Primary MALToma of the Prostate

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To Editor,

Mucosa-associated lymphoid tissue (MALT) lymphoma is a distinct disease with specific clinical and pathologic features that may affect diverse organs. MALT lymphoma of the prostate is rare, and only eight cases have been reported in the literature.

A 48-year-old male patient presented with urinary obstruction, pollacuria, and the sensation of burning during urination. Physical examination was normal, except for an enlarged prostate. CTs of the neck, thorax, and upper abdomen were normal, but lower abdominal CT confirmed the enlarged prostate. Serum prostate-specific antigen (PSA) level was 0.38 ng mL^{-1} (normal: <4 ng mL^{-1}) and urological evaluation revealed lower urinary tract obstruction. The patient underwent transurethral resection (TUR) of prostate with the diagnosis of lower urinary tract obstruction. The morphology and immunohistochemical staining of the prostatic specimen were in accordance with marginal zone lymphoma and the patient was diagnosed as the mature small B-cell lymphoid neoplasm of the prostate.

Microscopic sections of the prostate consisted of small to medium sized lymphoid cells of the centrocytic type with circular nuclei and narrow cytoplasm, showing diffuse infiltration. The results of immunohistochemical staining were as follows: CD20(+); CD3(–); CD5(–); CD23(–); cyclin D1(–); few lambda mature plasma cells(+), kappa (focal positive), (Figure 1). MRI showed that the dimensions of the prostate were 53 × 46 × 32 mm. Systemic PET showed increased FDG uptake (SUVmax: 7.3) in the prostate gland. Bone marrow biopsy showed no involvement. The patient was determined to be stage IAE. The patient received external beam radiation therapy to the prostate with a total dose of 40 Gy in 20 fractions, and became disease free within the following 6 months.

Although MALTomas cannot be precisely classified, they originate from mature B-cell lymphoproliferation. This mechanism is related to antigenic stimulation and is classified as extranodal marginal zone indolent B-cell non-Hodgkin's lymphoma. Even though most MALTomas occur in the stomach, occasionally they can be seen in other organs, including the prostate. Primary prostatic MALToma is extremely rare. MALTomas account for 0.1-8% of all lymphomas. Among prostatic neoplasms, lymphomas constitute less than 0.09%.[1] To the best of our knowledge only 8 cases of primary prostate MALToma were reported previously[2] (Table 1).

According to the literature the median age of the 8 reported patients with MALToma of the prostate was 73 years (range: 57-87 years)[2]. The presented case was comparatively younger that 48 years old. In 5 of the reported 8 patients urinary obstruction was observed, as in the presented case. Elevated PSA was reported in 3 of the previous 8 cases, whereas it was not elevated in the presented case. In all, 8 patients (including the presented case) underwent TUR, whereas 1 patient was diagnosed via fine needle biopsy. With the exception of 1 patient, in whom the epididymis and spermatic cord were involved, all other cases had localized stage 1AE disease (including the presented case).

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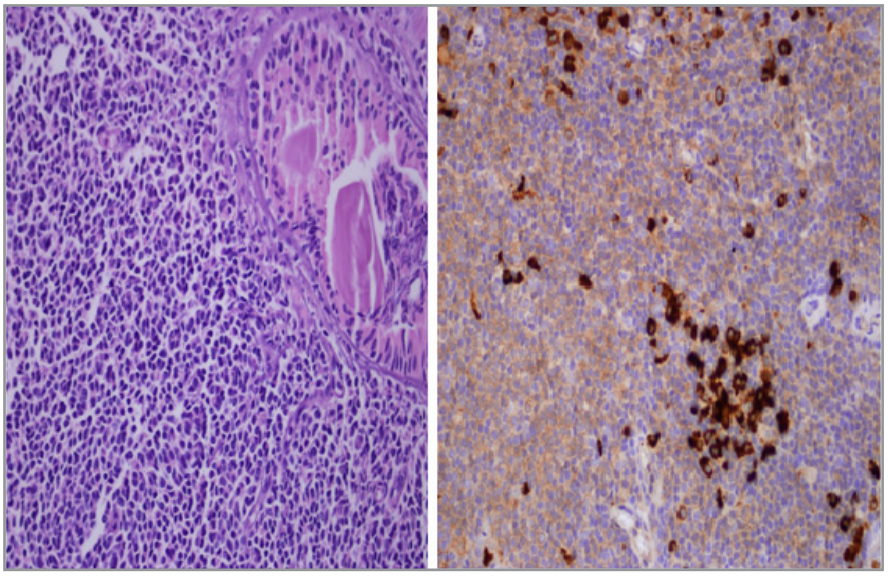

Figure 1. Microscopic sections of the prostate consisted of small to medium sized lymphoid cells of the centrocytic type with circular nuclei and narrow cytoplasm, showing diffuse infiltration

Most non-gastric MALToma cases are diagnosed during the localized disease stage.[3]

Most localized MALToma cases respond well to local therapy, including surgery and radiation therapy. MALToma of the stomach is highly sensitive to radiation therapy.[4] A prospective multicenter phase II study reported that among stage IAE non-gastric MALToma patients treated with radiation therapy at a median dose of 30 Gy, 92% had progression-free survival and 97% had local control.[5] A retrospective study of 70 patients with stage IE and IIE MALToma, including gastric MALToma treated primarily with involved-field radiation therapy only at doses of 25-35 Gy, reported that 5-years disease-free and overall survival rates were 76% and 96%, respectively.[1] Complete response was obtained in the presented case with a 40-Gy dose of radiation therapy.

Despite the fact that MALToma patients respond well to chemotherapy, the efficacy of chemotherapy in early-stage patients is unclear. The addition of anthracycline-based chemotherapy to radiation therapy in a randomized study including 98 previously untreated patients with stage IE primary orbital marginal zone B-cell lymphoma (MZL) did not improve the results obtained with radiation therapy alone.[6] Although concurrent fludarabine and rituximab regimen is highly effective in the treatment of MALTomas, its hematological and infectious toxicity prohibits its widespread use in this patient population.[2]

Among the 8 previously reported patients with primary prostate MALToma, 7 underwent TUR, of which 3 received additional chemotherapy and 2 received additional radiation therapy, whereas 1 patient received radiation therapy only. In the presented case, radiation therapy was given following TUR.

Non-gastric lymphomas have a median relapse time of 2.1-2.7 years. In patients with early-stage disease locoregional relapse and relapse in other MALTs were common. In patients with advanced-stage disease systemic relapse occurred and good results were obtained with salvage therapies.[2] Only 1 of the 8 previously reported prostate MALToma patients was lost due to another disease, and 1 patient relapsed 7 years after TUR and received salvage therapy. The presented case received radiation therapy at a total dose of 40 Gy and became free of disease within the following 6 months.

Non-gastric lymphomas are indolent tumors; 5-year disease-free survival is 70% and overall survival is 93%.[2] It can be concluded that despite the short follow-up periods (5-108 months), these tumors have a good prognosis in prostate patients. Primary prostate MALTomas are extremely rare and more case reports are needed to further elucidation their clinical course, treatment options, and prognosis.

Table 1. Clinical characteristics of 9 prostate MALToma cases

Author	Age	Symptom	Laboratory Finding (s)	Stage	Treatment	Response	Result	Follow-up (months)
Parsonnet et al (4)	67	Urinary tract obstruction	Elevated PSA	I	RT	TR	AW	15
Franco et al (8)	75	Hematuria						
pyuria	N	II	TUR, ChT	TR	AW	12		
Ferreri et al (9)	57	Urinary tract obstruction	N	I	TUR, ChT	TR	AW	18
Cerrani et al (10)	84	Urinary tract obstruction	Elevated PSA	I	TUR	TR	DOC	24
Roggero et al (11)	67	Prostatism	N	I	TUR, RT	TR	AW	36
Luppi et al (12)	87	Urinary tract obstruction	N	I	TUR	NR	NR	NR
Ascoli et al (13)	79	Urinary tract obstruction	N	I	TUR	TR	AW	108
Kawamoto et al (14)	70	Dysuria	Elevated PSA	I	TUR, ChT	TR	AW	5
Our case	48	Urinary tract obstruction		N	I	RT	TR	AW 6

AW: Alive and well; NR: not recorded; ChT: chemotherapy; PSA: prostate-specific antigen; RT: radiotherapy; TUR: transurethral resection; TR: total response; DOC: died of other causes

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