Transformation in Primary Cutaneous Follicle Center Lymphoma (T-PCFCL): Implications for the Indolence of PCFCL.

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Case Report

- Jan, 2007: 45 y/o man with 2 skin nodules of the chest & back, thought to be cysts.
- Dec, 2007: 3rd growing skin nodule over right scapula, punch biopsy in Feb, 2008: Diffuse large B-cell lymphoma (DLBCL).
Case Report

DLBCL: CD10(+), CD20(+), CD30(-), MUM1(-), BCL2(-), Ki-67(~90%+), no light chains detected by immuno, TdT(- ISH) & EBV(-ISH).

FISH analysis:
 (+)FISH for c-MYC rearrangement.\(^1,2\)
(-) FISH for IGH/MYC fusion.\(^1\)
(-) FISH for BCL2 and BCL6 translocations\(^2\) [no double hit].

1. Performed by Genzyme Laboratories.
2. Performed by Dr. Steven H. Swerdlow’s Laboratory, UPMC Health System.
DLBCL: High (~90%+) Ki-67 rate
Case Report

- CD10(+), BCL2(-), no light chains detected by immuno.
- (-) FISH for c-MYC rearrangement.¹

¹. Performed by Genzyme Laboratories.
Case Report

After acceptance of this case: PCR for clonality: DLBCL & FCL had clonal IgH rearrangements, but DIFFERENT clonal peaks (using BIOMED-2 primers).\(^1\) Sequencing with FR1-4 primers pending.

Possibilities: 1) Clonal evolution at different anatomic sites via somatic mutation or receptor revision. 2) A clonally unrelated DLBCL.

Working Dx: Possible T- PCFCL.

1. Performed by Genzyme Laboratories.
Follicular lymphoma, grade 3.
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Case Report

- Systemic work-up negative.
- 6 cycles of R-CHOP and 10 of XRT.
- 3 years, 8 months later, NO RECURRENCE.
**PCFCL & PCDLBCL (non-leg): Good Prognosis**

- PCFCL & PCDLBCL (non-leg): Usually localized area of head/neck or trunk with ~20% recurrence rate & infrequent dissemination.
- PCFCL & PCLBCL (non-leg): 5-year survival >90%.

Poor Prognosis of Transformed Nodal FCL

- Nodal FCL: Annual risk of transformation is ~3% per year X 15 years.¹
- After transformation, median survival: <1-2 years, worse than de novo DLBCL¹
- Nodal FCL and nodal DLBCL with c-MYC rearrangement have poor prognosis with median survival <1 year.²

Review of Transformation in Cutaneous Lymphoma

PubMed search: “transformation”, “cutaneous”, “lymphoma”: 268 cases

- T-PCFCL: 7 articles (total: 8 patients: 2 died of lymphoma, 2 disseminated lymphoma, 1 in remission & 3 no follow-up).
- T-PCMZL: 6 articles (total: 13 patients, 5 died of lymphoma).
- T-MF/SS: 46 articles (total: 358 patients).
Review of the 7 reports of T-PCFCL

1. 44 y/o male with >3 years of untreated PCFCL. Developed cutaneous & systemic DLBCL. 6 cycles of R-CHOP & other chemo: No improvement.
2. 44 y/o male with a BCL6 (+), MUM-1 (-) & BCL2 (-) scalp PCFCL transformed (?time) to PCDLBCL. 8 cycles of CHOP-R. In complete remission at 14 mos.
3. 30 y/o male with enlarging skin nodules of trunk & face x ~4 years, PCFCL: CD10(+), BCL-2(-). XRT => CR X 6 mos. Then DLBCL in neck nodes: CD10(+), BCL-6(+), BCL-2(-), t(14,18) negative. IgVH chain gene analysis: 2 DIFFERENT clones.
4. Study of 145 patients: 1 patient had a primary cutaneous low-grade centrocytic lymphoma transformed to a PCLBCL. No further details.
5. Patient with PCFCL, grade 3, and relapsed with DLBCL 166 months later.
6. Two patients initially diagnosed with PCMZL, reclassified as PCFCL by EORTC, developed T-PCFCL, both died of lymphoma.
7. “Young man with primary cutaneous low-grade centrocytic lymphoma developed a cutaneous immunoblastic lymphoma followed by a leukemic phase.” (Article in Hungarian.)
Corresponding References

5-year Retrospective Search for T-PCFCL at Colorado Kaiser & University of Colorado Dermatopathology Consultants (Drs. Jim Fitzpatrick & Wit High).

\[ \sim \frac{1}{2} \text{ million bxs: } \emptyset \text{ cases of T-PCFCL.} \]
Conclusions: T-PCFCL

- Transformation more common in nodal FCLs (25-35% cases) and MF/SS (20-25% cases).\(^1\)
- Time period for transformation of PCFCL: Varies considerably (0-166 months).
- T-PCFCL mutations: No reports (probable as PCDLBCL has aberrations in \(\frac{3}{4}\) cases)\(^2\)
- T-PCFCL: Few cases, ?adverse prognosis, frequently heralds dissemination and death. If T-PCFCL limited to skin, may be better (?this case).
- Needs further study: ?prognosis, clonality.
