Primary isolated amyloidosis of the urethra: a case report

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ABSTRACT

Background: Amyloidosis is a tissue protein deposition. Urethral amyloidosis is rare. Clinical presentation includes hematuria, obstructive urinary symptoms, urethritis-like symptoms, urethral discharge, or a palpable penile/urethral mass. The diagnosis is made by urethro-cystoscopy and biopsy.

Case Presentation: We report the case of a 38-year-old man with a medical history of recurrent urinary tract infections, urethral discharge, and a palpable bulbar urethral mass. Urethral biopsies confirmed the urethral amyloidosis. The treatment was endoscopic urethral dilatation.

Conclusion: Urethral amyloidosis is a rare condition and a challenging diagnosis. Urethroscopy is the best method for diagnosis. Biopsies should be done to exclude malignancy.

Keywords: Urethra, amyloidosis, genito-urinary amyloidosis, urinary tract infection, case report.

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Background

Primary localized amyloidosis of the urethra (UA) is the first entity described by Tilip [1]. It results from the deposition of misfolded proteins in fibrils leading to tissue compression. In the literature, there are around 50 cases, most of them are case reports.

The clinical presentation includes obstructive and/or irritative voiding symptoms, urethral discharge, urinary tract infections, hematuria, penile induration, or masses, which may mimic a urethral carcinoma. Diagnosis is made by urethroscopy with biopsies.

Treatment depends on the degree of urethral stricture and the associated symptoms. Management includes conservative, endoscopic, or reconstructive surgery.

We report a case of a primary localized urethral amyloidosis (UA) presenting with recurrent urinary tract infections and a bulbar urethral mass managed by an endoscopic approach.

Case Presentation

A 38-year-old man with a medical history of recurrent urinary tract infections, Still-Chaufard disease, and macroscopic hematuria upon penile trauma during sexual intercourse (2002) presented himself to the urology department for recurrent urinary tract infections and urethral discharge. The infections had started 3–4 years ago with a frequency of 3–4 episodes/year. No history of sexually transmitted disease.

He complained of obstructive lower urinary tract symptoms. On clinical examination, the abdomen was normal. On external genitalia: the scrotum is normal. On penile palpation, ventrally, there was a 4, 5 cm longitudinal unpainful mass at the junction of the penile and the bulbar urethra.

Uroflowmetry: 4 ml/seconds. Blood analysis was normal. Urinalysis was normal.

Urethro-cystoscopy showed a patent anterior urethra, at the junction of the penile and bulbar urethra, there was a whitish and fibrotic stenotic lesion (Figure 1). Differential diagnosis: urethral carcinoma, scar tissue.

The workup included an MRI that showed “a thickening of the peno-bulbar urethra with contrast enhancement and peri-urethral infiltration. The adjacent corpus spongiuosum is also enhanced” (Figure 2). The urethrography showed “a stenotic lesion 5 cm long at the penile-bulbar urethra.”

Under anesthesia, biopsies of the mass were taken and urethro-cystoscopy completed with urethral dilatation of the fibrotic tissue. Membranous and prostatic urethra: normal. Bladder: normal.

The histopathology report revealed “a UA positive on congo red (figure 3) and apple-green birefringence (figure 4) Under a polarizing microscope. There were no amyloid AL or AA chains. Negative cytology.”

The extensive internal medicine review for systemic amyloidosis was negative.

Discussion

Tilip [1] was the first to describe a case of UA in 1909. Since then, around 50 cases of isolated UA have been published in the literature, most of them are case reports.

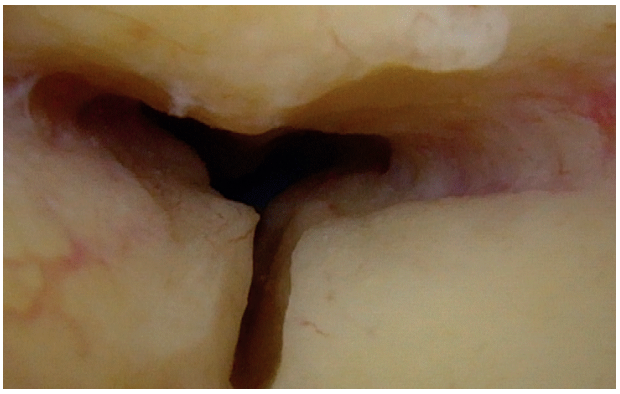

Figure 1. *Uretral whitish and fibrotic stenotic lesion.*

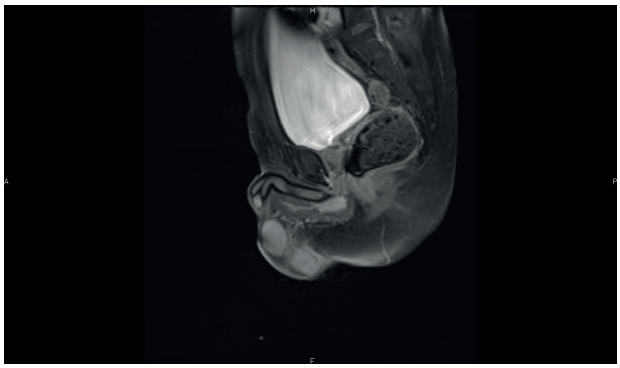

Figure 2. *MR thickening of the peno-bulbar urethra with contrast enhancement.*

Amyloidosis is a tissue deposition of extracellular homogeneous, eosinophilic and fibrillar proteins [2]. It might be primary with an amyloid deposit of immunoglobulins light chains (amyloid AL) or be secondary to chronic inflammation processes (Amyloid AA). More than 25 other proteins are known to cause amyloidosis. It might also be classified according to its anatomy as systemic (80%–90%) or localized (10%–20%) [7,8,10,14].

Possible predisposing factors are previous multiples urinary tract infections, gonococcal/chlamydial urethritis, chronic inflammations, and trauma [7]. Almost, all the cases of isolated UA have been in males. Only one case in a female has been reported.

The clinical presentation might be [3,5,9] hematuria, obstructive urinary symptoms, urethritis-like symptoms, and urethral discharge, and there might be a palpable penile/urethral mass.

The diagnosis is made by urethro-cystoscopy and biopsy. In endoscopy, one might see a submucosal plaque, erythema, fibrotic tissue, or ulceration. The differential diagnosis is a carcinoma of the urethra.

In this case, the patient had urethral leakage and recurrent urinary tract infections. On clinical examination, there was a ventral peno-bulbar urethral mass. Under endoscopy, he had a whitish fibrotic tissue occluding the bulbar urethra.

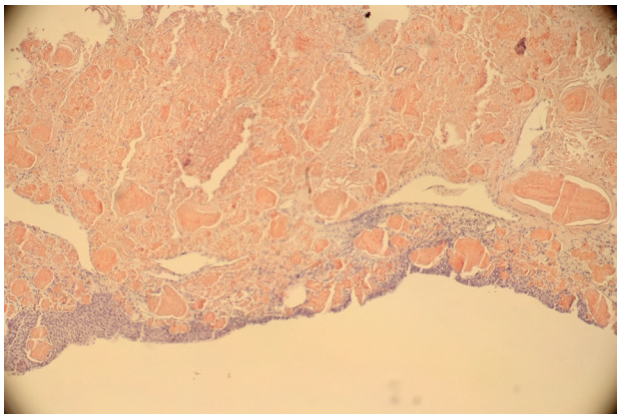

Figure 3. *Congo red.*

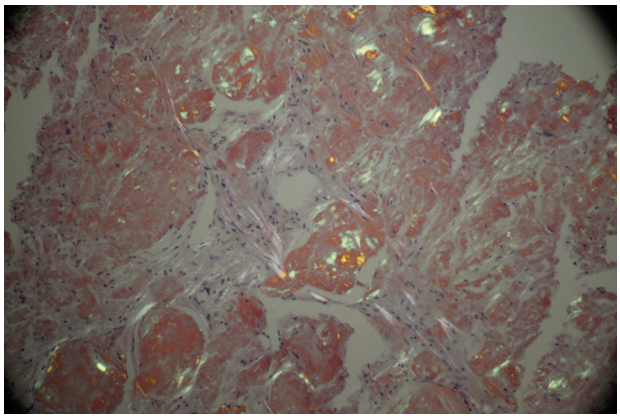

Figure 4. *Apple-green birefringence.*

While planning for surgery, MRI might be useful. Ichioka et al. [6] found UA to have a low signal intensity on T1 and T2 weighted images and good enhancement on gadolinium T1 weighted images, which helps to differentiate from urethral carcinoma [6,7]. Also, MRI findings may be useful to delineate UA before surgical resection and aid in planning surgery [7].

On penile ultrasound, the urethra and peri-urethral tissues have an increased echogenicity with a posterior shadowing within the corpus spongiosum [12].

A retrograde urethrogram shows a filling defect or an irregular stricture [2].

On histology examination, amyloid demonstrates a characteristic apple-green birefringence under a polarizing microscope after staining with Congo red [4,5,13]. Frequently, as in our case, no specific amyloid protein is identified. It is thought that the proteins are deposited through micturition [7].

A systemic workup must be done to exclude occult multi-systemic disease. In our case, internal medicine systemic workup was done. It included skin biopsy, medullar liquid exam, bone MRI, cardiac ultrasound, gastroscopy, and colonoscopy with biopsies. All were negative for amyloidosis.

The management depends on the degree of the urethral stricture and the severity of associated symptoms. It can be done by urethral dilatation, urethrotomy, transurethral resection, or urethroplasty [11]. Mangera et al. [7] did a review of the literature. He found few reports of successful treatment by urethral dilatation or resection. Most reports of conservative or endoscopic approach have a short follow-up. In his review, Mangera et al. found that repeated urethrotomy leads to symptomatic progression and a fibrous urethra. Urethroplasty with buccal mucosa or penile skin graft saw no recurrence at 18 and 6 months, respectively. The question of future amyloid deposits in the buccal or penile skin graft is still unanswered.

In the review by Crook et al. [8], of the 40 patients reviewed, two had a recurrence although the method of treatment was not specified, and the follow-up period was short.

After the urethral dilatation, our patient described an improved urinary stream, he had no more infections. In our case, unfortunately, the patient did not manage to carry out the uroflowmetry (did not need to pass urine during the consultations) and 16 months after surgery was lost.

The follow-up period that has been suggested is of 2 years [7,8]. UA can be a progressive disease, so the follow-up must be clinical and endoscopic.

In case of recurrence as has been suggested by Mangera et al. [7], the optimum treatment may be to completely resect the obvious pathology and reconstruct the urethra (urethroplasy).

Conclusion

In conclusion, the UA is a rare condition and a challenging diagnosis. It can mimic other conditions such as urethral carcinoma. Urethroscopy is the best method for diagnosis. Biopsies should be done to exclude malignancy.

Most cases are localized amyloidosis. Our case is a rare primary and isolated UA. Systemic workup is mandatory.

Endoscopic treatment can be successful although available information on follow-up is short term. In the case of recurrence, one has to consider a reconstructive approach.

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List of Abbreviations

UA Urethral amyloidosis

Consent for publication

Informed consent was obtained from the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report.

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Summary of the case

1	**Patient (gender, age)**	Male, 38Y
2	**Final diagnosis**	Localized urethral amyloîdosis
3	**Symptoms**	Recurrent urinary tract infection and urethral discharge
4	**Medications**	Generic
5	**Clinical procedure**	Urethral dilatation
6	**Specialty**	Urology