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Case Report

Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma in the practice of a dermatologist: A case report

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ABSTRACT

The article demonstrates a case of diagnosis of a rare variant of primary skin lymphoma, namely of primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma with an indolent course. The diagnosis of SMPTCL is based on the combination of the clinical picture, histological and immunomorphological data. A 70-year-old patient presented at the clinic with complaints about tumor-like formations on the skin of the face and back accompanied by moderate itching. The diagnosis was verified by pathomorphological and immunohistochemical studies of the biopsy of the affected skin. The presented clinical case emphasizes the importance of doctors' clinical oncological alertness and the need for clinical and laboratory examination of the patient using modern histological and immunohistochemical methods for studying the skin biopsy.

Key words: Primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma, disease pattern, pathomorphological and immunohistochemical studies

INTRODUCTION

According to the WHO-EORTC classification, primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma (SMPTCL) is a subtype of primary cutaneous peripheral T-cell lymphoma with an indolent course. The disease occurs more often in adults. The clinical picture is characterized by the presence of single plaques or tumors which are usually located on the face and neck or upper half of the trunk. Therefore, SMPTCL must be differentiated from primary cutaneous B-cell lymphoma, cutaneous anaplastic CD30+ large-cell lymphoma, tumor stage of mycosis fungoides, pseudolymphoma, which results from differences in therapeutic approaches and prognoses of the diseases [1 - 3].

We describe a clinical case of common clinical manifestations and pathomorphological diagnosis of primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma.

CASE REPORT

A 70-year-old patient B. presented at the clinic of the Ural Research Institute of Dermatovenerology and Immunopathology with complaints about tumor-like formations on the skin of the face and back accompanied by moderate itching. He had been ill for 5 years noting a gradual increase in the size of the neoplasm on the skin.

The general condition of the patient was satisfactory. No pathological changes were found in the internal organs.

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Status localis: Rounded tumor-like formations are localized on the skin of the parietal region, forehead, temples and cheeks. These formations represent tuberous-surfaced nodes and plaques, having sizes from 1.5 to 7 cm, densely elastic consistency, and clear boundaries with pink, pale-red and cyanotic shade, without peeling; they are painless on palpation. No excoriation is observed on the surface of the elements (Figs. 1a – 1b). Regional lymph nodes are enlarged, they have soft-elastic consistency and are mildly painful on palpation. Polymorphism of the clinical manifestations in the patient resulted in differential exclusion with the following diagnoses: Primary cutaneous lymphoma? Pseudolymphoma? Paraneoplasia?

No abnormalities are revealed in the clinical blood analysis, clinical urine test, and biochemical blood test. Blood tests for viral hepatitis, HIV and a complex of serological reactions to *Treponema pallidum* are negative.

Pathomorphological study of the skin biopsy. Hyperkeratosis, acanthosis, focal spongiosis are observed in the epidermis. There is a dense diffuse nodular infiltrate without signs of epidermotropism in the dermis. The infiltrate is represented by a mixture of cells, among which small- and medium-sized lymphoid cells with moderate signs of cellular atypia (irregular oblong hyperchromic nuclei) prevail. Lymphocytes of the usual type, histiocytes, including giant multinucleated cells, immunoblasts and plasmocytes are also found in significant numbers (Fig. 2a).

Immunohistochemical study of the skin biopsy. The immunophenotype of most infiltrate cells corresponds to T-lymphocytes: CD3+, CD4+, CD5+, CD7+, CD8-, CD30-, CD56- (Fig. 2b). In small quantity single CD8+ lymphocytes and B lymphocytes (CD20+) are visible. The expression of Ki-67 is approximately 10% in the T-cell infiltrate zone (Fig. 2c). Monoclonal rearrangement of the T-cell receptor of lymphocytes is detected.

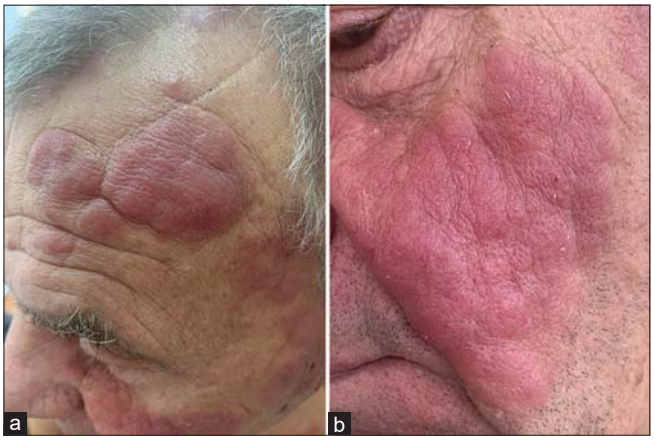

Figure 1: Patient B., polymorphism of clinical manifestations of primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma: a – rounded tumor-like formations on the forehead and temple, b – tumor-like formation on the skin of the cheek.

Based on the clinical, morphological and immunohistochemical data, the patient was diagnosed with the primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma (ICD-O code 9709/3). The patient was referred for treatment to an oncohematologist.

DISCUSSION

The diagnosis of SMPTCL is based on the combination of the clinical picture, histological and immunomorphological data. The morphological picture of the skin biopsy in SMPTCL is characterized by the presence of diffuse or nodular infiltration in the dermis with a tendency to penetrate into the subcutaneous tissue. The typical feature of the composition of the lymphocytic infiltrate is the predominance of small- and medium-sized pleomorphic T-cells. Large pleomorphic cells can also be present, with their number being no more than 30%. A considerable admixture of small reactive lymphocytes, histiocytes and eosinophils is possible. B-cells can account for 10 to 40% in the composition of the infiltrate in the form of discretely located large immunoblasts and nodules of small lymphocytes resembling primary follicles. Immunohistochemical study shows that SMPTCL tumor cells have a CD3 +, CD4 +, CD8-, CD30- phenotype, in most cases with the normal expression of T-cell antigens CD2, CD3, CD5, and CD7, and less often with the loss of pan-T-cell antigens. Ki-67 express no more than 10-30% infiltrate cells. Plasma cells express κ- and λ-light chains of immunoglobulins. In most cases, the PCR method reveals the clonal rearrangement of the T-cell receptor [2 - 5].

SMPTCL belongs to rare diseases, with the frequency of occurrence ranging from 2% to 5% in the structure of T-cell cutaneous lymphomas. In domestic and foreign literature, there are only a few publications on this disease which demonstrate mainly local skin lesions [4,6-9]. According to the data of the clinic of the Ural Research Institute of Dermatovenerology and Immunopathology, from 2010 to 2019, cutaneous small/medium-sized pleomorphic T-cell lymphoma was diagnosed in only 2 patients out of 561 patients with suspected lymphoproliferative skin diseases [7].

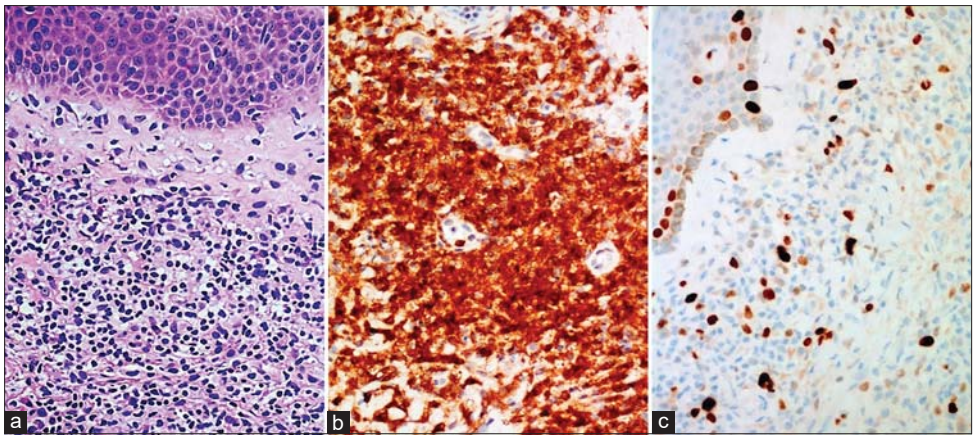

Figure 2: Pathomorphological changes in the patient's skin (with 400 x magnification): a – the presence of a dense diffuse nodular infiltrate with the predominance of small- and medium-sized lymphoid cells, stained with hematoxylin and eosin. b – expression of CD4+ by most cells of the infiltrate (immunophenotyping method). c – the presence of proliferating cells (Ki67+) in the epidermis and dermal infiltrate (immunophenotyping method).

CONCLUSION

The presented clinical case confirms the complexity of diagnosis of primary cutaneous CD4+ small/medium-sized pleomorphic T-cell lymphoma due to the rare occurrence of this nosology and the slow progression of the disease. It emphasizes the importance of doctors' clinical oncological alertness and the need for clinical and laboratory examination of the patient using modern histological and immunohistochemical methods for studying the skin biopsy.

Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that his name and initials will not be published and due efforts will be made to conceal their identity.

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