Primary Urethral Plasmacytoma: A Case Report and Literature Review

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Plasmacytomas of the urethra are extremely rare neoplasias; they may occur as isolated tumors or concomitantly with generalized multiple myeloma. Herein, we describe the clinical presentation and characteristics of a patient with primary plasmacytoma of the urethra. A 51-year-old man presented with terminal hematuria and a palpable penile mass. Magnetic resonance urethrography revealed a 3-cm long stenotic segment along which the urethral mucosa was found to be irregular. On urethroscopy, papillary mucosal projections extending to the presphincteric area were noted. Lesions were found to be composed primarily of neoplastic plasma cells capable of producing mainly lambda light chain. Upon diagnosis, the patient received external beam radiation therapy targeting the pelvic region. The lesion diminished in size progressively during the treatment course. He was disease-free after 6 months. Although it is a relatively rare disease, primary urethral plasmacystoma should be considered in the differential diagnosis of urethral tumors and radiation therapy should be an integral part of the treatment strategy.

Key Words: extramedullary plasmacytoma, multiple myeloma, radiotherapy, urethra
(*Kaohsiung J Med Sci* 2008;24:274–7)

Multiple myeloma is characterized by the disordered production of immunoglobulins by malignant plasma cells located in bone marrow. Localized form of malignant plasma cell tumors are known as extramedullary plasmacytomas. They represent unusual accumulation of neoplastic monoclonal plasma cells causing soft-tissue mass. Extramedullary plasmacytomas occur mainly in the upper respiratory tract. Deposits in the urogenital tract (bladder, kidney) have also been reported. However, plasmacytoma involving primarily urethral tissue is an extremely rare condition with only seven cases having been reported so far. Herein, we describe the clinical presentation and characteristics of a patient who was diagnosed with primary plasmacytoma of the urethra.

Received: Sep 4, 2007 Accepted: Nov 2, 2007
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Case Presentation

A 51-year-old man presented to our clinic with terminal hematuria and a palpable penile mass. Except for mild dysuria, the patient had no obstructive or irritative voiding symptoms. He had no history of recurrent urinary tract infection, calculi, or urinary incontinence. Physical examination revealed a palpable mass located on the ventral aspect of the penis that was painless, immobile, 1.5 cm in length and with indefinite borders. Routine biochemical and hematologic parameters were within normal ranges. Chest X-ray findings were normal. Microscopic examination of urine sediment revealed >15 red blood cells and 2 white

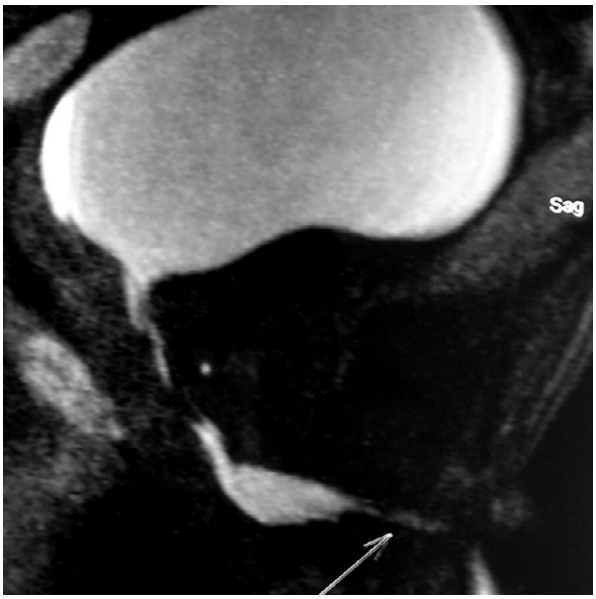

Figure 1. *Magnetic resonance urethrography shows a 3-cm long stenotic segment along which the urethral mucosa was found to be irregular (arrow).*

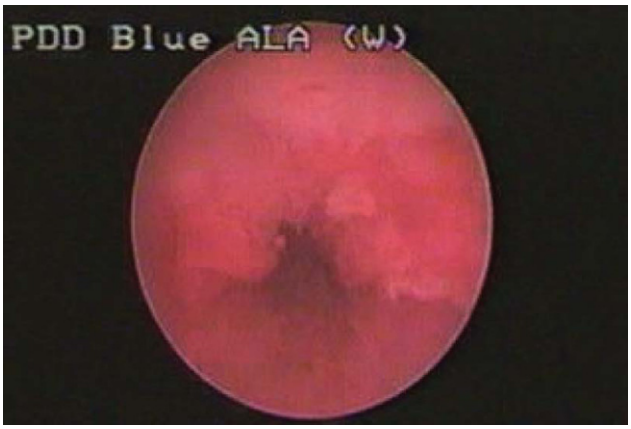

Figure 2. *Urethroscopic image of papillary mucosal projections partially obstructing the lumen.*

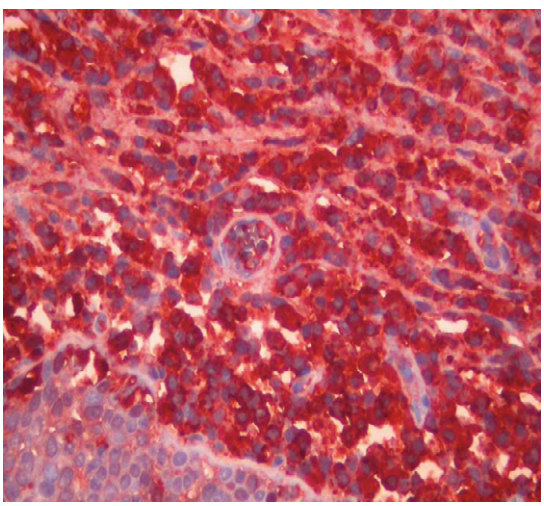

Figure 3. *Neoplastic plasma cell deposits (hematoxylin & eosin, 360×).*

blood cells per high power field. Ultrasonography, intravenous pyelography and computed tomography (CT) of the abdomen and pelvis excluded all possible pathologies. After these basic evaluations in the context of hematuria work-up, we decided to perform penile magnetic resonance imaging (MRI) since the patient had a palpable penile mass.

Penile MRI showed a 3.5-cm long, irregular bulbar urethral mass with contrast enhancement. No other space-occupying lesion was noted. In addition, a pathologically enlarged (2 cm in greatest dimension) lymph node was detected in the left inguinal region. MR urethrography revealed a 3-cm long stenotic segment along which the urethral mucosa was found to be irregular. This narrow segment was localized at the proximal end of the penile urethra (Figure 1). Similar findings were revealed by conventional retrograde urethrography. On endoscopic examination of the urethra, papillary mucosal projections originating from the level of the fossa navicularis, extending to the presphincteric area, were noted (Figure 2). The bladder was free of any neoplastic lesion. Multiple biopsy specimens from the papillary lesions were obtained using cold-cup biopsy forceps. On histopathologic examination, the lesions were found to be composed primarily of neoplastic plasma cells (Figure 3). Apart from plasma cells, AL type amyloid deposits were also noted. On antigenic analysis, 20% of the cellular population was CD20(+), which signified B lymphocyte phenotype. The rest of the population was composed of CD38(+) plasma cells. The majority of the plasma cells (90%) harbored lambda light chain. Diffuse AL type amyloid deposits which stained positively with Congo red were also of diagnostic value. This positive reaction persisted after the specimens were treated with permanganate, which was strongly suggestive of primary amyloidosis.

To ascertain the diagnosis of extramedullary plasmacytoma, bone marrow biopsy was performed. Plasma cell infiltration was not evident in bone marrow. Whole body bone scintigraphy was negative for metastatic osseous lesion. No osteolytic lesions typical of multiple myeloma was noted on X-ray evaluation

of the axial skeleton. Serum protein electrophoresis was negative for monoclonal "M" spike, which is one of the major diagnostic criteria of multiple myeloma. Bence-Jones proteinuria, which signifies the excess excretion of immunoglobulin light chains in urine, was not found in the patient. To confirm the neoplastic nature of the lesion, positron emission tomography was carried out. Fluorodeoxyglucose uptake was significantly higher along the penile shaft and left inguinal region. This finding demonstrated the active proliferative nature of the lesion. Based on these findings, a diagnosis of solitary extramedullary plasmacytoma was made.

On diagnosis, the patient received a total dose of 40 Gy (200 cGy/fr × 20fr) of external beam radiation therapy targeting the perineal region, which was tolerated well. His symptoms subsided after the initiation of radiotherapy. The lesion progressively diminished in size during the treatment period. This alteration was evident both clinically and radiologically. After a follow-up period of 6 months, he was disease-free. On control imaging studies, there was no sign of residual disease or progression.

Discussion

Multiple myeloma is a malignant hematologic disease of plasma cells that is characterized by the presence of monoclonal immunoglobulins or light chains in the serum and urine. Its main feature is bony destruction caused by focal plasma cell tumors known as plasmacytomas. Malignant plasma cells have the potential to disseminate and form metastatic deposits in extraosseous tissues. Dissemination of the neoplastic plasma cells should be ruled out by bone marrow aspiration/biopsy, X-ray study of the axial skeleton, immunoelectrophoresis of serum and urine before a diagnosis of primary extramedullary plasmacytoma can be made. Extramedullary plasmacytoma more commonly affects males between the ages of 50 and 70. Its main site of occurence is the upper respiratory tract (76%). Other sites include lymph nodes and spleen (6%), the lower respiratory tract (4%), skin and subcutaneous areas (3.5%), gastrointestinal tract (3%) and the thyroid gland (3%) [1]. The urethra is an extremely rare localization for this kind of tumor. To our knowledge, only seven cases of primary urethral plasmacytoma have been reported in the English literature [2–4].

In the urinary tract, bladder and kidney plasmacytomas are treated with various combinations of surgery, chemotherapy and radiotherapy. Involvement of the regional lymph nodes is a frequent finding in extramedullary plasmacytomas. This is the reason why regional lymph nodes should also be included in the radiation field for plasmacytomas [1]. Optimal treatment of renal plasmacytomas consists of radical nephrectomy and local irradiation of the tumor bed [5,6]. Meanwhile, plasma cell deposits involving the bladder can be managed by surgery with or without radiotherapy or radiochemotherapy [7].

The overall recurrence rate for extramedullary plasmacytomas was found to be 30% in a review of the literature [8]. In general, the 10-year survival rate of patients with solitary plasmacytoma is estimated to be in the range of 50–60% [8]. However, it was reported that plasmacytomas can potentially disseminate in 90% of cases up to 20 years after the initial diagnosis [9]. For this reason, lifelong follow-up of these patients is mandatory. Once dissemination is evident, chemotherapy may be a treatment alternative.

To date, six cases of urethral plasmacytoma were treated with surgery (transurethral resection) and adjuvant radiotherapy and one with only radiotherapy. Plasmacytomas are known to be radiosensitive neoplastic lesions. Our patient with urethral plasmacytoma received radiotherapy including the site of nodal involvement. Wiltshaw noted that only less than 2% of extramedullary plasmacytomas were refractory to radiation therapy with additional lymph node surgery [8]. Our case is the second in whom local control was achieved with solely radiotherapy during a follow-up period of 6 months. The first case was a 35-year-old man who received radiotherapy with a total dose of 45 Gy. He remained disease-free without any therapy-related side effects after a follow-up of 36 months [10]. However, radiotherapy cannot be regarded as the primary treatment modality for urethral plasmacytoma based on the outcome of these two cases with limited follow-up. Nonetheless, radiotherapy may be a curative tool for these patients with minimal morbidity.

In conclusion, owing to the radiosensitive nature of extramedullary plasma cell deposits, radiation therapy should be an integral part of the treatment strategy for urethral plasmacytoma. Since there is a continuing risk of distant metastasis throughout the natural course of this disease, lifelong follow-up is mandatory.

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