The management of progressive mycosis fungoides with overlapping presentation of sezary syndrome

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Introduction

Mycosis fungoides (MF) and Sezary syndrome (SS) are the most common clinical types of cutaneous T – cell lymphomas (CTCLs). MF and SS have overlapping presentations and are not distinguished in the WHO/EORTC staging criteria; however, they are considered separate entities. The WHO/EORTC and the International Society for Cutaneous Lymphomas consider SS to be a clinical syndrome presenting with erythrodermic skin and leukemic disease. In contrast, patients who initially present with classic skin lesions of MF (patches, plaques and tumours) and only later meet the staging criteria for SS are referred to as leukemic MF, SS preceded by MF, or secondary SS. The National Comprehensive Cancer Network (NCCN) considers patients with SS to be anyone who meets the criteria for a high blood burden of disease (B2 disease) [1].

Aim and Objective

A case report.

Materials and methods

A 78-year-old male presented with erythroderma and severe pruritus (VAS score – 8). Histopathological examination revealed a dermal infiltrate of atypical lymphocytes with irregular cerebriform nuclei, slight epidermotropism (Fig. 1). Immunohistochemical staining showed positivity for CD3, with elevated CD4:CD8 ratio (>10) (Fig. 2-4). Flow cytometry showed a presence of circulating atypical lymphocytes, increased CD4/CD8 ratio (15.71). Enlarged peripheral lymph nodes by ultrasonography were found. Based on these findings diagnosis of Sezary syndrome (cT4N1M0B2) was made.

Results

The extracorporeal photopheresis (ECP) together with topical corticosteroids and oral prednisolone (30 mg daily, gradually reducing the dose to 15 mg daily) were prescribed. The ECP was performed by Haemonetics MCS+ apheresis system (Haemonetics Corporation). Treatment comprised three phases: leukapheresis, photoactivation with 8 – methoxypsoralen (0,1 mg) with exposure to ultraviolet A radiation (2.01 J/cm2) and reinfusion of lymphocytes back to the patient. The procedure was repeated on two consecutive days, every 2–3 weeks. After this treatment positive response was obtained: BSA decreased from 90% to 10%, mSWAT decreased from 80 to 12, LDH decreased from 1177 U/l to 382 U/l. Additional treatment with oral 8 – methoxypsoralen plus ultraviolet A (systemic PUVA) was prescribed (6 procedures in total). However, after ECPs procedures (8 in total) multiple red plaques on the face, chest and thighs, as well as tumours in both armpits appeared (Fig. 5, 6). BSA together with mSWAT increased again, even though CD4/CD8 ratio decreased to 2.1 and LDH did not increased significantly – 426 U/l. Clinically and histologically abnormal axillary and inguinal lymph nodes were detected. The patient was treated with methylprednisolone 250 mg/d infusion (5 days). Due to rapid progression of the disease cyclophosphamide, doxorubicin, vincristine, etoposide and prednisone (CHOEP) regime was initiated. After one cycle of chemotherapy, patients condition continues to deteriorate: erythroderma is present again – BSA 95%, mSWAT increased to 136.5, patient has developed agranulocytosis and fever.

Conclusions

Patients with MF/SS have unfavourable prognosis and are in a grave need of effective treatment options and precise diagnostics. MF and SS diagnostics requires not only a clinical and pathological examination, but also molecular tests such as T – cell receptor (TCR) clonality in blood, skin and lymph nodes. The goal of treatment of MF and SS is to reduce symptomatic morbidity and limit progression of the disease. ECP combined with other treatment modalities is the treatment of the choice for erythrodermic MF and SS. For MF stage IV – polychemotherapy is often given, although as a first line treatment it is associated with a higher risk of death [2, 3]. Increasingly, hematopoietic stem cell transplantation is being considered for patients with advanced stages, it is the only possibly curative therapy.

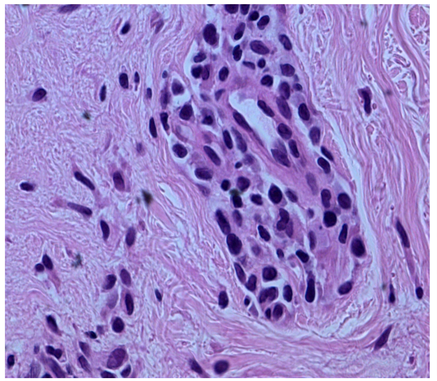

Figure 1. Histopathology findings H+Ex400: atypical (with cerebriform nuclei) lymphocytes; epidermotropism

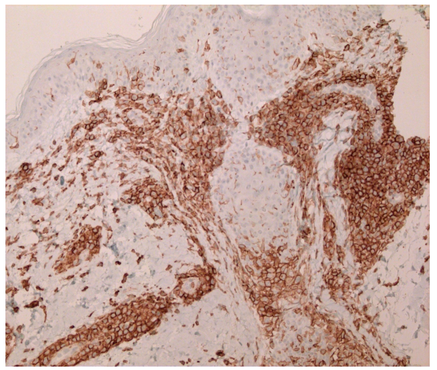

Figure 2. Imonohistochemistry findings x100: CD4 positive cells

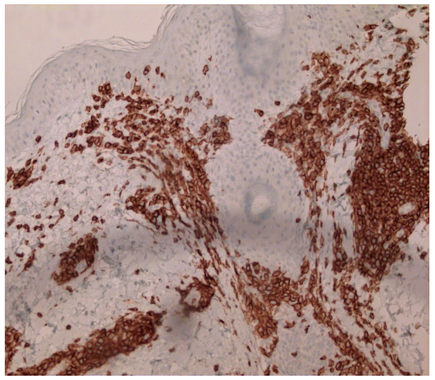

Figure 3. Imonohistochemistry findings x100: CD3 positive cells

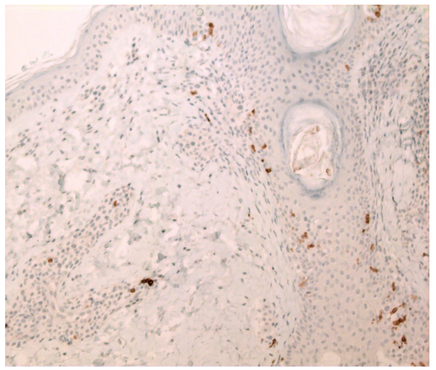

Figure 4. Imonohistochemistry findings x100: CD8 positive cells

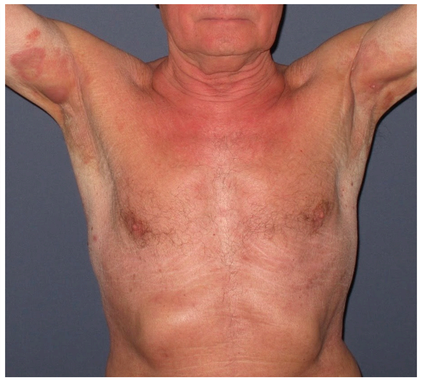

Figure 5. Clinical evaluation: after 6 ECP procedures

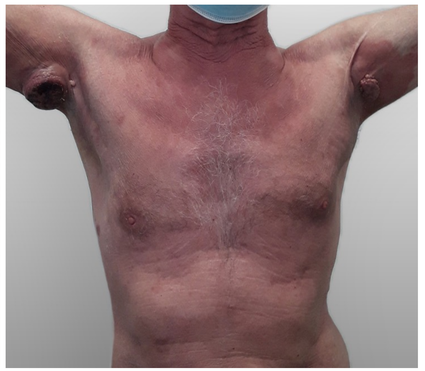

Figure 6. Clinical evaluation: after 8 ECP procedures

References

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3. Trautinger F, Eder J, Assaf C, Bagot M, Cozzio A, Dummer R, et al. European Organisation for Research and Treatment of Cancer consensus recommendations for the treatment of mycosis fungoides/Sézary syndrome - Update 2017. Eur J Cancer. 2017 May; 77:57-74.