CASE REPORT

Primary bladder amyloidosis as a cause of haematuria

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SUMMARY

We report the case of a 46-year-old man who initially presented with macroscopic haematuria. Although initially concerning for a malignancy in the bladder, histology demonstrated a primary bladder amyloidosis that has remained stable for 6 years since the initial diagnosis. Primary bladder amyloidosis is an important clinical entity that can mimic bladder malignancy on clinical history, radiological investigation and cystoscopic evaluation. Although uncommon, it should not be neglected as a possible diagnosis in patients presenting with haematuria.

BACKGROUND

Primary localised bladder amyloidosis is a rare disease with only approximately 200 cases reported in the literature to date.¹ The aetiology is poorly understood, although it has been hypothesised that chronic inflammation in the bladder recruits lymphoplasmacytic cells to the bladder submucosa.² It usually presents with painless haematuria and is an important diagnosis to be aware of as it can mimic bladder neoplasia clinically, cystoscopically and radiologically.³ This is an important distinction to make as primary bladder amyloidosis has a benign clinical course and is usually cured by complete resection.⁴

CASE PRESENTATION

A 46-year-old man was referred with a history of several episodes of intermittent gross painless haematuria. This was not associated with any other urinary symptoms, and urine culture showed no growth. He was a moderate smoker of Syrian descent, and had no other significant medical history. He did not report any environmental risk factors for urothelial malignancy.

INVESTIGATIONS

He underwent CT intravenous pyelogram and urine cytology, both of which showed no abnormality. Flexible cystoscopy demonstrated a suspicious-appearing erythematous area on the posterior bladder wall, as well as diffuse yellow-coloured bladder mucosa. Biopsies were taken which demonstrated evidence of 'AA type' amyloidosis (figure 1). Repeat cystoscopy and biopsy were undertaken 6 months later which again confirmed the diagnosis of amyloidosis.

During a repeat CT scan, he was found to have a new large bladder lesion (figure 2). He underwent a repeat cystoscopy which demonstrated a large area of erythema and yellow colouration on the posterior bladder wall. This was resected, and the histology again demonstrated amyloidosis.

DIFFERENTIAL DIAGNOSIS

The patient was subsequently referred for investigation of systemic involvement of amyloidosis. Although the patient did not report any other symptoms, it was thought a search for systemic involvement may reveal an underlying pathological process. This included referral to a haematologist, rheumatologist and gastroenterologist. Extensive investigation did not reveal any other sites affected by amyloidosis.

TREATMENT

The patient was discussed at a multidisciplinary meeting, and the consensus achieved was that this reflected an unusually behaving primary bladder amyloidosis. As he was asymptomatic, it was decided that he would be managed conservatively with periodic surveillance cystoscopies every 6–12 months to ensure no progression of disease.

Figure 1 Histological confirmation of diagnosis with amorphous eosinophilic appearance on H&E staining (A) and apple-green birefringence on Congo red stains under polarised light (B).

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Figure 2 Axial and coronal views of the CT scan, with arrows demonstrating site of bladder lesion.

Figure 3 Cystoscopic view of the posterior bladder wall, showing an atypical yellow-coloured mucosal lesion later confirmed to be amyloid on biopsy.

OUTCOME AND FOLLOW-UP

He is now 6 years since initial diagnosis and cystoscopies have demonstrated stable, persistent lesions (figure 3). He has developed no further urinary symptoms and remains well from a medical point of view. He has not required any specific treatment for his bladder amyloidosis and has continued to be followed up by the rheumatology unit to assess for any clinical changes.

DISCUSSION

Other treatment options have been reported including colchicine and intravesical dimethyl sulfoxide but these are usually not

required.⁴ Once a diagnosis of amyloidosis of the bladder is made, it is necessary to exclude systemic involvement that may point to another underlying pathology although there remains no definitive guideline as to what this involves. Systemic involvement with amyloidosis reflects a poorer prognosis and necessitates specialist management.⁵ Furthermore, ongoing surveillance of the bladder should be undertaken because such patients may develop urothelial carcinoma regardless of the presence of amyloidosis in the bladder.⁶

Our patient's clinical story reflected the typical history of primary bladder amyloidosis with the exception that he developed recurrent disease in his bladder following initial cystoscopic resection. However, since that time, the disease burden of amyloidosis in his bladder has remained stable. Although this is a rare entity, further developments in management of such patients could be made in the form of a treatment and follow-up protocol for primary localised bladder amyloidosis.

Learning points

- ▶ Primary bladder amyloidosis is a rare disease but can easily mimic clinically the more common and sinister diagnosis of bladder cancer.
- ▶ Diagnosis should trigger an investigation for evidence of systemic amyloidosis which portends a poor prognosis.
- ▶ Cases of primary bladder amyloidosis in the literature have shown it to be a benign disease, but there is consensus on treatment or surveillance pathways.

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