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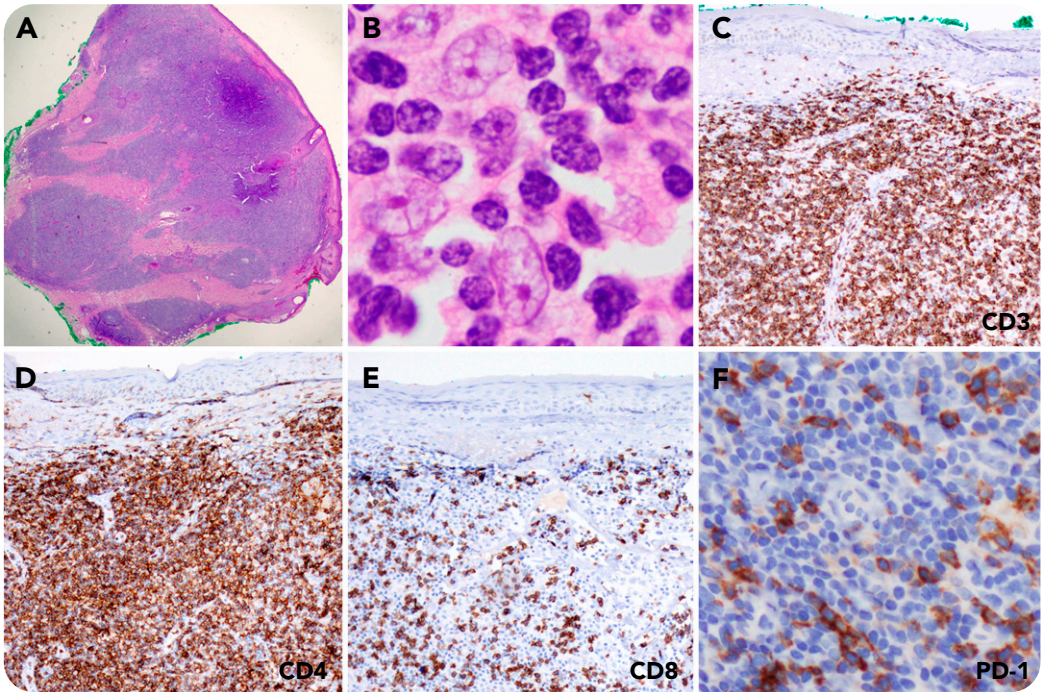
Primary cutaneous small/medium CD4$^+$ T-cell lymphoproliferative disorder

Jing Xu and Qin Huang, Cedars-Sinai Medical Center



A 51-year-old man presented with a solitary pink raised lesion (0.8 cm) on his right temporal scalp. No lymphadenopathy or other skin lesions were identified. Biopsy demonstrated a dense, nodular lymphoid infiltrate in the dermis (panels A-B; hematoxylin and eosin stain, original magnification ×4 [A], ×100 [B]). The infiltrates were predominantly small- to medium-sized CD4$^+$ pleomorphic T cells, admixed with CD8$^+$ T cells, CD20$^+$ B cells, and histiocytes (panels C-E; immunohistochemistry stain, original magnification ×10). Many CD4$^+$ T cells coexpressed PD-1, BCL-6, and CXCL-13 proteins (panel F; immunohistochemistry stain, original magnification ×20), indicating T-follicular helper (TFH) cell derivation (panel F). The clinical presentation, TFH cellular origin, and absence of prominent epidermotropism distinguished this lesion from mycosis fungoides. The Ki-67 index was low. Polymerase chain reaction demonstrated T-cell receptor β/γ gene rearrangements. Thus, primary cutaneous small/medium CD4$^+$ T-cell lymphoproliferative disorder was diagnosed.

This clinical lesion was initially introduced by 2008 World Health Organization classification as a provisional entity of primary cutaneous small/medium T-cell lymphoma. Because of its indolent behavior, the entity was modified in 2017 to primary cutaneous small/medium-sized T-cell lymphoproliferative disorder. As a cutaneous clonal proliferation of TFH cells, the lesion usually presents as a solitary plaque or nodule on the patient's face, neck, or upper trunk. The prognosis is very favorable with treatment modalities such as intraregional steroids, excision, or radiotherapy when the clinical presentation is localized or solitary.

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