Primary cutaneous CD4+ small to medium T-cell lymphoproliferative disorder in a 50-year-old man: A case report

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Abstract

Primary cutaneous CD4+ small to medium T-cell lymphoproliferative disorder is rare, accounting for only 2-3% of all primary cutaneous lymphomas. In this report, we describe the case of a 50-year-old man with an asymptomatic, small, round nodule on his temple area.

Introduction

Primary cutaneous CD4+ small to medium T-cell lymphoproliferative disorder is rare, accounting for only 2-3% of all primary cutaneous lymphomas. In 2006, the World Health Organization (WHO) revised the name of this condition from primary cutaneous CD4+ small to medium pleomorphic T-cell lymphoma to its current name to indicate its uncertain malignant potential. The disease is indolent and localized, rarely showing systemic involvement (1-3). The typical lesion is a solitary skin-colored papule or nodule involving the head or neck; it usually grows rapidly but remains asymptomatic, with most patients being in their 50s-60s (4-7). Histopathological findings show dense infiltration of CD4+ small or medium T-cell lymphocytes in the dermis or subcutis area (1, 2). Suggested treatments include an excisional biopsy, intralesional steroid injection, or radiotherapy; the prognosis is excellent (4, 7, 8). Herein, we report a rare case of primary cutaneous CD4+ small/medium T-cell lymphoproliferative disorder.

Case report

A 50-year-old man visited the outpatient dermatology clinic of Faghihi Hospital (Shiraz, Iran) in December 2021 with the chief complaint of a rapidly growing lesion on his temple area. The patient had noticed a small round lesion that appeared abruptly, with rapid growth during the last two months. There was no sign or symptom of pain, tenderness, pruritis, bleeding, discharge, or hypoesthesia. Skin examination revealed a solitary, round, firm, 1×1.2 cm erythematous nodule with a sharp, well-defined border on the patient's temporal area (Fig. 1). Neither lymphadenopathy nor any other systemic symptoms were found in the general examination. The patient had no specific history of surgery, rheumatoid arthritis, malignancy, or any other disease or medication usage. There was also no family history of similar cutaneous lesions or other diseases.

An excisional biopsy with a safety margin of 0.5 mm was taken under local anesthesia. The specimen was fixed in formalin and studied by an expert dermatopathologist. The histopathologic study revealed thinning of the epidermis with a dense partly diffuse partly nodular infiltration of small to medium-sized atypical

lymphocytes and a few large atypical lymphocytes, plasma cells, and histiocytes scattered in the entire dermis, extending to the subcutaneous fat. The infiltrate was separated from the dermis by the grenz zone (Fig. 2).

Immunohistochemical studies revealed that 90% of the lymphoid cells were CD4+, a few CD8+, and the majority of those cells were positive for CD3, CD5, and CD7, but negative for CD30, CD20, BCL6, CD10, PAX 5, and MUM1. Ki67 was 10% (Table 1) (Fig. 3). The final diagnosis was primary cutaneous CD4+ small to medium T- cell lymphoproliferative disorder. Further laboratory tests were normal, including a complete blood count and lactate dehydrogenase assay. The biopsy specimens were reviewed by a dermatopathologist, who confirmed the mentioned findings. No significant signs of recurrence were observed over two months of follow-up.

Discussion

Primary cutaneous CD4+ small to medium T-cell lymphoproliferative disorder is a sporadic disease that typically affects individuals aged 50-70, though it can also occur during childhood and even infancy (3, 7, 9). The typical lesion is an asymptomatic, rapidly growing, solitary nodule or plaque in the face, neck, or upper extremities (1, 7, 10, 11). Our patient's age, the shape of the lesion, and the area of involvement were compatible with previous studies. The greater prevalence of this disease in the elderly and also its greater presentation on sun-exposed areas can raise the suspicion of the effective role of sun exposure as a risk factor for this disease. Out histological findings were compatible with the literature, where most publications described the infiltration of CD4+, small to medium-sized T-cells in the dermis, with some infiltration of B-cells, plasma cells, and histiocytes (1, 10).

Most cases need surgical or medical interventions to be cured, but there are two case reports of spontaneous complete remissions. A case was also reported to regress three months after a local biopsy (10-12). For patients with localized lesions, surgical excision, intralesional steroids, or radiotherapy is recommended. Some studies revealed the effective role of cyclophosphamide or interferon-alpha for multiple or generalized skin involvement (4, 6-8, 13). Even though there are multiple choices for treatment, most studies consider surgical excision as the best choice for solitary lesions due to desirable outcomes, minimal complications, inexpensiveness, and rare recurrences. In our study, an excisional biopsy with a safety margin of 0.5 mm taken under local anesthesia yielded an excellent outcome.

In the literature, recurrence has been reported in 4.26% of cases. Most individuals showed no recurrence during 10-26 months of follow-up. A rare local recurrence was reported in the second month of follow-up in a patient who underwent local excision. In another patient, the progression of multiple cutaneous lesions was reported despite systemic chemotherapy and local excision (7, 12). Like most documented cases, no signs and symptoms of relapse were detected in our patient after two months of follow-up.

Patients with CD4+ small to medium-sized T-cell lymphoproliferative disorder rarely progress toward malignancies in the course of the disease. Recurrence and generalized involvement are also scarce. Even though novel studies have shown the uncertain potential of malignancy of the disease and indicated an excellent prognosis, many studies have reported a five-year survival rate of approximately 60-80% (1, 2, 7, 10, 13). The incompatibility between disease prognosis and five-year survival may be due to the use of the previous classification of disease (cutaneous CD4+ small to medium pleomorphic T-cell lymphoma) in most publications. This incompatibility may be resolved using updated criteria of classification.

The very low prevalence of CD4+ small to medium-sized T-cell lymphoproliferative disorder has greatly reduced its clinical suspicion, such that it may be overlooked in the list of differential diagnoses for rapidly growing lesions. It is important to remember that although the lesions appear abruptly and grow rapidly, they are not malignant and can be cured with simple, cheap, non- invasive treatments. Here, we reported this disease as a rare subtype of cutaneous T-cell lymphomas in a 50-year-old man to remind all physicians to keep this entity in mind and avoid unnecessary aggressive interventions.

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Conflict of interest

The authors declare no conflicts of interest.

Data availability statement

Data are available upon request to the corresponding author,

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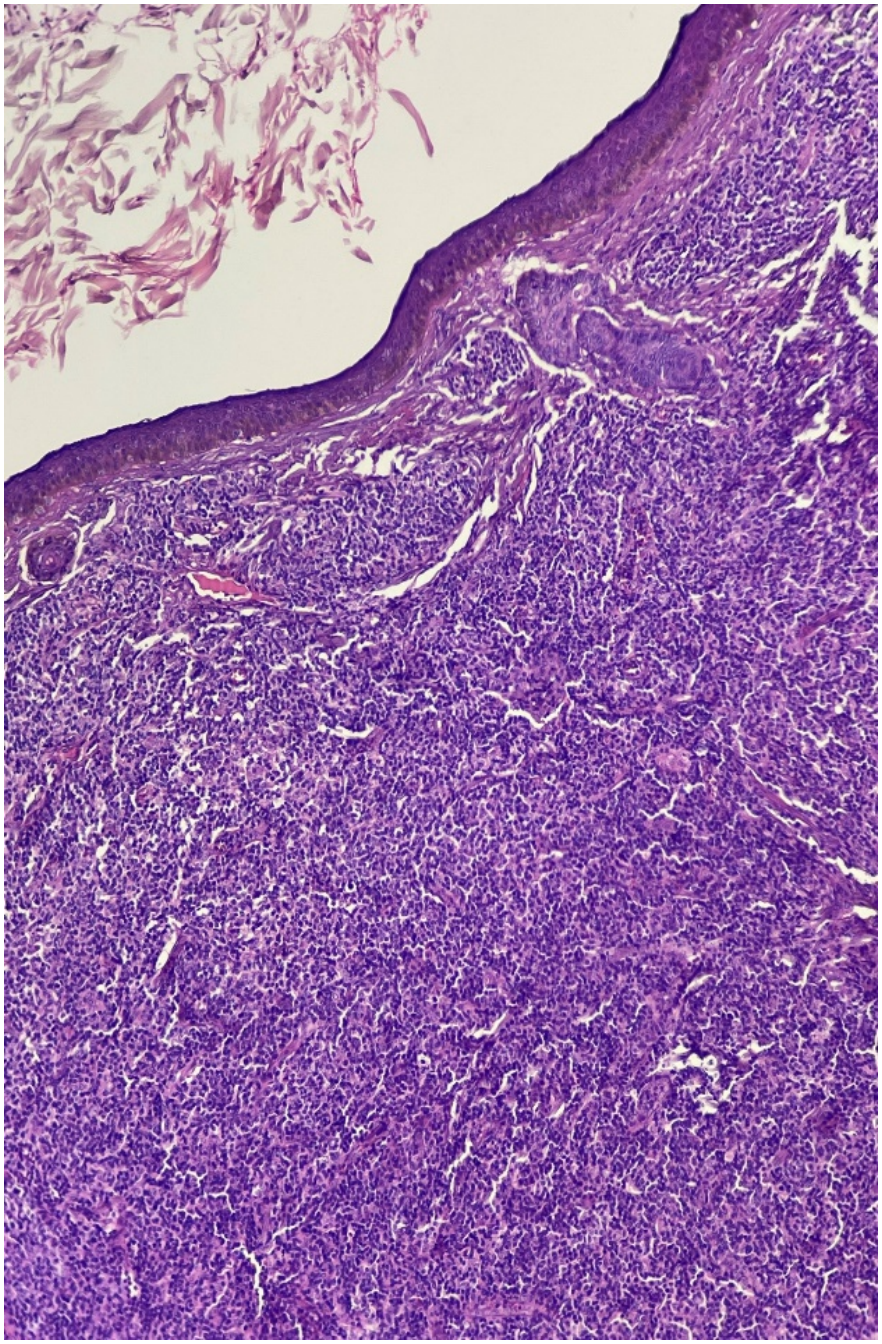
Table 1. Immunohistochemistry results

Marker	**Result**	**Marker**	**Result**
CD3	Positive	BCL2	Positive
CD5	Positive	BCL6	Negative
CD7	Positive	CD10	Negative
CD30	Negative	PAX5	Negative
Ki67	10%	MUM1	Negative
CD20	Negative	CD4	Positive (90%)
CD8	Positive (10%)		

Fig. 1. The patient's initial lesion: a skin-colored nodule on the temple area.



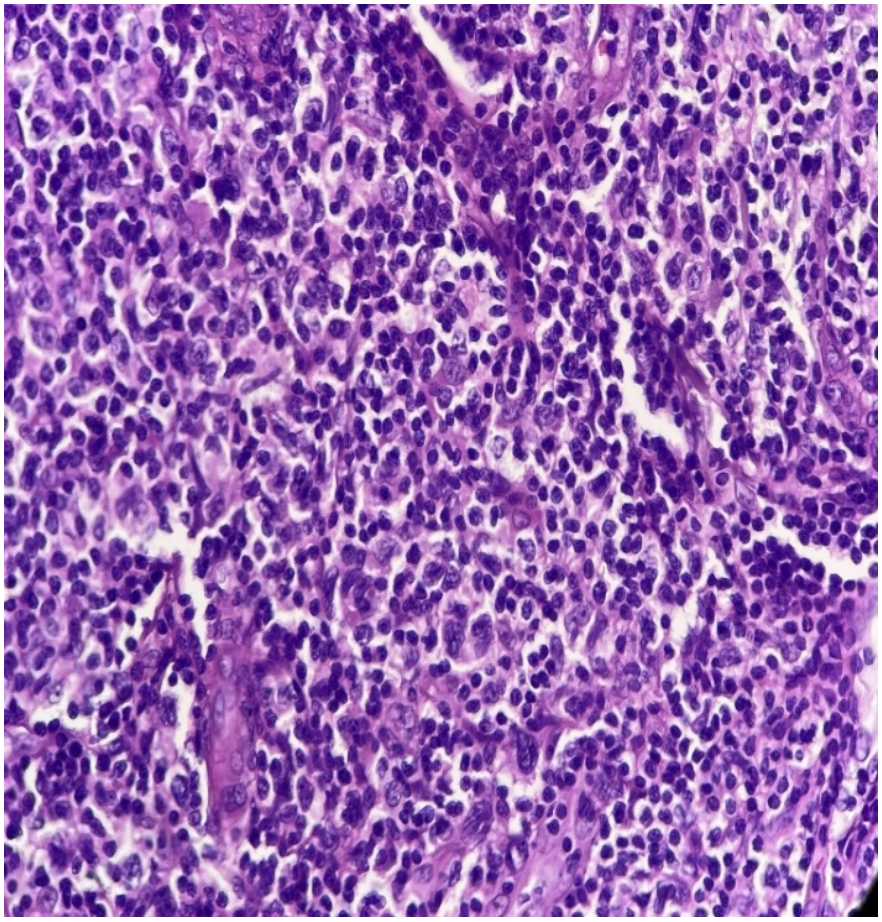


Fig. 2. There is thinning of the epidermis with a dense infiltration of small to medium sized atypical lymphocytes and a few large atypical lymphocytes, plasma cells, and histiocytes scattered in the entire dermis. The infiltrate is separated from the epidermis by a grenz zone (H&Ex100 & 400).

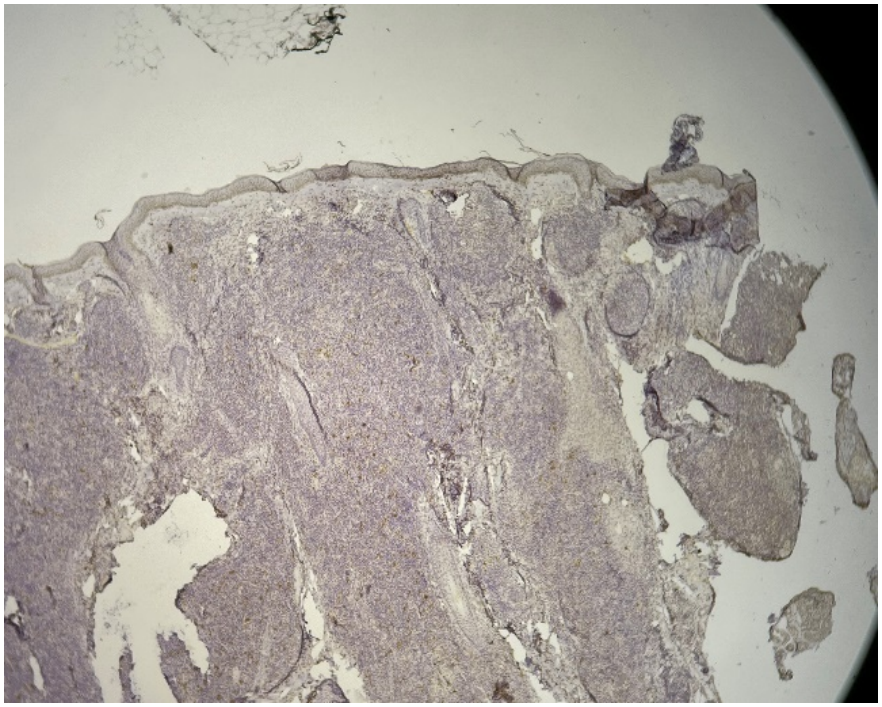


Fig. 3. Immunohistochemically, majority of the lymphocytes expressed CD3, CD5, & CD4, (A, B, C); a few CD8+ cells, low Ki67 (10%), & CD30 negativity (D,E,F) (x40).

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