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Clinical History:
The patient is a 6 year old male with an 18 month history of diffuse recurrent papulonodules and hypopigmented scaly macules. The patient initially presented with papules that evolved into eroded nodules over a period of 3 weeks prior to healing spontaneously. The hypopigmented macules were thought to be consistent with post-inflammatory pigment alteration. Two previous biopsies were interpreted as spongiotic dermatitis with superficial perivascular infiltrates. The diagnoses of pityriasis lichenoides et varioliformis acuta versus pityriasis lichenoides chronica were considered. The patient was treated initially with phototherapy (15 narrowband UVB treatments) and azithromycin without improvement. The patient was then switched to methotrexate, ultimately reaching a dose of 7.5 mg each week. Although there was some initial improvement, new papules continued to erupt with increasing frequency, albeit with decreased duration and severity. At the time of the current biopsies he was found to have several ill-defined 2 to 3 mm pink to erythematous papules studding the anterior thighs, buttocks, lower back, and upper chest. A similar, larger plaque was identified on the mid back. In addition, numerous subtle hypopigmented macules and patches with fine scale were present on the trunk and extremites. There was no evidence of scarring or palpable lymph nodes in the cervical, axillary or inguinal regions. Biopsies of a hypopigmented patch on the left flank (specimen #1) and the larger plaque on his mid back (specimen #2) were performed.

Biopsy Fixation Details:
Specimens #1 and 2 were fixed in formalin.

Description of Clinical Image if Any:
The image demonstrates scattered erythematous, slightly indurated papules without erosion or ulceration on the back set within a background of numerous subtle hypopigmented macules and patches with fine scale.

Details of Microscopic Findings:
Sections from specimen #1 show a slightly papillomatous epidermis with overlying basket weave orthokeratosis. Scattered throughout the epidermis are large haloed cells with smaller hyperchromatic lymphocytes at the dermal/epidermal junction. The epidermis lacks spongiosis, basal vacuolar change, or dyskeratotic keratinocytes. Within the dermis there is a superficial perivascular lymphocytic infiltrate. The lymphocytes lack cytologic atypia.

Sections from specimen #2 show an acanthotic epidermis with superficial necrosis, erosion, and overlying parakeratotic scale crust. Numerous lymphocytes extend into the epidermis, many of which are large with cerebriform nuclei and surrounding halos. Focally, these lymphocytes collect into intraepidermal nests. Within the dermis there is a superficial and deep perivascular lymphocytic infiltrate with extravasated red blood cells within the dermis.

Immunophenotyping by Immunohistochemistry and/ or Flow Cytometry:
In specimen #1, the large intraepidermal haloed cells highlight with CD1a. CD3 shows T-cells around the superficial dermal vessels and at the dermal/epidermal junction without significant

epidermotropism. There are some periadnexal lymphocytes. CD8+ T-cells outnumber CD4+ T-cells. CD30 is negative. CD20 highlights scattered B-cells.

In specimen #2, the majority of the intraepidermal cells are CD3+/CD8+ T cells. CD4 and CD20 highlight scattered T and B-cells, respectively. CD30 is negative.

Special Stains:
none

Cytogenetics:
none

Molecular Analysis:
T-cell receptor gamma (*TCRG*) chain gene rearrangement studies were performed on both specimens #1 and 2 using a mixture of ^{32}P-labeled joining region primers with 1 of 4 non-labeled variable primers (V1-V8; V9; V10; and V11) (see: Greiner et al. Am. J. Pathol. (1995) 146:46-55). Reaction products were analyzed by gel electrophoresis on a 6% denaturing polyacrylamide gel. Using the V1-8 and V9 primers, identical bands were identified from both specimens, consistent with identical biallelic *TCRG* rearrangements indicating a shared clonal population of cells in both specimens. However, the clonal bands are less prominent above a polyclonal background in specimen #1.

Interesting Feature(s) of Submitted Case:
This case is diagnostically challenging and demonstrates the varied clinical and histologic presentation of mycosis fungoides, raises the differential diagnosis of recurrent papules in the setting of mycosis fungoides and highlights the fact that mycosis fungoides may occur with other lymphoproliferative disorders. The patient was initially diagnosed with and underwent treatment for pityriasis lichenoides, a common clinical and histologic mimic of mycosis fungoides. The current biopsy from the hypopigmented patch showed very subtle histologic findings with a scant infiltrate of cytologically banal-appearing lymphocytes, numerous intraepidermal langerhans cells and minimal lymphocytic epidermotropism. The majority of the lymphocytes were CD8+, a common finding in pediatric hypopigmented mycosis fungoides. The presence of numerous intraepidermal langerhans cells is of unclear significance and is not a well-described histologic feature of mycosis fungoides. The second biopsy showed numerous CD8+/CD30- intraepidermal cytologically atypical lymphocytes in a pattern very suggestive of mycosis fungoides. An identical clone was detected in both specimens. Although the papular lesions could represent papular mycosis fungoides within the setting of hypopigmented disease or pityriasis lichenoides, given the clinical presentation and the history of recurrent self-resolving lesions we favor a diagnosis of hypopigmented mycosis fungoides with concomitant type B lymphomatoid papulosis.

Proposed Diagnosis:
CD8+ hypopigmented mycosis fungoides with type B lymphomatoid papulosis in a child.

Panel Diagnosis:
Pediatric mycosis fungoides with CD8+ phenotype.