A case of langerhans cell histiocytosis associated with mantle cell lymphoma

Mantle hücreli lenfoma ile birlikte gelişen bir langerhans hücreli histiyositozis olgusu

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Abstract

Langerhans Cell Histiocytosis (LCH), which is caused by neoplastic clonal proliferation of Langerhans cells, may be associated with various malignant neoplasms, especially lymphomas, in rare cases. In literature some studies observed an unexpected number of second malignancies, both prior to and after the diagnosis of MCL. Here, we report a case with concurrent LCH and mantle cell lymphoma (MCL) in the same lymph node. To the best of our knowledge, this is the second reported case of LCH associated with mantle cell lymphoma in a lymph node.

Keywords: Langerhans cell histiocytosis, Mantle cell lymphoma, Lymphoma

Öz


Anahtar kelimeler: Langerhans hücreli histiyositozis, Mantle hücreli lenfoma, Lenfoma

Introduction

Langerhans Cell Histiocytosis (LCH) is a rare disease of unknown etiology a neoplasm of the clonal neoplastic proliferation of Langerhans cells. The clinical spectrum ranges from indolent isolated lesions to a life-threatening systemic disease [1,2]. In rare occasions, LCH can be associated with a variety of malignant neoplasms, especially lymphomas [1, 4-12]. While classic Hodgkin's lymphoma is the most common associated lymphoma, other non-Hodgkin's lymphoma cases associated with Langerhans cell histiocytosis have been reported only sporadically in the literature [11]. However, due to the uncommon nature of this association, biological effects and prognostic significance have not been thoroughly investigated [4]. Here, we report a case with concurrent LCH and mantle cell lymphoma in a lymph node.

Case presentation

A 70-year-old man diagnosed with 17p del Chronic lymphocytic leukemia (CLL) at the external center came to our Hematology Clinic. A month after first application he had pleural effusion and pleural biopsy was performed. Microscopically pleural tissue was infiltrated by polymorphonuclear cells and lymphoid cells. Most of the lymphoid cells show positive staining with CD20 and CD5 (pale). CD23 and cyclin D1 were negative. Pleura biopsy diagnosed as “Small lymphoid neoplasm”. During this time, the patient continued to receive CLL therapy because of his first diagnosis. Due to his progression under the treatment, a right inguinal lymph node biopsy was done. In the sections of the lymph node, the normal structure was distorted in most areas and CD3 and CD5 positive T lymphocytes with diffuse patern were observed. Residual lymphoid follicles with few germinal centers (CD21 and CD23 positive) were seen.
The Langerhans cell histiocytosis (LCH) is currently defined as a clonal proliferation of antigen-presenting dendritic type Langerhans cells (LCs) that express CD1a, Langerin (CD207) and S100 protein, and demonstrate Birbeck granules by ultrastructural examination [1,2]. Despite the advances in molecular biology and genetics, etiology is still uncertain. Current literature suggest that LCH is a myeloid neoplasia with inflammatory properties [10]. Chromosomal instability and gene mutations play an important role in the development of this disease [2,10]. Recurrent BRAF V600E mutation has been reported in more than half of LCH patients [12,13]. However, we have not observed BRAF mutation in our case.

Mantle cell lymphoma (MCL) was termed a lymphocytic lymphoma of intermediate differentiation and characterized by atypical small lymphoid cells with wide mantles around benign germinal centers [14]. In literature some studies observed an unexpected number of second malignancies, both prior to and after the diagnosis of MCL. Reasons for this observation are unclear, but it suggests either a genetic predisposition or some other common cause for both tumor groups [15].

LCH is defined concurrently with other tumor types, before, after, or synchronous with other tumor types, especially malignant lymphomas [1,3-9]. Egeler et al. [5] reviewed 91 LCH cases associated with other malignant tumors, the most common being malignant lymphoma (39 cases). A total of 40 cases with association of LCH and Hodgkin Lymphoma (HL) were reported in literature [11]. Proliferation of Langerhans cells in association with HL does not appear to portend a worse prognosis in most reports [9]. West et al. [8] showed a clonal relationship between the FL and Langerhans cell neoplasm with using a combination of immunoglobulin gene rearrangement and fluorescence in situ hybridization studies. Pina-Oviedo et al. [10] reported seven cases of LCH associated with lymphomas included five classical HL, one MCL and one angioimmunoblastic T-cell lymphoma and this is the first report of LCH associated with MCL. In their study all cases were negative for BRAF V600E and MAP2K1 mutations. Therefore they suggested that lymphoma-related LCH is a clinically benign process.

LCH is a rare neoplasm of the clonal neoplastic proliferation of Langerhans cells. Even more rare, LCH can be associated with a variety of malignant neoplasms. To the best of our knowledge, this is the second reported case of LCH associated with mantle cell lymphoma in a lymph node.

References
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