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A CASE OF PRIMARY LOCALIZED AMYLOIDOSIS OF URETHRA

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Amyloidosis of the urethra is a rare disease. The clinical appearance resembles carcinoma of the urethra, so that biopsy is required to make the appropriate diagnosis. Once primary localized amyloidosis of the urethra has been diagnosed, selection of the appropriate treatment for each case becomes important.

We report a case of primary localized amyloidosis in the male anterior urethra. This case was treated successfully with urethral dilatation.

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Key words: Amyloidosis, Urethra, Urethral stricture

CASE REPORT

A 54-year-old man was referred with a chief complaint of dysuria and a painless induration of the penile shaft of several years duration. He had a history of an appendectomy and placement of a pacemaker for arrhythmia, but he had not had any sexually transmitted disease. Social and family histories were unremarkable.

Physical examination revealed a hard mass measuring 1.5×2 cm extending from the ventral aspect of the penile shaft down to the penoscrotal region. Examination of remaining area, including the external genitalia and prostate showed normal results. Preoperative laboratory examinations including complete blood cell count, coagulation profile, urinalysis, chest X-ray and intravenous urogram showed all normal results. Urine cytology indicated no malignant cells. Bence-Jones protein could not be detected in the urine. Serum protein electrophoresis and immunoglobulin assay showed no abnormalities, but the retrograde urethrogram showed an irregular narrowing at the site of the penile mass (Fig. 1). During urethroscopy, the

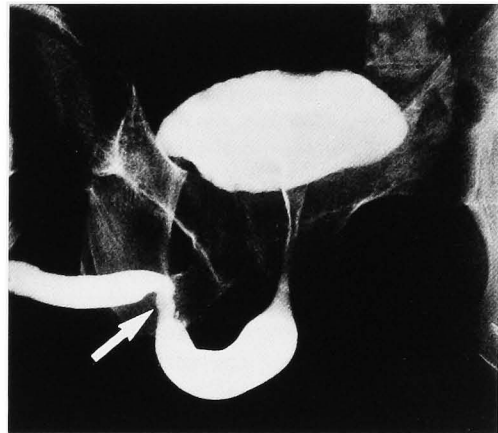


Fig. 1. Retrograde urethrogram demonstrates an irregular narrowing of anterior urethra (arrow).

narrow segment appeared as an erythematous urethral mucosa covered with whitish plaque. Cystoscopy could not be performed because of severe narrowing of the urethra.

On the basis of a preoperative diagnosis of urethral tumor or paraurethral tumor, transurethral and open biopsy of the penile mass were performed. Microscopic examination of surgical specimens showed sube-

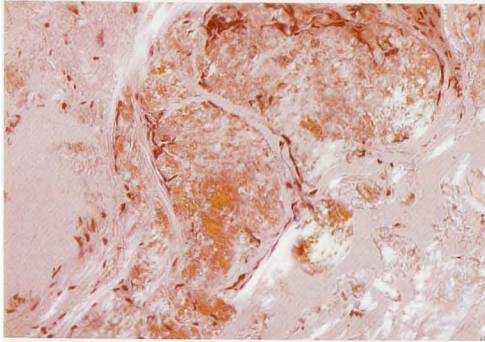


Fig. 2. Polarized light microscopic examination shows lime-green birefringence by Congo-red stain.

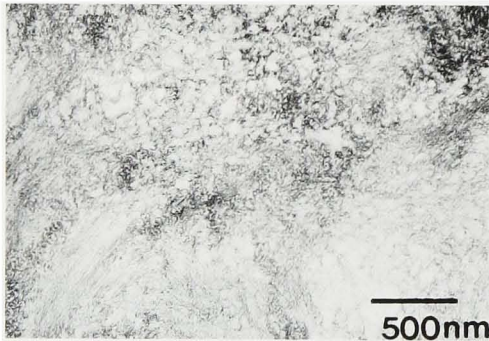


Fig. 3. Electron microscopic examination revealed the fibrillary structure characteristic of amyloid.

pithelial eosinophilic amorphous deposition in the mass of the lesion. This deposition stained markedly with Congo-red stain and glistened in response to depolarization, so that it was proved to consist of amyloid (Fig. 2). Since the reaction changed to Congo-red-negative after treatment with KMnO_4 , it was identified as an AA protein. Electron microscopic examination demonstrated the fibrillary structure characteristic of amyloid (Fig. 3). Further examinations revealed no signs of multiple myeloma, rheumatoid or collagen disease. Amyloid could not be identified in a rectal mucosal biopsy.

Urethral dilatation was performed on the basis of the diagnosis of primary localized amyloidosis of the urethra. The patient has since remained symptom-free on voiding while the penile mass has remained unchanged for one year without the need for any additional therapy.

DISCUSSION

Amyloidosis is characterized by extracellular deposition of a homogeneous, eosinophilic, and fibrillar protein in various tissues. Although the urinary tract is a common site for amyloid deposition, primary localized amyloidosis of the urethra is rare. Tilp¹⁾ reported the first autopsy case in 1909 and only 33 more clinical cases have been reported since then. Thirty-two of the previous cases were in men, and only one was in a woman. The ages of patients reported ranged from 21 to 82 years. Symptoms on presentation consisted of hemorrhage in the form of a bloody urethral discharge or of hematuria, dysuria, and urethral mass. The amyloid mass was located in the anterior urethra in 27 cases (81.8%), posterior urethra in 2 cases (6.1%), and the external urethral meatus in 4 cases (12.1%). Physical examination may reveal a palpable nodular mass along the urethra or at the external urethral meatus. A retrograde urethrogram may show a filling defect or an irregular stricture in the urethra, while endoscopic examination may show a submucosal plaque and an erythema or an ulcerative formation of the mucosa.

The clinical appearance resembles carcinoma of the urethra, so that the diagnosis must be based on adequate biopsy and appropriate staining of the specimen. Amyloid is eosinophilic, shows an amorphous deposition under light microscopy, stains positively with Congo-red, and shows lime-green birefringence under polarized light. Amyloid protein fibrils can be identified by electron microscopy. Given a histological diagnosis of amyloidosis and clinical indications of systemic amyloidosis or the presence of a predisposing disease, an examination should be performed to check for the presence of systematic amyloidosis or an other disease. In many cases of systematic amyloidosis, rectal mucosal biopsy reveals amyloid deposition.

Treatment of localized urethral amyloidosis depends upon the degree of urethral stricture and associated symptoms. The

management of previous cases has varied from no treatment to open surgery. Endoscopic procedures were selected in 22 cases, including transurethral biopsy alone, transurethral resection, optic internal urethrotomy, and urethral dilatation. On the other hand, open surgery was performed in 11 cases, including excision of the mass, segmental resection of the urethra combined with end-to-end anastomosis^{2,3)}, urethrectomy combined with urethrostomy⁴⁾. Dimethyl sulfoxide, the agent for systemic amyloidosis, was given in only three cases⁵⁾. In our case, the amyloid lesion was small, so that urethral dilatation was selected. As a result, symptoms of dysuria have improved.

We conclude that, once primary localized amyloidosis of the urethra has been diagnosed, the selection of an appropriate treatment for each case based on the view point of the degree of urethral stricture

and associated symptoms is essential for a satisfactory outcome.

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和文抄録

原発性限局性尿道アミロイドーシスの1例

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尿道アミロイドーシスは稀な疾患である。臨床的には尿道癌と類似しているため、適切な診断を下すには生検術が不可欠である。原発性尿道アミロイドーシスの診断がなされたなら、それぞれの症例に適した適切な治療法の選択が重要である。

われわれは男性の前部尿道に発生した原発性限局性アミロイドーシスの1例を報告する。自験例は尿道拡張にて経過良好である。

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