Case report Olgu sunumu

Coexistence of primary amyloidosis of the bladder and urothelial carcinoma *in situ*

Mesanede primer amiloidoz ve *in situ* ürotelyal karsinom birlikteliği

Şükrü Kumsar, Müge Ünlükaplan, Levent Peşkircioğlu, Hakan Özkardeş

Department of Urology, Başkent University School of Medicine, Ankara

Summary

A 64-year-old woman presented with painless gross hematuria and frequency. She had no other symptoms. Her past or familial medical history was insignificant. Physical examination was normal. Pelvic ultrasound demonstrated a localized thickening in the right posterolateral part of the bladder wall. Cystoscopy revealed an erythematous mucosa which was slightly protruding into the lumen. This area of about 3 cm in diameter covering the ureteral orifice was resected transurethrally and the orifice was exposed. Histopathological evaluation revealed urothelial carcinoma *in situ* and focal amyloidosis.

Key words: Amyloidosis; hematuria/etiology; urinary bladder neoplasms.

Özet

Altmış dört yaşında kadın hasta ağrısız bariz hematüri ve sıklık yakınmalarıyla başvurdu. Başka herhangi bir semptomu ve özgeçmişinde ya da soygeçmişinde özellik yoktu. Fizik muayenesi normaldi. Pelvik ultrasonda, mesane duvarının sağ posterolateral kısmında kalınlaşma görüldü. Sistoskopide, lümen içine hafifçe çıkıntı yapan eritematöz mukoza görüldü. Üreter orifisini çevreleyen ve yaklaşık 3 cm çapında olan bu alan transüretral olarak rezeke edildi ve orifis açığa çıkarıldı. Histopatolojik incelemede *in situ* ürotelyal karsinom ve fokal amiloidoz birlikteliği izlendi.

Anahtar sözcükler: Amiloidoz; kadın; hematuri/etyoloji; mesane neoplazileri.

Submitted (Geliş tarihi): February 13, 2008 *Accepted after revision (Düzeltme sonrası kabul tarihi):* December 16, 2008

Primary amyloidosis of the urinary bladder is a rare disease that can mimic bladder cancer on cystoscopic examination as well as with its clinical presentation of painless gross hematuria. To our knowledge, we report the first case of primary bladder amyloidosis associated with urothelial carcinoma *in situ*.

Case report

A 64-year-old woman presented with painless gross hematuria and frequency in February 2007. She was not a smoker. She had no history of urinary tract infection, urolithiasis, or any chronic inflammatory disease. Physical examination showed no abnormality. Urinalysis demonstrated numerous red blood cells and her urine was sterile. Complete blood count and biochemistry were within normal ranges. Pelvic ultrasound demonstrated thickening of the right posterolateral bladder wall.

On cystoscopy, an edematous and erythematous bladder mucosa covering the right periureteral orifice was noted. That region of the bladder wall was resected transurethrally and the orifice was exposed. Histopathological examination revealed urothelial carcinoma *in situ* (Fig. 1a) with focal amyloidosis (Fig. 1b). Subsequent investigations for evidence for systemic amyloidosis were all negative. Fifteen days after transurethral resection, she received weekly intravesical BCG for six weeks. Upon completion of this treatment, abdominal magnetic resonance imaging performed in the third month demonstrated prominent dilatation of the right ureter and renal collecting system (Fig. 2a) with thickening of the right lateral posterior bladder wall (Fig. 2b), suggesting an invasive bladder tumor. Control cystoscopy showed a bulging, erythematous flat mass of about 3 cm covering the right ureter. Following a deep transurethral resection, the right ureter was exposed and a double J stent was placed. Histopathological examination revealed only granulomatous inflammation and amyloid deposits in the stroma and vessel walls. The double J stent was removed after four weeks. Control cystoscopies performed at 6 and 9 months were totally normal and washout cytology was negative. The

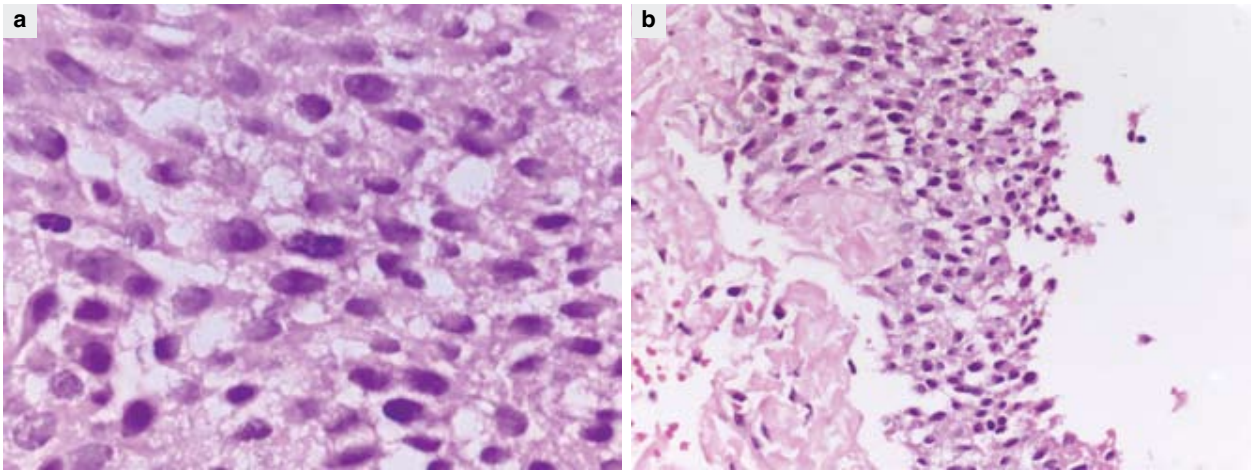

Figure 1 **(a)** Mitotic figure (at the center of the visual field) can be seen in the urothelial epithelium, together with minimal nuclear pleomorphism, polarity loss, and increment in stratification (H-Ex400). **(b)** Amyloid deposition in the subepithelial area is seen in the urothelial epithelium, together with minimal nuclear pleomorphism, polarity loss, and increment in stratification (H-Ex40).

patient had no urinary symptoms or pelvicalyceal dilatation.

Discussion

Amyloidosis is a disorder of protein metabolism characterized by extracellular deposition of abnormal protein fibrils. It may either be localized or systematically distributed throughout the body.[1] Primary amyloidosis of the urinary tract is a rare condition with less than 200 cases reported.[2] It mainly presents as intermittent gross hematuria. A few patients may present solely with irritative bladder symptoms. The onset of disease is often in the 6th to 8th decades of life similar to that of transitional cell carcinoma.[3] Clinical, radiological, and cystoscopic findings closely simulate a bladder malignancy. Therefore, a correct diagnosis of amyloidosis requires a histopathologic study of the bladder biopsy. Typically, in localized amyloidosis of the bladder, amyloid deposits are demonstrated in the lamina propria and muscularis propria; however, we and others[4] noted vessel wall involvement.

Bladder amyloidosis is generally treated conservatively and surgery, primarily endoscopic resection, has been successful.[3] Medical treatment methods such as intravesical dimethyl sulfoxide instillation and oral colchicine have also been used.[5] More aggressive procedures are at times necessary, such as

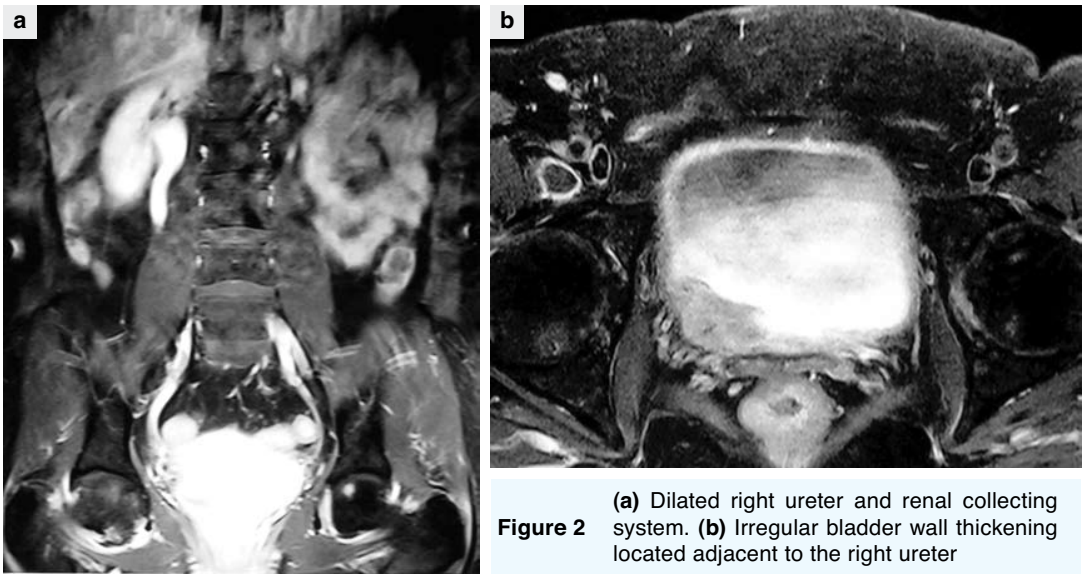

Figure 2 **(a)** Dilated right ureter and renal collecting system. **(b)** Irregular bladder wall thickening located adjacent to the right ureter

cystectomy or ligation of the internal iliac arteries, for control of massive hemorrhage. The association of amyloidosis with various malignancies, particularly multiple myeloma, medullary carcinoma of the thyroid, Hodgkin's disease, and renal cell carcinoma has been well documented.[6] To our knowledge, urothelial carcinoma *in situ* coexisting with primary bladder amyloidosis has hitherto not been reported. Therefore, the cause-effect relationship of localized bladder amyloidosis and urothelial carcinoma remains unclear.

References

1. Tan SY, Pepys MB. Amyloidosis. Histopathology 1994; 25:403-14.
2. Merrimen JL, Alkhudair WK, Gupta R. Localized amyloidosis of the urinary tract: case series of nine patients. Urology 2006;67:904-9.
3. Auge BK, Haluszka MM. Primary amyloidosis of the bladder. J Urol 2000;163:1867-8.
4. Zaman W, Singh V, Kumar B, Mandhani A, Srivastava A, Kumar A, et al. Localized primary amyloidosis of the genitourinary tract: does conservatism help? Urol Int 2004;73:280-2.
5. Akram CM, Al-Marhoon MS, Mathew J, Grant CS, Rao TV. Primary localized AA type amyloidosis of urinary bladder: case report of rare cause of episodic painless hematuria. Urology 2006;68:1343.e15-7.
6. Biyani CS, Fitzmaurice RJ, Upsdell SM. Localized amyloidosis of the urethra with transitional cell carcinoma of the bladder. BJU Int 1999;83:722-3.

Correspondence (Yazışma): Dr. Şükrü Kumsar. Mareşal Fevzi Çakmak Cad., 5. Sok., No: 48, 06490 Bahçelievler, Ankara, Turkey.
Tel: +90 312 212 68 68 e-mail: drkumsar@yahoo.com