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Case Reports

Anaplastic T-cell lymphoma of the urinary bladder with unspecific clinical and radiological characteristics – a unique case report

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Summary

Purpose: To report a case of extremely rare anaplastic T-cell lymphoma of the urinary bladder in an advanced stage, with completely unspecific radiological features and symptomatology mimicking necrotic myomas. *Case Report:* A 38-year-old women was admitted for pelvic pain, dysuria, enlarged uterus, subfebrile temperature, but without vaginal bleeding and hematuria. On ultrasound scan necrosis of the uterine myomas was suspected. Magnetic resonance imaging revealed an anteuterine tumor (9×9×11 cm) infiltrating the uterus and urinary bladder and retrouterine formation (7×5×7 cm) adhering to the posterior uterine wall and rectum, right kidney hydronephrosis, and parailiac lymphadenopathy. Cystoscopy confirmed the presence of the tumor on the posterior bladder wall and bladder roof infiltrating the right ureteral orifice. Histopathological and immunohistochemical analyses of tumor biopsy confirmed the presence of T-cell anaplastic ALK+ non Hodgkin's lymphoma of the urinary bladder. *Conclusion:* This case report shows that pelvic pain and dysuria alone can imply on urinary bladder tumors even in the absence of hematuria. Additionally, uniqueness of this case lies in the younger age of the female patient. Moreover, the authors showed for the first time that lymphomas could spread locally into both uterus and intestines, without systemic dissemination.

Key words: Urinary bladder; T-cell lymphoma; Pelvic pain; Dysuria; Necrotic uterine myoma.

Introduction

Out of all lymphomatoses, the extranodal ones account for 25-40% of all cases [1]. The most common locations of the primary extranodal lymphoma include gastrointestinal tract, connective tissue, and skin [1]. Primary malignant lymphoma of the urinary bladder is very rare lymphoma type originating in the urinary bladder with no known lymphoma elsewhere [2-4]. It represents 0.2% of primary lymphomas and is usually of B-cell origin [2-4].

This case report describes a unique case of the T-cell lymphoma of the urinary bladder in an advanced stage, with unspecific clinical and radiological features, in a female patient admitted for suspected necrosis of the uterine myomas. Diagnosis of primary T-cell lymphoma of the urinary bladder was made based on clinical, radiological, and histological findings.

Case Report

A 38-year-old female patient was referred to Clinic of Ob/Gyn due to pain in the small pelvis, primary localized in the suprapubic region, occurring intermittently for the past two months. During previous two weeks patient had dysuria without bacteriuria, self-treated only by herbal tea. She denied having urinary frequency or any other complaints that might imply urinary bladder pathology. Furthermore, the patient did not have hematuria or irregular vaginal bleeding. In the last 24 hours her body temperature had risen to 37.8°C. The patient had fatigue and dizziness and stated that she had lately lost substantial weight. Ultrasound scan, performed in primary healthcare institution, revealed enlarged uterus with sonographic texture indicative of necrotic changes within the uterine myomas.

Gynecological examination revealed a tumor formation in the small pelvis in the uterine projection, solid, with an uneven surface, immobile, tender, and approximately 15 cm in diameter. Another solid, smooth, tender, and restrictively mobile tumor was also palpated behind the uterus through the posterior vaginal fornix. Ultrasound examination of the small pelvis and abdomen confirmed that the uterus was enlarged and non-homogenous. Both ovaries were typically positioned with normal ultrasono-

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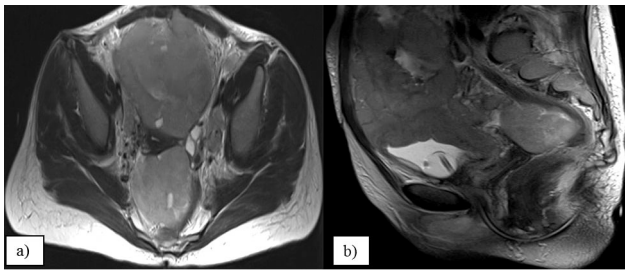

Figure 1. — MRI: a) axial image showing infiltrative urinary bladder lesion and tumor mass in the presacral region, as well as bilateral enlargement of the parailiac lymph nodes; b) sagittal image showing diffuse intestinal infiltration; catheter is placed into the bladder.

graphic characteristics. An anteuterine non-homogenous tumor formation sized 120×62 mm was seen in the projection of the urinary bladder. Another non-homogenous tumor formation sized 50 ×43 mm was present in Douglas pouch. Second degree hydronephrosis of the right kidney was also diagnosed.

Laboratory tests performed upon admission indicated increased levels of the white blood cells, sedimentation, C-reactive protein, platelets, fibrinogen as well as anemia. Urine sediment suggested presence of the fresh erythrocytes. Vaginal swab revealed physiologic flora. MRI showed an infiltrative urinary bladder lesion and tumor mass in the presacral region, as well as a bilateral enlargement of the parailiac lymph nodes and diffuse intestinal infiltration (Figure 1). An anteuterine tumor sized 9×9×11 cm infiltrating the uterus was observed, as well as a retrouterine mass sized 7×5×7 cm adhering to the posterior uterine wall and rectum. There were no disseminated metastases on liver, lungs, brain, bones, and other organs that could be visualized by MRI. Cystoscopy revealed a large tumor plate on the posterior wall, toward the bladder roof, of whitish color with infiltration of the right ureteral orifice. Tumor biopsy was performed and it was accompanied by excessive hematuria.

Histopathological and immunohistochemical examinations documented presence of T-cell anaplastic ALK+ non Hodgkin's lymphoma (Figure 2). The tumor was composed of large cells with abundant cytoplasm and pleomorphic, often horseshoe-shaped or kidney-shaped nuclei. Tumor cells were CEA-, CK-, vimentin+, actin-, desmin-, S-100-, CK7-, synaptophysin-, EMA+, LCA+, MPO-, NSE-, CD117-, Pax-5-, CD20-, CD3+, CD4+, CD5+/-, CD7-, CD8-, CD43+/-, CD45RO+/-, CD56-, CD30+, ALK-1+, CD68-, bcl-2-, EMA+, CK-, NSE-, Pax-5-, CD34-, Granzyme B, CD138-, and Ki 67+ in 80% of tumor cells.

On the 14th hospitalization day, the patient developed generalized lymphadenopathy, abdominal distension, as well as edema of the lower extremities without venous thrombosis. Patient's general condition exacerbated and renal insufficiency developed. In spite of regularly performing dialysis as well as all supportive medications applied in intensive care unit lethal outcome ensued on the 20th hospitalization day.

Discussion

This unique case report is important for proper diagnosis of pelvic tumors, as this very rare primary T-cell lymphoma of urinary bladder even in an advanced stage might present with unspecific clinical and radiological features.

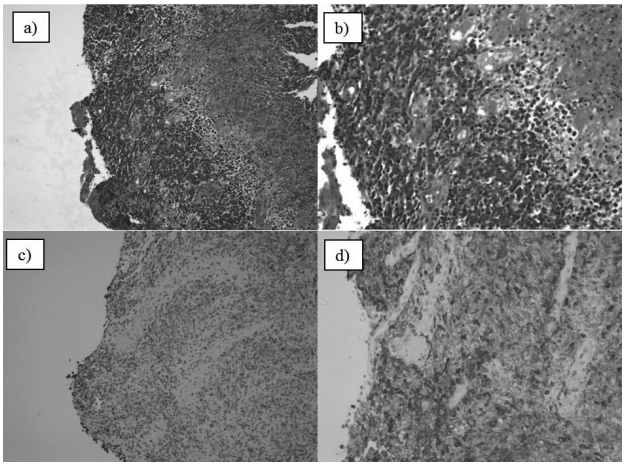

Figure 2. — Histopathological findings of the tumor biopsy showing diffuse infiltration of urinary bladder wall with large pleomorphic T-cells. a) Hematoxylin and Eosin staining, magnification ×10; b) Hematoxylin and Eosin staining, magnification ×20; c) negative cytokeratin staining, magnification ×10; d) positive leukocyte common antigen staining, magnification ×20).

The most common lymphomatoses of the urinary bladder are mucosa-associated lymphoid tissue (MALT) and diffuse large B-cell lymphomas [3, 4]. Bladder lymphomas can be of primary (rare form of non-Hodgkin's lymphoma that is confined only to the urinary bladder) or secondary origin (metastatic, disseminated or recurrent disease) [5, 6]. Primary bladder lymphoma is very rare comprising ≤ 5% of non-urothelial tumors of the lower urinary tract, occurring in 1-2/1000 patients [2-4]. Secondary involvement of the bladder as part of advanced stage systemic lymphomatoses can occur in up to 50% of cases [1, 6, 7]. Lymphomas of the urinary bladder (especially MALT) have a female predominance, and most cases of lymphoma of the urinary bladder occur in middle-aged or older (≥ 60 years) women [7]. A review of the current literature data identified 1,264 patients with primary urinary tract lymphomas, out of which less than 100 had primary lymphoma of the urinary bladder and only few of the T-cell type [3-9]. Anaplastic large cell lymphoma (ALCL) is even more rarely found in urinary bladder and is usually associated with young age at presentation, male predominance, and extranodal involvement [10, 11]. It is typically diagnosed in an advanced stage and the disease is rapidly progressive. Bladder involvement is a sign of widely disseminated disease and it is associated with a very poor prognosis [10, 11]. This was true for the present patient as well.

Major symptoms of any bladder tumor, including lymphomas, are intermittent hematuria, dysuria, and urinary frequency [7-9]. Literature data show that in all previously published cases of urinary bladder lymphomas, visible hematuria was present, while dysuria and ureterohy-

dronephrosis were found in just few cases each [7-9]. The present patient did not have macrohematuria prior to admission and complained only of dysuria and pelvic pain, mostly suprapubic, regardless of the advanced stage of the disease. Only weight loss might have implied on grave malignancy. Moreover, she had right ureterohydronephrosis due to obstruction of the right ureteral orifice by the tumor. All these characteristics make the presented case one of a kind, with exceptional presentation and unusual symptomatology of primary T-cell lymphoma of the bladder. This atypical clinical presentation resembling necrotic myomas, caused incorrect patient referral to gynecology rather than urology department. Therefore, this case is clinically unique as, according to available literature data, this is the first case of bladder lymphoma that infiltrated both uterus and intestines mimicking gynecologic pathology, and without hematuria.

The diagnosis of bladder lymphomas is usually a diagnosis of exclusion, due to the fact that it is so rarely encountered and with unspecific symptomatology. The lesions are usually diagnosed as urothelial carcinoma, while differential diagnosis also encompasses gynecological tumors [6, 9]. Pelvic imaging (ultrasound, MRI, CT, and PET) is essential in preoperative assessment [6, 11]. T-cell lymphoma is most commonly radiologically seen as sessile solitary mass (66% of cases), multiple sessile mass (14%) or polypoid mass (10%) [7, 12]. A case of T-cell lymphoma containing calcification within it was diagnosed using MRI [11]. In the present patient, ultrasonography was misleading. MRI scan revealed the tumor of infiltrative nature, which, based on its characteristics, could not be differentiated from the transitional cell carcinoma (TCC).

Tissue biopsy obtained by cystoscopy and immunohistochemical tests are usually necessary in order to determine the anaplastic type of lymphoma, since clinical and radiological characteristics are similar as in any urinary bladder carcinoma [12-14]. Immunohistochemical staining of urinary bladder lymphomas varies by the tumor subtype. However, all lymphomas of the urinary bladder stain negatively with Pan-keratin, vimentin, CK20, and CK7 [7, 9]. Tissue samples of the present patient were also CK7 and CD20 negative, which determined the final diagnosis.

Treatment of primary lymphoma of the urinary bladder is not uniform and the procedures employed comprise radical cystectomy, radiation therapy, and chemotherapy [13, 14]. Radiotherapy is the treatment of choice for the MALT type of lymphoma of the urinary bladder [7]. Due to a rapid deterioration in overall condition the authors had a chance to only apply symptomatic therapy. Death occurred in less than one month after admission to hospital, as the patient already had advanced disease.

Conclusion

In conclusion, it could be stated that this is a unique case of anaplastic T-cell lymphoma of the urinary bladder with completely unspecific clinical characteristics. It was proven for the first time that pelvic pain and dysuria can imply on urinary bladder tumors even in the absence of hematuria. Moreover, the authors showed that this tumor type could spread locally into uterus and intestines, without systemic dissemination. Therefore, lymphomas, although rarely found, should always be considered under differential diagnosis of bladder tumors. The present case report is significant for proper diagnosis of pelvic tumors in females, as this very rare primary T-cell lymphoma of urinary bladder in an advanced stage might be presented by unspecific radiological features. When assessing similar cases MRI is far more reliable than ultrasound, but the definite diagnosis is based on histopathological analysis of bladder tumor biopsy. Therefore, all female patients with pelvic pain and masses need to be checked by both gynecologist and urologist.

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