

pr 6, Volume 53, pages 421-423 DOI: 10.5603/AHP.a2022.0057 ISSN 0001-5814 e-ISSN 2300-7117

Mycosis fungoides with orbital involvement and COVID-19 — case study

Adrian Juda^{1*} • Waldemar Tomczak¹ • Aleksandra Tomczak², Anna Toczek², Agnieszka Gerkowicz³ • Dorota Krasowska³ • Marek Hus¹

¹Chair and Department of Hematooncology and Bone Marrow Transplantation, Medical University in Lublin, Lublin, Poland ²Students Scientific Association, Chair and Department of Dermatology, Venerology and Pediatric Dermatology, Medical University in Lublin, Lublin, Poland

³Chair and Department of Dermatology, Venerology and Pediatric Dermatology, Medical University in Lublin, Lublin, Poland

Introduction

Mycosis fungoides is a cutaneous lymphoma which originates from mature T lymphocytes. Symptoms include macular and papular eruptions or ulcerations. We herein present a case of progression of mycosis fungoides characterized by orbital involvement. After treatment with a monoclonal antibody (brentuximab vedotin), a partial clinical response was obtained, although the patient was re-infected with the severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2), which forced a discontinuation of the treatment. Secondary immune disorders occurring in hematological patients determine the specific course of coronavirus disease 2019 (COVID-19).

Clinical case

A 71-year-old female patient diagnosed two years previously with mycosis fungoides in its nodular form was admitted to the Department of Hematooncology due to disease progression after multiple lines of treatment (Table I). On admission, the patient's condition was average. Nodular and infiltrative changes on the skin of the trunk, upper limbs and thighs were seen. A tumor with central ulceration localized in the right orbital area was also observed (Figure 1). This mass involved the eyelids, had infiltrated the orbit and was displacing the eyeball. Material for histopathological examination was collected. Morphological signs suggested large-cell transformation of mycosis fungoides. The expression of CD30 antigen was found on the neoplastic cells.

Table I. Previous treatment and clinical response

Treatment	Response
Acitretin	Partial response
Methotrexate	Progression of disease
Four cycles of CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine, prednisone)	Progression of disease
Two cycles of BGD chemotherapy (bendamustine, gemcitabine, dexamethasone)	Progression of disease

The clinical staging before the start of treatment according to the International Society for Cutaneous Lymphomas/ /European Organization for Research and Treatment of Cancer (ISCL/EORTC) was assessed at stage IIB (T3N0M0B0), and the mSWAT (modified Severity-Weighted Assessment Tool) ratio was 42.5. It was decided to start therapy with a monoclonal antibody called brentuximab vedotin (BV) at a dose of 1.8 mg/kg body weight, given every 21 days. Three doses of the drug were administered. After treatment, regression of skin lesions and reduction of the orbital tumor were observed (Figure 2). Response to treatment was assessed using the mSWAT scale. Radical radiotherapy was performed on the area of the right orbit, eyelids and cheek (total dose 30 Gy; 15 fractions of 2 Gy each). Due to progressive neoplastic cachexia, additional parenteral nutrition was used. The patient was consulted by an ophthalmologist and a laryngologist and was qualified for right orbital exenteration, which was postponed due to her general condition.

*Address for correspondence: Adrian Juda, Department of Haematooncology and Bone Marrow Transplantation, Staszica 16, 20–081 Lublin, Poland, e-mail: adrianjuda@op.pl

Received: 20.06.2022

Accepted: 27.09.2022



Copyright © 2022

The Polish Society of Haematologists and Transfusiologists, Insitute of Haematology and Transfusion Medicine.

All rights reserved.

This article is available in open access under Creative Common Attribution-Non-Commercial-No Derivatives 4.0 International (CC BY-NC-ND 4.0) license, allowing to download articles and share them with others as long as they credit the authors and the publisher, but without permission to change them in any way or use them commercially.



Figure 1. Before treatment with brentuximab vedotin

In the third month of hospitalization, a SARS-CoV-2 virus antigen test was performed, the result being positive. Re-infection was diagnosed. The patient had not been vaccinated against COVID-19. Symptoms included weakness and a periodic mild dry cough. Due to the risk of severe course related to the general condition of the patient (neoplastic cachexia, CMV infection), the treatment included plasma from convalescents, remdesivir, passive oxygen therapy, and enoxaparin in anticoagulant prophylaxis. After three weeks, the presence of the SARS-CoV-2 virus antigen was still observed. The patient was transferred to the Department of Infectious Diseases in order to continue the therapy. Treatment with BV was discontinued. Inflammatory changes in the lungs and progressive respiratory failure were observed. Sudden cardiac arrest occurred on day 32 of the COVID-19 infection. The patient died.

Discussion

Diagnosis of mycosis fungoides is based on the typical clinical manifestation and histopathological examination [1]. Skin lesions are most often located in places sheltered from the sun [2]. Extramedullary manifestations of MF are rare. The risk of involvement of internal organs increases in the case of large-cell transformation [3]. The most common systemic locations are: lymph nodes, liver, spleen and lungs [4]. Cases of mycosis fungoides involving the central nervous system [5], larynx [6] and



Figure 2. After three cycles of brentuximab vedotin

eye tissues [7] have also been reported. Lymphopenia, commonly observed in patients with primary cutaneous lymphomas, is associated with a worse prognosis in viral infections, including SARS-CoV-2 [8]. The United States Cutaneous Lymphoma Consortium made a stratification of the relative frequency and risk for COVID-19 complications in patients with primary cutaneous lymphomas [9]. According to this, patients in IIB clinical stage have an intermediate/high risk of COVID-19 complications. Brentuximab vedotin treatment is also associated with a worse course of infection prognosis. Lymphocyte T and natural killer cells express CD30, which take part in CD8 lymphocyte T activation. An anti-CD30 drug could lessen cell immune response [10]. Due to the advanced disease seen in this patient, less aggressive therapy was not recommended. More trials are needed to describe risk factors and management in patients with COVID-19 and mycosis fungoides.

Authors' contributions

AJ, WT — manuscript preparation. AG, AT — data collection. AT — language edition. MH, DK — final approval.

Conflict of interest None.

none.

Financial support

None.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; EU Directive 2010/63/EU for animal experiments; uniform requirements for manuscripts submitted to biomedical journals.

References

- Larocca C, Kupper T. Mycosis fungoides and Sézary syndrome: an update. Hematol Oncol Clin North Am. 2019; 33(1): 103–120, doi: 10.1016/j.hoc.2018.09.001, indexed in Pubmed: 30497668.
- Hodak E, Amitay-Laish I. Mycosis fungoides: a great imitator. Clinics in Dermatology. 2019; 37(3): 255–267, doi: 10.1016/j.clindermatol.2019.01.004.
- 3. Bunn PA, Huberman MS, Whang-Peng J, et al. Prospective staging evaluation of patients with cutaneous T-cell lymphomas. Demonstration of a high frequency of extracutaneous dissemination. Ann Intern Med. 1980; 93(2): 223–230, doi: 10.7326/0003-4819-93-2-223, indexed in Pubmed: 6967710.
- Burg G. Systemic involvement in mycosis fungoides. Clin Dermatol. 2015; 33(5): 563–571, doi: 10.1016/j.clindermatol.2015.05.008, indexed in Pubmed: 26321404.
- Nguyen GH, Mohler A, Leppert M, et al. Index case of cutaneous follicular mycosis fungoides with central nervous system involvement

- and review ofliterature. Am J Dermatopathol. 2018; 40(3): 185-190, doi: 10.1097/DAD.000000000000955, indexed in Pubmed: 29470304
- Bauman TM, Wichterman CM, Musiek AC, et al. Hoarseness as a presentation of mycosis fungoides infiltrating the larynx. BMJ Case Rep. 2017; 2017, doi: 10.1136/bcr-2017-221531, indexed in Pubmed: 28993359.
- Jusufbegovic D, Char DH. Clinical variability of ocular involvement in mycosis fungoides. JAMA Ophthalmol. 2015; 133(3): 341–343, doi: 10.1001/jamaophthalmol.2014.5223, indexed in Pubmed: 25541838.
- Mateeva V, Patsatsi A. Management of primary cutaneous lymphomas during the COVID-19 pandemic. Clin Dermatol. 2021; 39(1): 64– -75, doi: 10.1016/j.clindermatol.2020.12.014, indexed in Pubmed: 33972055.
- Zic JA, Ai W, Akilov OE, et al. United States Cutaneous Lymphoma Consortium recommendations for treatment of cutaneous lymphomas during the COVID-19 pandemic. J Am Acad Dermatol. 2020; 83(2): 703-704, doi: 10.1016/j.jaad.2020.04.049, indexed in Pubmed: 32305443.
- Maschmeyer G, De Greef J, Mellinghoff SC, et al. European Conference on Infections in Leukemia (ECIL). Infections associated with immunotherapeutic and molecular targeted agents in hematology and oncology. A position paper by the European Conference on Infections in Leukemia (ECIL). Leukemia. 2019; 33(4): 844–862, doi: 10.1038/s41375-019-0388-x, indexed in Pubmed: 30700842.