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Mycosis fungoides (MF) is a presentation of primary cutaneous T-cell lymphoma with adverse prognosis for patients with advanced stages of the disease. Refractory disease and advanced-stage disease require systemic therapy. We reported a rare case of an atypical predominantly CD8+ disease. Refractory disease and advanced-stage disease require systemic lymphoma with adverse prognosis for patients with advanced stages of the disease.

Patients with advanced cutaneous lymphoma have a poor prognosis with a median survival of 1–5 years [1]. Current guidelines recommend tailoring treatment based on disease staging [2]. Allogeneic haematopoietic stem cell transplant (HSCT) may be used to treat eligible patients with late stage disease, aggressive disease or those with poor prognostic factors who achieve a durable remission [2]. At our centre, the pre-transplant conditioning regimen adopted in 2013 consists of Total Skin Electron Beam Therapy/Total Nodal Irradiation and anti-thymocyte globulin (TSE/TNI/ATG), with Sézary syndrome (SS) patients receiving additional extracorporeal photopheresis (ECP) (3). 19 patients including 9 males and 10 females, with a median age of 49 at the time of HSCT (range 35 to 67 years old) have since been transplanted. We present longer term follow up of these patients [3].

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Background: Patients with primary cutaneous lymphoma receive immunosuppressive therapy for long term disease control. Both cutaneous lymphoma and immunosuppressive treatment can contribute to the development of more severe COVID-19 complications. The real challenge during the COVID-19 pandemic remains the management of the advanced and aggressive forms of cutaneous lymphomas, including late-stage mycosis fungoides (MF) and Sézary syndrome (SS). Extracorporeal photopheresis (ECP) is one of those treatments. ECP is considered high risk therapy according to the United States Cutaneous Lymphoma Consortium recommendations for treatment of cutaneous lymphomas during the COVID-19 pandemic because it may require travel to the clinic or hospital.

Methods: In this cross-sectional retrospective study, data of patients with MF or SS who received ECP treatment were collected. ECP consisted of a two-session cycle every two to four weeks. In our group we did not carry out prophylactic interruption of the therapy, once started. In patients with stable disease (SD) or partial response (PR), ECP was administered every 4 weeks, until a 6-week maximum interval was reached, and the response maintained. During the study period, the frequency of treatments was decreased, especially for patients with severe comorbidities and/or older age. The associated therapy was considered individually, depending on the extension of the disease, comorbidities, and adverse effects of each agent.

Results: 16 patients with cutaneous lymphoma received ECP (9 (56%) with SS and 7 (44%) with MF). Their median age at diagnosis was 63 (57–67) years. The median number of treatments before ECP was 2 (1–3), which was typically either phototherapy or systemic corticosteroids. Regarding the associated treatment during ECP and pandemic, INP and retinoids were the first-choice treatments. In the MF group 3 patients (2 PR and 1 relapse) required additional therapy with acitretin and topical corticosteroids. In the SS group 5 patients (1 PR, 3 progressive disease and 1 relapse) received concomitant treatment with INP alfa 2b, bexarotene and electron beam; the relapsed patient underwent mono chemotherapy. Three patients were infected with SARS-CoV-2, all of them were from the SS group, the contagion was outside the hospital environment. Two of the patients who developed COVID infection died.

Conclusion: For patients with advanced forms of LCCT, who usually have multiple risk factors for a severe course of SARS-CoV-2 infection therapy. The critical patient subset includes those with advanced disease, who require treatment with polychemotherapy, ECP and checkpoint inhibitors. Treatment decisions should be made on an individual basis. In our experience, the continuous use of ECP during the pandemic did not increase the risk of contagion.

Keywords: Extracorporeal photopheresis, COVID-19 pandemic

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Tre-P-22
The primary cutaneous CD30-positive lymphoproliferative diseases; the own observations of the center, prospects of therapy
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Primary cutaneous T-cell lymphomas are the heterogeneous group of T-cell lymphoproliferative diseases (LPD), which are developed mainly in the skin and characterized by diagnostic features, clinical course and therapeutic approach. Primary cutaneous anaplastic lymphoma (c-ALCL) and lymphomatoid papulosis (LyP) are diagnosed in a quarter of cases of all T-cell skin lymphomas. The tumor cells in primary cutaneous CD30-positive skin lymphomas express CD30 in more than 75% of cases. The majority of patients with cutaneous CD30+ LPD have an indolent course with a favorable prognosis, the resistant course of disease develops in about 30% of cases and fatal cases from lymphoma are registered in 8% of cases. Most commonly, the treatment of these forms of LPD includes the surgical removal, radiation therapy or small doses of methotrexate, the systemic chemotherapy is used to generalize the process. Recently, monoclonal antibodies have been included in clinical practice for the treatment of skin lymphomas, one of which is brentuximab vedotin, the use of which has shown a rather high efficiency in the treatment of CD30+ skin lymphomas: more than 2/3 of the patients responded to the treatment, despite the many lines of therapy (median of prior effect is 3.1 for c-ALCL).

The general group of patients with cutaneous T-cell lymphomas and who received the consultative-diagnostic or inpatient treatment at the National Research Centre of Hematology includes 328 patients. Among them the CD30-positive cutaneous LPD were verified in 33 patients (10%): 16 patients with LyP, 17 patients with c-ALCL. The eruptions were regressed on its own without specific therapy in 75% of patients with LyP and 4 patients received the treatment. As well as one patient with the relapsing course of disease, with a long period of self-regression of the papules, with the lack of the treatment effect (phototherapy and low-dose methotrexate) received brentuximab vedotin (BV). The complete clinical response was achieved after the first cycle of monotherapy with BV and persists for more than a year.

Patients with c-ALCL more frequently needed the specific therapy (76% of patients), only 4 patients were observed with the self-regression of the papules within 6–9 weeks. Most patients received the interferon alpha therapy with the complete clinical response; the rest patients - local radiation therapy, the small doses of methotrexate. The systemic chemotherapy was done to the 5 patients. Considering the common process, however 2 of them died from infectious complications during the chemotherapy, 2 patients had early recurrence and only 1 patient is in the long-lasting complete remission after the high-dose of chemotherapy.

In summary, the primary cutaneous CD30+ lymphoproliferative diseases cover a spectrum of benign condition with the so-called “malignant” phenotype. LyP and c-ALCL have a favorable prognosis, 5-year survival rate is exceeding 95%, photo- and immunotherapy are successfully used in the treatment, while the use of systemic chemotherapy should be avoided (shortening of the remissions duration, progression of the infectious complications). The use of targeted therapy (brentuximab vedotin) increases the therapeutic ability in the situations where the routine treatment options are ineffective.

Keywords: primary cutaneous CD30+ lymphoproliferative diseases, brentuximab vedotin, systemic chemotherapy

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Tre-P-23
Papuloerythroderma of Ofuji successfully treated with oral retinoids
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Papuloerythroderma of Ofuji (PEO) is a rare skin disorder, which affects predominantly older males with male to female ratio 4:1. It is characterized by pruritic erythematous papules, which might progress to erythroderma. Exanthema typically spares the skin folds (deck-chair sign). Common laboratory findings include lymphopenia, peripheral eosinophilia and elevated serum IgE. Histological image generally shows non-specific inflammatory reaction and needs to be correlated with clinical and laboratory findings. PEO has been associated with cutaneous T-cell lymphomas (CTCL). However, this relationship remains poorly understood. Diagnosis