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Unusual presentation of skin nodule in a child: a case report of primary cutaneous CD4$^+$ small/medium T-cell lymphoproliferative disorder and literature review

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Consent for publication: informed consent was given by the patient's father for the publication of this case report and any accompanying images.

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Abstract

Primary cutaneous $CD4^+$ small/medium T-cell lymphoproliferative disorder (SMPLPD) is a rare, definite type of cutaneous T-cell lymphoma (CTCL) that accounts for 6% of all primary CTCLs. Characteristically, this lymphoproliferative disorder presents as a solitary nodule, plaque, or tumor, generally on the face, neck, or upper trunk. We report a case of SMPLPD presenting as a facial nodule in a 6-year-old Saudi girl. The patient was successfully treated with complete excision and showed full resolution, with no recurrence at 6-month follow-up.

Introduction

Primary cutaneous T-cell lymphomas (CTCLs) are a rare heterogeneous group of cancers characterized by the clonal proliferation of malignant T lymphocytes. For decades, primary cutaneous $CD4^+$ small/medium T-cell lymphoproliferative disorder (SMPLPD) has been considered a provisional CTCL entity.[1] In the 5th Edition of the World Health Organization Classification of Hematolymphoid Tumors: Lymphoid Neoplasm (WHO-HAEM5), SMPLPD was upgraded to a definite entity.[2] Clinically, SMPLPD is characterized by solitary erythematous papules or nodules on the head, upper extremities, or upper trunk. Its histological features include small-to-medium-sized CD4+ lymphocytes in the dermis, with a nodular or diffuse pattern. Proliferative cells include reactive B cells, plasma cells, eosinophils, and histiocytes.[1] This case report describes a 6-year-old Saudi girl with primary cutaneous $CD4^+$ SMPLPD.

Case Report

A 6-year-old Saudi girl presented to our dermatology clinic at King Fahad Specialist Hospital in Dammam with a 3-month history of an erythematous nodule on the left cheek. The lesion began as a single, painless, nonpruritic pink papule that grew slowly for 3 months. She had no discharge, bleeding, trauma, or ulceration of the nodule. She had no history of constitutional symptoms such as fever, night sweats, weight loss, appetite loss, or generalized body pruritus.

On examination, the lesion was a single, well-demarcated, shiny, erythematous nodule with telangiectasia and a firm consistency. The nodule, measuring 1.0×0.8 cm in diameter, had no ulceration and tenderness (Figure 1). General examination revealed no other skin abnormalities, hepatosplenomegaly, or lymphadenopathy.

Histopathological examination of the skin biopsy specimen revealed extensive and dense dermal infiltrates mainly composed of small-to-intermediate-sized lymphocytes (Figure 2A). Plasma cells and histiocytes were also observed in the background. The epidermis showed mild spongiosis with no significant lymphocytic epidermotropism. Immunostaining was performed using the appropriate

controls. The lymphocytic infiltrate was composed of small-to-intermediate-sized lymphocytes highlighted by CD3 and CD4 (Figure 2 B,C). Medium-to-large-sized atypical T cells expressed programmed death 1 (PD-1) (Figure 2D).

Additional workup was performed, including a complete blood count; liver and renal function tests; a lactate dehydrogenase level test; a peripheral blood smear analysis; chest radiography; computed tomography of the chest, abdomen, and pelvis; and positron emission tomography, with normal and unremarkable results.

The clinical and histopathological findings confirmed the diagnosis of primary cutaneous $CD4^+$ SMPLPD. Thus, complete surgical excision of the nodule on the patient's left cheek was performed under general anesthesia by a plastic surgeon. No complications and recurrences were found within a 6-month follow-up period.

Discussion

Primary cutaneous $CD4^+$ SMPLPD is a definite and rare entity of CTCL, accounting for 6% of all primary CTCLs. The disease has a favorable course and prognosis, with an estimated 5-year survival rate of 100%. However, the potential for malignancy remains unknown; hence, the disease has been reclassified as a lymphoproliferative disorder rather than a lymphoma.[1,2] We identified 15 case reports, 7 case series, and 3 systematic reviews, which are summarized in Table 1.[3-17]

A literature review suggested that most cases exhibited a solitary, well-defined, asymptomatic erythematous nodule, and only one case presented with multiple papules and plaques.[17] Most lesions occurred on the face, especially on the forehead and cheeks. However, in some cases, the lesion was observed on the chest, abdomen, or lower limb. The lesions were associated with mild symptoms such as pain and mild pruritus. The median age of onset was approximately 54 years, with most cases occurring in male patients. In our case, the patient had a slightly unusual age of occurrence, which was 6 years of age.

Typically, histopathological features include diffuse and dense dermal infiltration of atypical small- to medium-sized pleomorphic T lymphocytes, along with histiocytes, plasma cells, and B cells, without epidermotropism. Immunohistochemical staining showed $CD3^+$, $CD4^+$, PD-1^+, and $CD8^-$ T-cell phenotypes. Some patients tested positive for Bcl-6. The rate of proliferation, characterized by Ki-67, ranged from 10% to 30%.[18]

All reported cases showed no extracutaneous manifestations, with most cases exhibiting normal results in laboratory tests, including complete blood count, liver and renal function tests, blood glucose analysis, and measurement of inflammatory markers. Radiological workups, including chest radiography, computed tomography, and positron emission tomography, revealed no abnormalities.

However, one case exhibited bilaterally enlarged jugulodigastric lymph nodes, which showed no neoplastic cells on biopsy.[11]

No consensus has been reached regarding the optimal treatment for primary cutaneous $CD4^+$ SMPLPDs. The available treatment options include surgical excision, intralesional corticosteroid therapy, laser CO_2 ablation, and localized radiotherapy.[4,19] Most patients who undergo surgical excision demonstrate complete resolution of the lesion, as in our case.[19] Finally, to initiate proper and adequate treatment of the skin nodule, high clinical suspicion and a wide range of disorders in the differential diagnosis are needed.

Conclusions

This case report describes an unusual presentation of SMPLPD in a 6-year-old girl that was managed successfully with surgical excision and showed no relapse at 6-month follow-up.

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Figure 1. A firm, shiny, telangiectatic erythematous nodule on the patient's left cheek.

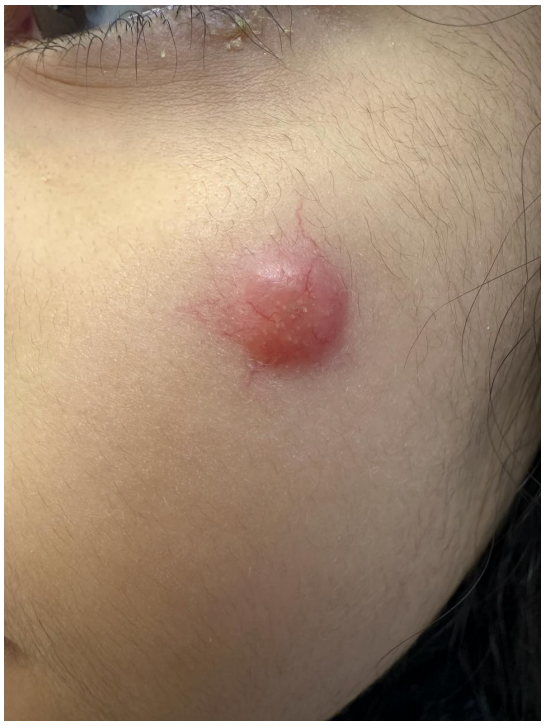

Figure 2. (**A**) (400×) Small-to-medium-sized lymphoid cells with mild pleomorphism infiltrating the deep dermis on high-power resolution (hematoxylin and eosin staining). Immunohistochemical stains show lymphoid cells expressing CD4 (**B**), CD3 (**C**). The tumor cells are also positive for programmed death-1 (PD-1) (**D**).

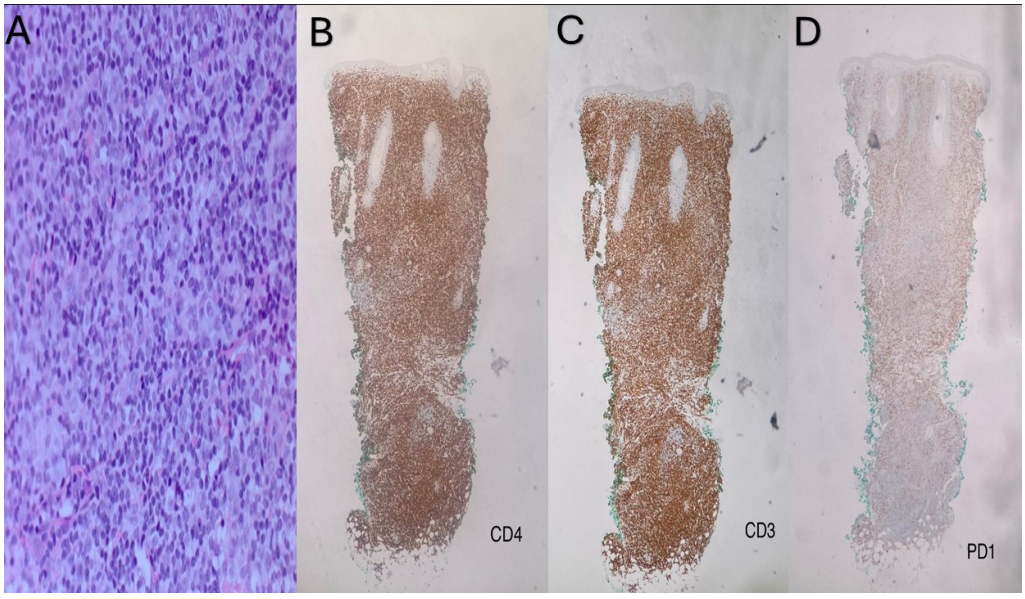

Table 1. Summary of the clinical data of reported SMPLPD cases.

Reference	Age at onset	Sex	Clinical presentation	Location of the lesion	Initial treatment	Relapse
Concilla *et al.*[3]	39 years	Female	Solitary, erythematous, and itchy nodule	Left nasal ala	Topical corticosteroids	None
Zengarini *et al.*[4]	83 years	Male	Violaceous, painless nodule	Neck	Laser CO_2 ablation	None
Kazan *et al.*[5]	64 years	Male	Erythematous patches and papules	Left medial thigh	Radiotherapy	None
Kim *et al.*[6]	51 years	Male	Protruding red mass	Forehead	Surgical excision	None
Valentim *et al.*[7]	36 years	Female	Solitary erythematous nodule with telangiectasia	Left nasal ala	Potent occlusive steroid	None
Jain *et al.*[8]	79 years	Male	Gradually growing ulcerated nodule	Forehead	None	NA
Koper *et al.*[9]	55 years	Male	Single large tumor	Lower extremity	Cyclophosphamide, methotrexate, and radiation	2 weeks after treatment
Keeling *et al.*[10]	47 years	Male	Solitary nodule	Right chest	N/A	NA
Micković *et al.*[11]	30 years	Male	Asymptomatic tumor	Forehead	None	NA
Topal *et al.*[12]	58 years	Female	Solitary, firm, and indurated tumor	Left preauricular area	Radiation therapy	None
Li *et al.*[13]	6 months	Male	Solitary nodule	Left cheek	Surgical excision	None

Volks *et al.*[14]	12 years	Female	Mildly pruritic swelling	Right supraorbital area	Intralesional triamcinolone acetonide	None
Zhang *et al.*[15]	44 years	Female	Solitary papule	Left cheek	Local radiotherapy (total dose of 36 Gy)	None
Toberer *et al.*[16]	21 years	Female	Slowly growing tumor	Right cheek	Oral doxycycline monohydrate	13 months after treatment
Choi *et al*.[17]	63 years	Female	Multiple erythematous nodules and plaques	Face, abdomen, and lower limb	Local radiotherapy (total dose of 45 Gy)	1 month after treatment