) comprise a heterogeneous group of hematological neoplasms originating from post-thymic T-cells at different stages of differentiation. Due to several reasons clinical management of PTCL probably represents one of the most challenging tasks in hematologic oncology:

- Patients often present at higher stages and with reduced performance status
- PTCL do not respond well to chemotherapy
- PTCL are rare

The lack of knowledge of the epidemiology and biology of this rare disease and above all, the urgent clinical need for improved therapies for PTCL are the primary motivations for this registry and biobank.

Design

This registry is a prospective as well as retrospective, observational, multicentre research initiative. Data will be collected from all sites in Austria willing to participate.

Within the registry biomaterial should be collected in the AGMT biobank. Given the low incidence of PTCL, only the establishment of a substantial biobank can lay the foundations for scientifically meaningful and internationally competitive translational research.

Inclusion Criteria (selected)

- Diagnosis of peripheral T-cell lymphomas (PTCL) according to WHO classification 2008
- Written informed consent (deceased patients may also be included without written consent)
- Age > 12 years. For inclusion of patients between >12 years and <18 years of age, additional written informed consent has to be obtained by the patient's parent(s) or guardian.

Recruitment (as of March 2019)

No	Site	Patients
01	PMU Salzburg / Innere Medizin III	87
02	UK Innsbruck / Innere Medizin V	58
03	Klinikum Wels Grieskirchen / Innere Medizin IV	9
04	LKH Feldkirch / Innere Medizin II - Interne E	18
05	LKH Steyr / Innere Medizin II	1
06	Kaiser-Franz-Josef Spital Wien / 3. Medizin	5
07	Ordensklinikum Linz/ Interne I	6
08	Kepler UK, Med Campus III./ UK Hämatologie	1
09	UK Graz / Innere Medizin - Hämatologie	0
10	AKH MUW / Innere Medizin I - Hämatologie	115
11	LKH-Hochsteiermark / Dep. Hämato- Onkologie	31
	TOTAL	**331**

Results (as of March 2019)

Epidemiologic data review (before start of first line therapy)	
Median age at first diagnosis, years (range, IQR) n=321	62 (16-92, 45-73
Sex n=331	
Men	181 (55%)
Women	150 (45%)
Diagnosis n=326	
Peripheral T-cell lymphoma (PTCL), NOS	119 (37%)
Angioimmunoblastic T-cell lymphoma (AITL)	40 (12%)
Anaplastic large cell lymphoma (ALCL), ALK-positive	34 (10%)
Anaplastic large cell lymphoma (ALCL), ALK-negative	32 (10%)
Enteropathy-associated T-cell lymphoma (EATL)	8 (3%)
T-cell prolymphocytic leukemia (T-PLL)	8 (3%)
Other	68 (20%)
Initial disease presentation n=266	
Nodal	203 (76%)
Leukemic	15 (6%)
Cutaneous	38 (14%)
More than one	10 (4%)
Stage n=267	
Stage I, stage II	83 (31%)
Stage III, stage IV	184 (69%)
Extra nodal disease presentation n=238	121 (51%)
Bone marrow involved n=212	70 (33%)
ECOG performance status n=91	
0, 1	73 (80%)
2, 3, 4	18 (20%)
CD30 positive n=173	122 (71%)

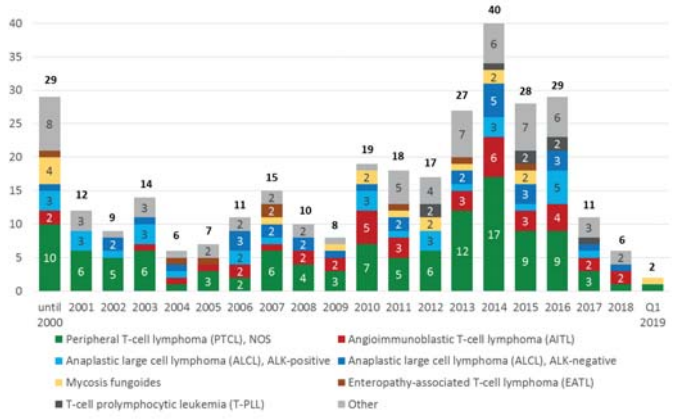

Figure 1: Documented subtypes per year of first diagnosis; subtypes with ≥ 3% in total are listed, less frequent are grouped in others

Induction therapy is actually documented for 320 patients: 138 patients received only 1st line therapy so far, 82 patients had maximum 2 lines of therapy, 100 patients underwent 3 or more therapy lines. The median number of lines per patient was 2 (range 1-14, IQR 1-3).
Most common first line induction therapy was CHOP or CHOP-like therapy (195 of 320 patients). In 2nd line treatment with DHAP or DHAP-like therapy regimen was most common (32 of 182 patients) followed by Brentuximab vedotin (24 of 62 patients). Treatment within a clinical trial was performed in only 11 out of 212 patients in the 1st line setting and 6 out of 117 patients in the 2nd line setting.

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An academic registry
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Clinical-Scientific Director: R. Greil