

CHINESE MEDICAL SCIENCES JOURNAL

CASE REPORT

Amyloidosis of the Unilateral Renal Pelvis, Ureter and Urinary Bladder: a Case Report

Dong-liang Pan and Yan-qun Na*

Wujiaping Urology Medical Center of Peking University, Department of
Urology, Peking University Shougang Hospital, Beijing 100144, China

Key words: amyloidosis; bladder; renal pelvis

AMYLOIDOSIS of more than two urinary organs happened in one person is very rare. Here we reported a patient with amyloidosis of the left renal pelvis, ipsilateral ureter as well as urinary bladder occurring successively.

CASE DESCRIPTION

A 70-year-old man, presented to the outpatient department of urology complaining of a tumor in his left renal pelvis without abnormality of ureter and bladder first found by color ultrasonography and successively confirmed by abdominal computed tomography (CT) scan in September, 2008, who suffered an operation of partial nephrectomy of the right kidney for an angiomyolipoma in August, 2007 when no other lesion was found in his urinary system and underwent regular CT scan once per six months since then. No any symptoms such as pain, urgency, frequency and hematuria and no abnormal signs of physical examination for other systems were noted, and no positive findings of blood and imaging examinations for systemic amyloidosis were found, however red blood cell count was always more than 3 per high power field in urine tests. He had past history of wine drinking (more than 500 mL/d) for fifty years and primary hypertension for more than twenty

years and no past history of other significant illness such as immune abnormality.

A midstream sample of urine was collected once daily for three days continuously and only one sample was reported to show microscopic hematuria. Exfoliative cytology of 24-hour urine once per day found no malignant cells for three times persistently. Urine culture showed no growth of microorganism. Plasma chemistry, full blood count and immunological profile were all normal.

CT scan in September, 2008 showed multiple soft-tissue dense lesions which maximum diameter is about 3 cm in his left renal pelvis and ipsilateral upper and middle calyces and no obvious change of density after enhancement (Fig. 1) with high concentration of fluorodeoxyglucose at positron emission tomography-CT. In order to show more clearly the outline of the tumor and the relationship of this tumor and renal pelvis and to make more certain whether similar lesions existed in ureters, intravenous pyelogram was performed and presented filling defect of pelvicalyceal system only and no hydronephrosis at the left kidney. Further cystoscopy found no abnormality in bladder. Urine of the left renal pelvis was obtained by means of catheterization inserted into the left renal pelvis to get more precise information about cytology which was still normal. The following retrograde pyelogram showed the same image as intravenous pyelogram.

Ureteroscopy was performed and found one reddish tumor of 3 cm in diameter with smooth surface. Biopsy of

Received for publication January 10, 2011.

*Corresponding author Tel: 86-10-88297266, E-mail: nayanqun@cuan.cn

lesion was taken successfully and a piece of specimen was gotten. At the same time the ipsilateral ureter showed smooth surface of lumen wall. Histological examination showed urothelial hyperplasia and islands of amorphous eosinophilic material (Fig. 2) in epithelium of some regions and small blood vessels. Staining with Congo red showed a characteristic birefringence.

Nevertheless, the patient declined any further surgical procedure and determined to accept intensive observation using regular CT scan because of benignity of amyloidosis and fear of surgical damage to his left kidney.

Abdominal CT scan in February, 2009 found no change of the left renal amyloidosis. On August 14, 2009, CT showed the same image of the left renal amyloidosis as before, but found an irregular tumor of 1 cm in diameter at the posterior wall of urinary bladder. Enhanced CT scan seemed to show nodular enhanced lesion in the bladder with a thin pedicle to combine with the posterior wall of the bladder. Sonography of bladder showed a mid-echo nodule about 2.4 cm×2.2 cm×1.9 cm protruding to the cavity of bladder with blood current signal at its basement. Subsequently, cystoscopy found a tumor with smooth surface at posterior wall of bladder just like a group of grapes with diameter of 1 cm (Fig. 3). And midstream urine test presented no abnormality.



Figure 1. Computed tomogram of coronary position in September, 2008 shows tumor in the left renal pelvis (arrow).

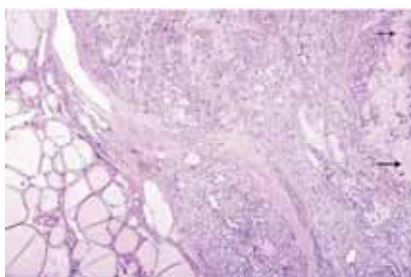


Figure 2. Haematoxylin and eosin staining of specimen section from the tumor in the left renal pelvis shows islands of amorphous eosinophilic material (arrows). ×10

Transurethral resection of bladder tumor was performed successfully and a F3 catheter was inserted into left renal pelvis to collect urine at an interval of two hours. Cytological results of four pieces of urine sample of the left renal pelvis were negative. HE staining showed the bladder tumor was islands of amorphous eosinophilic material (Fig. 4), suggesting amyloidosis of urinary epithelium, involved in inherent lamina of mucosa and small blood vessel wall, with positive staining of Congo red. The patient refused any postoperative prophylactic medicine therapy to prevent recurrence of bladder amyloidosis.

In November, 2010, CT scan showed the same image of the left kidney as before but three new papillary tumors in his bladder. Cystoscopy confirmed lesion of the bladder and this patient underwent transurethral resection of bladder tumor again. Before this operation, he received soft ureteroscopy to observe the lumen wall of the ureter and at the same time to perform biopsy to make certain the pathological profile of tumor in the renal pelvis once more. Unfortunately, a papiloma with thin pedicle was found in upper segment of the ureter and resected by 2-μm laser. The tumor in the left renal pelvis and papiloma of the ureter were pathologically diagnosed as amyloidosis again.

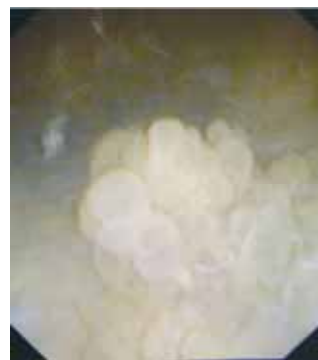


Figure 3. Cystoscopy in August, 2009 shows a tumor at the posterior wall of the urinary bladder, just like a group of grapes.

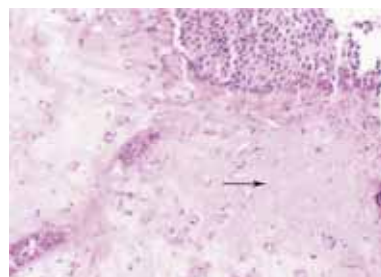


Figure 4. Haematoxylin and eosin staining of specimen section of urinary bladder tumor shows amyloid substance (arrow). ×10

DISCUSSION

Amyloidosis of urinary system is a rare condition and few cases are with simultaneous or successive multi-organ urothelial amyloidosis. According to the involved frequency rate of urinary organ with amyloidosis, bladder is the commonest, then ureter and renal pelvis, urethra the least. About 200 cases with amyloidosis of urinary system were reported in the literatures including 10 patients with more than two urinary organs involved (4 cases with renal pelvis and ureter, 4 with ureter and bladder, 2 with bilateral ureter),¹⁻¹³ but there was no case with renal pelvis and urinary bladder involved successively or simultaneously. Here we reported 1 patient with amyloidosis involved unilateral renal pelvis, bladder and ureter successively.

Until now, the etiology of amyloidosis of urinary system is still uncertain. One popular hypothesis is that amyloidosis is a process of self-immunity reaction with regard to abnormal metabolism of body protein. The phenomenon of amyloid deposits in the individual organ induced by local metabolic abnormality of protein is often called local amyloidosis. On the contrary, amyloid deposits in systematic organs is called systematic amyloidosis. Another opinion is that chronic urinary tract infection or repeat inflammation of mucosa and submucosa might lead to retrograde reflux and infiltration of adjacent monoclonal plasmacytes, which secrete immunoglobulin that is hydrolyzed into undissolvable fibrosis which deposits in urinary tract and forms amyloid tumor. It was of interest that the case we described had no past history of urinary tract infection and systematic amyloidosis and no evidence of other correlative immunological system diseases, so his etiology is also unclear.

This patient had past history of an angiomyolipoma in his right kidney before amyloidosis of the left renal pelvis and bladder occurred. Then, is there a relationship between angiomyolipoma and amyloidosis? The answer is no. Because the individual etiology of them is different. Angiomyolipoma is a benign clonal neoplasm consisting of varying amounts of mature adipose tissue, smooth muscle, and thick-walled vessels and is most likely derived from the perivascular epithelioid cells, and its growth may be hormone dependent, as suggested by its female predominance and rarity before puberty. But amyloidosis may be a process of self-immunity reaction with regard to formation of self-antigen and self-antibody induced by inflammation.

The clinical presentation of urothelial multiple amyloidosis is not particular of its own. The most common presenting complaints are microscopic haematuria without pain, or symptoms from urinary tract obstruction which

differ as obstructive site varies such as urethral amyloidosis leading to dysuria and proximal segmental ureter extension and pyelectasis resulting from ureteral amyloidosis. Nevertheless, massive haemorrhage or stimulative symptoms, the direct result of amyloid deposits in the bladder, has commonly been reported. Therefore, all the clinical presentations mimic other benign and malignant neoplasm of urinary epithelium, for example, papiloma, inflammatory granuloma, urothelial carcinoma, adenocarcinoma, etc. And it is very difficult to make differential diagnosis only according to ultrasonography, intravenous pyelography and CT scan. The diagnosis of amyloidosis is confirmed by performing ureteroscopy and obtaining tissue biopsy. And the patient we presented was finally diagnosed by biopsy with pyeloureteroscopy and cystourethroscopy. In order to avoid misdiagnosis, specimens should be taken from multiple sites and deep site because amyloidosis may coexist with renal pelvis carcinoma.⁸

The management of urothelial amyloidosis varies, depending largely on the clinical characteristics and the degree of pathological changes of the lesion as well as conditions associated with functional deterioration of urinary organ. Patients with primary localized urothelial amyloidosis or no severe symptoms and complications only need intensive observation and follow-up. Considering that our patient had history of surgery at the right kidney, he refused to undergo another operation at the left kidney as well as the biopsy findings demonstrated amyloidosis, we decided to use intensive observation. Our observation indicated the left renal lesion had no change one year later. But clinical intervention is necessary when dysuria, pyeloureterectasis, renal function failure or severe stimulative symptoms of bladder occur. Surgical resection is considered as the first-line therapeutic method and transurethral resection is chosen. Pharmacologic therapy can be used when lesion is difficult to completely resect, or when patients can not tolerate surgery. Colchicine^{14, 15} or dimethyl sulfoxide^{16, 17} may be beneficial to control the progression or recurrence of amyloidosis.

In conclusion, a biopsy is needed to make a definitive diagnosis of amyloidosis of urinary system and the entire urinary tract should be manipulated in a careful manner during follow-up because amyloidosis could occur at multiple organs of urinary system successively and be easy to recur.

REFERENCES

1. Slavov Ch, Vlahova A, Khristova S, et al. Amyloidosis of the urinary tract-case report and review of the literature.

- Khirurgiia (Sofia) 2006; 2:47-9.
2. Nagasaka T, Togashi S, Watanabe H, et al. Clinical and histopathological feature of progressive-type familial amyloidotic polyneuropathy with TTR Lys54. *J Neurol Sci* 2009; 276:88-94.
 3. DeSouza MA, Rekhi B, Thyaviahally YB, et al. Localized amyloidosis of the urinary bladder, clinically masquerading as bladder cancer. *Indian J Pathol Microbiol* 2008; 51:415-7.
 4. Jain M, Kumari N, Chhabra P, et al. Localized amyloidosis of urinary bladder: a diagnostic dilemma. *Indian J Pathol Microbiol* 2008; 51:247-9.
 5. Patel S, Trivedi A, Dholaria P, et al. Recurrent multifocal primary amyloidosis of urinary bladder. *Saudi J Kidney Dis Transpl* 2008; 19:247-9.
 6. Hajji K, Martin L, Devevey JM, et al. Rheumatoid arthritis-induced pseudotumoral AA amyloidosis of the bladder with vesico-peritoneal fistula. *Clin Nephrol* 2007; 67:38-43.
 7. Gómez García I, González Chamorro F, Fernández Fernández E, et al. Hematuria and sharp renal failure as debut of secondary bladder amyloidosis. *Actas Urol Esp* 2005; 29:603-6.
 8. Kirkpantur A, Baydar DE, Altun B, et al. Concomitant amyloidosis, renal papillary carcinoma and ipsilateral pelvicalyceal urothelial carcinoma in a patient with familial Mediterranean fever. *Amyloid* 2009; 16:54-9.
 9. Eccher A, Brunelli M, Gobbo S, et al. Subepithelial pelvic hematoma (Antopol-Goldman lesion) simulating renal neoplasm: report of a case and review of the literature. *Int J Surg Pathol* 2009; 17:264-7.
 10. Merrimen JL, Alkhudair WK, Gupta R. Localized amyloidosis of the urinary tract: case series of nine patients. *Urology* 2006; 67:904-9.
 11. Korkmaz C, Kebapci M. Addison's disease associated with widespread abdomino-pelvic visceral calcification due to secondary amyloidosis: a case report. *Acta Radiol* 2004; 45:800-2.
 12. Savareux L, Guy L, Essamet W, et al. Pyelo-ureteric amyloidosis. *Prog Urol* 2004; 14:406-10.
 13. Iida S, Chujyo T, Nakata Y, et al. A case of amyloidosis of the renal pelvis. *Hinyokika Kyo* 2003; 49:423-6.
 14. Heras M, Sánchez R, Saiz A, et al. Renal amyloidosis in a female with familial Mediterranean fever: clinical response to treatment with colchicine and infliximab. *Nefrologia* 2009; 29:373-5.
 15. Azzabi S, Ben Hassine L, Cherif E, et al. Renal amyloidosis during Behcet's disease, Study of one case. *Tunis Med* 2009; 87:213-4.
 16. Summerton DJ, Hall IS, Tulloch DN. Primary amyloidosis of the urinary bladder and ureters. *Br J Urol* 1998; 81:935.
 17. Mccammon KA, Lentzner AN, Moriarty RP, et al. Intra-vesical dimethyl sulfoxide for primary amyloidosis of the bladder. *Urology* 1998; 52:1136-8.