

An Unusual Presentation of Cutaneous B-Cell Marginal Zone Lymphoma/Lymphoproliferative Disorder and Successful Treatment with Rituximab

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BACKGROUND

- Primary Cutaneous B-Cell Marginal Zone Lymphoma/Lymphoproliferative Disorder (PCMZL/LPD) is the second most common subtype of primary cutaneous B-cell lymphoma (PC-BCL), representing 24-31% of cases, though rare overall.¹ Diagnosis is often challenging, as there is often clinical and histopathologic overlap with atypical reactive B-cell infiltrates.²
- Prognosis is usually excellent (5-year survival rate 99%), though recurrence occurs in 50% of cases.¹

CASE PRESENTATION

- A 67-year-old woman presented with a 2-month history of a pruritic facial eruption refractory to topical corticosteroids, calcineurin inhibitors, and intralesional triamcinolone, with prior biopsy interpreted as cutaneous lymphoid hyperplasia.
- Exam: firm, irregular red-to-violaceous papules, plaques, and nodules involving the medial eyebrows, frontal hairline, right preauricular cheek, nasal tip, left lower eyelid, and left conchal bowl (Figure 1).
- Initial morphology of the eruption was favored to represent cutaneous sarcoidosis. Repeat biopsy instead demonstrated an atypical lymphoid infiltrate involving the deep dermis and subcutis (Figure 2).

DIAGNOSTIC WORK-UP

- Immunohistochemistry: CD20 and BCL2 stained positively in atypical cells; CD10 and BCL6 stained negatively in atypical cells (Figure 3).
- Clonal immunoglobulin heavy-chain gene rearrangement supported a clonal B-cell proliferation.
- PET/CT showed hypermetabolic cutaneous lesions and multi-station bilateral cervical lymphadenopathy, suspicious for nodal involvement.

TREATMENT

- The patient received two four-dose courses of IV rituximab 375 mg/kg, separated by six months.
- The patient had an excellent clinical response, with near-complete resolution of facial papules and erythema (figure 4, figure 5).

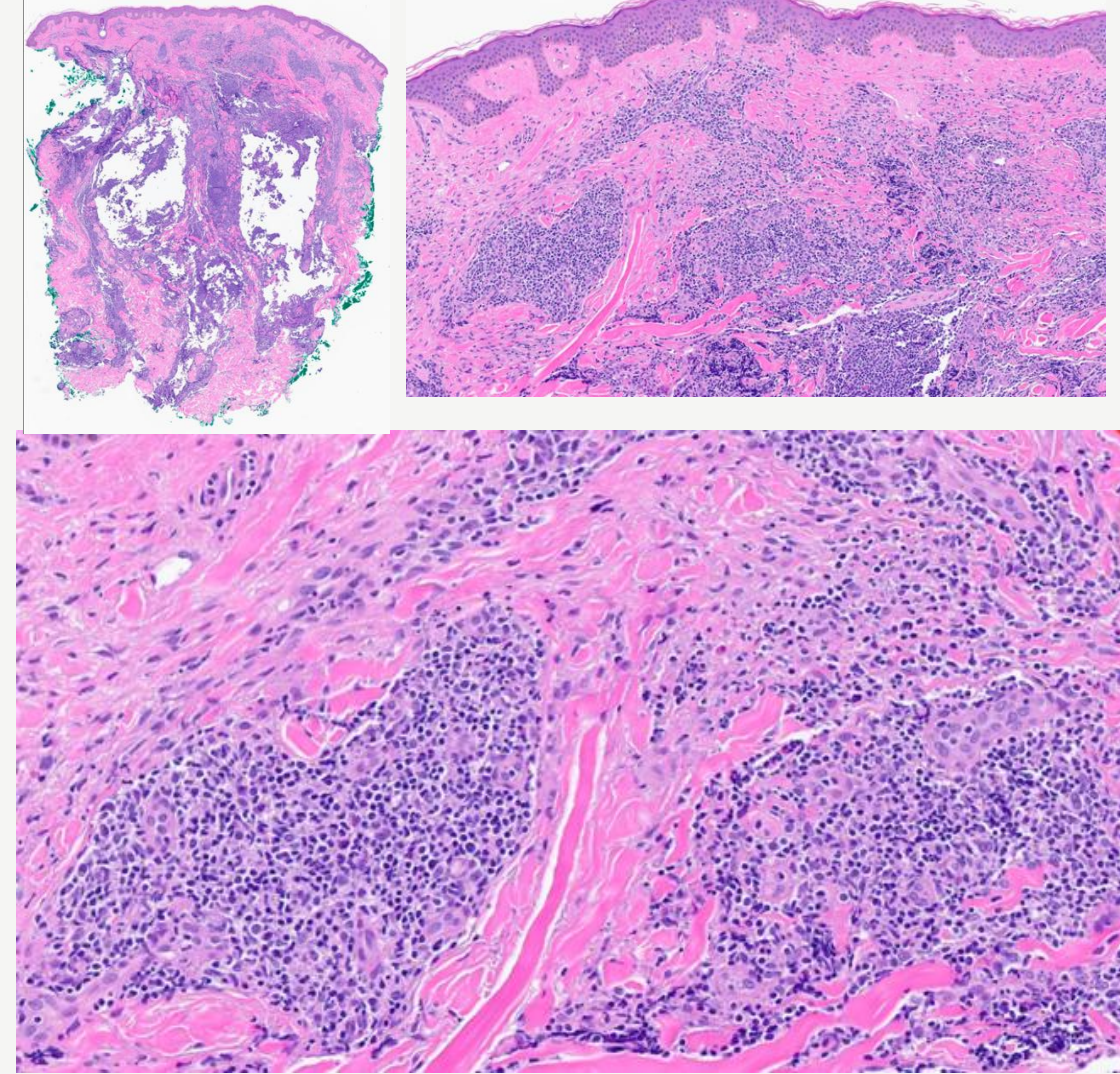
CLINICAL & HISTOPATHOLOGIC FINDINGS

Figure 1. Presentation



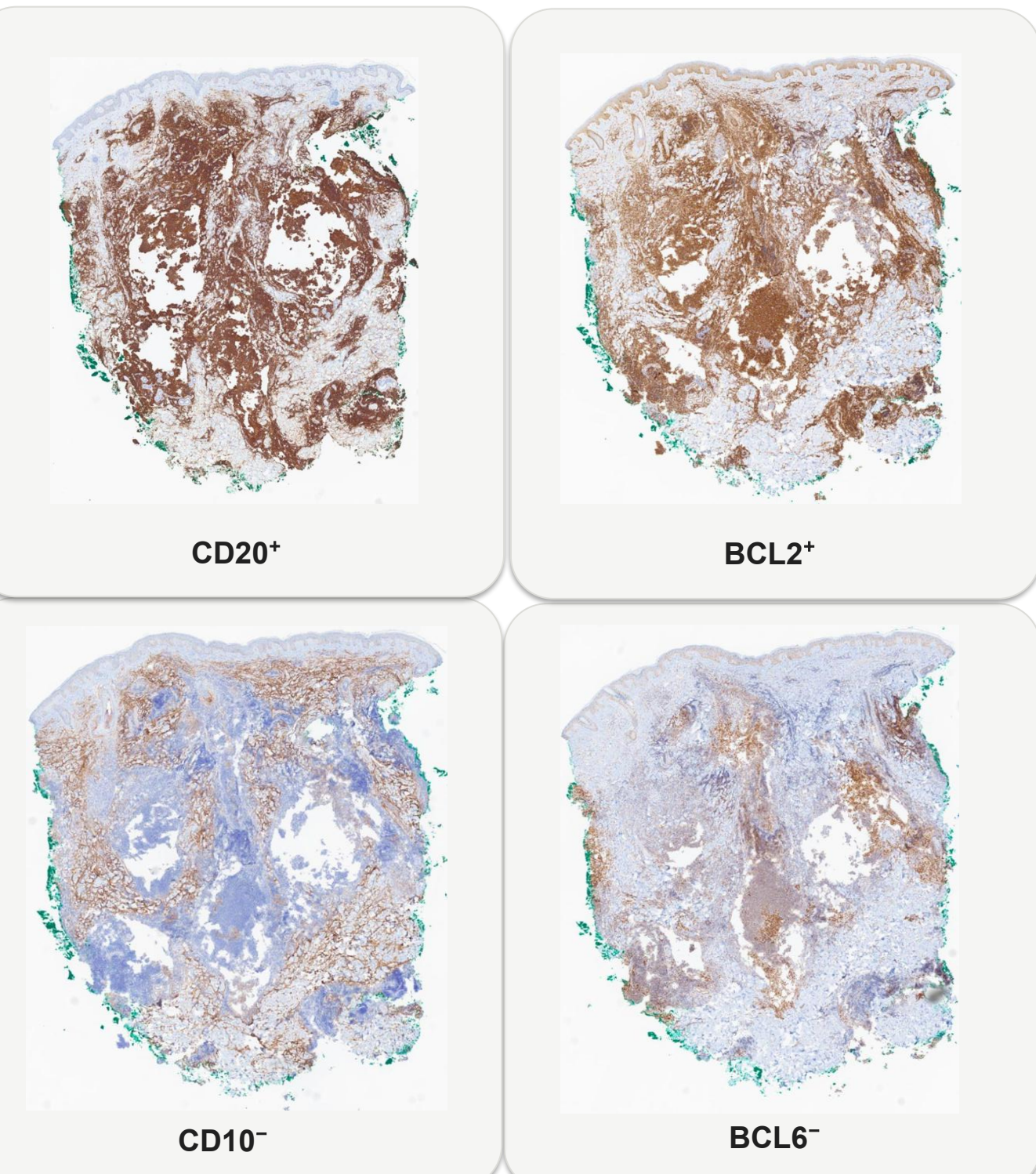
Clinical photographs obtained during initial patient visit demonstrating firm, irregular, red-to-violaceous papules, plaques, and nodules involving the bilateral medial eyebrows, frontal hairline, and nasal tip.

Figure 2. Histopathology H&E



H&E-stained sections demonstrate an atypical lymphoid infiltrate with a vaguely nodular pattern involving the deep dermis and subcutaneous tissue.

Figure 3. Immunohistochemistry

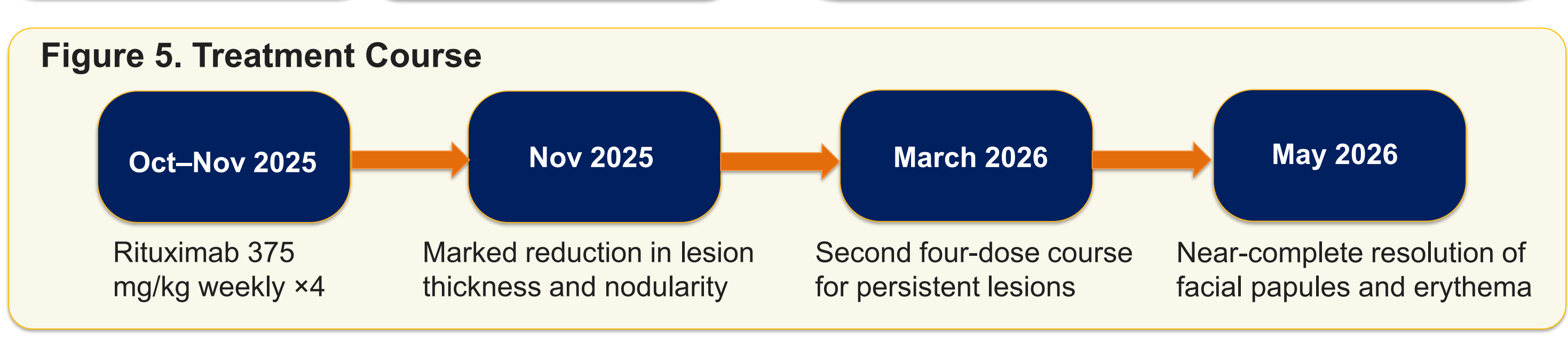


CD20⁺ BCL2⁺
CD10⁻ BCL6⁻

Figure 4. Response to Rituximab



Left: November 2025, demonstrating marked reduction in lesion thickness and nodularity. **Right:** March 2026, demonstrating sustained improvement with slight residual erythema.



DISCUSSION

- This case demonstrated diagnostic uncertainty, with the morphology of the facial eruption initially favored to represent sarcoidosis and prior outside biopsy interpreted as cutaneous lymphoid hyperplasia.
- Repeat histopathologic evaluation, immunophenotyping, and gene clonality studies established a clonal B-cell process consistent with PCMZL/LPD.
- Staging identified cervical lymphadenopathy suspicious for nodal involvement, prompting systemic treatment with rituximab. The patient achieved near-complete clinical resolution, consistent with prior reports demonstrating treatment success with rituximab.³
- Other reported therapies include skin-directed therapies, intralesional steroids, local involved-site radiation therapy (ISRT), or other combination chemotherapy regimens for extensive/refractory disease.¹

KEY TAKEAWAYS

- PCMZL/LPD can be difficult to diagnose and multiple biopsies may be needed to establish diagnosis.
- Clinicopathologic correlation was essential after an initial outside biopsy was interpreted as cutaneous lymphoid hyperplasia.
- Rituximab was associated with substantial clinical improvement in this patient and may be considered in patients with PCMZL/LPD and evidence of generalized or extracutaneous disease.

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