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LETTER TO THE EDITOR

Primary mediastinal large B-cell lymphoma that occurs in an unexpected localization

Beklenmedik bir lokalizasyonda ortaya çıkan primer mediastinal büyük B hücreli lenfoma

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To the Editor,

Primary mediastinal large B-cell lymphoma (PMLBCL) is a mature aggressive large B-cell lymphoma of putative thymic B-cell origin arising in the mediastinum, with distinctive clinical, immunophenotypic, genotypic, and molecular features. Cases that arise outside the mediastinum are very uncommon[1]. The vast majority of patients with PMLBCL present with a localized anterior mediastinal mass in the thymic area. The mass is often bulky and frequently invades adjacent structures, such as the lungs, pleura, and pericardium. Patients present with cough, dyspnea, chest pain, and superior vena cava syndrome[2].

A 27-year-old woman presented with weakness and loss of sensation in her legs. On examination, muscle strength was 1/5 in lower extremities, hypoesthesia under T6, and loss of pain sensation were detected. Thoracal MRI showed a mass originating from the posterior mediastinum. This mass had an extramedullary, extradural component at the right T5-6 foramina indenting to the spinal canal, extending craniocaudally from the middle mediastinum to the level of the aortic hiatus. There was significant spinal cord compression. There was also involved in the T4 and T5 vertebrates and fourth-fifth right costovertebral joints. These findings suggest a mass with a neurogenic origin. However, lymphoma should be thought of in the differential diagnosis. Total laminectomy was performed and spinal cord compression was corrected. Pathological examination of the mass revealed atypical lymphoid cells with medium-large size, eosinophilic and clear cytoplasm. There were alveolar clusters within marked sclerosis. Immunohistochemical analysis showed CD20, CD30, LCA, Pax5, CD23, and MAL expression. Diffuse Large B-cell Lymphoma (DLBCL), NOS, which we expect to see more frequently, is included in the differential diagnosis of a lymphoma located in the posterior mediastinum and invading the spinal cord. In our case CD23 and MAL expression with morphological findings supported PMLBCL. The sensitivity and specificity of MAL expression ranged between 58–72% and 97–100%, respectively[2,3]. CD23 expression is also useful in distinguishing PMLBCL from DLBCL[3,4].

The presence of PMLBCL cases without anterior mediastinal disease raises the question of the origin of PMLBCL as in our case. There are 2 hypotheses about the localization of these lesions: 1- some cases arise from an ectopic thymus, 2-there is a small, undetectable lymphoma in the thymus, but this is difficult to substantiate[4]. In our case, there was no ectopic thymus residue. PMLBCL should be distinguished from DLBCL, due to the different biology and outcome of PMLBCL[5].

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In conclusion, PMLBCL may be seen with abnormal localization and clinical presentation. Immunohistochemical profile including MAL is helpful to detect these cases.

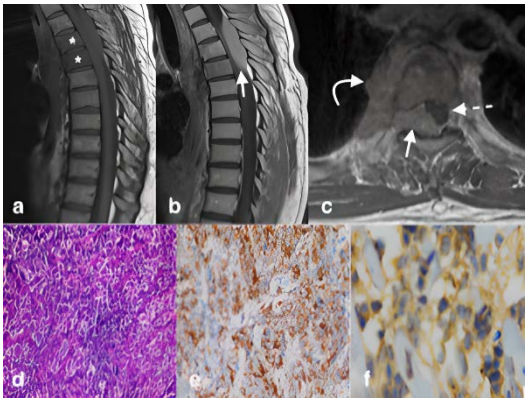

Figure 1. Sagittal pre-contrast T1-weighted images revealed the involvement of the T4 and T5 vertebrae (asterisksin a).Sagittal (b) and axial (c) planes post-contrast T1-weighted images revealed the intraspinal (straight arrow) and posterior mediastinal (curved arrow) components of the mass, and spinal cord compression (dotted arrow). d) Sheets of medium-large cells with abundant pale cytoplasm, separated by alveolar fibrosis (H&E x200). e) Most tumor cells express CD23 (x200). f) Tumor cells also express MAL, with a cytoplasmic accentuation in the Golgi region (x400).

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