CASE REPORT

Primary amyloidosis of the bladder treated with partial cystectomy

Nejd F. Alsikafi, MD,¹ R. Corey O'Connor, MD,¹ Ximing J. Yang, MD,² Gary D. Steinberg, MD¹

¹Section of Urology, University of Chicago Pritzker School of Medicine, Chicago, Illinois, USA ²Department of Pathology, University of Chicago Pritzker School of Medicine, Chicago, Illinois, USA

ALSIKAFI NF, O'CONNOR RC, YANG XJ, STEINBERG GD. Primary amyloidosis of the bladder treated with partial cystectomy. The Canadian Journal of Urology. 2003;10(4):1950-1951.

A 56-year-old man presented with a 1-year history of intermittent gross, painless hematuria. Extensive evaluation revealed primary localized amyloidosis of the urinary

Introduction

Amyloidosis is a poorly understood condition that encompasses a wide range of diseases characterized by the extracellular tissue deposition of eosinophilic proteinaceous material. The disorder most commonly affects the synovium, skin and subcutaneous tissue, nerves, and liver. The cause is unknown but usually occurs in association with blood cell dyscrasias, systemic inflammatory conditions or infections. The presence of amyloid is diagnosed pathologically by its characteristic green birefringence when stained with Congo red and viewed under a polarized light. Localized primary amyloidosis of the bladder is a rare clinical entity with several cases described in the literature.¹ Initially, it is often confused with transitional cell carcinoma of the bladder. We report a case of primary amyloidosis of the urinary bladder treated with partial cystectomy after failed conservative management.

Accepted for publication December 2002

The Canadian Journal of Urology; 10(4); August 2003

bladder. Despite several endoscopic resections and fulgurations, the patient continued to have episodes of significant hematuria due to recurrent amyloid deposition. He was then successfully treated with partial cystectomy. At 60 months following surgery, the patient remains free of recurrent or systemic disease.

Key Words: amyloidosis, bladder, partial cystectomy

Case report

A 56-year-old otherwise healthy man presented with a 1-year history of intermittent gross, painless hematuria. He denied lower urinary tract symptoms or previous urinary tract infections. Urinalysis was significant for gross hematuria without pyuria or bactiuria. An intravenous pyelogram was unremarkable. Cystoscopic examination revealed a 4 cm x 5 cm ulcerative mass at the dome of the bladder. Transurethral resection was negative for malignancy but positive for amyloid deposits extending into the musculature Figure 1. Over the next 6 months the patient underwent several repeat transurethral resections/fulgurations for recurrent bleeding. He was then referred to our institution for further management.

Initially, the patient received an extensive systemic evaluation including whole body computerized tomography, a bone marrow biopsy, and serum and urine protein electrophoresis. All tests were negative for evidence of systemic disease. The patient was subsequently brought to the operating room and underwent partial cystectomy Figure 2. Pathologic examination showed amyloid deposits invaded into, but not through the muscularis mucosa. Surgical margins were negative for amyloid. The patient

Address correspondence to Nejd F. Alsikafi MD, Section of Urology, University of Chicago Hospitals, 5841 S. Maryland Ave, MC 6038, Chicago, IL 60637 USA

Primary amyloidosis of the bladder treated with partial cystectomy



Figure 1. A Congo red stained section showing extensive perivascular involvement of amyloid, and its characteristic "apple-green" birefringence (arrows).

tolerated the procedure well and was discharged on post-operative day #2. He remains asymptomatic with an unremarkable cystoscopy 60 months after surgery.

Discussion

The diagnosis of primary localized amyloidosis of the bladder is rare and can only be made after an extensive examination fails to reveal other sites of amyloid deposition, myeloma, or predisposing systemic disorders.² The vast majority of patients present with painless hematuria or irritative voiding symptoms.³ Cystoscopic examination reveals a broad-based, ulcerative or polypoid lesion with areas of yellow surface discoloration. Histologically, amyloid deposits are seen in the submucosa and muscularis mucosa. Occasionally, the amyloid also involves the subendothelial vessels. To date, there have been no



Figure 2. Photograph of the resected amyloid mass.

reports of malignant degeneration of localized amyloidosis in the urinary tract.

No set treatment of amyloidosis of the urinary tract has been established. Endoscopic management with close follow-up is reasonable for many localized lesions.^{2,4,5} Medical management with oral colchicine,⁶ intravesical dimethyl sulfoxide⁷ or nitrofurazone⁸ has also been found to be beneficial. In this case, we believe that a more aggressive strategy was warranted due to continued rapid recurrence into the bladder muscle with hematuria despite several transurethral resections.

Although the prognosis of primary amyloidosis of the bladder is favorable, close follow-up is mandatory due to the high rate of recurrence. With a median follow-up of 10 years, Tirzaman et al reported that only 6/24 (25%) patients with primary amyloidosis of the bladder did not have a recurrence.³ Most recurrent lesions are detected within 2 years after treatment,³ however, the longest published delay before recurrence was 14 years.⁹

Conclusion

Primary amyloidosis of the bladder is a rare disorder. Most cases can be treated conservatively with transurethral resection and/or medical management. We describe a case of recurrent, muscle invasive primary amyloidosis of the bladder successfully treated with partial cystectomy after failed endoscopic management.

References

- 1. Ferch R, Haskell R, Farebrother T. Primary amyloidosis of the urinary bladder and ureters. *Br J Urol* 1997;80(6):953-954.
- Caldamone AA, Elbadawi A, Mostagi A, Frank IN. Primary localized amyloidosis of the urinary bladder. *Urology* 1980;15(2):174-180.
- 3. Tirzaman O, Wahner-Roedler DL, Malek RS, et al. Primary localized amyloidosis of the urinary bladder: a case series of 31 patients. *Mayo Clin Proc* 2000;75(12):1264-1268.
- 4. Lehtonen T, Makinen J, Wikstrom S. Localized amyloidosis of the urinary bladder. *Eur Urol* 1991;20(2):113-116.
- 5. Boorjian S, Choi BB, Loo MH, et al. A rare case of painless gross hematuria: primary localized AA-type amyloidosis of the urinary bladder. *Urology* 2002;59(1):137iii-137iv.
- 6. Livingstone RR, Sarembock LA, Barnes RD, et al. Colchicine therapy in primary amyloidosis of the bladder. *J Urol* 1989;142(6):1570-1571.
- 7. Tokunaka S, Osani H, Morikawa M, et al. Experience with dimethyl sulfoxide treatment for primary localized amyloidosis of the bladder. *J Urol* 1986;135(3):580-582.
- 8. Auge BK, Haluszke MM. Primary amyloidosis of the bladder. *J Urol* 2000;163(6):1867-1868.
- 9. Ruffion A, Valignat C, Champetier D, et al. Long-term recurrence of primary amyloidosis of the bladder. Urology 2002;59(3):444i-444ii.