Erythema chronicum migrans like- T cell lymphoma: First description in the medical literature. Successful management with methotrexate and methylprednisolone aceponate!

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Abstract

Background: The group of cutaneous T-cell lymphomas (CTCL) or the so-called Mycosis fungoides (MF) is also referred to as a “great imitator” and in some cases may be misinterpreted as benign inflammatory skin disorders. Difficulties could arise both clinically and histopathologically, especially in the early stages of the disease. For this reason, it is recommended that the diagnosis of MF to follow a three-stage pattern of behavior and be performed on the basis of clinic, histopathology and immunohistochemistry.

Case report: We present a 90-year-old man with an erythematous-brownish plaque lesion in the left thoracic region, a pink-red plaque in the right thigh area with signs of initial atrophy and disseminated erythematous macular skin rash on the trunk, upper and lower extremities. The changes have about a 2-3 months duration. Based on the clinical data, the presence of erythema chronicum migrans was suspected. ELISA for Borrelia burgdorferi was performed, with serological testing being negative for antibodies against B. burgdorferi. Histological and immunohistochemical examination of biopsy material from erythema chronicum migrans-like lesion and the dense lesion was performed, which revealed the presence of completely similar histopathological changes by the type of mycosis fungoides. Immunohistochemical data were also indicative of mycosis fungoides with an abundance of CD3 positive cells, partial CD7, single CD8 and CD20 positive expression in the lesional material. Conclusion staging was performed for a patient with mycosis fungoides stage IB (T2bN0M0). Immunosuppressive therapy with methotrexate 2x7.5mg / weekly was performed, combined with topical application of methylprednisolone aceponate 0.1% cream x 2 / daily with good clinical response within the first three weeks of the treatment.

Conclusion: Borrelia burgdorferi is cited as one of the possible etiological factors for the development of mycosis fungoides. We describe a case of a patient with clinical data for erythema chronicum migrans and negative serology for B. burgdorferi. The possibilities of 1) the presence of Lyme disease in seronegative results are discussed, as well as the probability of 2) Borrelia-induced MF in the absence of antibodies against the agent within the ELISA assay and 3) PCR positivity for Borrelia burgdorferi in a negative serology.

Keywords: Mycosis fungoides; Erythema chronicum migrans like- T cell lymphoma; Borrelia burgdorferi; ELISA; PCR;

Introduction

In the literature, the group of cutaneous T-cell lymphoma (CTCL) is defined and interchanged with the term Mycosis fungoides (MF) [1]. In turn, MF is classified into four stages, with the system form being defined as Sézary Syndrome [2]. Certain authors have identified the disease as a “great imitator” because it can be presented as benign inflammatory skin disorders either clinically, both clinically and histopathologically [3]. For this reason, it is recommended that the diagnosis for this type of disease to be based on the correlation between clinical presentation, histopathology, and T-cell monoclonality detected by molecular studies [4].

Case Report

We present a 90-year-old man with a complaint for an erythematous itchy rash on the skin of the entire body surface with a duration of 2-3 months. Additionally, the patient reported the appearance of a red-brown plaque in the left thoracic region, as well as the presence of a pink-red plaque in the right femoral area. During the dermatological examination, an erythematous-brownish plaque lesion over 10 cm in diameter, located in the left thoracic region, was visualized above the axillary area (Fig. 1a). Additionally, an oval lesion with signs of initial atrophy clinically suspected for Morbus Bowen (Fig. 1b) was found on the lateral part of the right thigh. Disseminated erythematosus macular rash on the skin of the trunk, upper and lower extremities (Fig. 1c-d) was registered as concomitant changes. Based on the clinical data, it was suspected that this was a patient with erythema chronicum migrans. An ELISA assay for the qualitative determination of Borrelia burgdorferi was performed and no antibodies against B. burgdorferi were available within the serological study. In parallel, biopsy specimens were taken from the affected cutaneous areas for histological examination and
**Fig. 1a:** Erythematous-brownish plaque lesion located in the left thoracic region, below the axillary fossa.

**Fig. 1b:** Clinical view of a pink-erythematous oval lesion with signs of initial atrophy in the area of the lateral part of the right thigh.

**Fig. 1c-d:** Disseminated erythematous macular rash on the skin of the trunk.

**Fig. 2a-d:** Dense infiltrate of lymphocytes around vessels in the superficial plexus, grouped intraepidermal presence of small lymphoids, scarce spongiosis, follicular mucinosis.
immunohistochemistry. The histological result of the erythema chronicum migrans-like lesion in the left thoracic region and the dense lesion in the right lateral femoral area revealed the presence of a dense lymphocyte infiltrate around the vessels in the superficial plexus, scarce spongiosis, follicular mucinosis (Fig. 2a-d), with the conclusion for the presence of completely similar changes for mycosis fungoides. So the biopsy material from the lesion in the right thigh area rejected the possibility of morbus Bowen. The immunohistochemical study confirmed the diagnosis of mycosis fungoides by finding evidence of epidermal “upholstery” with irregularly alternating acanthosis and atrophic changes, combined with the grouped intraepidermal presence of small lymphoids with dense chromatin, as well as abundance of CD3 positive cells, CD7 (+) cells in the cell composition, single CD8 and CD20 positive expression. No clinical and ultrasound data on pathologically enlarged lymph nodes were available. Abdominal ultrasound revealed evidence of chronic cholecystitis, and radiography of the lungs and heart found data for enhanced striated bilateral perihilar pattern, with dilated hilus shadows, a thickened and expanded aortic arch. Staging was performed according to which it was found to be a patient with mycosis fungoides stage IB (T2bN0M0). Methotrexate immunosuppressive therapy was started according to the regimen: 2x7.5mg / weekly, combined with topical application of methylprednisolone aceponate 0.1% cream x 2 / daily, resulting in good clinical response within the first three weeks.

Discussion

Rare forms of mycosis fungoides that present a diagnostic challenge include folliculocutaneous MF granulomatous slack skin, and pagetoid reticulosis [5]. Usually the histopathological definition includes the presence of epidermotropic infiltrate of atypical lymphocytes with lining up of lymphocytes along the junctional zone and formation of intraepidermal clusters of atypical lymphocytes, the so called Pautrier microabscesses [5]. However, clinically, mycosis fungoides (MF) or the initial forms of cutaneous T-cell lymphomas (CTCL) can often present as inflammatory erythematous patches or plaques and mimic benign skin disorders [5]. Interesting is the role of Borrelia burgdorferi, which according to the literature can be associated with certain forms of primary cutaneous lymphomas [6]. A case-control studies suggests that the persistence of multiple infectious agents may cause the long-term antigenic stimulation contributing to the malignant transformation of T lymphocytes, and this group also include Borrelia burgdorferi [7]. On the other hand, according to some authors, it refers to the group of agents that may cause so-called cutaneous pseudolymphomas (PSLs), which histopathologically and / or clinically simulate lymphomas [8]. This includes also the induction of pseudo-mycosis fungoides (pseudo-MF) [8].

We present a patient with initial clinical data for erythema chronicum migrans and negative serology for Borrelia burgdorferi. According to the available literature, the presence of chronic Lyme disease may be available according to these studies, which is evidence of infection [10]. PCR is recommended as an additional method of detecting Lyme disease, indicating a higher sensitivity of Borrelia PCR from tissue specimens against to body fluids [11]. PCR has proven to be an important alternative method of investigation since the case of PCR positivity and absence of a serologic response to Lyme borreliosis has been described [12].

A case of cutaneous t-cell lymphoma-like nodule in a patient with erythema chronicum migrans has been described [13]. In the case presented, we pose the question whether B. burgdorferi can cause the development of cutaneous lymphoma without having positive antibodies against it or is it actually a case of pseudo-erythema chronicum migrans-like T cell lymphoma?

References
