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Primary amyloidosis of the bladder

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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.[1] It is usually a systemic disorder, but it may rarely present in a localised organ.[2] Urinary tract amyloidosis is rare and bladder primary bladder amyloidosis has approximately 200 cases reported worldwide.[3] The cause is unknown, but it is thought to be linked to chronic inflammation and cystitis.[4] Bladder amyloid classically presents with visible haematuria and storage bladder symptoms, mimicking bladder malignancy.[1–4]

Cystoscopic evaluation is important but often appearances are difficult to interpret with a variation of appearances ranging from ulcerated masses to diffuse thickening.[2–4] 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.[2–4]

Once localised amyloid has been diagnosed, it is important to exclude systemic amyloidosis and referral to a specialist amyloid

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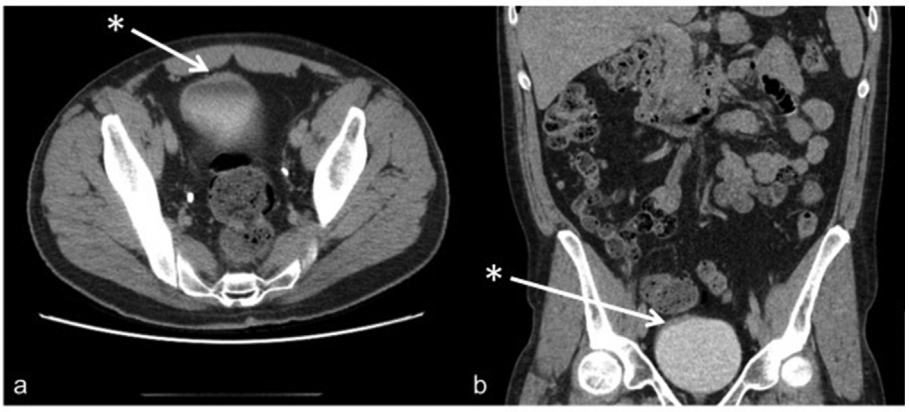

Fig. 1. CT Urogram demonstrating thickened anterior superior bladder wall in axial (a) and coronal (b) planes.

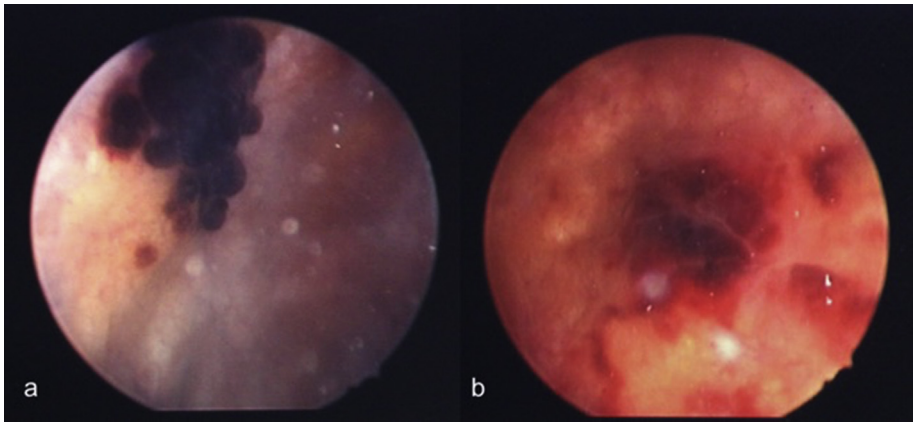

Fig. 2. Cystoscopic views (a) and (b). Both demonstrating thickened haemorrhagic lesions.

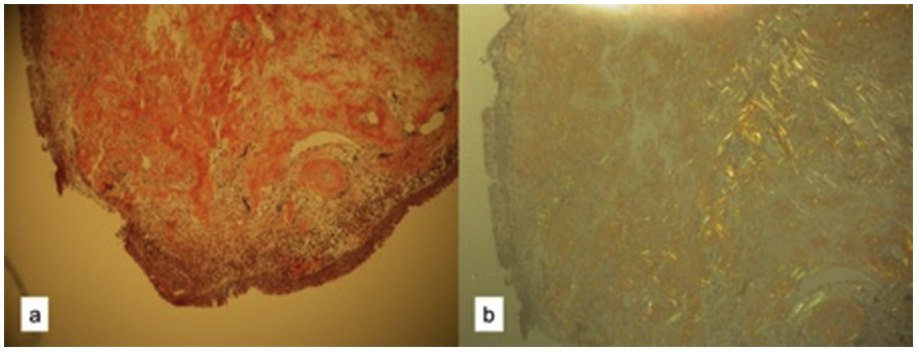

Fig. 3. Histology slides of the biopsied lesions showing uptake of Congo red stain (a) and apple-green birefringence when viewed under polarised light (b) which confirmed the presence of amyloid.

centre is advised.[4]

Treatment of localised bladder amyloid is by transurethral resection and subsequent cystoscopic follow up is recommended due to a reported recurrence rate of 50%.[3] Although there are no clinical guidelines regarding the length of follow up, previous authors have suggested follow up with cystoscopy every 1–3 years.[3]

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.

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