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Clinical History:
43 year old male who at age 40 presented with 8x4mm firm violaceous plaque on his left arm that had been present for 6 months. Biopsy showed an abnormal lymphoid infiltrate with inconclusive immunohistochemical stains and too little material for molecular studies. The otherwise asymptomatic patient presented again 3½ years later at age 43 with an 8mm bluish red nodule that had been present for two months on the right shoulder ("pseudo-lipoma" vs B-cell lymphoma) – case submitted for review. About 6 weeks later, the patient presented for reexcision of the right shoulder lesion and also had biopsy of new erythematous papule on right posterior shoulder/upper arm. The excision showed some residual lymphoma that was completely excised plus a cicatrix. The biopsy had an infiltrate at its edge considered compatible with the lymphoma but it was not further evaluated. The patient had an unremarkable H/H, LFTs, CMP, and LDH. CT and PET/CT showed no pathologic adenopathy/no evidence of malignancy. There was mild prominence of the palatine tonsils. He returned for follow-up about 2 weeks later and numerous (10-12) barely palpable blue to violaceous macules and papers were noted on the upper back (see clinical image). Two punch biopsies were performed and both showed a similar histologic appearance. Immunohistochemical stains performed on one of the biopsies confirmed the presence of his lymphoma which is presumably what the second biopsy also represented. The patient was subsequently treated with IV Rituximab.

Biopsy Fixation Details:
Right shoulder punch biopsy fixed in formalin

Description of Clinical Image if Any:
Image of patient's back shows the multiple macules/papules when his disease recurred (see clinical history above).

Details of Microscopic Findings:
The histologic sections of the submitted biopsy demonstrate a dense dermal infiltrate with scattered follicles that include some germinal centers and a somewhat pale-appearing polymorphous infiltrate that includes many small lymphocytes with round to irregular nuclei and pale cytoplasm, some transformed cells, histiocytes and foci, often at the periphery of the infiltrate, with many plasma cells.

Immunophenotyping by Immunohistochemistry and/ or Flow Cytometry:
IHC highlights the CD20+ B-cell follicles with their IgM+IgD+CD23+CD21+ mantle zones and CD10-BCL6+BCL2- germinal centers plus scattered CD20+ B-cells. The B-cells appear to be CD5-, CD10-, CD43- and are cyclin D1-. CD3 demonstrates numerous interfollicular positive cells with CD4>CD8. The plasma cells are CD138+, IgG+, IgM-, IgD-, IgA-, Kappa+, Lambda largely – (ie, monotypic IgG Kappa). Scattered CD123+ cells lying singly and in clusters and an occasional loose aggregate are also present. A moderate number of CD68+ cells are also present. Ki-67 shows many positive cells in the germinal centers plus scattered positive cells.

Special Stains:
A treponema IHC stain was negative.

Cytogenetics:
None performed.

Molecular Analysis:
Attempted on recurrent lesion but failed.

Interesting Feature(s) of Submitted Case:
Classic primary cutaneous marginal zone lymphoma – T-cell rich, IgG+ with plasma cells mostly at periphery of lesion, many of the B-cells present are not a part of the neoplasm, common CD10-BCL6+ GC (see Am J Surg Pathol. 2010 Dec;34(12):1830-41 – the submitted case was not included in this study).

Demonstrates different clinical appearances with solitary lesion and also multiple much more subtle lesions (as in submitted clinical image). Highlights how some cutaneous marginal zone lymphomas may be barely detectable. We have reviewed another case, also not a part of the AJSP series, where a recurrent cutaneous marginal zone lymphoma was present in normal-appearing and slightly erythematous skin.

Submitted biopsy has clearcut mass lesion but recurrent lesions had pathologically more subtle disease consistent with more subtle clinical lesions.

Raises question as to how cases with recurrent disease should be treated.

Proposed Diagnosis:
Primary cutaneous marginal zone lymphoma/extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma).

Panel Diagnosis:
Primary cutaneous marginal zone lymphoma.

Comments:
All biopsies can be illustrated.