Session 4 Summary

Cutaneous B-Cell
Lymphoma/Lymphoproliferative
Disorders

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Approach

- Discussion divided into diagnostic categories or their differential diagnosis:
  - Marginal zone lymphoma (MZL)
  - Follicle center/follicular lymphoma (FCL/FL)
  - Diffuse large B-cell lymphoma (DLBCL)
  - Other B-cell neoplasms
  - B-cell lymphoma/lymphoproliferative disorders in immunocompromised patients
- Apologize for minimal description of some cases
- Remember, the panel did not always agree, so some cases are controversial

68 cases in 20 minutes---

Marginal Zone Lymphoma
and Differential Diagnosis

16 cases
Primary Cutaneous Marginal Zone Lymphoma: Classical Features

- Case 4, Swerdlow, Pittsburgh
- Case 88, Leeborg, Fan, Oregon (staging pending)
- Case 6, Song and Oetama, UCLA

IgG kappa

Primary Cutaneous Marginal Zone Lymphoma: Classical Features

- Case 6, Song and Oetama, UCLA

BCL2 CD3 CD20 CD5 CD10 CD21

Difficult Cases

- Normal B-cells outnumber the tumor cells
- Plasma cells may not be numerous or clonal
- More T-cells than B-cells

Where are the tumor cells?
Primary Cutaneous MZL: Diagnostic Difficulties

- Case 70, Epuri, Shet, Tata Mem Hosp, Mumbai; 80 yo F, thickened scalp w/ alopecia; IgH+, plasma cells not clearly clonal

Panel stains: Plasma cells appear polytypic; IgH similar to BCL-2 c/w mantle cells

CD3
CD20
CD21

Panel dx: "Worrisome for MZL"

Primary Cutaneous MZL: Descriptive Diagnosis

- Case 58, Venkatraman et al, Belfast; 60 yo F, persistent lesion for 17 mo
  - Biopsied 3 times, IgM, same clone in two
  - Called "low-grade B-cell lymphoma, probable pc MZL" (cytogenetics)
  - Excision 6 mo later appeared reactive; no clonal IgH

Panel stains: kappa+ lambda; no Dutcher bodies; few plasma cells present; partially disrupted FDC network

Primary Cutaneous MZL: T-Cells May be Predominant

- Case 307, Robu et al, Fox Chase Cancer Ctr.; plasma cells clonal, IgH clonal
- Case 143, Morgan, Leith, WI; ClL B M, w/ recurrent nodular lesions, evolution over 3.5 yrs from T-cell rich CLH (IgH-) to MZL (T-cell rich) (IgH+)

Panel dx: MZL

Panel stains: kappa >> lambda, kappa+ Dutcher bodies and Mott cells

Primary Cutaneous MZL: T-Cells May be Predominant

- Case 217, Merzianu, Roswell Park; 28 M lesions on buttock and arm x 5 yr; Two bx: Numerous T-cells; IgH+, but different clones

Panel dx: Controversial, CLL vs. MZL with ? multiple clones over time

Panel stains: few pc, kappa+ lambda, rare lambda+ Dutcher bodies
Primary Cutaneous MZL: T-cells are Predominant and Clonal

- Case 53, Shet, Mumbai: 40 yo F; solitary 3 x 2 cm lesion; large numbers of histiocytes and T-cells differential dx TCHRBCL, MZL, abnormal CLH; plasma cells appear polytypic. Lesion recurred, patient refused re-biopsy

Panel dx: Controversial, ML. Ddx: PC small-medium TCL vs. MZL

Primary Cutaneous MZL: Diagnostic Difficulties, IgH+ and TCR+, T-cells are Predominant

- Case 207, Lopategui/Nathwani, Cedars-Sinai LA: 42 M 3 x 3 cm cheek lesion; clonal TCR beta, TCR gamma, IgH, IgK

Other stains: CD10-, CD21 no meshwork, BCL-6-, EBER-
Primary Cutaneous MZL: Diagnostic Difficulties, IgH+ and TCR+, T-cells are Predominant

- Case 207, Lopategui/Nathwani, Cedars-Sinai LA: 42 M 3 x 3 cm cheek lesion; clonal TCR beta, TCR gamma, IgH, IgK

Panel dx: MZL + TCL vs. TCL with clonal plasma cells

MZL-like Lesions in AITL

- Case 9, Bayerl et al. Penn State, 66 yo M, abn LN 7 mo prior to skin lesions; skin lesions dx as MZL, ~18 mo later AILT

Variable EBER staining in the skin biopsies

Original LN, 18 mo

Identical T-cell clones in two skin and LN bx at 18 mo

Variable IgH clones in 2nd LN and skin bx

Panel dx: AITL with monoclonal plasma cells and different B-cell clones
Possible Etiologies of Abnormal Lymphoid Infiltrates with Clonal B-Cells and T-Cells

- Indolent CLH due to an abnormal immune response/background; ? Precursor to MZL
- MZL with restricted T-cell response or a pseudoclonal
- Primary cutaneous CD4+ small/medium TCL (or pseudolymphoma or CLPUS)
- Rule out secondary cutaneous involvement by AITL or PTCL, NOS with clonal B-cells

Primary Cutaneous versus Systemic MZL

- Case 80, de Mascarel, Bordeaux; 70 yo F bilateral infiltration upper eyelid; PB, molecular IgH+ eyelid location
- Case 11, Banks, Carolinas Med Ctr; 86 yo F, BM/blood involvement 6 yrs prior to cutaneous lesion; amyloid present
- Case 155, Gabali et al., MI; 11 yo M lower lip/minor salivary gland; amyloid present

Panel dx: ML w/plasmacytic differentiation, favor MZL and amyloid; cannot determine if the two processes are clonally related (both lambda+)

Primary Cutaneous Marginal Zone Lymphoma: Unusual Manifestations

- Case 292, Pawade, Bristol, UK; 30 yo M with h/o HL, 2008; lung bx showed sarcoidosis

Panel dx: Probable MZL

Panel stains: Disrupted meshwork, many aggregates of IgD+ (mantle) cells; possible focal kappa light chain restricted pc
Primary Cutaneous MZL: Recommended Panel

- CD20 and CD3 to determine predominant cell and distribution
- CD5, CD10, cyclin D1
- CD21, to determine if colonized meshwork
- IgD to delineate normal mantle cells and clonality
- BCL-6 to look for weak expression in B-cells or expression in T-cells
- CXCL13, PD-1 to detect primary cutaneous small-medium T-cell lymphoma/LPD/CLPUS

Follicle Center or Follicular Lymphoma

15 cases

Primary Cutaneous Follicle Center Lymphoma

- Case 35, Olson, Lansing, MI; 62 yo F, scalp lesion: PCFCL often shows numerous LCC
- Case 309, Raza et al., Detroit, multiple lesions over time, IgH negative, but classical histology, and immunophenotype (CD10+, BCL6+, BCL2-)

Primary Cutaneous FCL vs. Reactive

- Case 310, Klepets, Hasserjian, MGH, Boston; 87 yo M “LPD” cervical LN; 8 yr h/o intermittent skin lesions; IgH repeatedly negative

Panel dx: Bx 1, probable PCFCL; Bx 2, PCFCL

Panel dx: Abnormal B-cell infiltrate, probable PCFCL
Problem cases with fragmented/colonized germinal centers

- Part of the spectrum of PCFCL?
- Should you worry about MZL?
- A form of cutaneous PTGC?

**Primary Cutaneous Follicle Center Lymphoma?**

- Case 22, Klimkowska, Simanaitis, Karolinska; 53 yo M enlarged solitary, draining inguinal LN, "in-situ FL" (BCL2-) resembling PTGC, flow 5% clonal CD10+, kappa+ B-cells: IgM and IgG clonal; multiple skin lesions buttock, arms; IgH not done

**Panel dx:** LN, FL: skin FL vs MZL

**Panel stains:** CD10 negative; kappa and lambda not definitive; CD21 some disrupted FDC

**Case 144, Roullet, Franzman, Virginia Beach; 73 yo M, persistent scalp lesion**

Panel stains: Small numbers of kappa/lambda plasma cells, rare kappa DB

**Panel dx:** PCFCL vs. MZL

**Case 77 Jiang, Cappel, Mayo Clinic; 71 yo M, persistent nasal lesion**

Rare lambda+ Dutcher bodies; CD10 in clusters

**Panel dx:** PCFCL vs. MZL
Cutaneous Lymphoid Hyperplasia with Progressive Transformation of Germinal Centers

- CLH with fragmentation of germinal centers may represent PTGC
- B-cells in the nodules are IgD+, cyclin D1-, and CD57+ T-cells are present


Primary Cutaneous Follicle Centre Lymphoma: Associated DLBCL

- Case 182, Chorny, Launder, Denver: PCFCL + DLBCL, MYC+, BCL2-, BCL6-

Panel 1: First Biopsy
Panel 2: Second Biopsy, a few weeks later

PCFCL with Progression to Diffuse Growth Pattern

- Case 61, Venkatraman et al., Belfast 48 yo M, 24 yr course, multiple relapses, evolution from nodular to diffuse growth


PCFCL: Complex Karyotype

- Case 316, Vakil, UCLA, 55 yo F, slowly growing scalp lesion x 6 mo; staging -; BCL2+, complex karyotype with IgH/BCL2 fusion + 8q24 (MYC), -14q, +18q and -4, -6, -10, +12; IgH PCR -
Secondary Cutaneous Follicular Lymphoma

- Several cases were secondary FCL in the skin/subcutaneous tissue:
  - Case 15, Jiang, Cappel, Mayo Clinic, FL
  - Case 17, Rollins-Ravel, Felgar, Pittsburgh, likely nodal
  - Case 97, Juntilla et al., Penn
  - Case 296, Sramek et al. UNM, 77 yo F, large multilobate cells resembling HRS cells; patient has h/o extranodal B-NHL, epidural and orbital masses

Cutaneous B-Cell Lymphoma vs. Hodgkin Lymphoma

- Case 94, Yue et al. US Labs, Irvine; 34 yo male w/ back lesion for 3 yr
  - BCL6- or wk+, CD10 NT, EBER-, IgH-, TCR-, Oct-2 variable; BOB.1+
  - Panel dx: Cutaneous B-cell lymphoma, unclassifiable vs CHL, difficult case

PCFCL with Clonal TCR: A targeted immune response or a pseudoclone?

- Case 200, Kansal et al. Cedars-Sinai, LA; 66 yo M, posterior auricular lesion

Diffuse Large B-Cell Lymphoma

- 11 cases:
  - 1 DLBCL, secondary
  - 1 DLBCL versus BL, transplant pt, “double hit”
  - 3 DLBCL, leg type
  - 3 Primary effusion lymphoma, extracavitary
  - 2 Intravascular LBCL
  - 1 PTCL (NOS vs. AILT) + EBV+ BCL with PCD

Panel dx: PCFCL
**DLBCL**

- Case 21, Sohani et al. MGH, Boston, secondary DLBCL, CD10-, BCL6+, BCL2-, MUM1-
- Case 32, Bailey et al. U MZ, Ann Arbor: 57 yo M, s/p renal tx 17 yrs prior, "double hit" with complex karyotype; stage IV, BM+

**Primary Cutaneous DLBCL, Leg Type**

- Case 253, Boyer, Ferry, MGH, Boston
- Case 99, Hutchison et al. SUNY, 85 yo F, 1.9 cm enlarging facial lesion, XRT and 3 cycles RCHOP, NED at 10 mo.

**DLBCL, Leg type vs. Cutaneous B-Lymphoblastic Lymphoma/Leukemia**

- Case 31, Sohani MGH and Abu-Jawdeh Salem MA, 83 yo F, isolated skin lesion with development of second lesion, 2 cycles CHOP, lesions resolved, NED at 1 yr

**DLBCL, Extracavitary PEL**

- Case 104, Ochs et al. Penn, 49 yo M HIV+ w/ KS, jaw lesion and lung, LN disseminated lesions, CD30+, CD45RB+, EBER+, HHV8+, dim CD4
- Case 110, Franck et al. Brussels, 51 HIV+ M w/disseminated KS, tx for KS started, 2 mo later multiple skin lesions + lymphadenopathy, dyspnea

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Panel dx: BCL unclassifiable between DLBCL and BL, "double hit"
**DLBCL, Extracavitary PEL**

- Case 96, Banki et al., SUNY, 41 yo M, left groin and right gluteal mass; found to be HIV+; DOD at 6 mo.

**Intravascular Large B-Cell Lymphoma**

- Case 41, Sidhu et al., Brown, IVL in hemangioma.
- Case 312, Czader, Nassiri, Indiana; 62M erythematosus rash, plaques, negative staging, treated with bimonthly Rituxan alone with NED at 5 yrs.

**PTCL, NOS with Some AITL-like Features and EBV+ BCL with PCD**

- Case 166, Siddig et al. Duke, 62 yo M w/maculopapular rash and diffuse lymphadenopathy.

**Other B-Cell Neoplasms**

10 cases:
- 1 LPL
- 4 CLL/SLL
- 3 B-Lymphoblastic lymphoma/leukemia (1 primary cutaneous and 2 BM+)  
- 1 MCL with unusual features
- 1 Plasma cell myeloma
Secondary Cutaneous Involvement by Other B-cell Neoplasms

- Case 234, Cosk, Cleveland Clinic, LPL
- Case 5, Song UCLA, Lai, Dieran, ulcerated skin lesion, CL with PCD
- Case 139, Geng et al, Thomas Jefferson Univ, CLL/SLL, aGCH
- Case 242, Shihshahara et al., Penn, B-CLL, leukemia cutis, predilection for inflammatory sites, such as rosacea; no adverse prognosis unless large cell infiltrate
- Case 252, Sohani et al. MGH, Boston, 82 yo F, CLL dx in 2006

Lymph node bx and blood 2006, dim CD20+ , CD5+ , CD23+ , kappa B-cell
FISH: del (13q)(14.3); IgH locus 14q13 rearranged

Multiple skin bx, 2010, CD5, CD10+ , BCL6 variable, EBER- B-cells; CD21 did not reveal meshwork; IgH+ in skin with similar clones

Panel dx: CLL w/ transformation (Richter syndrome) w/ change in phenotype

Cutaneous Involvement by B-Lymphoblastic Lymphoma/Leukemia

- Case 180, Ortonne, Crétiel, 78 F, left leg nodule, primary cutaneous B-LBL, CD19+, CD20-, CD10-, BCL6+, BCL2+, TdT+, CD34+, BM+
- Case 245, Antelo et al. Harbor-UCLA, 9 yo M scalp swelling, CD10+, CD19+, CD30 ak-, TdT+, CD34+, BM+

Bone marrow

Cutaneous Involvement by B-Lymphoblastic Lymphoma/Leukemia

- Case 184, Grant, Univ W Virginia, 4 yo F w/Down Syndrome thigh swelling, erythema, TdT+, CD34-, BM+

TdT

Plasma Cell Myeloma with Cutaneous Relapse

- Case 98, Nelson et al., Caris Life Sciences, Phoenix, 68 M PCM, chemotherapy, BM- at 2 mo, multiple skin lesions at 9 mo.

Panel studies: CD10-, MYC rearrangement-, multiple MYC signals, aneuploidy/polyploidy

Panel dx: PCM w/ MYC amplification
Secondary Cutaneous Involvement by Mantle Cell Lymphoma
- Case 198, Soma, Sacramento, purpuric lesions on the face: BM+

Lymphocytes are CD20+ and lack CD5, CD43; MIB-1~5%; FISH + IgH/BCL1+

Immunodeficiency-Related Lymphoproliferative Disorders
16 cases:
11 Iatrogenic
- 1 Remicade associated
- 10 Methotrexate-associated:
  - 6 MCU type, 2 DLBCL, 1 CHL, 1 HL-like
  - 1 MCU arising in an elderly patient
  - 1 EBV+ DLBCL of the Elderly
  - 2 EBV+ DLBCL w/PCD ? Related to EBV+ DLBCL of the Elderly
  - 1 EBV+ Gingival lesion in a pt. with tumor stage MF

Immunodeficiency Related Cutaneous B-Cell Proliferations: Biologic Response Modifier Drugs
- Case 187, Al-Khatib, Beaty, Wake Forest, 77 F Crohn Dz + Remicade and ? Imuran, 2 yrs later multiple, tender dermal nodules; lesions resolved with discontinuation of therapy + 4 cycles Rituximab.

Methotrexate Associated Cutaneous B-Cell Proliferations
- Case 129, Dojcinov, Cardiff; Jaffe NCI 80 F RA + MTX, ulcerated solitary arm lesion; withdrew MTX, NED at 5 yrs. polymorphous infiltrate with HRS-like cells; features of lymphomatoid granulomatosis

Other studies: CD15-, Oct-2+, BOB.1+, MUM1+, TCR+ clonal restricted response; EBER negative

Panel de: MTX-associated LPD, EBV+, MCU type
Methotrexate-Associated Cutaneous B-Cell Proliferations

Case 18, Vasef, Reichard, Univ NM, 75 F RA + MTX, 2 yrs earlier MTX-associated LPD with features of HL and LBCL: ulcerated solitary chin lesion

CD20
CD3
PAX5/CD30
EBER
Panel dx: MTX-associated LPD, EBV+, MCU type

Methotrexate-Associated Cutaneous B-Cell Proliferations

Case 81, Gruver, Hsi, Cleveland Clinic, 63 M RA + MTX + hydroxychloroquine, shallow suprapubic ulcer, remitted with d/c of MTX

Other stains: BCL6+, EBER+, CD10-, MUM1-, ALK1-

Panel dx: MTX-associated LPD, EBV+, MCU type

Methotrexate-Related Cutaneous B-Cell Proliferations

Case 251, Perry et al., Nebraska, 79 F RA + MTX, 18 yrs; ulcerated draining foot lesion with additional leg lesions; lesions resolved with d/c MTX and Rituxan x 8 doses

PAX5
BCL2
MUM1
EBER
FOXP1
Resembles DLBCL, Leg Type
Panel dx: MTX-associated LPD, EBV+, MCU type

Methotrexate-Associated Cutaneous B-Cell Proliferations

Case 325, de Leval, Lausanne, 78 M dx w/MF 2006; evolving Sézary syndrome 2006; tx w/clorambucil, interferon, MTX; 2009 ulcerated skin nodules on the legs

Panel dx: MTX-associated LPD, EBV+, MCU type

Other stains: CD15-, BCL6, BCL2, MUM1, BCL2/-, IgM/Lambda, CD30/-, Ki67
Methotrexate-Associated Cutaneous B-Cell Proliferations
• Case 260, Summers, Jaffe, NCI; 81 F RA + MTX, ? outcome
  Panel dx: MTX-associated DLBCL, EBV- (~25% DLBCL are EBV-)
  Other stains: CD10 wk +; BCL6 wk +

Methotrexate-Associated Cutaneous B-Cell Proliferations
• Case 301, Nathwani BN et al, Cedars-Sinai, Los Angeles; 83 M RA + MTX + prednisone skin lesion on face; Pt died of pulmonary embolus 11 days after skin bx
  Panel dx: MTX-associated DLBCL, EBV+
  Panel studies: kappa and lambda not clearly clonal; FISH for MYC negative
  Other studies: Large cells CD10-, BCL6-, CD21-, EBER-

Methotrexate-Associated Cutaneous B-Cell Proliferations
• Case 171, Alobeid et al. Columbia; 51 F dermatomyositis + MTX, lower back subcutaneous mass
  Panel dx: MTX-associated LPD, CHL-type (fibroblastic appearance)
  Panel stains: Oct-2+, BCL1-, CD10 blush

Methotrexate-Associated LPD, CHL versus Hodgkin-like
• Case 173, Alobeid et al. Columbia: 55 F RA + MTX, upper back non-healing lesion
  Panel dx: MTX-associated LPD, MCU type vs. Hodgkin-like
  Panel stains: OCT-2+, BCL1-, CD10 blush

Panel dx: MTX-associated DLBCL, EBV-
**Methotrexate-Associated LPD, Hodgkin-like**
- Case 228, Hoehn et al. MDACC, 25 F dermatomyositis + MTX + steroids + thalidomide, multiple scalp nodules and systemic symptoms; staging negative.
- Panel dx: MTX-associated LPD, Hodgkin-like
- CD30
- EBV, small and large cells
- Panel stains: Oct-2+, BOB.1+ in some areas

**Methotrexate-Associated Cutaneous B-Cell Proliferations**
- 7 cases were ulcerated and considered MCU type
  - 5 DCLL morphology
  - 2 mixed LPD-like
- 1 case with DLBCL histology was ulcerated but EBV-
- 2 cases in patients with dermatomyositis had more CHL or Hodgkin-like lesions (1 CHL was EBV+)
- EBER+ in 80%
- CD15+ in 2/6 tested (1 CHL, 1 Hodgkin-like); 4 MCU were CD15-
- Follow-up is limited: 4 cases
  - 3 MCU resolved (one who received Rituxin)
  - 1 HL-like with NED 6 yr after XRT+ ABVD
- 1 died of unrelated causes and 5 have unknown outcomes

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**EBV+ DLBCL of the Elderly**
- Case 92, Climent et al. Barcelona, 84 M with skin lesions and enlarged inguinal LN
- CD20
- CD3
- lambda
- kappa
- EBER
- IgH and TCR beta clonal

**EBV+ Mucocutaneous Ulcer in the Elderly**
- Case 246 Liu, Jaffe et al. NCI, 79 M with a rapidly growing, ulcerated right cheek lesion which resolved w/o therapy
- CD20
- CD3
- CD30
- EBER
- Panel dx: EBV+ mucocutaneous ulcer
EBV+ ML with Plasmacytic Differentiation in Elderly Patient

- Case 295, Shackley et al. UCLA: 81 F w/ plaque-like lesion on the left shin

IgH and TCR not tested

CD20
CD3
kappa
lambda

Panel dx: EBV+ B-cell lymphoma with plasmacytic differentiation, possibly related to EBV+ DLBCL of the elderly

FISH: no MYC translocation

EBV+ ML with Plasmacytic Differentiation in Elderly Patient

- Case 311, Moore et al. Penn: 70 M multiple subcutaneous masses for several mo.; SPEP negative; developed paraspinous mass

IgH and IgK clonal; and TCR gamma oligoclonal; no MYC translocation

CD10-, CD20-, CD56-, CD79a+, MUM1+, HHV8-, LMP1-

lambda ISH

Panel dx: EBV+ B-cell lymphoma with plasmacytic differentiation, possibly related to EBV+ DLBCL of the elderly

Time to stop!

EBV-associated LP in Patient with a History of MF

- Case 206, Simons, Head, Vanderbilt: 53 M h/o MF (original material not available) w/ progression to tumor stage (DRT, bexarotene); now with gingival lesion

Panel dx: EBV associated lymphoid proliferation

EBV+ ML in Patient with a History of MF

- Case 206, Simons, Head, Vanderbilt: 53 M h/o MF (original material not available) w/ progression to tumor stage (DRT, bexarotene); now with gingival lesion

Panel dx: EBV associated lymphoid proliferation
### Primary Cutaneous Marginal Zone Lymphoma: Take Home Lessons

- If molecular results and morphology don’t agree, be descriptive; follow the patient
- When T-cells are predominant and considering MZL, have to exclude T-cell lymphoma or CLH; do TCR, CXCL13, PD-1
- Clonal/restricted plasma cells may be present in reactive and several neoplastic processes
- Colonized/disrupted FDC may be a useful marker to help distinguish MZL from TCL with clonal plasma cells

### Primary Cutaneous Follicle Centre Lymphoma: Take Home Lessons

- Monomorphic LCC, extension of CD10+/BCL-6+ cells outside the germinal center are useful dx features
- Weak expression of CD10, BCL-2 and negative IgH may make the diagnosis difficult
- When fragmented germinal centers are present, differential dx includes PTGC and PMZL:
  - Clonal plasma cells favor PMZL
  - IgD and CD57 may be useful to distinguish PTGC
- May show progression to diffuse growth with increased large cells

### Primary Cutaneous DLBCL Take Home Lessons

- PC DLBCL, NOS is rare
- DLBCL, leg-type occurs at non-leg sites and should be considered with tumor nodules with centroblasts and immunoblasts that strongly express BCL2, MUM1, and express IgM
- If B-cells have a blastic appearance and lack CD20 consider B-ALL/LBL
- CD30+ tumors in the skin rarely include extracavitary PEL in the DDx; correlate with clinical history/HIV; if immunoblastic or plasmablastic, do HHV8 and EBV
- Always look for CD20+ B-cells in the vessels
- DLBCL or BCL with plasmacytic differentiation can arise in or be associated with TCL


- Three cases were limited to the skin
- 1 resolved, CR 20+ mo: 1 CHOP, CR x 43+ mo; 1 RT, CR, relapsed, then RT + etoposide, 2nd CR
- 1 LCL, 2 polymorphic LPD
- Angiocentricity, 2/3; RS-cell-like giant cells, 2/3; extensive necrosis 2/3
- CD15-, CD30+, EBER+, EBNA2+ 1/3
- IgH monoclonal

- Indolent course related to limited skin disease
**FINAL SUMMARY**

- 21% of cases were BCL or DDx
- 45% MZL, PCFCL, or DDx
- Primary cutaneous DLBCL are predominantly leg type
- Always rule-out secondary cutaneous involvement
- Remember, T-cell lymphomas can have clonal B-cell proliferations (large B-cells; plasma cells)

**FINAL SUMMARY**

- Skin lesions are frequent in immunocompromised patients and have variable morphology (DLBCL, CHL, HL-like, polymorphic, and plasmablastic)
- Test for EBV
- Get clinical history
- If lesions are limited to the skin, conservative therapy appears be indicated, particularly with features of “mucocutaneous ulcer”

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**Primary Cutaneous MZL: What to do When T-Cells are Predominant**

- If IgH+, plasma cells are clonal, FDC meshwork colonized \(\rightarrow\) MZL, “T-cell rich” (? Different etiology)
- If IgH+, plasma cells are not distinctly clonal, FDC meshwork colonized \(\rightarrow\) do TCR, if negative \(\rightarrow\) favor MZL
- If IgH-, plasma cells are not distinctly clonal, T-cells dysplastic \(\rightarrow\) do PD-1, CXCL13 and TCR, if negative CLH, if positive, possible primary cutaneous CD4+ sm/med TCL vs. AILT, do EBER and staging