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Case report Open Access

Primary Cutaneous CD4+ Small/Medium T-cell Lymphoproliferative Disease: A Case Report

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Abstract

Primary cutaneous lymphoma is a heterogeneous group of diseases. Primary cutaneous CD4+ small/medium T-cell lymphoma (CD4+ SMTCL) is one of the rare subtypes; representing only 2-3% of all cutaneous lymphomas. It was recently described as lymphoproliferative disorder because of its indolent clinical behavior and many similarities with cutaneous pseudolymphoma. This disease is a provisional entity according to latest World Health Organization (WHO) classification of hematolymphoid tumors. CD4+ SMTCL has a n excellent 5-year survival of 60-85%. CD4+ SMTCL usually present with a solitary skin lesion of the head and neck region, and most of reported cases were treated with local managements only. The common clinical presentations and histological features are still not well understood, and no optimal therapy is established. We report a case of primary cutaneous CD4+ small/medium T-cell lymphoma, our patient presented with a facial skin lesion that was treated with excision only.

Keywords: Primary cutaneous lymphoma; Skin tumor; T cell; Small/medium T cell; CD4 cell cutaneous lymphoma

Introduction

Primary cutaneous lymphoma is a group of heterogeneous clinicopathological entities that describe lymphomas that involve the skin primarily without evidence of extracutaneous disease at diagnosis [1]. Different classification systems can be used to describe and classify the diverse entities of primary cutaneous lymphoma. The most important classification system is the World Health Organization-European Organization for Research and Treatments of Cancer classification (WHO-EORTC) published in 2005, the 2008 WHO classification of tumors of hematopoietic and lymphoid tissue and the revision of the later in 2016 [2,3].

The most common types of primary cutaneous T cell lymphomas are mycosis fungoids (MF), Sezary syndrome and CD30+ primary cutaneous lymphoma [3].

One of the rare and controversial types is CD4+ small/medium cell T-cell cutaneous lymphoma (CD4+ SMTCL) which is considered a provisional entity according to the latest WHO classification, it accounts for 2-3% of all cutaneous T cell lymphoma. CD4+ SMTCL is composed of predominantly small to medium sized CD4-antigen positive cells that lack features of mycosis fungoides. CD4+ SMTCL has an indolent course of controversial clinical significance, and because of that the preferred term is lymphoprolifrative disorder instead of lymphoma [4]. CD4+ SMTCL manifests most commonly with a solitary skin lesion of head and neck region or the upper extremities, with minimal local symptoms [5]. We report a 56-year-old gentleman with the diagnosis of CD4+ SMTCL.

Case

A 56-year-old male patient with no significant past medical history was referred by the dermatology service to our plastic surgery clinic with a facial skin lesion, the lesion was noticed by the patient 1 week prior to his visit. The patient reported itching with no bleeding or discharge. No similar lesions were noticed.

No significant weight loss or night sweets were reported. There was no family history of malignancies.

At presentation, a 1-cm, brownish, oval skin lesion on the right side of the forehead, 2 cm away from hair line was identified with regular borders. The adjacent skin was normal with no scaling or ulcerations. The mass lesion was indurated and located deep to the skin with no tenderness and no fixation to the underlying tissues. No lymphadenopathy was identified. An excisional biopsy was performed. Histopathological examination of the lesion revealed a vaguely nodular infiltration of the dermis with small to medium size lymphocyte with moderately clear cytoplasm, oval-to-irregular nuclei, with dense chromatin and indistinct nucleoli as shown in Figure 1. A few large atypical cells were also noted. No epidermotropism was seen. A background of reactive lymphocytes and histiocytes was noted. A panel of immunohistochemical stains was employed to further characterize the tumor. The tumor cells were positive for CD4 (figure 2), CD3, CD7, CD2, and CD5 while negative for CD8, CD30, CD63, CD20, TDT and CD56 as shown in Figures 3 and 4. Final histopathological diagnosis was primary cutaneous T-cell lymphoma with features of CD4+ small/medium T- cell lymphoma.

Discussion

CD4+ SMTCL is a rare entity representing 2-3% of all cutaneous T cell lymphoma. Virmani et al. described 22 cases with features of CD4+ SMTCL; the median follow up period was 32 months, data were collected between January 1981 and August 2015 [5,6]. Local management included surgical excision, radiotherapy or intralesional steroids was sufficient with no recurrence in 18 patients. Only 4 patients had a recurrence of skin lesions [6].

Another series showed similar results with local management alone [5] Toberer report a case of CD4+ SMTCL that achieved a remission

with a course of doxycycline; others used antimetbolites such as cyclophosphamaide with similar results [7,8].

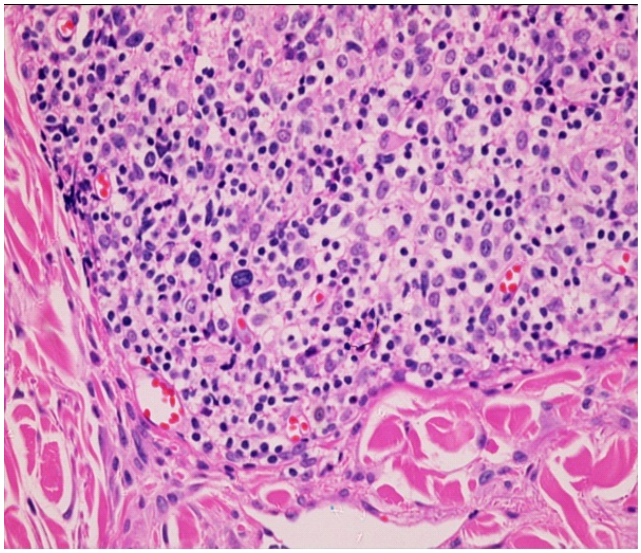

Figure 1: Diffuse Lymphocytic Infiltrates containing Small and medium sized cells, Tumor cells showing significant atypia (Hematoxylin and eosin, X100).

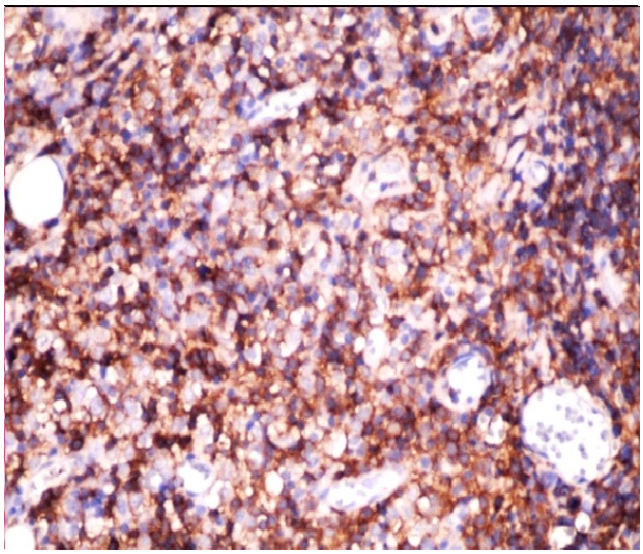

Figure 2: Immunohistochemistry of the tumor cells, strongly positive for CD4.

CD4+ SMTCL is described histologically as an intradermal infiltrate with predominant small to medium sized CD4+ T-cells without features of Mycosis Fungoides [3]. Dense nodular to diffuse dermal infiltrate with infiltration of subcutaneous zones is typical.

The cells are predominatly small/medium sized T cells, scattered large atypical cells are noted, but, by definition should be less than 30%. The background cells include small reactive CD8+ cells, B cells, and histocytes [9]. Immunophenotypically, CD4+ SMTCL is CD3+, CD4+, CD8-, and CD30-; similar to idiopathic T cell pseudolymphoma. Pan-T cell markers (CD7 and to a lesser extent CD5) loss is uncommon and no cytotoxic proteins are expressed [10].

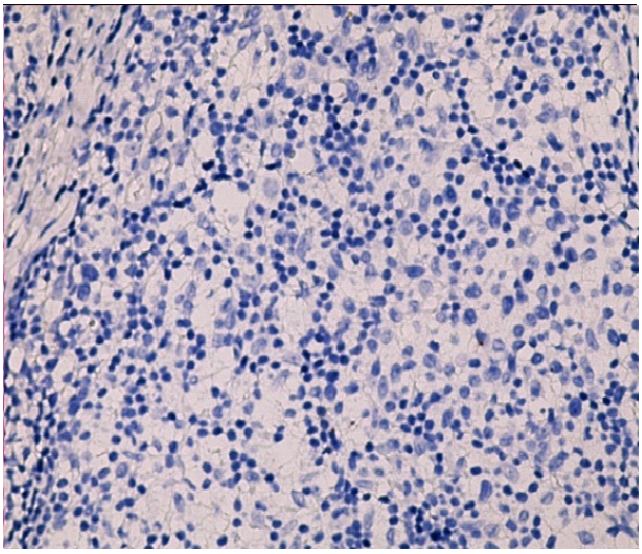

Figure 3: Immunohistochemistry of the tumor cells negative for CD30.

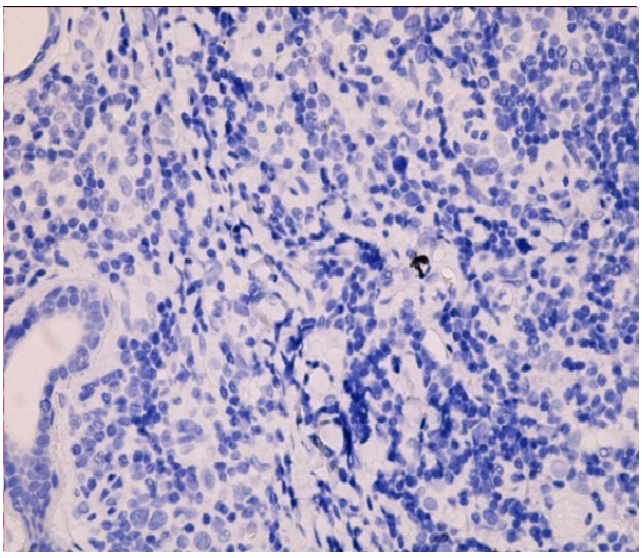

Figure 4: The tumor cells are negative for CD8.

The proliferation rate is low between 5-20%. Epstein-Barr Virus testing is negative as in our case. Genetic features of *TCR* genes rearrangement are clonal in most cases [10].

Diagnosis of CD4+ SMTCL is based on clinical presentation with histopathological and immunophenotypic analysis [9,10] . Excisional or incisional biopsies can be used depending on the size of the lesion. Excision of lesions can be sufficient for CD4+ SMTCL, re-excision or intra-lesional steroids can be used if lesions persist. Rarely,

radiotherapy can be part of the treatment [5]. Local recurrence is rare, and patients have an excellent long term prognosis with a 5-year survival of 60-80% [6,11]. As it can be part of a systemic disease, treating physicians must be aware of the possibility of being part of systemic disease that needs further managements [12].

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