Cervical Gray Zone Lymphoma: Case Report

Servikal Gri Zon Lenfoma

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ABSTRACT The differential diagnosis of a neck mass includes many etiologies. Cervical gray zone lymphoma is one of these. Gray zone lymphoma is a very rare disease and represents a range of tumors processing characteristics of both nodular sclerosis classical Hodgkin lymphoma and diffuse large B-cell lymphoma. These lymphomas have been reported using different terms, such as borderline lymphomas, B- cell lymphomas unclassifiable, atypical Burkitt lymphoma, Burkitt-like lymphomas or gray zone lymphomas. Cases reported as gray zone lymphoma in the literature especially are diagnosed in the area of mediastinal region, but not in the cervical region. Here we report a patient who is 47 years old woman with a neck mass for a four year history and diagnosed as cervical gray zone lymphoma which is rare in the literature.

Key Words: Hodgkin disease; head and neck neoplasms

ÖZET Boyundaki kitlenin ayırıcı tanısı birçok etiyolojiyi içermektedir. Servikal gri zon lenfoma bunlardan biridir. Gri zon lenfoma çok nadir görülen bir hastalıktır ve hem nodüler skleroz klasik Hodgkin lenfoma hem de yaygın büyük B hücreli lenfoma özelliklerini içeren tümör aralığını göstermektedir. Bu lenfomalar sınırda lenfomalar, sınıflandırılamayan B- hücreli lenfomalar, atipik Burkitt lenfoma, Burkitt-benzeri lenfomalar veya gri zon lenfomalar gibi farklı terimler kullanılarak bildirilmiştir. Literatürde gri zon lenfoma olarak rapor edilen vaka sunumları servikal bölgede değil, özellikle mediastinel bölgede teşhis edilmişlerdir. Burada dört yıldır boyunda kitle hikayesi olan ve literatürde çok nadir görülen servikal gri zon lenfoma tanısı alan 47 yaşında bir kadın hastayı sunmaktayız.

Anahtar Kelimeler: Hodgkin hastalığı; baş ve boyun tümörleri

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The neck mass is a common clinical entity in almost every age group. Although the differential diagnosis of many diseases, mainly congenital, inflammatory and neoplastic causes should be considered. Gray zone lymphoma is a very rare disease.[1-4] A complete head and neck examination is essential to get to the differential diagnosis. Endoscopic nasal cavity, nasopharynx, oropharynx, hypopharynx and larynx should be displayed. Serological tests and imaging modalities should be performed in case of need. Fine-needle aspiration biopsy or by the clinical case of necessity, excisional biopsy can be facilitated for the diagnosis. Neoplastic causes of neck masses are mainly squamous cell carcinomas, thyroid carcinomas, lymphomas and metastatic carcinomas.

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In this article we report a case who attended to our clinic with a neck mass and diagnosed as cervical gray zone lymphoma which is rare in the literature.

CASE REPORT

A 47 year old female patient presented with a four year history of neck mass and pain. No weight loss, night sweats or fever was reported. Neck examination by digital palpation revealed a 3x4 cm solid mass in the tail section of left parotid region. There was soft tissue on the middle cervical region of the left neck. Her systemic examination including respiratory, cardiac, abdominal region and central nervous system were normal. Routine investigations: hemogram, urine analysis, were normal. Liver enzymes like ALT, AST was high. Serology for HIV, HBV and CMV was negative. EBV IgM was negative but IgG was positive.

A neck ultrasound revealed multiple lymph nodes on the left parotid gland, submandibular gland, anterior and posterior cervical chain, and supra-clavicular region. The largest of these lymph nodes was 20x31 mm. The patient's previous interventional tru-cut biopsy from left parotid gland reported as small lymphoid tissue samples containing loose groups of eosinophil polymorphs and small clusters of histiocytes. Light microscopic examination of sections revealed preserved follicles, histiocytic aggregates, and dispersed cells with large nuclei, prominent nucleoli. Some of these cells were bi or multinucleated. In immunhistochemical study these cells were positive for LCA, CD20, CD30, CD15 and PAX-5; while CD3, CD4, CD5, CD57, CD43, CD45RO, CD23, MUM-1, EBV LMP-1, CD68, bcl-2, cyclin D1 were negative (Figure 1-4).

The patient was extensively investigated for other sites of involvement. Bone marrow aspiration, PET scan was performed. PET CT reported by sections of the head and neck region, lymph nodes on the left pre-auricular, infra-aurikuler, parotid, the left cervical chain level 1B,2A, 2B, 3, 4, 5A, and supra-clavicular localization with increased FDG uptake (SUVmax: 9.8) and the largest one of these hypermetabolic lymph nodes was 32x25mm in diameter (Figures 5, 6). No other sites in the body

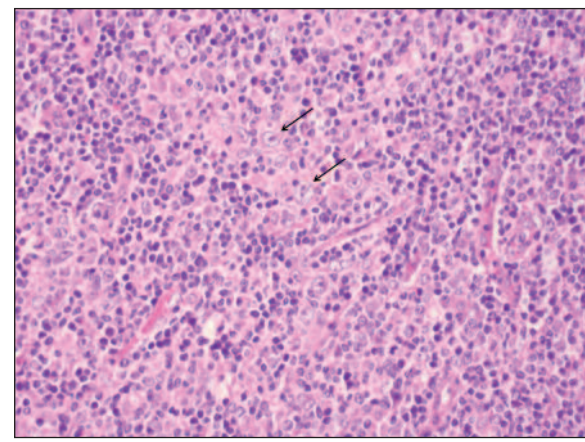

FIGURE 1: Cells indicated with arrows which have huge nucleus, distinct nucleolus were positive for LCA, CD 20, CD 30 and CD 15 and were diagnosed as gray zone lymphoma (x400).

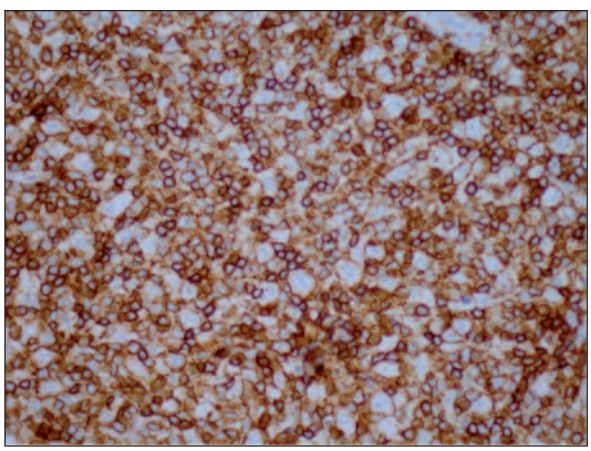

FIGURE 2: Cells positive for LCA (x400).

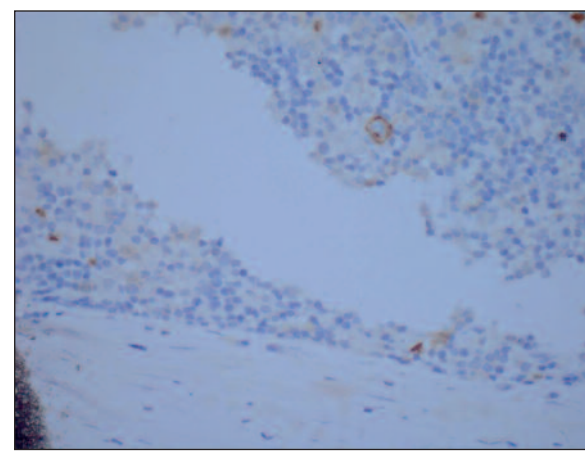

FIGURE 3: Cells positive for CD 30 (x400).

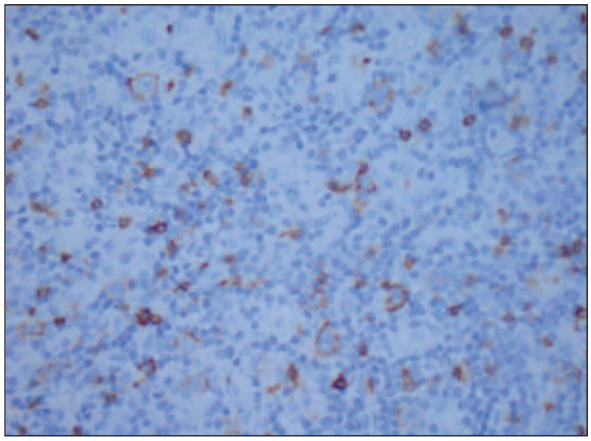

FIGURE 4: Cells positive for CD 20 (x400).

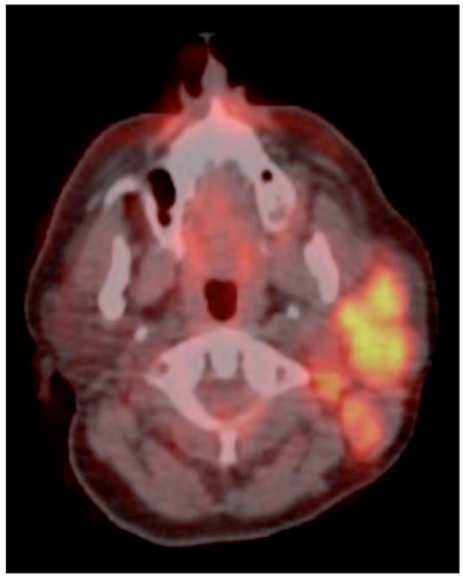

FIGURE 5a: PET-CT scan of the patient that shows increased 18F-FDG uptake on the left pre-auricular, infra-auricular, parotid localization.

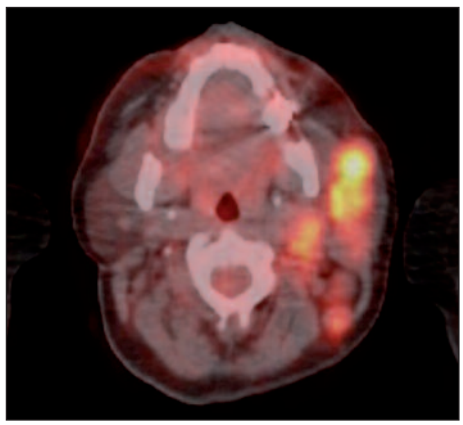

FIGURE 5b: PET-CT scan of the patient that shows increased 18F-FDG uptake on the left cervical chain level.

were found to be affected by the disease. Due to the concurrent, strong expression of LCA, CD20, CD30, CD15 in neoplastic cells combined with the morphological features, the case is diagnosed as "B cell lymphoma, unclassifiable, with features intermediate between diffuse large B cell lymphoma and classical Hodgkin lymphoma" according to WHO 2008 classification. The patient's treatment was planned by Medical Oncology Department with chemotherapy which consisted of R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, Prednisone).

DISCUSSION

The differential diagnosis of a mass in the neck is broad, extensive, and includes both serious and benign etiologies. Accurate diagnosis of a neck mass is critical. In the present study we described a patient with Cervical Gray Zone Lymphoma that exhibited different histological features. Cervical Gray Zone Lymphoma is a new disease entity of lymphoma, and there is no previous report in the literature. The cases usually occur in the mediastinum previously reported. Our case is important due to its localization in the neck.

The current classification of lymphoid neoplasms is based on clinical information morphology, immunophenotype and molecular genetic characteristics. Most lymphomas can be accurately classified. However, some lymphomas present with features transitional between diffuse large B- cell lymphomas (DLBCL) and classical Hodgkin lymphoma (CHL) or DLBCL and Burkitt lymphoma. These lymphomas have been reported in the literature using different terms, such as borderline lymphomas, B- cell lymphomas unclassifiable, atypical Burkitt lymphoma, Burkitt-like lymphomas or gray zone lymphomas.[5] The term "gray zone lymphoma" for borderline cases of Hodgkin lymphoma was introduced for the first time in the proceedings of the "Workshop on Hodgkin's disease and related diseases" in 1998.[6] The updated 2008 World Health Organization (WHO) classification of tumors of hematopoietic and lymphoid tissues recognized this problem and introduced two new provisional categories of B-cell lymphoma unclas-

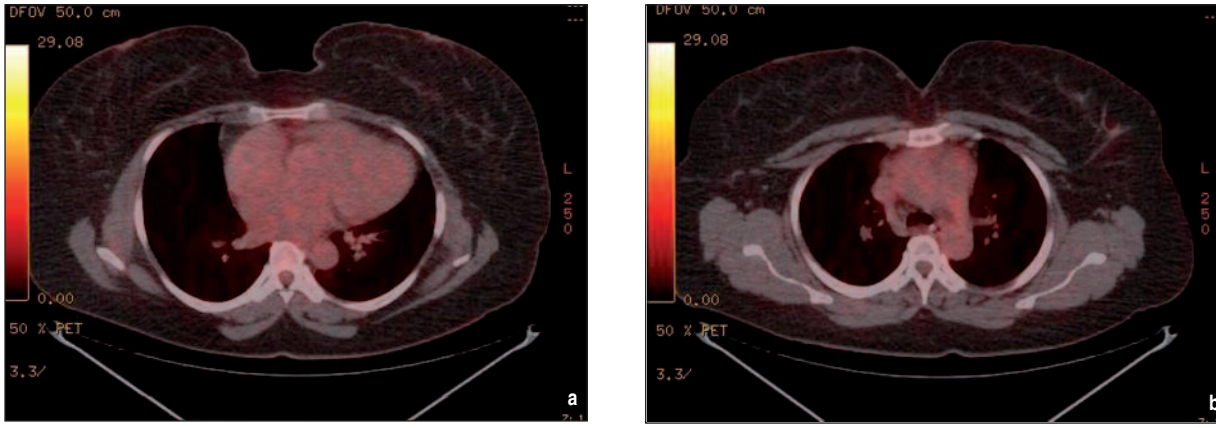

FIGURE 6a,b: PET-CT scan of the patient that shows no 18F-FDG uptake on mediastinal level.

sifiable; one, B- cell lymphoma unclassifiable with features intermediate between DLBCL and CHL and other, B-cell lymphoma unclassifiable with features intermediate between DLBCL and Burkitt's lymphoma.[1-6] The cases usually occur in the mediastinum previously reported.[7-9] Another phenomenon related to Mediastinal Gray Zone Lymphoma (MGZL) is the occurrence of DLBCL and CHL as composite or sequential lymphomas. MGZL, in contrast to DLBCL and CHL, is more common in young men and has a more aggressive clinical course and poorer outcome than either CHL and DLBCL, which emphasizes the importance of keeping them separate with the hope that further studies will reveal whether these cases represent biologically true borderline cases or whether they can be assigned to a specific entity. The treatment approaches for MGZL are yet to be established. Due to rareness of the disorder and the lack of uniform diagnostic criteria, it is improbable that such a trial can be performed, at least in the near future.[7]

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