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Clinical History:
A 68 year old man presented in Sept. 2002 with a papule on right foot that was thought be an insect bite. A second lesion developed on the right leg. The possibility of a drug reaction was considered and he stopped his glucosamine. The lesions resolved spontaneously but returned 2 months later and were up to 3 cm leading to a Nov. 2002 biopsy that was subsequently reviewed. At this time the patient underwent computed tomographic imaging which failed to show any evidence of systemic disease and the lesions regressed without further treatment.

He did well until late 2003 when cutaneous papules began to recur (1-2 cm lasting 3-4 weeks on legs and earlobes). He otherwise was well. A bone marrow was performed and was normal (not submitted). A second skin biopsy of a papule on the right axilla was performed at Cleveland Clinic in Jan. 2004. Lesions regressed again without treatment and the patient was followed expectantly. Lesions recurred again in Dec. 2004 and a repeat biopsy of the axilla showed the same process (images submitted).

The patient has remained well without therapy through last follow-up (Dec. 2010) and without additional lesions, 8 years from original presentation.

Biopsy Fixation Details:
Formalin

Description of Clinical Image if Any:
None

Details of Microscopic Findings:
All biopsies showed similar histopathologic features. There was a dense dermal infiltrate of large atypical cells with vesicular chromatin. The cells had round to indented or convoluted nuclei with variably prominent nucleoli.

Immunophenotyping by Immunohistochemistry and/ or Flow Cytometry:
2002: Immunostaining showed the cells were CD3+, CD30+, CD20-, and ALK1-.
Jan 2004: Immunostaining showed that the cells were CD3+, CD4-, CD8-, CD7-, CD30+, CD20-, ALK1-, MUM1+, □F1-

Special Stains:
None

Cytogenetics:
None

Molecular Analysis:
PCR for T-cell receptor beta was performed and was positive for a monoclonal pattern in all three biopsies. Comparison of the amplicon shows an identical size, suggesting clonal identity.

FISH testing in the biopsy from Jan 2004 using a breakapart probe for chromosome 6p25.3 translocation was positive (BAC probes flanking both *IRF4/DUSP22*; CTD-2308G5 telomeric and

RP11-164H16 centromeric). Recent study of the biopsy from 2002 also showed the same abnormality was present at presentation.

Interesting Feature(s) of Submitted Case:
This patient has a primary cutaneous CD30+ T-cell lymphoproliferative disorder that was monoclonal. While the histopathologic features of sheets of CD30+ large cells suggest cutaneous anaplastic large cell lymphoma (ALCL), the clinical features are more in keeping with lymphomatoid papulosis (LyP), leading to the latter as the favored interpretation. Interestingly this patient harbors the recently described translocation at 6p25.3 (*IRF4/DUSP22*) present in approximately 20% of primary cutaneous ALCL. To our knowledge, this represents the only case to date of LyP with this translocation.[1] The ability to study this over time suggests that 1) the translocation may be an early event in the molecular pathogenesis; and 2) the translocation does not portend an aggressive clinical course and should not be used as evidence of lymphoma in this setting.

Proposed Diagnosis:
Primary cutaneous CD30+ T-cell lymphoproliferative disorder, consistent with lymphomatoid papulosis, type C with translocation at 6p25.3 (*IRF4/DUSP22*).

Panel Diagnosis:
Primary cutaneous CD30+ T-cell lymphoproliferative disorder, lymphomatoid papulosis, type C.

Reference:
(1) Wada DA, Law ME, Hsi ED et al. Specificity of IRF4 translocations for primary cutaneous anaplastic large cell lymphoma: a multicenter study of 204 skin biopsies. Mod Pathol. 2010.