Clinical Case Report

Medicine®

OPEN

A 92-year-old man with primary cutaneous diffuse large B-cell non-Hodgkin's lymphoma manifesting as a giant scalp mass

A case report

Chenlong Liao, MD, PhD, Min Yang, MD, Pengfei Liu, MD, PhD, Wenchuan Zhang, MD, PhD*

Abstract

Rationale: Primary cutaneous non-Hodgkin's lymphoma (NHL) is an uncommon entity, representing 10% of all extranodal NHLs. Among all cutaneous sites, the scalp is a rare site of representation.

Patient concerns: A 92-year-old Chinese man visited our hospital with a multiple-nodular huge scalp mass on the right parieto-occipital regions. The mass was of 7-month duration and progressively enlarging in size.

Diagnoses: On the basis of the result of biopsy, diffuse large B-cell NHL was diagnosed.

Interventions: The mass was partially resected by surgery and no further treatment was conducted due to the advanced age and poor physical status.

Outcomes: The tumor relapsed in situ after 6 months and the patient died after 2 years.

Lessons: This case highlighted the limited access to standard treatment options in patients with advanced age. A thorough examination is necessary to decide upon the treatment for the primary cutaneous lymphoma.

Abbreviations: CBCL = cutaneous B-cell lymphomas, CTCL = cutaneous T-cell lymphomas, EORTC = European Organization for Research and Treatment of Cancer, NHL = non-Hodgkin's lymphoma, WHO = World Health Organization.

Keywords: diffuse large B-cell lymphoma, non-Hodgkin's lymphoma, primary cutaneous lymphoma, scalp mass, surgery

1. Introduction

Primary cutaneous non-Hodgkin's lymphoma (NHL) is an uncommon entity, representing 10% of all extranodal NHLs. It is characterized by skin involvement without signs of systemic disease at initial presentation and 6 months after identification.[1] Skin can be involved in systemic lymphomas as secondary cutaneous lymphomas, which have a completely different clinical behavior and prognosis from primary cutaneous lymphomas, and thus require different treatments.[2] Therefore, primary cutaneous lymphomas were classified as separate entities in different classification systems, such as the European Organization for Research and Treatment of Cancer (EORTC) classification,[3] the World Health Organization (WHO) classification,[4] and the recent WHO-EORTC classification (Table 1).[2]

Primary cutaneous B-cell lymphomas typically present as smooth, well-circumscribed, erythematouis to violaceous, non-ulcerated nodules predominantly located on the trunk, head, and neck regions.[5] Among all cutaneous sites, the scalp is a rare site of representation.[6,7] The conventional therapeutics include surgery, chemotherapy, and radiotherapy. In elder patients, however, the available options were limited due to the deteriorative physical condition. We present a rare case of primary cutaneous diffuse large B-cell NHL with a mass lesion of scalp in an elderly patient.

2. Case report

A 92-year-old Chinese man visited our hospital with a multiple-nodular huge scalp mass on the right parieto-occipital regions. The mass was of 7-month duration and progressively enlarging in size. No history of trauma was presented. The patient complained no weight loss, night sweats, or fever. On physical examination, a firm, painless, nonpulsating subcutaneous multiple-nodular huge scalp mass measuring about 8 cm × 12 cm was evident (Fig. 1A). The surface tension of the local scalp was high. Neither lymphadenopathy nor hepatosplenomegaly was detected. Neurological examination revealed no abnormalities and all the serum chemistry profile were within the normal values. Serological tests for human immunodeficiency virus, Epstein–Barr virus, and human T-cell lymphotropic virus were negative. Cerebral computed tomography (CT) excluded the invasion of skull (Fig. 1B). Magnetic resonance imaging demonstrated a

Editor: Mauro Alaibac.

The study was approved by Xinhua Hospital Medical Ethics committee. The next of kin has consented to submission of this case report to the journal.

The authors report no conflicts of interest.

Department of Neurosurgery, XinHua Hospital affiliated to Shanghai JiaoTong University School of Medicine, Shanghai, P.R. China.

** Correspondence: Wenchuan Zhang, No. 1665 Shanghai Kongjiang Road, Yangpu District, Shanghai 200092, China (e-mail: zhangwench88@hotmail.com).*

Medicine (2017) 96:10(e6270)

Received: 15 September 2016 / Received in final form: 3 February 2017 / Accepted: 9 February 2017

http://dx.doi.org/10.1097/MD.0000000000006270

Table 1

WHO-EORTC classification of cutaneous lymphomas with primary cutaneous manifestations.

Cutaneous T-cell and NK-cell lymphomas
Mycosis fungoides
MF variants and subtypes
Folliculotropic MF
Pagetoid reticulosis
Granulomatous slack skin
Sézary syndrome
Adult T-cell leukemia/lymphoma
Primary cutaneous CD30 lymphoproliferative disorders
Primary cutaneous anaplastic large cell lymphoma
Lymphomatoid papulosis
Subcutaneous panniculitis-like T-cell lymphoma
Extranodal NK/T-cell lymphoma, nasal type
Primary cutaneous peripheral T-cell lymphoma, unspecified
Primary cutaneous aggressive epidermotropic CD8 T-cell lymphoma (provisional)
Cutaneous/T-cell lymphoma (provisional)
Primary cutaneous CD4 small/medium-sized pleomorphic T-cell lymphoma (provisional)
Cutaneous B-cell lymphomas
Primary cutaneous marginal zone B-cell lymphoma
Primary cutaneous follicle center lymphoma
Primary cutaneous diffuse large B-cell lymphoma, leg type
Primary cutaneous diffuse large B-cell lymphoma, other
Intravascular large B-cell lymphoma
Precursor hematologic neoplasm
CD4/CD56 hematodermic neoplasm (blastic NK-cell lymphoma)

NK = natural killer.

multiple-nodular huge scalp mass with heterogeneous signals with low priority on T1-weighted imaging (Fig. 1C) and heterogeneous signals on T2-weighted imaging. Bone marrow aspiration and CT scan of the chest, abdomen, and pelvis failed to identify any other evidence of systemic lymphoma. Lumbar puncture was not attempted. The patient was treated with surgery for partial removal of the scalp mass. Straight incision was made at the base of the tumor on the parietal part. The tumor was turned over and resected. Subtotal resection of the tumor was performed and the surface tension of the local scalp was decompressed. The pathological diagnosis was diffuse large B-cell NHL (Fig. 1D). Immunohistochemistry showed that the lymphoma cells were positive for CD 20 and Bcl-2. Allowing for the advanced age, physical status, and the expectation of the patient and the family, no additional treatment such as radiotherapy or chemotherapy was conducted. The tumor relapsed in situ after 6 months and the patient died of dyscrasia after 2 years.

3. Discussion

Only 15% to 25% of cutaneous lymphoma show extracutaneous manifestation at the time of diagnosis. However, primary cutaneous NHL of scalp may eventually progress to involve the underlying skull and dura.[6,8,9] In such case, the origin of lymphoma should be carefully detected and the differential diagnosis between primary cutaneous lymphoma and primary NHL of the cranial vault should be made, as the therapeutic

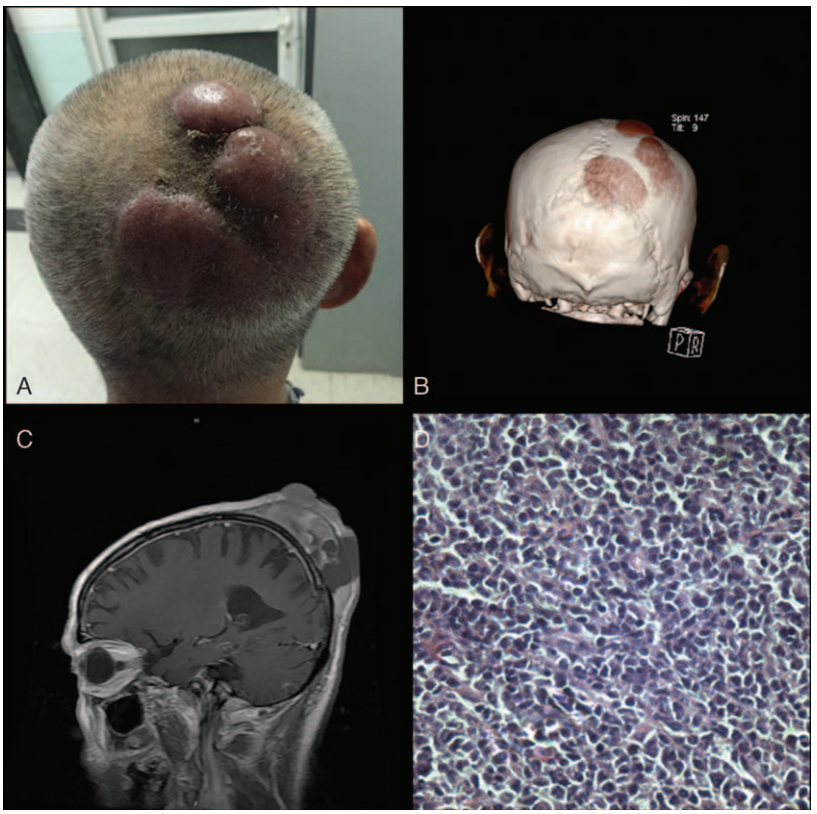

Figure 1(A) Physical examination showing a firm, nonpulsating, subcutaneous, multiple-nodular huge scalp mass measuring about 8 cm × 12 cm. (B) Three-dimensional reconstructive CT scan of the head displaying the location of the multi-nodular scalp mass and excluding the invasion of the skull. (C) Enhanced T1-weighted sagittal magnetic resonance imaging demonstrating a multiple-nodular huge scalp mass with heterogeneous signals. (D) Histological features showing the tumor comprising diffuse large B-cell. HE stain, ×400.

approaches and the prognosis were different.[10] The prognosis of cutaneous lymphoma was reported to be relatively good, as the average survival time from the diagnosis is 12 to 14 years.[11] When compared with primary cutaneous lymphoma, shorter duration of symptomatology, early onset of focal neurological deficits, and extensive osteolytic lesions were presented in primary NHL of the cranial vault.[12]

Diffuse large B-cell lymphoma is a heterogeneous group of neoplasms in which previous morphological, phenotypic, genetic, and molecular studies have not been able to identify well-defined disease entries with clinical and therapeutic relevance.[13] It seems that demonstration of bcl-12 expression should always raise suspicion of a systemic lymphoma involving the skin secondarily and unfavorable course of disease.[2,7] However, as seen in the present case, the disease ran an indolent course with no intracranial invasion despite of bcl-12 expression. There is a possibility that this indicates transformation of large B-cell lymphoma to follicular lymphoma, but this could not be proved by available clinical and morphopathologic studies.

Even though a well-accepted therapeutic algorithm of primary cutaneous NHL of scalp has not been defined, radical surgical removal associated with radiotherapy and chemotherapy was suggested to be the best option. Notwithstanding, the most difficult part is to decide on the sequence of treatment.[6] According to the concept of personalized medicine, individual condition including age and physical status should also be taken into consideration. As in the present case, the patient was 92 years old and the course of the disease was 6 months. The surgery was performed in order to relieve the high surface tension of the local scalp and reduce the tumor burden for possible subsequent treatment such as radiotherapy and chemotherapy. However, allowing for the advanced age, physical status, and the expectation of the patient and the family, no additional treatment such as radiotherapy or chemotherapy was conducted. It could be thus inferred that the adjuvant treatments were of great importance in preventing relapse of primary cutaneous diffuse large B-cell NHL.

The present study described a rare case of primary cutaneous diffuse large B-cell NHL with multiple-nodular huge scalp mass. To the best of our knowledge, this is the oldest patient with primary cutaneous NHL of scalp that has been reported. This case highlighted the limited access to standard treatment options in patients with advanced age. A thorough examination is necessary to decide upon the treatment for the primary cutaneous lymphoma. Further accumulation of data for scalp lymphoma in elder patients is needed to improve the treatment modality and the prognosis.

References

[1] Condarco T, Sagatys E, Prakash AV, et al. Primary cutaneous B-cell lymphoma in a child. Fetal Pediatr Pathol 2008;27:206–14.
[2] Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. Blood 2005;105:3768–85.
[3] Willemze R, Kerl H, Sterry W, et al. EORTC classification for primary cutaneous lymphomas: a proposal from the Cutaneous Lymphoma Study Group of the European Organization for Research and Treatment of Cancer. Blood 1997;90:354–71.
[4] Jaffe ES, Harris NL, Stein H, editors. World Health Organization Classification of Tumors: Pathology and Genetics of Tumours of Hematopoietic and Lymphoid Tissues. Lyon, France: IARC Press; 2001.
[5] Watsky KL, Longley BJ, Dvoretzky I. Primary cutaneous B-cell lymphoma. Diagnosis, treatment, and prognosis. J Dermatol Surg Oncol 1992;18:951–4.
[6] Yadav BS, Bansal A, Sharma SC, et al. A 60-year-old Indian male with altered sensorium and extensive lymphoma of the scalp. Semin Oncol 2013;40:e9–21.
[7] Colovic M, Vidovic A, Colovic N, et al. Primary cutaneous large B-cell non-Hodgkin lymphoma in first-degree relatives. Biomed Pharmacother 2012;66:425–7.
[8] Mongia S, Shukla D, Devi BI, et al. Primary cranial vault non-Hodgkin's lymphoma. Neurol India 2003;51:293–4.
[9] Thurnher MM, Rieger A, Kleibl-Popov C, et al. Malignant lymphoma of the cranial vault in an HIV-positive patient: imaging findings. Eur Radiol 2001;11:1506–9.
[10] Magina S, Mesquita-Guimaraes J, Resende C, et al. Scalp tumour as a sign of systemic B-cell lymphoma. J Eur Acad Dermatol Venereol 2004;18:196–8.
[11] Willemze R, Beljaards RC, Rijlaarsdam U. Classification of primary cutaneous large cell lymphomas. Dermatol Clin 1994;12:361–73.
[12] Martin J, Ramesh A, Kamaludeen M, et al. Primary non-Hodgkin's lymphoma of the scalp and cranial vault. Case Rep Neurol Med 2012;2012:616813.
[13] Abdelhamid T, Samra M, Ramadan H, et al. Clinical prognostic factors of diffuse large B cell non-Hodgkin lymphoma: a retrospective study. J Egypt Natl Canc Inst 2011;23:17–24.