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PRIMARY LOCALIZED AMYLOIDOSIS OF THE RENAL PELVIS COEXISTING WITH TRANSITIONAL CELL CARCINOMA: A CASE REPORT

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Primary localized amyloidosis of renal pelvis is very rare, and only 12 cases have been reported. More than half of the reported cases were treated by nephrectomy, and the preoperative diagnosis could not be made correctly because of its resemblance to renal pelvic tumor. The ureteroscopy is a useful diagnostic means in such a condition. This case is unusual in that a papillary transitional cell carcinoma was present coincidentally.

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Key words: Amyloidosis, Renal pelvis, Transitional cell carcinoma

INTRODUCTION

Primary localized amyloidosis is a relatively rare disease of obscure etiology characterized by extracellular deposition of amorphous eosinophilic fibrillar protein in various organs. This primary pathological change is uncommon and formed only 9% of 236 cases of amyloidosis¹⁾. The most common sites involved were bladder, lung, skin and larynx. Among the genitourinary tract, the bladder is the most frequently affected, and involvement of the upper urinary tract is less common. Herein, we report the 13th case of primary renal pelvic amyloidosis along with a review of the relevant literature.

CASE REPORT

A 63-year-old woman visited our outpatient clinic on October, 1986, complaining of left flank pain and gross hematuria. Her previous medical history was uneventful, and physical examination revealed no significant abnormalities. On cystoscopic examination, there were no abnormal findings in the bladder. Urine cytology was negative. The renal ultrasonogram was grossly normal. An excretory urogram (IVP) demonstrated the irregular margin of left renal pelvis and calyces (Fig. 1A).

No stone shadow was recognized. The flank pain improved without treatment and gross hematuria discontinued. During eight months of follow up painless gross hematuria recurred intermittently in spite of negative urine cytology, so that she was admitted for further evaluation in June, 1987.

Laboratory data showed no abnormal findings, except for microscopic hematuria, counting 40 to 50 red blood cells per high power field. A retrograde pyelogram (RP) revealed left hydronephrosis and obstruction at the outlet of renal pelvis and the upper ureter (Fig. 1B). There was no tumor in the bladder, and ureteral brushing cytology was not significant. Ureteroscopic examination was carried out to confirm the presence of a tumor. A 12.5 F ureteroscope was inserted into the left ureter with ease. There was no abnormal finding in the lower or middle ureter. A small papillary tumor was seen at the outlet of the renal pelvis and was biopsied with a cold punch. The specimen showed papillary proliferation of well differentiated transitional cells with mild nuclear atypia, which was diagnosed as transitional cell carcinoma grade I (Fig. 2).

In July, 1987, left nephroureterectomy was performed. There was conspicuous

inflammatory adhesion around the renal pelvis and the upper ureter. No lymph-node swelling was recognized.

On gross examination of resected specimen, marked hemorrhage was present in the renal pelvis. Its surface was covered with dark brown "jelly" like material continuing to the pyeloureteric junction (Fig. 3). On microscopic examination, massive amyloid deposition in the submucosa of the renal pelvis was observed. Atypical hyperplasia of transitional epithelium was noted (Fig. 4). The amyloid was positive for Congo-red staining and immunohisto-

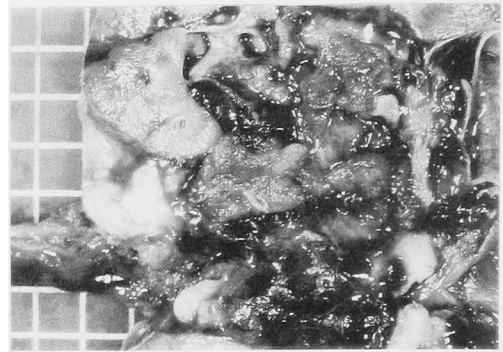
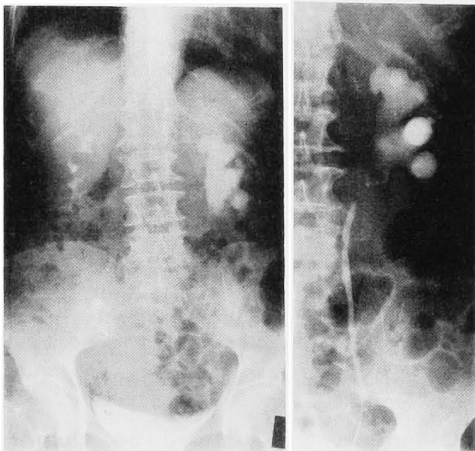


Fig. 3. The surface of the renal pelvis was covered with fragile jelly-like material.



1 A

1 B

Fig. 1 A: IVP showed left hydronephrosis and obstruction at the outlet of renal pelvis. B: Left retrograde pyelogram demonstrated the stricture between the outlet of renal pelvis and the upper ureter.

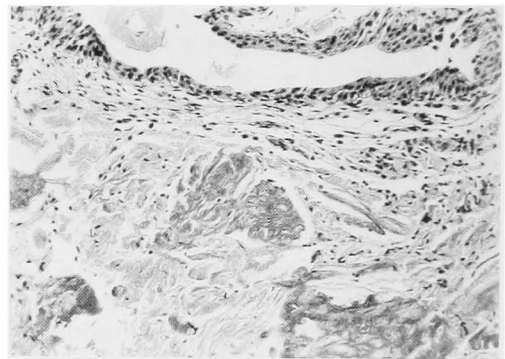


Fig. 4. Microscopic examination of resected specimen demonstrated marked deposit of amyloid in the renal pelvic mucosa. Congo-red stain.

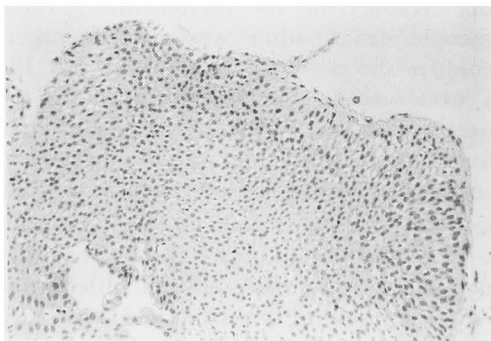


Fig. 2. Histological section of biopsy specimen showed papillary proliferation of well differentiated transitional cells with mild nuclear atypia. HE stain.

chemically positive for lambda chain, but negative for kappa chain and AA protein (biochemically related to and acute phase alpha-1 globulin and associated with chronic inflammatory condition). No deposit of amyloid was noted in the artery or glomeruli in the renal parenchyma.

Postoperative course was uneventful. Subsequent rectal biopsy was negative for amyloid. Postoperatively she has continued to do well.

DISCUSSION

Amyloidosis represents the clinical expression of extracellular deposits of the eosinophilic and amorphous proteinaceous material, which may be systemic or localized in distribution. The etiology and pathogenesis of primary localized amyloidosis remains obscure. A recent study using

Table 1. Analysis of 12 reported cases of primary localized amyloidosis of renal pelvis

Author year	Age, Sex	Symptom	Preoperative diagnosis	Operative procedure
Akimoto 1927	N.A.	N.A.	N.A.	N.A.
Gilbert et al 1952	52, F	flank pain	upper ureteral lesion intraepoic calculus	nephrectomy
Sato 1957	37, M	hematuria flank pain	renal pelvic tumor	nephrectomy
Chisholm et al 1967	66, M 58, F	hematuria none	N.A. staghorn calculus	exploration pyelotomy
Gardner et al 1971	39, F	hematuria flank pain	UPJ obstruction	exploration
Ullman 1973	58, F	hematuria flank pain	N.A.	nephrectomy
Dias et al 1979	67, F	hematuria	UPJ obstruction	nephrectomy
Gelbard et al 1980	67, M	hematuria flank pain	UPJ extravasation	nephrectomy
Fujihara et al 1981	66, M	hematuria	renal pelvic tumor	nephrectomy
Fox et al 1984	81, M	hematuria	renal pelvic tumor	nephrectomy
Murphy et al 1986	76, F	hematuria flank pain	renal pelvic tumor	nephrectomy

N.A. : not available

UPJ : ureteropelvic junction

the unlabelled immunoperoxidase method in combination with specific antisera against different chemical types of amyloid fibril proteins (AA, A lambda and A kappa), has shown the presence of A lambda antigenic fibril determinants in localized genitourinary amyloidosis and suggested that the fibrils are of immunocytic origin²⁾. Similarly, in our case, the amyloid deposit showed positive staining with an immunoglobulin light chain (lambda type) in immunohistochemistry.

Primary localized amyloidosis of the genitourinary tract has been reported in less than 100 cases, in which the bladder is the most frequently affected site²⁾. Localized amyloidosis of the renal pelvis is extremely rare, and only 12 cases were reported³⁻¹²⁾ (Table 1). The most common symptoms were gross hematuria and flank pain. Gross hematuria was seen in almost all cases, and half the cases complained of flank pain on the affected side. Every case showed hydronephrosis and irregular defect of renal pelvis on IVP. These clinical features resemble the malignant tumor of

the upper urinary tract. The final diagnosis has been made by following histopathologic assessment, such as exploratory surgery or nephrectomy. Because an amyloid lesion of the genitourinary tract is rare, a correct diagnosis is difficult to make preoperatively and may be confused with a neoplasm. These reported cases reaffirm the importance of considering this condition in the differential diagnosis of pelvic disorders.

There have been no reports about ureteroscopy to confirm the diagnosis of renal pelvic amyloidosis as we did preoperatively. In our case, if a deeper layer specimen had been taken by ureteroscopic biopsy, the preoperative diagnosis of amyloidosis could have been established. Ureteroscopy and biopsy should be extremely helpful procedures in establishing the accurate diagnosis of benign ureteral lesions such as amyloidosis.

Another interest which deserves special mention in this case is the coexistence of a papillary tumor and amyloidosis in one renoureteral unit. The papillary tumor

was very small. The ureteral mucosa covering the main lesion of amyloid deposit was apparently benign. Though the causal relationship between amyloid deposition and papillary tumor is not clearly understood, some inflammatory process may have occurred in the renal pelvis, and may have induced antigenic stimuli that produced amyloid deposition and reactive atypical hyperplasia of the urothelium which accelerated to cell dysplasia. In our case the amyloidosis was considered to be a primary, rather than secondary reaction to transitional cell carcinoma because the malignant lesion was small and was localized in a portion different from the amyloid deposit.

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和文抄録

移行上皮癌が合併した原発性限局性腎盂アミロイドーシスの1例

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腎盂の原発性限局性アミロイドーシスは稀な疾患であり、現在までに12例が報告されているに過ぎない。半数以上の報告例は腎摘によって治療されており、腎盂腫瘍との類似性のため術前診断をえることは難し

い。尿管鏡が診断に役立つと考えられる。本報告例は乳頭状移行上皮癌が合併していた点において稀である。

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