Letter to the Editor

DOI: 10.4274/tjh.2013.0432

Central Nervous System Involvement in Primary Adrenal Non-Hodgkin Lymphoma

Primer Böbrek Üstü Bezi Hodgkin Dışı Lenfomasında Merkezi Sinir Sistemi Tutulumu

Padhi Somanath[1], Sahoo Jayaprakash[2]

[1]*Pondicherry Institute of Medical Sciences (P.I.M.S), Department of Pathology, Pondicherry, India*

[2]*Jawaharlal Institute of Postgraduate Medical Education and Research, Department of Endocrinology and Metabolism, Pondicherry, India*

To the Editor,

We read with great interest the case report of central nervous system (CNS) involvement in primary adrenal lymphoma (PAL) in an elderly, HIV-seronegative male patient by Aydın et al. in the December issue of your journal [1]. In spite of initial partial regression of the CNS lesion, the patient succumbed to progressive CNS disease after rituximab-based chemotherapy and whole-brain radiotherapy. Though the pathogenesis and therapeutic aspects of this lymphoma at both anatomic sites were highlighted, there was a lack of precise information regarding adrenal function (prior autoimmune adrenalitis) and detailed immunophenotype of PAL (germinal center or non-germinal center), which could have also influenced the clinical outcome in this patient.

PAL is an enigmatic entity with nearly 200 cases reported in the world literature up to 2013 [2,3]. Out of all parameters studied, adrenal insufficiency, high lactate dehydrogenase (LDH), B symptoms, and initiation of chemotherapy have been reported to be the significant independent predictors of poor prognosis in PAL. Secondary CNS involvement is known to occur in 2%-10% cases of diffuse large B-cell lymphoma (DLBCL) and confers a poor prognosis [3]. Of all reported cases of PAL (1980-2013, including the case by Aydın et al.), 18 patients had CNS involvement [7 (39%) at presentation, 11 (61%) at relapse (within 6 months of diagnosis)]. Their mean age was 63.8 years (range: 42 to 82 years), 17/18 (94.5%) were male, 16/16 (100%) had bilateral PAL, 10/13 (77%) had a mean lesion size of 5 cm or more, 3/18 (16.6%) had disseminated disease at presentation, 1/18 (5.5%) had coexistent secondary involvement of thyroid, 9/11 (82%) had adrenal insufficiency, 11/13 (84.6%) had elevated LDH, and 11/14 (78.5%) had B symptoms. Thirteen of 18 (72%) had DLBCL, 2 had peripheral T-cell lymphoma, 1 had Burkitt-like lymphoma, and the remaining 2 (11%) had non-Hodgkin lymphoma unclassified [2,3] (Table 1). Though patients with PAL are at risk of CNS involvement, there has been no consensus, at present, regarding CNS-directed prophylaxis in these patients. As most of the reported CNS events in PAL cases occurred prior to the rituximab era, larger in-depth prospective studies in the post-rituximab era will, hopefully, throw more light on this topic in future.

Conflict of Interest Statement

The authors of this paper have no conflicts of interest, including specific financial interests, relationships, and/or affiliations relevant to the subject matter or materials included.

Key Words: Primary adrenal non-Hodgkin lymphoma, Central nervous system, Prognosis, Therapy

Anahtar Sözcükler: Primer adrenal non-Hodgkin lenfoma, Merkezi sinir sistemi, Prognoz, Terapi

Address for Correspondence: Padhi SOMANATH, M.D.,
Pondicherry Institute of Medical Sciences (P.I.M.S), Department of Pathology, Pondicherry, India
Phone: +591-413-2025212

Received/*Geliş tarihi* : December 26, 2013
Accepted/Kabul tarihi : December 30, 2013

Table 1. Central nervous system involvement in primary adrenal non-Hodgkin lymphoma (PAL): review of literature (1980-2013, n=18).

Case no., year, place	Age (years)/ sex	Adrenal lesion	Mean size (cm)	AI/LDH†/B symptoms			CNS involvement	Histology	Therapy	Outcome
1, 1983, USA	58/M	B/L	NA	NA	+	+	At presentation	NHL, widespread	None	Death
2, 1983, Japan	44/M	B/L	NA	NA	NA	-	At presentation	NHL, widespread	None	Death
3, 1986, USA	74/M	B/L	<5	+	NA	+	At presentation	PTCL, widespread	None	Death
4, 1996, Canada	42/M	B/L	NA	+	+	+	At relapse (5 months)	PTCL	CHOP, IF, E, MTx	Death (9 months)
5, 1998, Japan	55/M	B/L	10.5	+	+	+	At relapse (4 months)	DLBCL (thyroid +)	BACOD-E, CHOP	Death (7 months)
6, 2000, USA	82/F	B/L	8.5	NA	+	+	At relapse (10 months)	DLBCL	CHOP	Death (10 months)
7, 2001, Israel	60/M	B/L	5	NA	NA	+	At relapse (6 months)	DLBCL	CHOP; MTx, XRT at relapse	Death (14 months)
8, 2002, Korea	61/M	B/L	5.8	+	+	-	At relapse (2 months)	DLBCL	CEOP; XRT at relapse	CR (6 months)
9, 2003, Australia	55/M	B/L	9	+	+	-	At relapse (6 months)	DLBCL	CT	Death (6 months)
10, 2003, Canada	67/M	NA	NA	NA	+	NA	At relapse (6 months)	DLBCL	CT	Death (6 months)
11, 2003, Canada	70/M	NA	NA	NA	+	NA	At relapse (3 months)	DLBCL	R-CHOP	Death (3 months)
12, 2004, Greece	80/M	B/L	9.5	+	+	+	At relapse (2 months)	Burkitt-like lymphoma	R-CNOP	Death (2 months)
13, 2005, France	51/M	B/L	4.5	+	NA	NA	At presentation	DLBCL	MVBP, IT-MTx, ACVBP	Death (16 months)
14, 2008, China	74/M	B/L	6.8	+	NA	NA	At relapse (6 months)	DLBCL	CHOP	Death (6 months)
15, 2010, Japan	58/M	B/L	10	+	+	+	At relapse (6 months)	DLBCL	R-CHOP, XRT	Death (8 months)
16, 2013, USA	81/M	B/L	3	-	-	+	At presentation	DLBCL	None	Death (3 months)
17, 2013, Turkey	75/M	B/L	5.3	-	-	+	At presentation	DLBCL	R-CHOP, XRT	Death (6 months)
18, 2013, Japan	62/M	B/L	6.3	NA	+	+	At presentation	DLBCL	R-MPV, WBRT, IT-MTx	Alive (40 months)

AI: adrenal insufficiency, LDH: serum lactate dehydrogenase, †: level above normal reference range, CNS: central nervous system, M: male, F: female, B/L: bilateral, NA: data not available, +: present, -: absent, NHL: non-Hodgkin lymphoma, PTCL: peripheral T-cell lymphoma, CHOP: cyclophosphamide, doxorubicin, vincristine, prednisone, IF: ifosfamide, E: etoposide, MTx: methotrexate, DLBCL: diffuse large B-cell non-Hodgkin lymphoma, BACOD-E: bleomycin, doxorubicin, cyclophosphamide, vincristine, dexamethasone-etoposide, XRT: radiotherapy, CEOP: cytoxan, epirubicin, vincristine, prednisolone, CR: complete remission, CT: chemotherapy, R: rituximab, CNOP: cyclophosphamide, mitoxantrone, vincristine, prednisolone, MVBP: methotrexate, etoposide, BCNU, prednisone, IT: intrathecal, ACVBP: adriamycin, cyclophosphamide, vindesine, bleomycin, prednisone, MPV: high-dose methotrexate, procarbazine, vincristine, WBRT: whole-brain radiotherapy. Cases 7, 8, 12, 13, 14, and 17 were reviewed by Aydın et al. [1].

References

1. Aydın K, Okutur K, Bozkurt M, Aydın Ö, Namal E, Öztürk A, Pilancı KN, Küçükkaya RD, Demir OG. Primary adrenal lymphoma with secondary central nervous system involvement: a case report and review of the literature. Turk J Hematol 2013;30:405-408.
2. Rashidi A, Fisher SI. Primary adrenal lymphoma: a systematic review. Ann Hematol 2013;92:1583-1593.
3. Ichikawa S, Fukuhara N, Inoue A, Katsushima H, Ohba R, Katsuoka Y, Onishi Y, Yamamoto J, Sasaki O, Nomura J, Fukuhara O, Ishizawa K, Ishinohasama R, Harigae H. Clinicopathological analysis of primary adrenal diffuse large B-cell lymphoma: effectiveness of rituximab-containing chemotherapy including central nervous system prophylaxis. Exp Hematol Oncol 2013;2:19.