Hepatosplenik T cell Lenfoma

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Hepatosplenik T hücreli lenfoma

- Lenfomalar; tüm kanserlerin %3'ü,
- Lenfomaların ~%75'ini non-Hodgkin (NHL), %25'ini Hodgkin lenfoma oluşturmaktadır*, ülkemizde % 80'e, % 20.
- Periferik T hücre lenfomaları NHL'ların %15'inden daha az
- Hepatosplenik T hücre lenfoma (HSTCL) ilk kez 1990 yılında tanımlanmış ve insidansının periferik T hücreli lenfoma tüm alt tiplerinin% 1'inden az olduğu belirtilmiştir*
- ABD'de HSTCL'ların T hücre lenfomaları içinde oranı ise % 3**

* Nat Rev Clin Oncol 6:707-717, 2009
** Clin Lymphoma Myeloma Leuk. 2013;13(4):360–369.

Table 1. Major Lymphoma Subtypes by Geographic Region

Subtype	%		
	North America	Europe	Asia
PTCL-NOS	34.4	34.3	22.4
Angioimmunoblastic	16.0	28.7	17.9
ALCL, ALK positive	16.0	6.4	3.2
ALCL, ALK negative	7.8	9.4	2.6
NKTCL	5.1	4.3	22.4
ATLL	2.0	1.0	25.0
Enteropathy-type	5.8	9.1	1.9
Hepatosplenic	3.0	2.3	0.2
Primary cutaneous ALCL	5.4	0.8	0.7
Subcutaneous panniculitis-like	1.3	0.5	1.3
Unclassifiable T-cell	2.3	3.3	2.4

Abbreviations: PTCL, peripheral T-cell lymphoma; NOS, not otherwise specified; ALCL, anaplastic large-cell lymphoma; NKTCL, natural killer/T-cell lymphoma.

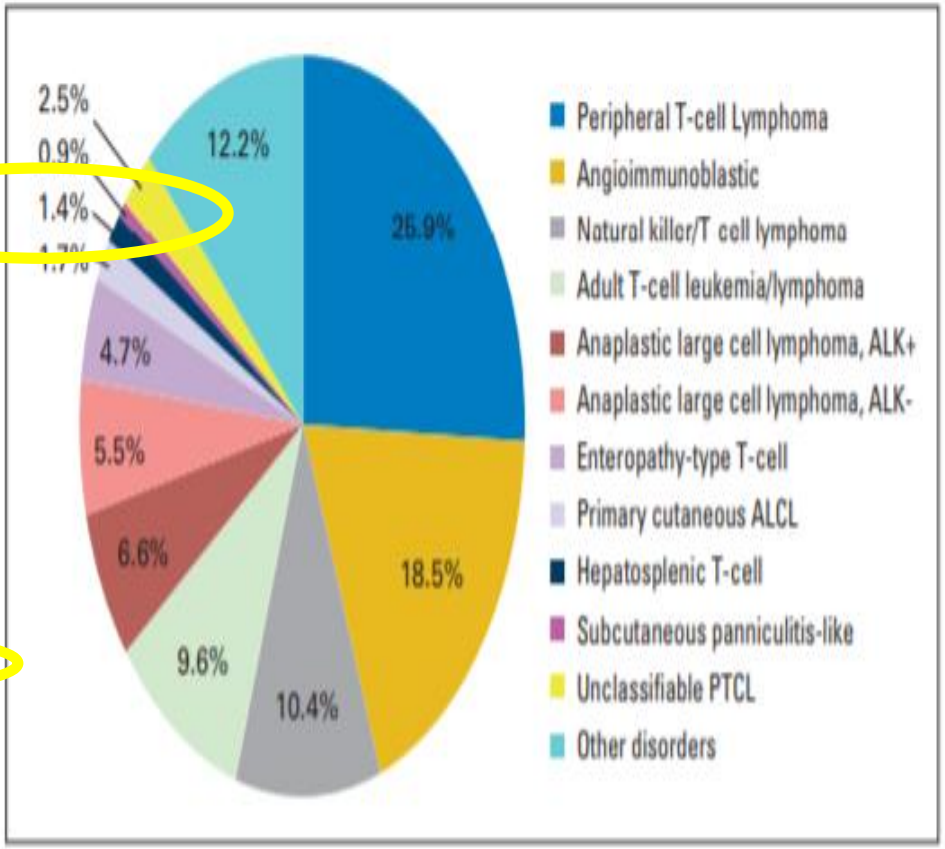

Fig 1. Distribution of 1,314 cases by consensus diagnosis. NOS, not otherwise specified; ALCL, anaplastic large-cell lymphoma; PTCL, peripheral T-cell lymphoma.

Vose J[1], Armitage J, Weisenburger D; International T-Cell Lymphoma Project.J Clin Oncol. 2008 Sep 1;26(25):4124-30.

Lenfoma Sınıflamalarının Tarihi

LENFOMA

Gall-Mallory	(1940)	Morfoloji
Rappaport	(1960)	
B.N.L.I	(1973)	
Lukes-Collins	(1974)	
Lennert	(1974)	İmmunfenotip
WF	(1980)	Morfoloji
REAL	(1994)	Klinikopatolojik antite
WHO	(2001)	
WHO	(2008)	
WHO	(2017)	

The International Lymphoma Study Group

Nancy Harris - Boston
Elaine Jaffe - Bethesda
Harald Stein - Berlin
Peter Banks - San Antonio
John Chan - Hong Kong
Michael Cleary - Stanford
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Chris De Wolf-Peeters - Leuven
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Kevin Gatter - Oxford
Thomas Grogan - Tucson
Peter Isaacson - London
Daniel Knowles - Cornell
David Mason - Oxford
Konrad Müller-Hermelink - Würzburg
Stefano Pileri - Bologna
Miguel Piris - Toledo
Elizabeth Ralfkiaer - Copenhagen
Roger Warnke - Stanford

PERSPECTIVE

A Revised European-American Classification of Lymphoid Neoplasms: A Proposal From the International Lymphoma Study Group

By Nancy Lee Harris, Elaine S. Jaffe, Harald Stein, Peter M. Banks, John K.C. Chan, Michael L. Cleary, Georges Delsol, Christine De Wolf-Peeters, Brunangelo Falini, Kevin C. Gatter, Thomas M. Grogan, Peter G. Isaacson, Daniel M. Knowles, David Y. Mason, Hans-Konrad Muller-Hermelink, Stefano A. Pileri, Miguel A. Piris, Elisabeth Ralfkiaer, and Roger A. Warnke

Lenfosit biyolojisi ve lenfomagenez moleküler mekanizmalar ve sonuçları

REAL Classification (1994)	→ Change	WHO Classification (2001)
Precursor cell lymphoma		***Precursor cell lymphoma***
Lymphoblastic lymphoma, T cell B cell		Lymphoblastic lymphoma, T cell B cell
Peripheral B-cell neoplasms		***Peripheral B-cell neoplasms***
B-chronic lymphocytic leukemia/small lymphocytic lymphoma/prolymphocytic leukemia	→	B-chronic lymphocytic leukemia/small lymphocytic lymphoma B-prolymphocytic leukemia
Lymphoplasmacytoid lymphoma	→	Lymphoplasmacytic lymphoma
Mantle cell lymphoma		Mantle cell lymphoma
Follicular center cell lymphoma	→	Follicular lymphoma
Marginal zone B-cell lymphoma – Extranodal – Nodal (*Provisional entity*)	→	Extranodal marginal zone B-cell lymphoma of MALT type Nodal marginal zone B-cell lymphoma
Splenic marginal zone B-cell lymphoma (*Provisional entity*)	→	Splenic marginal zone B-cell lymphoma
Hairy cell leukemia		Hairy cell leukemia
Diffuse large B-cell lymphoma		Diffuse large B-cell lymphoma
Burkitt lymphoma High grade B-cell lymphoma, Burkitt-like (*Provisional entity*)	→	Burkitt lymphoma (including Burkitt-like lymphoma)
Plasmacytoma/plasma cell myeloma		Plasmacytoma/plasma cell myeloma
Peripheral T and NK cell neoplasms		***Peripheral T and NK cell neoplasms***
T-cell chronic lymphocytic leukemia/ prolymphocytic leukemia	→	T-prolymphocytic leukemia
Large granular lymphocyte leukemia (T or NK cell)	→	T-cell granular lymphocytic leukemia Aggressive NK cell leukemia
Mycosis fungoides/Sezary syndrome		Mycosis fungoides/ Sezary syndrome
Peripheral T-cell lymphoma unspecified		Peripheral T-cell lymphoma, not otherwise characterized
Angioimmunoblastic T-cell lymphoma		Angioimmunoblastic T-cell lymphoma
Angiocentric lymphoma	→	Extranodal NK/T cell lymphoma, nasal and nasal-type
Intestinal T-cell lymphoma	→	Enteropathy-type T-cell lymphoma
Hepatosplenic $\gamma\delta$ T-cell lymphoma (*Provisional entity*)	→	Hepatosplenic $\gamma\delta$ T-cell lymphoma
Subcutaneous panniculitis-like T-cell lymphoma (*Provisional entity*)	→	Subcutaneous panniculitis-like T-cell lymphoma
Anaplastic large cell lymphoma, T/null cell	→	Anaplastic large cell lymphoma, T/null cell, primary systemic type Anaplastic large cell lymphoma, T/null cell, primary cutaneous type
Adult T-cell lymphoma/leukemia (HTLV1+)		Adult T-cell lymphoma/leukemia (HTLV1+)

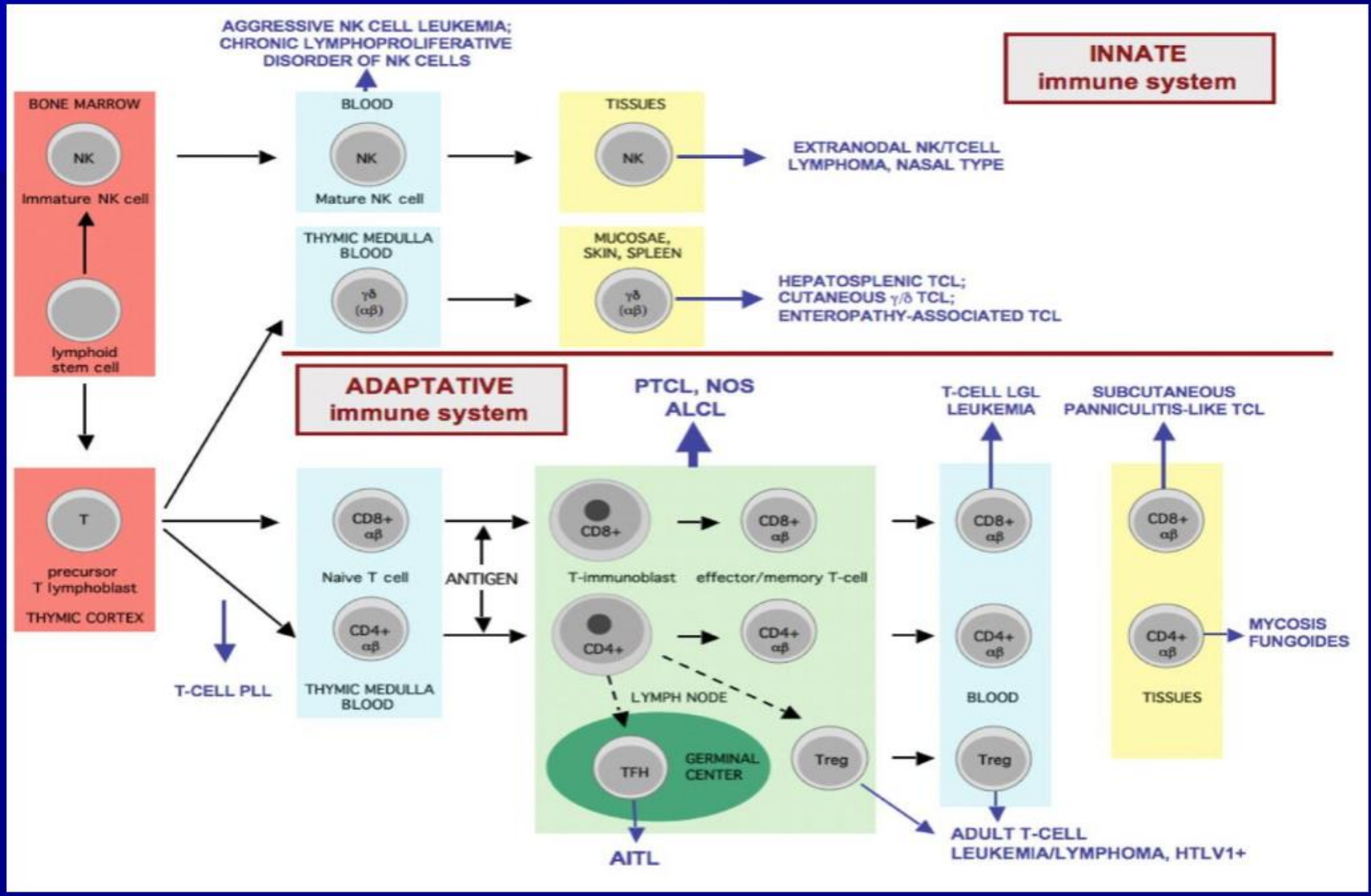
*Anaplastic large cell lymphoma, Hodgkin-like, considered a provisional entity in the REAL classification, has been deleted from the new WHO classification.

TABLE 3 Mature T and NK-cell neoplasms within the 2008 and revised 2016 WHO classification

2008 WHO classification	2016 revision	Comments
T-cell prolymphocytic leukemia	T-cell prolymphocytic leukemia	- no major changes
T-cell large granular lymphocytic leukemias	T-cell large granular lymphocytic leukemias (T-LGL)	- *STAT3* and *STAT5B* mutations in a subset of cases
	Chronic lymphoproliferative disorder (LPD) of NK cells[a]	- new provisional entity - NK-cell counterpart of T-LGL
Aggressive NK-cell leukemia	Aggressive NK-cell leukemia	- no major changes
Systemic EBV$^+$ T cell lymphoproliferative disorder (LPD) of childhood	Systemic EBV$^+$ T cell lymphoma of childhood*	- change in nomenclature due to the fulminant clinical course and monoclonal proliferation - Hemophagocytic syndrome usually present
Hydroa vacciniforme-like lymphoma	Chronic active EBV infection[a] - systemic form - cutaneous form - Hydroa-vacciniforme-like LPD[a] - severe mosquito bite allergy[a]	- new category to encompass the EBV$^+$ T and NK-cell LPD in the pediatric age - can be monoclonal - change in nomenclature. Umbrella term that covers the entire spectrum of the disease - new subgroup.
Adult T-cell leukemia/lymphoma	Adult T-cell leukemia/lymphoma	- no major changes
Extranodal NK/T-cell lymphoma, nasal type	Extranodal NK/T-cell lymphoma, nasal type	- no major changes
Enteropathy-associated T-cell lymphoma, type I	Enteropathy-associated T-cell lymphoma	- no major changes
Enteropathy-associated T-cell lymphoma type II	Monomorphic epitheliotropic intestinal T-cell lymphoma[a]	- change in nomenclature - lack association with celiac disease - mostly derived from γδ T cells - *STAT5B* mutations in 36% the cases - recurrent *SETD2* alterations
	Indolent T-cell LPD of the GI tract[a]	- new provisional entity
Hepatosplenic T-cell lymphoma	Hepatosplenic T-cell lymphoma	- no major changes
Subcutaneous panniculitis-like T-cell lymphoma	Subcutaneous panniculitis-like T-cell lymphoma	- no major changes
Mycosis fungoides	Mycosis fungoides	- no major changes
Sezary syndrome	Sezary syndrome	- no major changes
Primary cutaneous CD30+ LPD - Lymphomatoid papulosis (LyP) - anaplastic large cell lymphoma	Primary cutaneous CD30+ LPD - Lymphomatoid papulosis (LyP) - anaplastic large cell lymphoma	- new morphological subtypes
Primary cutaneous γδ T-cell lymphoma	Primary cutaneous γδ T-cell lymphoma	- needs to be distinguished from other γδ T-cell cutaneous disorders mainly LyP
Primary cutaneous CD8+ aggressive epdermotropic cytotoxic T-cell lymphoma	*Primary cutaneous CD8+ aggressive epdermotropic cytotoxic T-cell lymphoma*	- no major changes - differential diagnosis with LyP type D
	Primary cutaneous acral CD8$^+$ T-cell lymphoma[a]	- new provisional entity
Primary cutaneous CD4+ small/medium T-cell lymphoma	*Primary cutaneous CD4+ small/medium T-cell LPD*[a]	- change in nomenclature. Indolent disorder indistinguishable from clonal drug reactions
Peripheral T-cell lymphoma, NOS	Peripheral T-cell lymphoma, NOS	- molecular subgroups recognized
Angioimmunoblastic T-cell lymphoma	Angioimmunoblastic T-cell lymphoma - *Follicular T-cell lymphoma*[a] - *PTCL with TFH phenotype*[a]	- frequent *TET2*, *RHOA* and *IDH2* mutations - new subgroups with THF phenotype

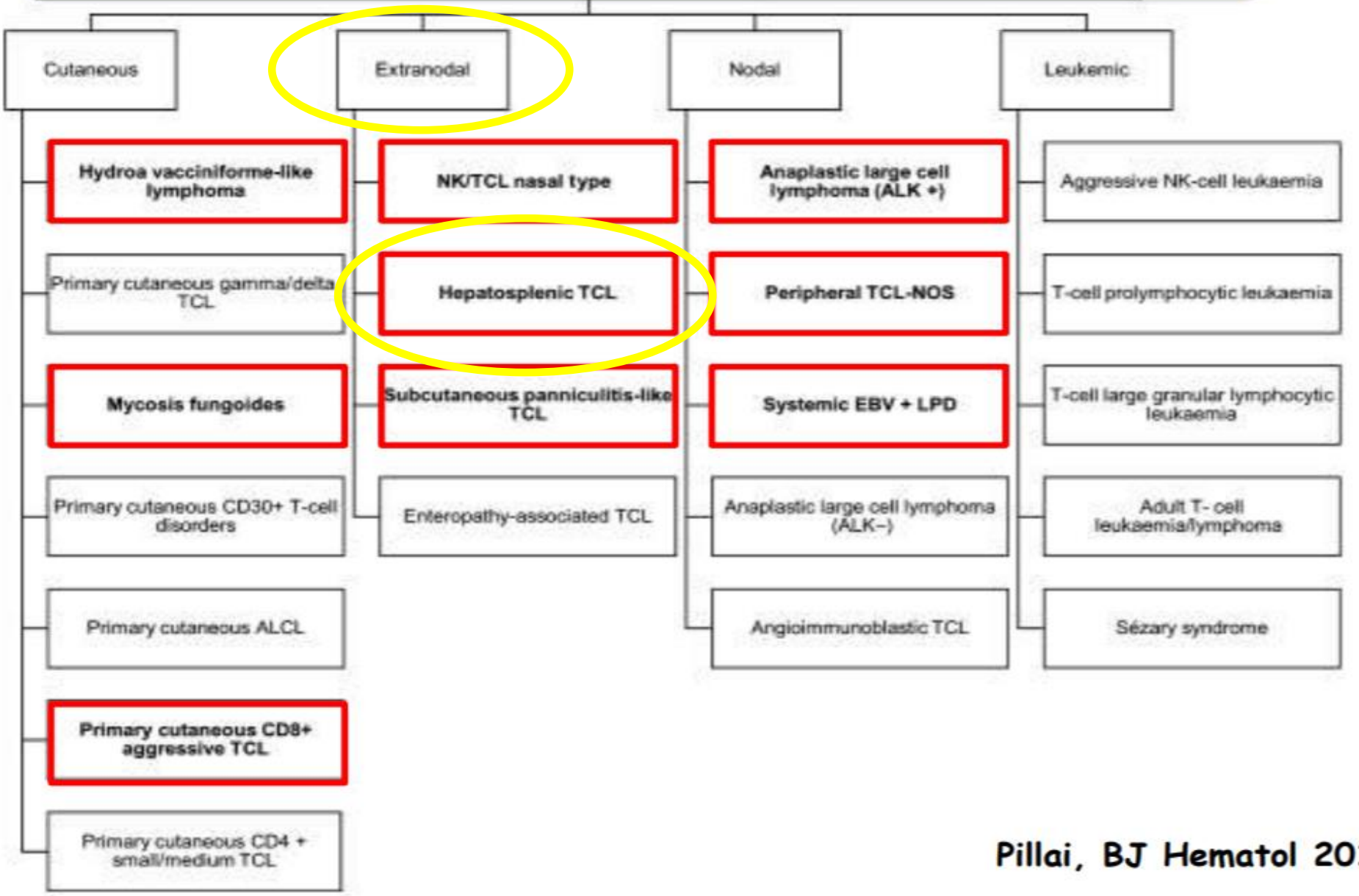
(Continues)

Overview of T-cell and NK-cell differentiation and the putative histogenetic derivation of the major subtypes of mature T- and NK-cell neoplasms



Hematology 2008;2008:491-506

Olgun T hücreli lenfomalar



Pillai, BJ Hematol 2016

Tanım ve epidemiyoloji;

- Hepatosplenik T-hücreli lenfoma, dalak, karaciğer ve kemik iliği tutulumu karakterize agresif klinik seyirli,
- Sistemik, ekstranodal periferik T hücreli bir lenfomadır.
- Bulky lenfadenopati nadir görülür.
- Ortanca yaşı 35 olan genç erkekleri etkiler.
- Vakaların % 20'si immün düzensizlik veya immün baskılanma sonucunda ortaya çıkar.

Hepatosplenik T hücreli lenfoma

İmmunosupresyonun İncelenmesi

- Parakkal ve ark. tarafından yapılan bir çalışmada immünosupresif tedavi alan hastalar arasında 25 HSTCL vakası tespit edildi.
- 22 hastada (%88) İBH ve 3 hastada RA vardı.
- Dört hasta (%16) kadındı ve dört hasta 65 yaşın üzerindeydi.
- Yirmi dört hasta (yüzde 96) ayrıca bir immünomodülatör (azatiyoprin, 6-merkaptopürin veya metotreksat) aldı.
- İki hasta yalnız adalimumab aldı.

herein review all cases of HSTCL reported to the Food and Drug Administration (FDA) in patients receiving TNF-α inhibitors.

METHODS: Individual reports from the FDA Adverse Event Reporting System database for lymphomas from the biological agents - infli adalimumab, certolizumab, natalizumab, and etanercept were downloaded and analyzed with Microsoft Access. Full reports for all ider HSTCL cases were obtained from the FDA.

RESULTS: Twenty-five cases of HSTCL were identified. Twenty-two (88%) patients had inflammatory bowel disease and three had rheumatoid arthritis. Four cases (16%) were in women and four patients were above 65 years of age. Twenty-four cases (96%) also re an immunomodulator (azathioprine, 6-mercaptopurine, or methotrexate). Two patients received adalimumab alone.

CONCLUSION: HSTCL is no longer restricted to the previously identified risk group of young male patients, but can also occur in patie rheumatoid arthritis, females and older adults receiving TNF-α inhibitors and immunomodulators. Improved disease outcomes using combination therapy should be tempered by the risk of developing HSTCL.

İmmunosupresyonun İncelenmesi

Am J Gastroenterol. 2013 Jan;108(1):99-105. doi: 10.1038/ajg.2012.334. Epub 2012 Oct 2.

T-cell non-Hodgkin's lymphomas reported to the FDA AERS with tumor necrosis factor alpha

- Deepak ve ark. FDA sisteminden (2003-2010) toplam 3.130.267 rapor indirildi. FDA AERS'de TNF-a inhibitörleri alan 91 T hücreli NHL vakası tanımlandı 38 RA, 36 Crohn,11 psöriyazis, 9 ÜK ve 6 AS
- Olguların 68'i hem TNF-a inhibitörü hem de bir immünomodülatör (azatiyoprin, 6-merkaptopürin, metotreksat, leflunomid veya siklosporin) kullanıyordu.
- Hepatosplenik T hücreli lenfoma (HSTCL) en sık bildirilen alt tipti, mikoz fungoidleri / Sezary sendromu ve HSTCL, TNF-a-inhibitör maruziyetinde daha yaygın olarak tanımlandı.

fungoides/Sezary syndrome and HSTCL were identified as more common with TNF-α inhibitor exposure compared with SEER-17 registry. Nineteen cases of T-cell NHL with thiopurines were identified in the FDA AERS and one additional case on MEDLINE. Reported risk of T-cell NHL was higher with TNF-α inhibitor use in combination with thiopurines (95% confidence interval (CI) 4.98-354.09; P<0.0001) and thiopurines alone (95% CI 8.32-945.38; P<0.0001) but not with TNF-α inhibitor use alone (95% CI 0.13-10.61; P=1.00).

CONCLUSIONS: Risk of T-cell NHL is increased with TNF-α inhibitor use in combination with thiopurines but not with TNF-α inhibitors alone.

Klinik ve fizik muayene;

- Hastalar sıklıkla ***sistemik semptomlar(ateş, halsizlik), hepatosplenomegali***, ***sitopeniler*** ve bazen de hemofagositik lenfohistiyositoz (HLH) ile kendini gösterir.
- B semptomları % 80 hastada bulunur.
- Klinik seyir ilerleyici olup bir yıllık medyan sağkalım
- Erken tanı ve uygun tedavi ile potansiyel daha iyi sonuçlar için çok fazla belirsizlik vardır.
- Sadece kemoterapiyle (antrasiklin bazlı) sonuçlar zayıf
- Otolog veya allojenik transplantasyon, klinik çalışmalara hasta alımı ile birlikte düşünülmelidir.

Hepatosplenik T hücreli lenfoma

Tanı;

- Kemik iliği ve / veya karaciğer biyopsisi veya splenektomi ya da dalak biyopsisine histolojik olarak bakılır.
- Eksizyonel veya insizyonel biyopsi gerekli
- Kemik iliği histolojisinde neoplastik T hücrelerinin tanımlanması zor olabilir ve tanı için immünohistokimya !!!
- Ayırıcı tanı için, akış sitometrisi, *IGHV* ve *TCR* gen yeniden düzenlemeleri için PCR ,karyotip ve FISH.

Hepatosplenik T hücreli lenfoma

National Comprehensive Cancer Network®

NCCN Guidelines Version 2.2019
Hepatosplenic Gamma-Delta T-Cell Lymphoma

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DIAGNOSIS[a,b]

ESSENTIAL:

- Review of all slides with at least one paraffin block representative of the tumor should be done by a hematopathologist with expertise in the diagnosis of T-cell lymphomas. Rebiopsy if consult material is nondiagnostic.
- A core or incisional biopsy of bone marrow, liver, or spleen is required for diagnosis. Bone marrow aspirate, FNA of liver or spleen, or peripheral blood studies may be helpful but are not alone sufficient for diagnosis.
- Adequate immunophenotyping to establish diagnosis[c,d]
  - IHC panel may include CD20, CD3, CD10, Ki-67, CD5, CD30, CD2, CD4, CD8, CD7, CD56, EBER-ISH, TCRβ, TCRδ, TIA-1, or granzyme B
  with
  - Cell surface marker analysis by flow cytometry may include kappa/lambda, CD45, CD3, CD5, CD19, CD10, CD20, CD30, CD4, CD8,CD7, CD2; TCRα/β, or TCRγ/δ

USEFUL UNDER CERTAIN CIRCUMSTANCES:

- Molecular analysis to detect[c] clonal *TCR* gene rearrangements or other assessment of clonality (karyotype, array-CGH, or FISH analysis to detect somatic mutations or genetic alterations)[e]
- Karyotype to establish clonality and investigate the presence of isochromosome 7q and trisomy 8.
- FISH analysis for isochromosome 7q and trisomy 8.
- Genomic analysis for *STAT3*, *STAT5B*, *PIK3CD*, *SETD2*, *INO80*, *TET3*, and *SMARCA2*.

Uzman hematopatolog

Kemik iliği/dalak/ karaciğer biyopsisi

İmmunhistokimyasal panel

(HGDTL-3)

Flow sitometri

TCR gen rearanjmanı

Karyotip ve FISH: 7q ve trizomi 8

Genomik analizler

NCCN Guidelines Version 2.2019
Hepatosplenic Gamma-Delta T-Cell Lymphoma

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WORKUP

ESSENTIAL:

- H&P exam; full skin exam; attention to node-bearing areas, including Waldeyer's ring; evaluation of size of liver and spleen, nasopharynx
- Performance status
- B symptoms
- CBC with differential
- Bone marrow biopsy ± aspirate
- LDH
- Comprehensive metabolic panel
- HLH workup
- Uric acid
- PET/CT scan[f] and/or C/A/P CT with contrast of diagnostic quality
- Echocardiogram or MUGA scan if anthracycline or anthracenedione-based regimen is indicated
- Pregnancy testing in women of child-bearing age (if chemotherapy or RT planned)
- HLA typing

USEFUL IN SELECTED CASES:

- Neck CT with contrast
- Head CT or MRI with contrast
- HIV testing
- Hepatitis B and C testing
- Consider quantitative EBV PCR
- Discussion of fertility issues and sperm banking

→ See Primary Treatment (HGDTL-4)

NCBI Resources How To

PubMed.gov
US National Library of Medicine
National Institutes of Health

PubMed

Advanced

Format: Abstract

Send to

- 21 hasta
- Ortanca yaş 34
- 6 hasta başta tanı alamadı
- 21 hastada (%100) splenomegali, 15 hastada (%72) hepatomegali, 20 hastada (%95) trombositopeni
- Ortanca sağkalım 16 ay
- 13 hastanın 9 unda izokromozom 7 q pozitif saptandı.
- 2 hasta hariç tüm hastalar pekiştirici veya kurtarma amaçlı YDKT e rağmen ex oldu

characteristics. Bone marrow biopsy with combined phenotyping is sufficient for diagnosis, and splenectomy is therefore unwarranted. Current treatment modalities appear to be ineffective in most patients.

PMID: 12907441 DOI: 10.1182/blood-2003-05-1675

[Indexed for MEDLINE] Free full text

Tedavi;

- HSTCL tanısı doğrulandıktan ve evreleme çalışması tamamlandıktan sonra, hastalık oldukça hızlı ilerleyebildiğinden derhal tedavi başlatılmalıdır.
- Standart bir tedavi mevcut değil; ancak, diğer agresif lenfomalarda yapılan çalışmalara dayanılarak kemoterapi rejimleri getirilmiştir.
- Hematopoetik kök hücre nakli ve klinik çalışmalara katılım, göz önünde bulundurulan seçenekler arasında olabilir.

NCCN Guidelines Version 2.2019
T-Cell Lymphomas

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Updates in Version 2.2019 of the NCCN Guidelines for T-Cell Lymphomas from Version 1.2019 include:

Peripheral T-Cell Lymphomas

TCEL-B 1 of 5

- First-line therapy for ALCL
  - The qualifier "ALK+ histology" was removed.
  - "Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisone)" was added as a preferred regimen with a category 1 designation.
- First-line therapy for other histologies (PTCL, NOS; AITL; EATL; MEITL; nodal PTCL, TFH; and FTCL)
  - Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisone) for CD30+ histologies was added as a preferred regimen with a category 2A designation.

Breast Implant-Associated ALCL

BIAA-2

- Extended disease (stage II-IV)
  - "Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisone)" was added as a systemic therapy option with a category 2A designation.

Adult T-Cell Leukemia/Lymphoma

ATLL-B 1 of 2

- Initial chemotherapy
  - "Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisone) for CD30+ cases" was added as an option with a category 2A designation.

Hepatosplenic T-Cell Lymphoma

HGTL-4

- Primary treatment
  - "Brentuximab vedotin + CHP (cyclophosphamide, doxorubicin, and prednisone) for CD30+ cases" was added with a category 2A designation. A corresponding footnote was added, "Patients with HGDTL were eligible for the ECHELON-2 study (Horwitz SM, Connor OA, Pro B, et al. The ECHELON-2 trial: results of a randomized, double-blind, active-controlled phase 3 study of brentuximab vedotin and CHP (A+CHP) versus CHOP in the frontline treatment of patients with CD30+ peripheral T-cell lymphomas [abstract]. Blood 2018;132:Abstract 997) but no patients were enrolled."

Continued

The ECHELON-2 Trial: Results of a Randomized, Double-Blind, Active-Controlled Phase 3 Study of Brentuximab Vedotin and CHP (A+CHP) Versus CHOP in the Frontline Treatment of Patients with CD30+ Peripheral T-Cell Lymphomas

Steven M. Horwitz, Owen A. O'Connor, Barbara Pro, Tim M. Illidge, Michelle A. Fanale, Ranjana H Advani, Nancy L. Bartlett, Jacob Haaber Christensen, Franck Morschhauser, Eva Domingo-Domenech, Giuseppe Rossi, Won Seog Kim, Tatyana A. Feldman, Anne Lennard, David Belada, Árpád Illés, Kensei Tobinai, Kunihiro Tsukasaki, Su-Peng Yeh, Andrei R. Shustov, Andreas Hüttmann, Kerry J Savage, Sam Yuen, Pier Luigi Zinzani, Zhaowei Hua, Meredith Little, Shangbang Rao, Joseph Woolery, Thomas Manley, and Lorenz Trümper

Blood 2018 132:997; doi: https://doi.org/10.1182/blood-2018-99-110563

- ✓ Toplam 452 hasta
- ✓ BV + CHP: brentuksimab vedotin 1.8 mg / kg + siklofosfamid 750 mg / m^2 , doksorubisin 50 mg / m^2 , ve prednizon Gün 1-5, 100 mg (n = 226)
- ✓ CHOP: siklofosfamid 750 mg / m^2 , doksorubisin 50 mg / m^2 , vinkristin 1,4 mg / m^2 gün 1-5 ve prednizon, 100 mg (n = 226)
- ✓ Medyan izlem süresi 35.2 ay, 3 yıllık PFS ve OS, % 52.9 (% 95 CI: 47.7-57.7) ve% 73.1'dir (% 95 CI: 68.3-77.2).
- ✓ Febril nötropeni A+CHP (%41)/CHOP(%33)
- ✓ Periferik nöropati A+CHP (%52/CHOP(%55)
- ✓ Ölümcül olay A+CHP (%3)/CHOP(%4)

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Hepatosplenic Gamma-Delta T-Cell Lymphoma

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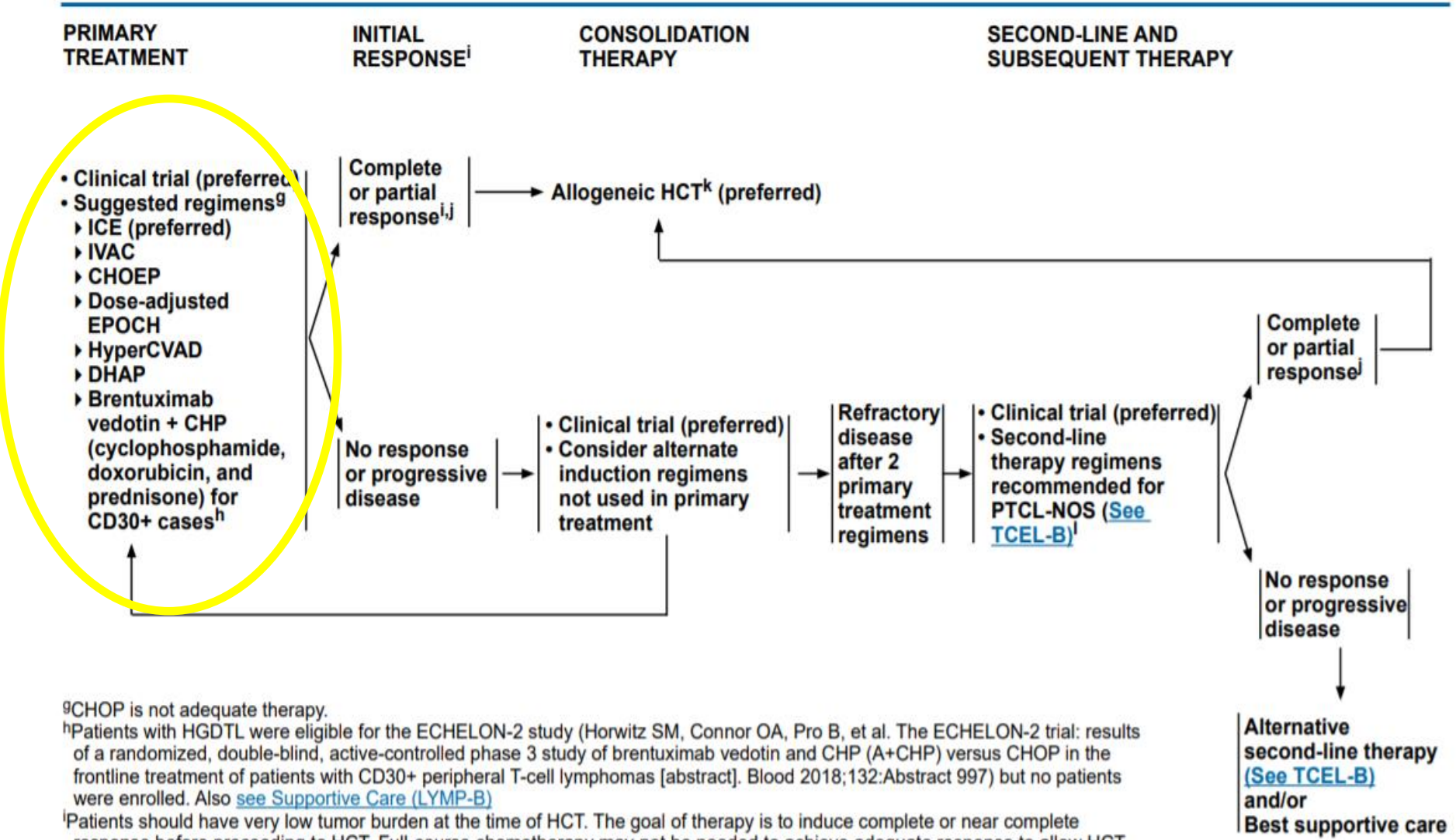


[g]CHOP is not adequate therapy.

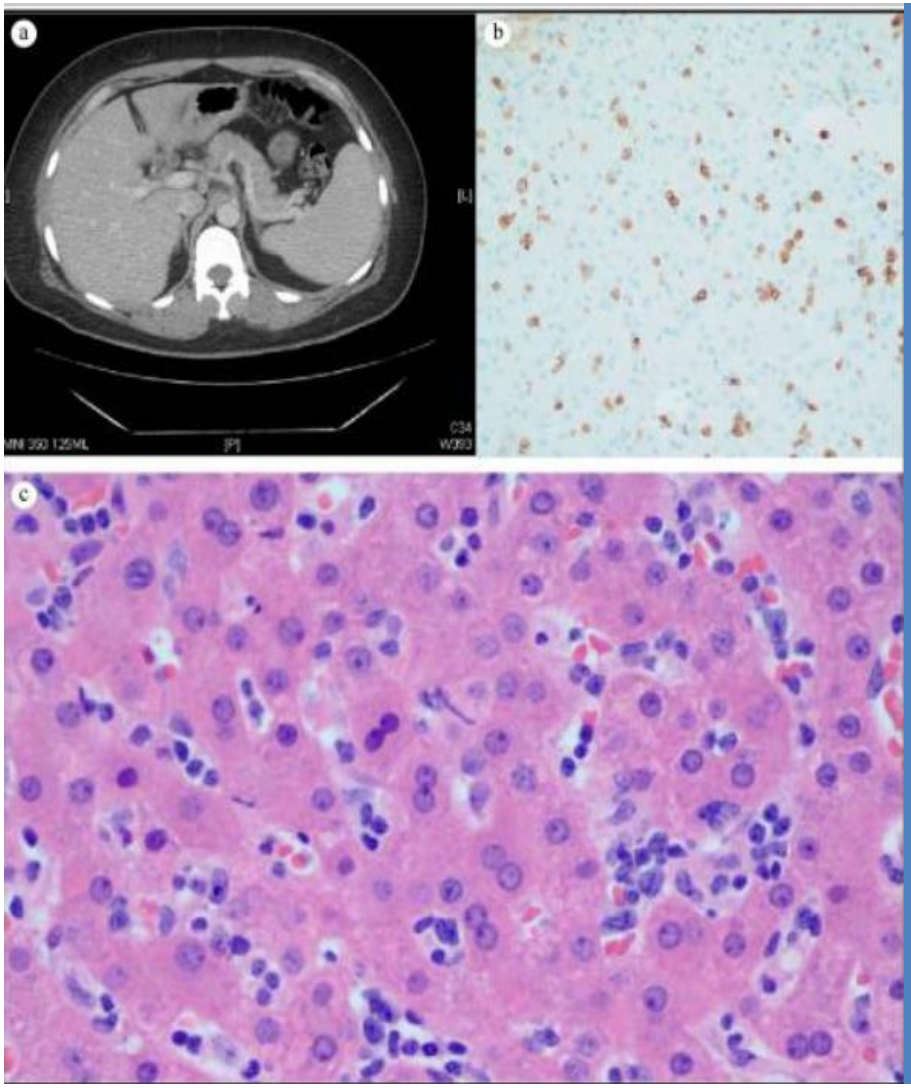
[h]Patients with HGDTL were eligible for the ECHELON-2 study (Horwitz SM, Connor OA, Pro B, et al. The ECHELON-2 trial: results of a randomized, double-blind, active-controlled phase 3 study of brentuximab vedotin and CHP (A+CHP) versus CHOP in the frontline treatment of patients with CD30+ peripheral T-cell lymphomas [abstract]. Blood 2018;132:Abstract 997) but no patients were enrolled. Also see Supportive Care (LYMP-B)

[i]Patients should have very low tumor burden at the time of HCT. The goal of therapy is to induce complete or near complete response before proceeding to HCT. Full-course chemotherapy may not be needed to achieve adequate response to allow HCT.

[j]PET scan alone is inadequate for response assessment. PET-negative response should be confirmed by bone marrow biopsy. HGTL is non-nodal and Lugano response criteria do not apply.

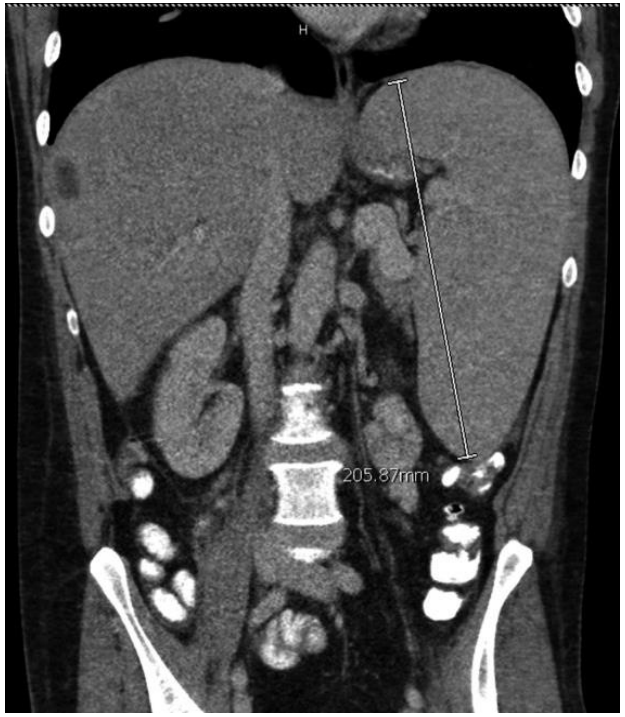
[k]Consider HDT/ASCR if unfit or lacking a suitable donor.

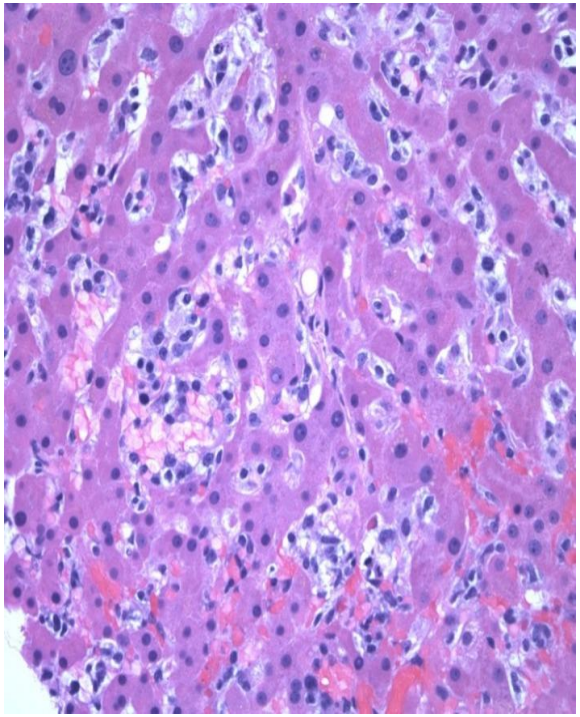
[l]Responses have been observed with alemtuzumab, pralatrexate, and ESHAP.

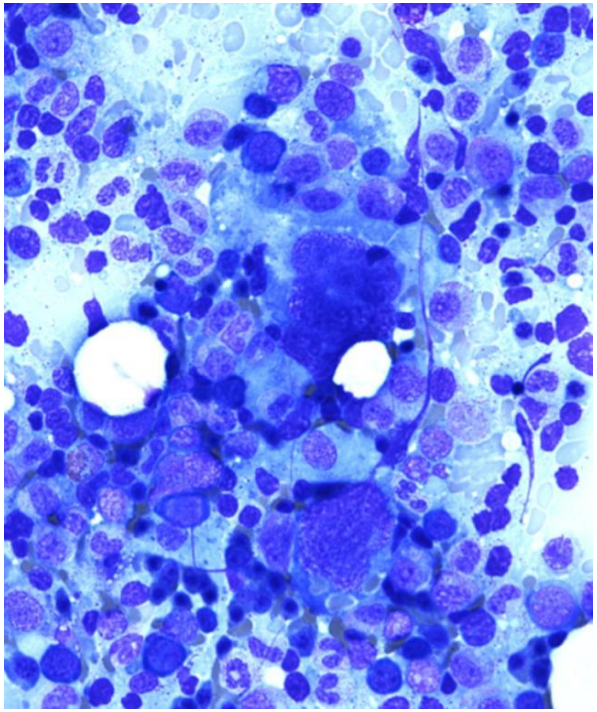


- Kliniğimizde baş ağrısı, ateş, yüksek karaciğer fonksiyon testleri ve hepatosplenomegali ile başvuran 47 yaşında bir kadın.
- Bir karaciğer biyopsisinde çarpıcı bir hepatik intrasinüzoidal lenfosit infiltratı saptandı.
- Bunların immünohistokimyasal boyamalar ($CD8^{+}$, $CD4^{-}$, $CD3^{+}$, $CD20^{-}$T hücreleri olduğu doğrulandı.).
- Moleküler çalışmalar, bir T-hücresi gama reseptörü gen yeniden düzenlemesini göstermiştir.
- Dalak 14,4 cm

- Daha önce azatiyoprin ve adalimumab ile immünmodülatör tedavide Crohn hastalığı öyküsü olan 55 yaşında bir beyaz erkek, subakut ateş, burun kanaması ve uyuşukluk şikayeti ile başvurdu.
- O pansitopeni ve hepatomegali, ve BT ile splenomegali (21 cm)
- Kemoterapi ile sistemik tedavi başlatıldı; ancak, CHOEP (siklofosfamid, doksorubisin, vinkristin, etoposid ve prednizon), ICE (ifosfamid, karboplatin, etoposid), romidepsin, kladribin ve alemtuzumab'a karşı dirençliydi.
- Sitopenileri steroidlere cevap verdi.
- Sonunda tekrarlayan ateşler, kötüleşen hepatosplenomegali ve ilerleyici hastalık nedeniyle öldü.



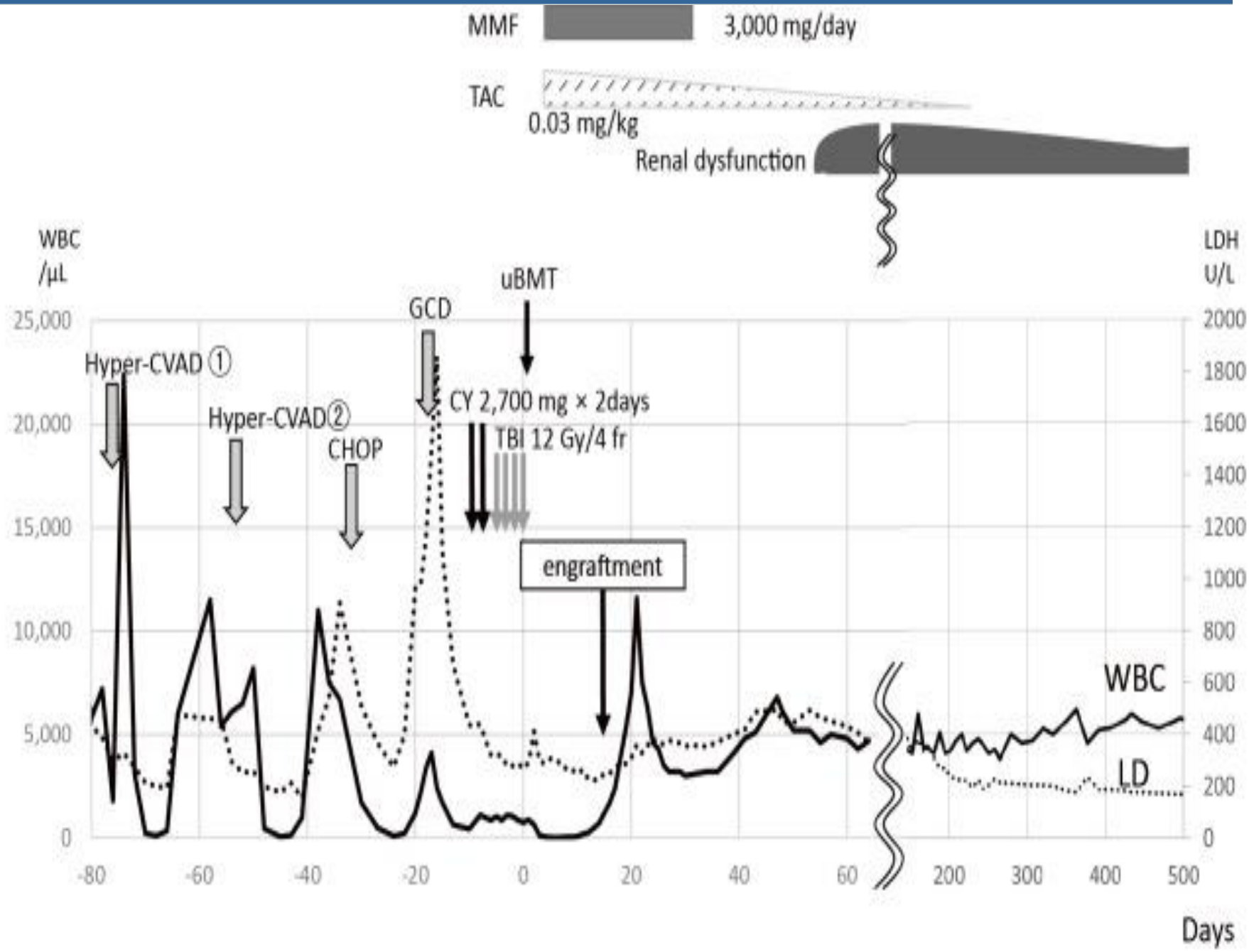




Otolog ve Allojenik Nakil;

- Hematopoetik kök hücre transplantasyonunun HSTCL için konsolidasyon stratejisi olarak tam rolü henüz belirlenememiştir.
- Allojenik transplantasyon ile sonuçların iyileşme şansı verildiğinde daha iyi olabileceği ve transplantasyona geçme süresinin sonuçların daha iyi olabileceği gözlenmiştir.*’**
- İndüksiyon sonrası CR elde eden hastaların bile kısa remisyon sonucu medyan sağkalım süresinin zayıf olduğu gösterilmiştir.
- İndüksiyon tedavisi ile bir CR elde eden hastalar, otolog kök hücre nakli için aday olabilirler.

*Blood 102:4261-4269, 2003
**Blood 122:3032, 2013



Blood Cancer Journal

Blood Cancer J. 2015 Jun; 5(6): e318.
Published online 2015 Jun 5. doi: 10.1038/bcj.2015.43

PMCID: PMC4648481
PMID: 26047388

Outcomes of allogeneic stem cell transplantation in hepatosplenic T-cell lymphoma

A Rashidi[1] and A F Cashen[1,*]

- 54 hasta, ortanca yaş 34 ve %73 erkek
- Lenfadenopati % 35, hepatosplenomegali %100 ve kemik iliği tutulumu %82
- Ölüm nedeni % 68 hastada nüks olmayan mortalite (NRM) ve % 32'de nüks idi.
- Tahmini 3 yıllık NRM,% 34 idi
- Kadın cinsiyet koşullandırma yoğunluğundan bağımsız olarak daha uzun RFS'nin belirleyici belirleyicisi olarak kaldı.

Am J Hematol. 2017 Jun;92(6):E99-E101. doi: 10.1002/ajh.24711. Epub 2017 Apr 29.

Incidence, clinical findings, and survival of hepatosplenic T-cell lymphoma in the United States.

Durani U[1], Go RS[2,3].

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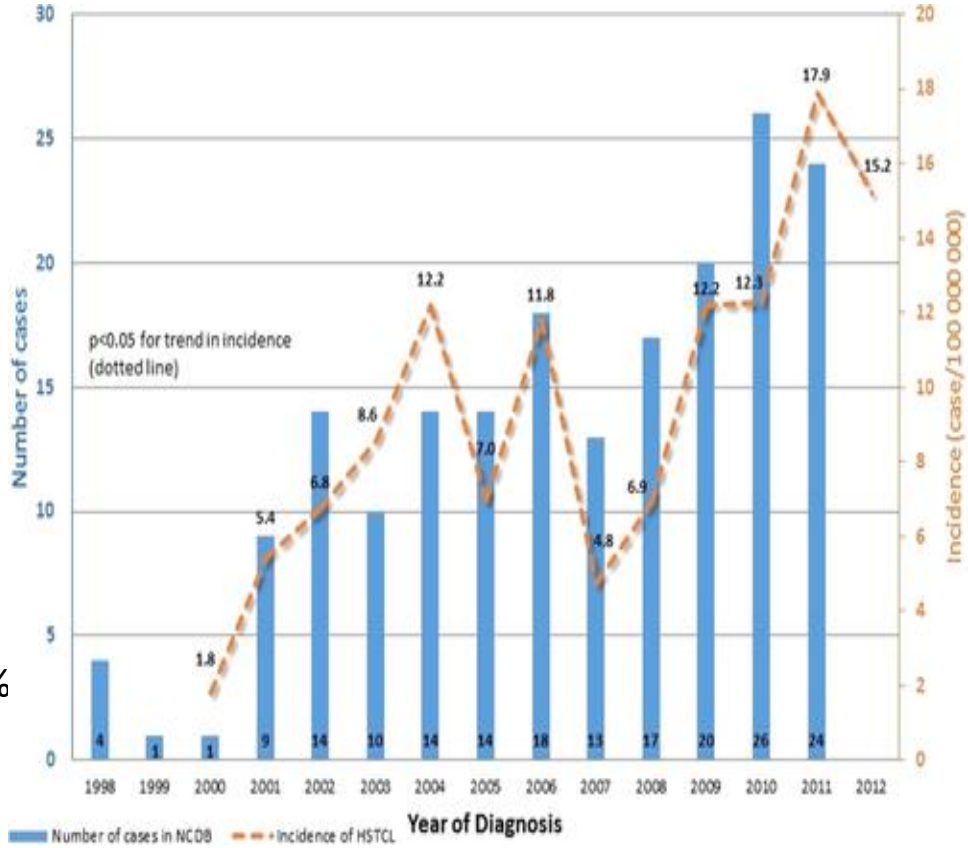
PMID: 28263402 DOI: 10.1002/ajh.24711

- HSTCL insidansı 2000 yılında 1.8'den 2012 yılında 15.2'ye yükseldi (*P* <.05) (Şekil 1).
- Siyahlar en yüksek insidans oranına (18.9), ardından Asyalılar / Pasifik Adalılarına (11.4) ve Beyazlara (6.1) sahipti.
- 1998 ve 2011 yılları arasında NCDB'de yeni tanı konmuş HSTCL'li 185 hasta tanımlandı.
- Analiz sırasında 149 hasta (% 81) öldü.
- Hayatta kalanlar için ortanca takip süresi 56.4 ay, ortanca yaş 47.0 ve % 68.6 erkekti.
- 130 hasta (% 73,5) birinci basamak tedavi olarak tek başına kemoterapi alırken, 12 (% 6,5) konsolidasyon amaçlı ÖTV (dört otolog, yedi allojenik ve bir bilinmeyen tip) aldı.
- Bir ve beş yıllık sağkalım oranları sırasıyla% 40.4 (% 95 CI: 33.3-47.4) ve% 14.6 (%
- Çalışmamız, çalışılan on yıl boyunca HSTCL hastaları için OS'de anlamlı bir iyileşme bulamadı.



Soru ve katkılarınız…