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Q1 Case Report and Review

Intraventricular treatment of secondary central nervous system lymphoma – Case study and Q2 literature overview

Q3 Dariusz Szczepanek [a], Ewa Wąsik-Szczepanek [b], Agnieszka Szymczyk [b,c,*], Tomasz Gromek [b], Ewelina Grywalska [d], Monika Podhorecka [b], Marek Hus [b]

[a] Chair and Department of Neurosurgery and Paediatric Neurosurgery Medical University of Lublin, Poland
[b] Chair and Department of Haematooncology and Bone Marrow Transplantation Medical University of Lublin, Poland
[c] Independent Clinical Transplantology Unit Medical University of Lublin, Poland
[d] Chair and Department of Clinical Immunology Medical University of Lublin, Poland

ARTICLE INFO

Article history:
Received 24 August 2017
Accepted 27 January 2018
Available online xxx

Keywords:
Primary central nervous system lymphomas
Secondary central nervous system lymphomas
Ommaya reservoir
Intraventricular treatment

ABSTRACT

Secondary nervous system lymphoma (SCNSL) is a rare extranodal form of non-Hodgkin lymphoma (NHL). This applies to a particular form of lymphoma that does not originally derive from the central nervous system (CNS); it can be both an isolated form of relapse or a systemic part of disease progression. Due to poor prognosis and a lack of established algorithms of therapeutic procedures, it is a big challenge for physicians from many specializations. In our study, we present an interesting case of a patient with a relapsed form of SCNSL for whom a unique form of treatment was used – intraventricular administration of rituximab and methotrexate.

1. Introduction

Lymphomas of central nervous system (CNS) both in a primary (PCNSL) and secondary form (SCNSL) are a rarely diagnosed extranodal location of non-Hodgkin lymphomas (NHL) usually with poor prognosis. SCNSL is a particular type of lymphoma, which does not initially derive from the CNS; it can present both as an isolated form of relapse or a systemic part of disease progression [1]. SCNSL is usually detected within a few weeks or months after a diagnosis of systemic lymphoma. It affects both the brain and cerebral meninges [2]. PCNSL is treated with chemotherapy based on high doses of methotrexate (MTX) [3]. The selection of optimal therapy for recurrent and resistant CNS lymphomas, is a particularly difficult challenge and creates many more doubts. In this study, we present a case of a patient with a relapsed form of a secondary CNS lymphoma for

* *Corresponding author at*: Chair and Department of Haematooncology and Bone Marrow Transplantation, Medical University of Lublin, Staszica 11 Str., 20-081 Lublin, Poland.
E-mail address: agnieszka.szymczyk.med@wp.pl (A. Szymczyk).
https://doi.org/10.1016/j.pjnns.2018.01.007

Please cite this article in press as: Szczepanek D, et al. Intraventricular treatment of secondary central nervous system lymphoma – Case study and literature Q2overview. Neurol Neurochir Pol (2018), https://doi.org/10.1016/j.pjnns.2018.01.007

whom a unique form of treatment was used – intraventricular administration of rituximab and methotrexate.

2. Case presentation

In March 2013, diffuse large B-cell lymphoma (DLBCL, NOS; CD20+, CD5+/−, CD10+, BCL2+, BCL6+, MUM1−/CD43-, Ki-67++ approx. 50% of cells) at stage IV B was diagnosed in a 74-year-old man on the basis of a histological examination of a pancreas sample and the surrounding lymph nodes. A total of 8 cycles of R-CHOP immunochemotherapy was ordered. In result complete remission (CR) was obtained. At the same time, the patient was receiving a hormone therapy due to prostate cancer. In September 2015 (with CR lasting 21 months) an isolated recurrence of lymphoma in the CNS, affecting the meninges and ependyma, was discovered and radiotherapy was started. A dose of 3000 cGy radiation was used on the brain and the medulla oblongata. A check-up PET examination revealed complete regression of lymphomatous lesions in the CNS and no relapse of the tumour of lymphoma in other parts of the body. After a 4-month observation, the patient's general condition deteriorated, and progressive psychomotor slow-down and memory disorders were observed. After another two months, a generalised epileptic seizure occurred. In April 2016 an MRI scan of the head showed a lymphomatous infiltration of the brain, the lateral ventricular walls, the septum pellucidum and the 3rd ventricle (Fig. 1 presents contrast enhancement of the tissue infiltrating the right lateral ventricle wall). An examination of the cerebrospinal fluid (CSF) did not reveal the presence of lymphomatous cells. The test for human immunodeficiency virus (HIV) was negative, however the test for the Epstein–Barr virus (EBV) was not performed.

The patient was qualified for the Ommaya reservoir implantation to administer chemotherapy. Such a decision was made due to the patient's advanced age, general condition (ECOG-3) and numerous co-existing diseases (prostate cancer, distal pancreatic resection in case history, degenerative spine disease, hypothyroidism, hypertension, diabetes). At the same time the patient did not consent to systemic chemotherapy. The treatment regimen consisted of simultaneous administration of rituximab (25 mg) and methotrexate (10 mg) twice a week for 4 weeks (8 doses in total). During the entire uneventful course of the intraventricular immuno-chemotherapy, steroid medications were not used, however considerable improvement in the psychomotor condition of the patient occurred. After therapy completion, an MRI check-up revealed considerable regression of changes in the CNS (Fig. 2). On treatment cycle end the patient was transferred to the Long-Term Care Unit. Unfortunately, he did not consent to further treatment. The documented period of the stable neurological state confirmed with CT was 7 months. Information about the later patient's medical history is not known.

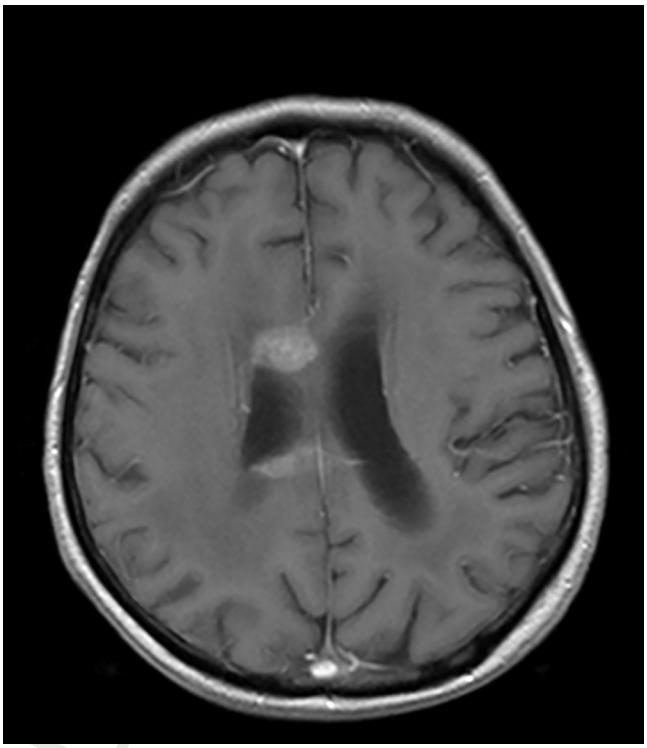

Fig. 1 – T1-weighted contrast-enhanced MR imaging presents contrast enhancement of the tissue infiltrating the right lateral ventricle wall before treatment.

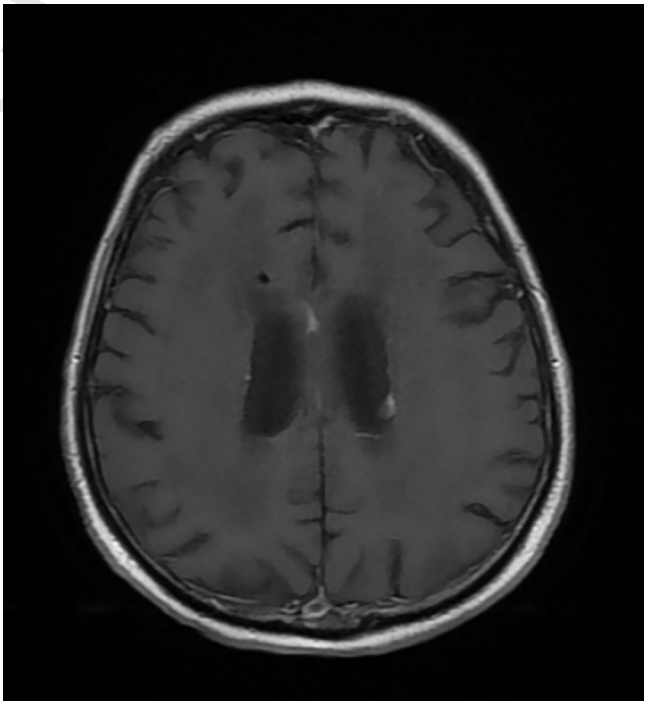

Fig. 2 – T1-weighted contrast-enhanced MR imaging presents radical regression of infiltrating changes in the right lateral ventricle wall after treatment.

3. Discussion

Diffuse large B-cell lymphomas (DLBCL) are one of the most often diagnosed types of non-Hodgkin lymphomas [4]. The use of regimens containing rituximab, vincristine, cyclophosphamide and prednison (R-CHOP) has made it possible to cure 50–60% of patients [5]. However, therapy of subjects with recurrent disease, especially affecting the central nervous

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