Case Report / Olgu Sunumu

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Primary Mucosa-Associated Lymphoid Tissue (MALT) Lymphoma of the Urinary Bladder: A Rare Case Report

Mesanenin Primer Mukoza-İlişkili Lenfoid Doku (MALT) Lenfoması: Nadir Bir Olgu Sunumu

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Abstract / Özet

Primary lymphoma of the urinary bladder is rare and comprises about 2% of all extranodal lymphomas. We here aimed to report a fifty six year-old man with complaints of dysuria, pollakuria having a primary urinary bladder lymphoma of mucosa-associated lymphoid tissue (MALT) type. Its differential diagnosis with severe chronic cystitis and other lymphoma types is important and may cause diagnostic problems. Immunohistochemistry is important in these cases.

Key Words: MALT lymphoma, primary urinary bladder lymphoma, extranodal marginal zone lymphoma

Primer mesane lenfoması oldukça nadirdir ve tüm ekstranodal lenfomalar arasında %0,2'den daha az sıklıkla izlenir. Pollaküri, dizüri yakınmaları olan elli altı yaşında erkek hastada mesanede yaygın yerleşimde tanı konan primer mesane MALT tipi B hücreli lenfoma olgusunu, histopatolojik ve immunhistokimyasal bulguları ile birlikte sunmaktayız. Mukoza-ilişkili lenfoid doku lenfoması (MALT lenfoma) ile mesanenin diğer nadir lenfomaları ve yoğun lenfoid infiltrasyon gösteren reaktif durumları ayırıcı tanı güçlüğü oluşturabilir. Bu durumda, immunohistokimyasal yöntem kesin tanı için önem kazanır.

Anahtar Kelimeler: MALT lenfoma, primer mesane lenfoması, ekstranodal marginal zon lenfoma

Introduction

Primary urinary bladder lymphoma is quite rare and represents 0.2% of all primary extranodal lymphomas (1). It is usually of extra nodal marginal zone B-cell lymphoma of mucosa associated lymphoid tissue (MALT) (2, 3) and less frequently of diffuse large B-cell type (4). It is usually seen in the sixth decade. Women are affected 6.5 times more than men (3). Here we present a case with primary low-grade MALT lymphoma of the urinary bladder and review the relevant literature on this rare entity.

Case Report

A fifty-six year old man was admitted to our Urology Outpatient Clinics with complaints of dysuria, pollakiuria and periumblical pain. Cystoscopy revealed diffuse thickening of the bladder mucosa. Biopsies from the neck, base, right and left wall of the bladder were obtained. Histopathologically, diffuse infiltration of atypical lymphoid cells were seen within the mucosal epithelium (Figure 1a-c). This infiltrate was mainly composed of centrocyte-like cells, monocytoid cells and a few centroblast/immunoblasts. Immunohistochemically, the diffuse atypical lymphoid infiltrate stained strongly positive for CD20 and less intensely for bcl-2 (Figure 2a). Reactive non-neoplastic T cells were positive for CD3 (Figure 2b). The proliferation index shown with Ki-67 was 20% (Figure 2c). CD30, CD5 and cyclin D1 were negative. The final diagnosis of extranodal marginal zone lymphoma of MALT was made according to the WHO/REAL System for lymphoid neoplasms (5). After an extensive systemic screening including the hematological, biochemical tests, imaging and bone marrow biopsy revealed no abnormality, thus the case was accepted as primary bladder lymphoma.

Discussion

During the course of systemic non-Hodgkin's lymphoma, the urinary bladder may be involved in 17% of the cases (6). However, a primary lymphoma of the urinary bladder is very rare and it represents 0.2% of all primary lymphomas (1). It tends to occur in the fifth to seventh decades of life with female preponderance (7). The most common presenting symptoms are hematuria, nocturia and dysuria (7). The history of chronic cystitis was reported in some cases (3, 7, 8). Cystoscopically, the most common finding is well-defined intravesical mass at the dome or lateral walls (2, 3, 8, 9) rather than diffuse thickening of the bladder wall (4). Our patient was a fifty six-year-old man with cystoscopic findings of diffuse involvement of the whole bladder and without any remarkable history of chronic cystitis.

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Marginal zone lymphomas are divided into three groups in the WHO/REAL System as 1) Nodal marginal zone lymphoma, 2) Splenic marginal zone lymphoma and 3) Extra-nodal marginal zone lymphoma (5). The most common type of primary bladder lymphoma is extra-nodal marginal zone lymphoma of MALT described in 1990 (1). Since then, more than eighty new cases have been reported (2, 3, 7, 9, 10). The cause of lymphoma in urinary bladder still remains unclear. Since there is no naturally occurring lymphoid tissue in the bladder, it is possible that preexisting chronic cystitis can induce MALT development and may further initiate the lymphoma. However; chronic cystitis is reported to precede in only 20% of MALT lymphomas in the bladder (7, 8). Besides, the lymphoepithelial lesions, a common histopathological feature of MALT lymphoma might also rarely be observed in the areas of chronic inflammation (2, 3). Thus, the presence of lymphoepithelial lesions does not always mean that the lesion is the MALT lymphoma of the bladder. In our case, we found typical lymphoepithelial lesions, both in the neoplastic and nonneoplastic areas. So the immunhistochemistry determined the definitive diagnosis. Other rare types of the lymphoma involving the bladder such as mantle cell lymphoma and follicular lymphoma should also be ruled out. Mantle cell lymphoma cells are immunoreactive for CD5 and CD23, whereas MALT lymphoma cells are not. Similarly, follicular lymphoma cells are positive for CD20 and bcl-2.

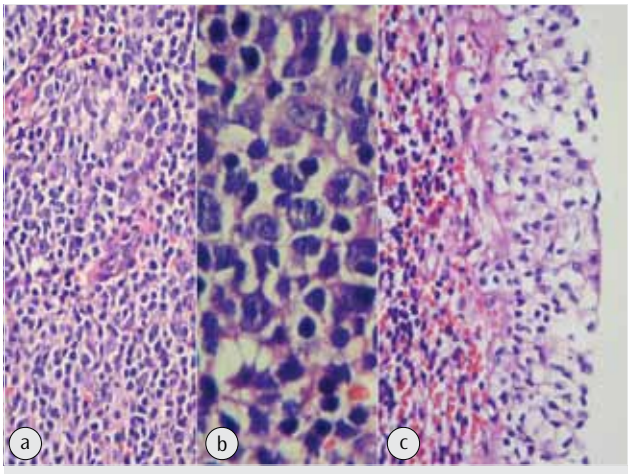

Figure 1. a) Lymphoepithelial lesions of MALT lymphoma, H&E,X200, b) Centrocyte-like cells and immunoblast/centroblasts, H&E,X400, c) Transitional epithelium of the bladder mucosa is also infiltrated with atypical lymphocytes, H&E,X200

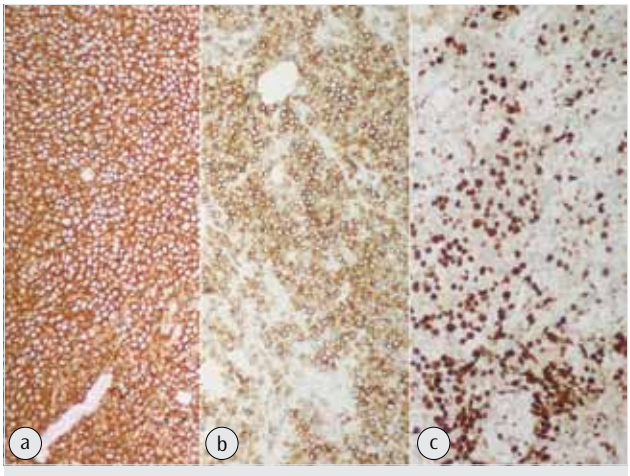

Figure 2. Strong CD20 (a) and bcl-2 (b) positivity, X100, (c) Ki-67 proliferative index was 20%, X100

The prognosis of MALT lymphoma is thought to be favorable with the selection of various treatment modalities (2). The most effective therapeutic procedure remains unclear. In previous reports, chemotherapy, radiotherapy or surgerywere chosen as single or combination therapy. The most frequently used chemotherapy regimen was R-CHOP (10). Radiotherapy might be a choice for low grade disease confined to the bladder. The role of surgery is controversial as it is possible that the lymphoma may recur in other organs and cystectomy definitely reduces the quality of life.

Our patient was not operated on but given 6 cycles of R-CHOP and was in complete remission at his last follow up over 5 years later.

Conclusion

Primary urinary bladder lymphoma is usually of MALT lymphoma and remains localized with a favorable prognosis. Immunohistochemistry is an important diagnostic tool to distinguish it from chronic cystitis and other lymphoma types. Before accepting it as primary, a careful systemic search should be made to rule out the secondary involvement of the bladder with systemic lymphoma which is actually a much more common situation.

Conflict of Interest
No conflict of interest was declared by the authors.

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Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Author Contributions
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Çıkar Çatışması
Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

Hakem değerlendirmesi: Dış bağımsız.

Hasta Onamı: Yazılı hasta onamı bu çalışmaya katılan hastalardan alınmıştır.

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