

PRIMARY LOCALIZED AMYLOIDOSIS OF THE URETER: CASE REPORT

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Primary localized amyloidosis of the urinary tract, especially that of the ureter, is rare. Nine cases of the ureteral amyloidosis have been reported in the world (Table).¹⁻⁹⁾ We have recently experienced the case which was preoperatively diagnosed as right ureteral amyloidosis, and its electron micrograph was obtained.

CASE REPORT

A 52-year-old woman visited our outpatient department on Dec. 1, 1973, complaining of dull pain in low back on the right side and gross hematuria. Her past history was noncontributory. Urological X-ray films revealed right hydronephrosis

without any stone. She was admitted for the further examinations. A papillary and edematous tumor protruded from right ureteral orifice on cystoscopic examination when the retrograde pyelography had been tried, and it was resected through trans-urethral approach. The histological examination showed neither malignancy nor inflammation. After this she was discharged and followed up at intervals. The tumor was resected again on Dec. 3, 1974. The hematoxylin-eosin preparation of this specimen demonstrated interstitial deposits of eosinophilic and amorphous substance. They were judged as amyloid, because they were well stained by congo red and showed

Table 1. Summary of the reported cases of primary ureteral amyloidosis.

No.	Authors	Sex & Age	Presenting Symptoms	Localization of Lesion	Preoperative Diagnosis	Treatment
①	Lehmann (1937)	f. 67		lower third portion of left ureter	postmortem	
②	Higbee & Millett (1956)	f. 71	right loin pain pyrexia	lower third portion of right ureter	not stated	nephroureterectomy
③	Andreas & Oosting (1958)	f. 12	left loin pain total hematuria	lower third portion of left ureter	? T.B. ureteritis ? neoplasm	nephroureterectomy
④	Konrath & Möbius (1960)	m. 55	left renal colic	lower third portion of left ureter	? stenosis	nephroureterectomy
⑤	Johnson & Ankenman (1964) (bilateral)	m. 17	right loin pain total hematuria	distal end of right ureter	not stated	ureteroneocystostomy
			left loin pain	distal end of left ureter	amyloidosis	ureteroneocystostomy
⑥	Yalowitz & Kelalis (1966)	f. 73	left loin pain, dysuria frequency	lower third portion of left ureter	? carcinoma	nephroureterectomy
⑦	Magri & Atkinson (1970)	f. 44	painless total hematuria	lower two thirds portion of left ureter	? endometriosis ? carcinoma	nephroureterectomy
⑧	Nagata & Takaha (1971)	m. 37	left lower abdominal pain	upper third portion of left ureter	ureteral tumor	nephroureterectomy
⑨	Klotz (1975)	f. 65	hematuria	lower third portion of left ureter	ureteral tumor	nephroureterectomy
⑩	Present case	f. 52	right loin pain hematuria	almost whole portion of right ureter	amyloidosis	nephroureterectomy

birefringence on congo red preparation and metachromasia in toluidine blue staining (Fig. 1).

She was readmitted on Jan. 6, 1975 for the operation, because of progression of right hydronephrosis and persistent right loin pain (Fig. 2).

Results of physical examinations were normal except for right CVA tenderness. Macroglossia, goiter, hepatosplenomegaly and enlargement of lymph nodes could not be found. Chest X-ray film and electrocardiogram did not reveal any particular finding. All of the laboratory data, including routine hematological studies, liver function test and serological examinations were within normal limit. Urinalysis showed occasional hematuria but no proteinuria.

The operation was performed on Jan. 13, 1975. Right nephroureterectomy was chosen rather than transureteroureterostomy or ureterocystoneostomy, because the involved portion of the ureter was too broad and the degree of the hydronephrosis was remarkable. The whole of right ureter was dilated. Its mucosa became darkly red in appearance, and was easily stripped from the muscular layer. This change of the mucosa extended from the site 1 cm below the ureteropelvic junction to the distal end (Fig. 3). We obtained same histological findings as before. On electron microscopic study, non-branching amyloid fibrils showing felt-like structure were found in intercellular space and cytoplasm of smooth muscle cells. The isolation of smooth muscle cell was also



Fig. 1. Congo red staining.



Fig. 2. Retrograde pyelography showed submucosal extravasation of contrast medium.



Fig. 3. Photograph of the specimen.



Fig. 4. Electron micrograph, $\times 10000$.

observed (Fig. 4). Histological investigation of the kidney showed dilatation of tubules, but amyloid deposition could not be detected in any part of the renal tissue. Her postoperative course was uneventful and she has been well for 36 months.

COMMENT

Amyloidosis was divided into four groups¹⁰: 1) primary, systemic or localized, 2) secondary, 3) heredofamilial and 4) amyloidosis associated with multiple myeloma. Our case was thought as primary localized amyloidosis of the ureter, because there was no evidence of coexisting or predisposing diseases and of amyloid deposition of the kidney which is said to be more frequently affected in systemic disorder than the rectum according to a study of Blum et al.¹¹ We did not perform congo red test because of the high frequency of the false negative case.

Nine cases have been reported in the world, but the operations were carried out with the tentative diagnosis of the ureteral tumor in many of them. Conservative operations were done on two cases. In our case, the extensive involvement of the ureter and a little fear whether we could completely rule out the malignancy led us to choose the nephroureterectomy.

SUMMARY

The tenth case of the primary localized

amyloidosis of the ureter was reported and its electron microscopic finding was demonstrated.

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和文抄録

原発性尿管アミロイドーシス 一症例報告一

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われわれは原発性尿管アミロイドーシスの1症例を最近経験した。

症例は52歳女子で肉眼的血尿を主訴として当科外来を受診。IVPで右水腎症，膀胱鏡で右尿管口より乳頭状かつ浮腫の強い腫瘤を認めたので生検をおこなった。

初診より1年後の再度の生検で尿管アミロイドーシスの診断を得た。右水腎症が亢進し，疼痛持続するため，右腎尿管全摘術を施行した。摘出標本の電顕像が得られたので，若干の文献的考察を加えて報告した。