Pathology – Research and Practice 208 (2012) 679–682

Contents lists available at SciVerse ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp

Teaching cases

Nodal marginal zone B cell lymphoma with prominent follicular colonization with deletion of chromosome 13

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ARTICLE INFO

Article history:
Received 30 May 2012
Received in revised form 23 July 2012
Accepted 24 July 2012

Keywords:
Marginal zone B-cell lymphoma
Follicular colonization
Chromosome 13
Prognosis

ABSTRACT

Nodal marginal zone B cell lymphoma is a rare type of malignant lymphoma and appears to be heterogeneous. Here we report a 60-year-old woman with stage I splenic type of nodal marginal zone B cell lymphoma with prominent follicular colonization. She was treated only by radiation therapy, and remained free of disease on examination for 4 years. The lymph node cells showed an abnormal chromosome of deletion 13, although neither bone marrow cells nor peripheral blood cells demonstrated the same abnormal chromosome. This type of chromosomal abnormality has not been previously reported and may be related to good prognosis in the present case.

Introduction

Nodal marginal zone B-cell lymphoma (NMZBL), initially recognized as monocytoid B cell lymphoma and parafollicular lymphoma, is a rare type of malignant lymphoma, accounting for only 1.0% of all lymphoid neoplasms in Japan [8]. It is thought to be derived from postgerminal center memory B-cells and appears to be heterogeneous [10]. Here we report a rare case of NMZBL with prominent follicular colonization with a chromosomal abnormality, the deletion of chromosome 13.

Case report

Clinical history

A 60-year-old woman was referred to the division of hematology from her gynecologist because she was suffering from left inguinal lymph node (LN) swelling. She had been diagnosed as having squamous cell carcinoma (SCC) of the uterine cervix (preoperative stage IIa). Since the stage could worsen to IV and the treatment strategy would have to be changed if the inguinal LN was invaded by SCC, an LN biopsy by resection was performed to confirm the histological diagnosis of the swollen LN.

Pathologic findings

At a low power field, the lesion was characterized by a follicular and diffuse growth pattern with a marginal zone component with an incomplete mantle zone (Fig. 1a). Trabecular fibrosis compartmentalized the LN parenchyma. At a high power field, the tumor cells were composed of medium-sized lymphocytes with a moderate amount of clear cytoplasm, irregular nuclei, and small nucleoli (centrocyte-like cells [CCL-cells]) (Fig. 1b). Colonized lymphoid follicles were occupied by neoplastic cells with various numbers of residual follicular center cells (Fig. 1c).

An immunohistochemical study demonstrated that both of the tumor cells exhibiting follicular and diffuse growth patterns with a marginal zone component were CD5− (Fig. 1d), CD10−, CD20+, CD43−, CD79a+, cyclinD1−, Bcl-2−, Bcl-6− (Fig. 1e), surface(s)IgM+ and sIgD−. Residual non-neoplastic follicular center B-cells were CD10+, CD20+, CD79a+, Bcl-2−, and Bcl-6+ (Fig. 1e). Various numbers of CD5+, CD10+, and Bcl-6+ immunostains demonstrated various numbers of germinal center T-cells (Fig. 1e). A CD23 immunostain demonstrated a disrupted follicular dendritic cell pattern, which is characteristic of follicular colonization in

Abbreviations: NMZBL, nodal marginal zone B-cell lymphoma; LN, lymph node; SCC, squamous cell carcinoma; CCL-cells, centrocyte-like cells; MZBL, marginal zone B-cell lymphoma; MALT, mucosa-associated lymphoid tissue; IgH, immunoglobulin heavy chain; BM, bone marrow; PHA, phytohemagglutinin; FISH, fluorescence *in situ* hybridization; PFC, prominent follicular colonization; CLL, chronic lymphocytic leukemia.

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http://dx.doi.org/10.1016/j.prp.2012.07.009

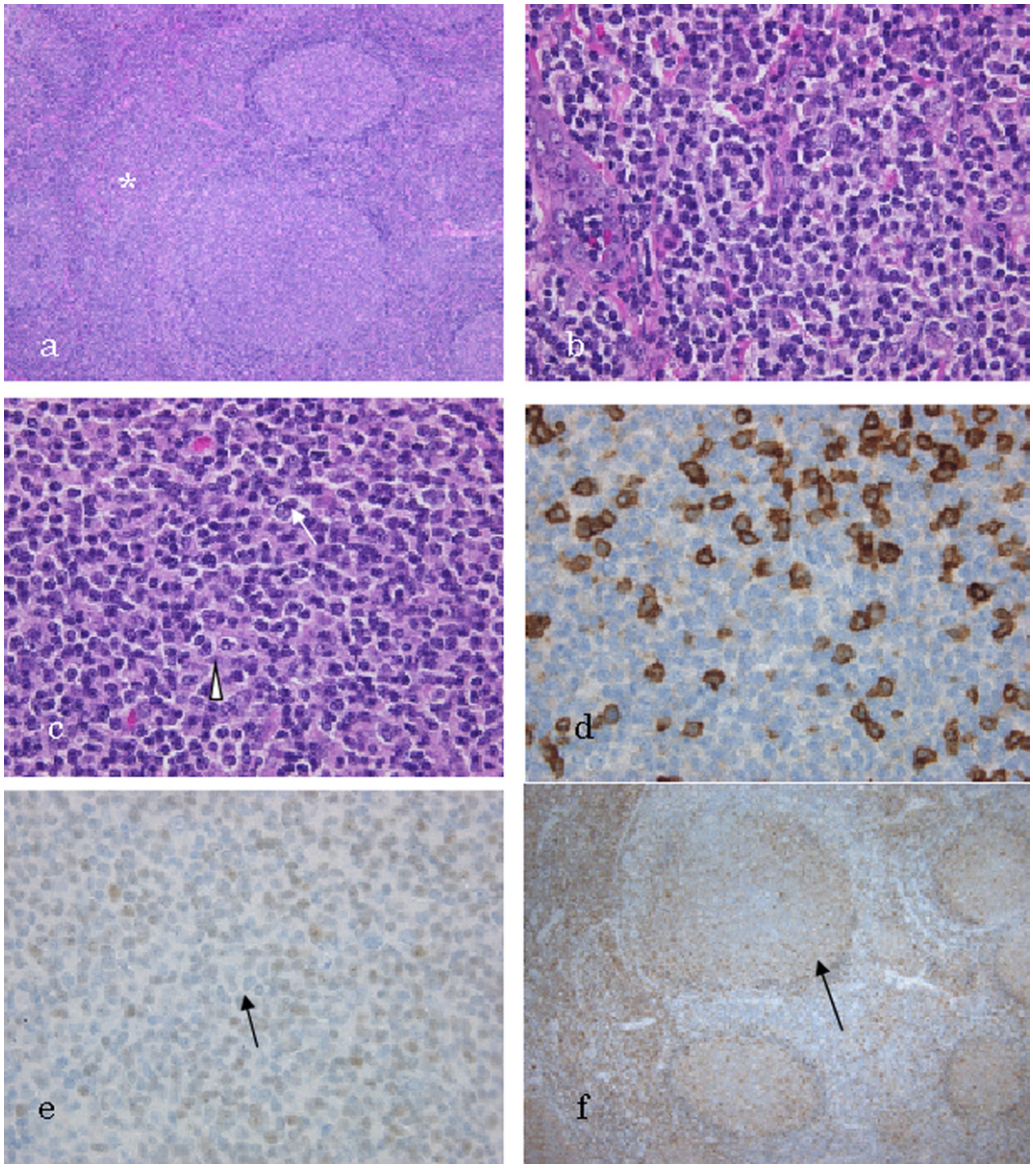

Fig. 1. Histological and immunohistochemical findings of the resected lymph node. (a) At a low power field, the affected lymph node was occupied by irregularly shaped, lymphoid follicular, and diffuse growth patterns with an incomplete mantle zone. Note the marginal zone component (*) HE 10×. (b) In the interfollicular area, tumor cells were composed of medium-sized lymphocytes with a moderate amount of clear cytoplasm, irregular nuclei, and small nucleoli. HE 40×. (c) At a high power field, the colonized lymphoid follicle was occupied by large neoplastic cells (arrow head) with residual germinal center cells (arrow). HE 40×. (d) An immunohistochemical study demonstrated that the residual germinal center T-cells were CD5+, whereas CCL-cells were CD5−. 40×. (e) Tumor cells were Bcl-6− (arrow). Note the Bcl-6+ residual germinal center cells. 40×. (f) CD23 immunostain demonstrated a disrupted follicular dendritic cell pattern characteristic of follicular colonization (arrow). 50×.

extranodal marginal zone B-cell lymphoma (MZBL) of the mucosa-associated lymphoid tissue (MALT) type (Fig. 1f).

Southern blot analysis

The monoclonal expansion of B cells was confirmed by an immunoglobulin heavy chain (IgH) rearrangement band demonstrated by Southern blot hybridization obtained from LN biopsy specimens (Fig. 2a), without the involvement of bone marrow (BM) (Fig. 2b).

Chromosomal analysis and fluorescence in situ hybridization (FISH) analysis

Chromosomal analysis demonstrated −13 in 4 of 10 cells from the resected LN (Fig. 3a). On the other hand, a different chromosomal abnormality, del(20)(q1?), was observed in 15 of 20 BM cells (Fig. 3b). A normal karyotype was observed in peripheral blood cells cultured with phytohemagglutinin (PHA) (data not shown). The fusion of IgH-BCL2, specific for follicular lymphoma, was 0% by fluorescence *in situ* hybridization (FISH) analysis in LN cells (data not shown). Using formalin-fixed, paraffin-embedded tissue, the presence or absence of trisomy 3 and trisomy 18 was examined by FISH [5,11]. However, trisomy 3 and 18 were absent (data not shown, kindly performed by Dr. Hiroshi Inagaki, Nagoya City University).

Clinical course

The patient was diagnosed to have MZBL [3] with prominent follicular colonization (PFC) [9]. Splenomegaly was not observed, and FDG-PET/CT demonstrated that the left inguinal LN was the only site affected (SUVmax = 5.12). The patient showed none of the B symptoms, such as fever, body weight loss, or night sweats. Ultimately, she was diagnosed with stage IIa uterine cervical SCC