Primary amyloidosis of the bladder

Adrian Ho a,*, Melissa C. Davies b, Raluca Guran c, James Brewin b

a Urology CT2, Salisbury District Hospital NHS Foundation Trust, Salisbury, UK
b Consultant Urologist, Salisbury District Hospital NHS Foundation Trust, Salisbury, UK
c Consultant Histopathologist, Salisbury District Hospital NHS Foundation Trust, Salisbury, UK

Introduction

Bladder amyloidosis is a rare condition of extracellular protein deposition. The aetiology is largely unknown and the case presentation often mimics bladder malignancy.

Case presentation

A 39 year old Caucasian male presented with a 10 day history of visible painless haematuria. He had one previous episode of haematuria ten months prior to this following ozone therapy for Lyme’s disease. His past medical history included alopecia and chronic fatigue syndrome. He took no regular medications. There were no risk factors for transitional cell carcinoma (TCC) of the bladder.

An ultrasound scan of the urinary tract did not demonstrate any renal masses and there was a smooth walled bladder. Flexible cystoscopy demonstrated multiple raised haemorrhagic lesions in the dome of the bladder and malignancy was suspected. A subsequent CT urogram showed a thickened anterior-superior bladder wall but normal upper tracts (Fig. 1). No other significant pathology was found on the CT.

A general anaesthetic cystoscopy was performed and the lesions were biopsied (Fig. 2). There was no palpable abnormality with bimanual examination under anaesthetic.

Histology confirmed the absence of malignancy, but demonstrated acellular eosinophilic amorphous material in the lamina propria and blood vessels. On Congo red staining there was apple-green birefringence with polarised light which confirmed the presence of amyloid (Fig. 3). The patient was subsequently referred to the national amyloid centre for further management, where additional immunohistochemical staining of the amyloid deposits determined the amyloid to be of lambda subtype. On further investigation by the national centre with echocardiogram and Serum Amyloid P component (SAP) scintigraphy, there was no evidence of systemic amyloidosis. Therefore, the diagnosis was confirmed to be localised amyloidosis of the bladder of the lambda subtype. The patient’s symptoms resolved without further treatment and the patient is under cystoscopic follow up.

Discussion

Amyloidosis is a rare disease of disordered protein metabolism characterised by extracellular protein deposition of monoclonal light chains, which can either be lambda or kappa light chains. It is usually a systemic disorder, but it may rarely present in a localised organ. Urinary tract amyloidosis is rare and bladder primary bladder amyloidosis has approximately 200 cases reported worldwide. The cause is unknown, but it is thought to be linked to chronic inflammation and cystitis. Bladder amyloid classically presents with visible haematuria and storage bladder symptoms, mimicking bladder malignancy.

Cystoscopic evaluation is important but often appearances are difficult to interpret with a variation of appearances ranging from ulcerated masses to diffuse thickening. This highlights the importance of histological tissue to successfully diagnose the condition, which will be confirmed by the presence of apple-green birefringence on Congo red staining when viewed with polarised light.

Once localised amyloid has been diagnosed, it is important to exclude systemic amyloidosis and referral to a specialist amyloid...
centre is advised. Treatment of localised bladder amyloid is by transurethral resection and subsequent cystoscopic follow up is recommended due to a reported recurrence rate of 50%. Although there are no clinical guidelines regarding the length of follow up, previous authors have suggested follow up with cystoscopy every 1–3 years.

**Conclusion**

This case of bladder amyloidosis highlights the importance for urologists to appreciate alternative diagnoses in patients who have had visible haematuria. Whilst malignancy is always a concern, awareness of this condition will allow patients to receive the appropriate investigation and management of this rare condition.

**Author disclosure statement**

No competing financial interests exist.

**References**

1. Bardapure M, Namastivayams SK, Rogawski K. Primary localized amyloidosis of...
bladder: is there a need for cystoscopic surveillance? Urol Ann. 2013;5:309–311.

2. Zhou F, Lee P, Zhou M, Melamed J, Deng FM. Primary localized amyloidosis of the urinary tract frequently mimics neoplasia: a clinicopathologic analysis of 11 cases. Am J Clin Exp Urol. 2014 Apr 5;2(1):71–75.

3. Wilkinson M, Fanning DM, Flood H. Primary bladder amyloidosis. BMJ Case Rep. 2011 Jul 20;2011.

4. Kobayashi T, Roberts J, Levine J, Degrado J. Primary bladder amyloidosis. Intern Med. 2014;53(21):2511–2513.