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Multifocal primary amyloidosis of the bladder presenting with gross hematuria: A case report and review of literature

Jaisukh Kalathia [a,*], Giriraj Vala [b], Bharti Talreja [a], Kaushal Patel [c], Ayush Khetarpal [d], Prathan Joshi [e]

[a] Department of Urology and Kidney Transplantation, Fortune Urology Clinic, India
[b] Department of Urology and Kidney Transplantation, KIMS Hospital, India
[c] Department of Urology and Kidney Transplantation, Nephron Kidney Hospital, India
[d] Department of Urology and Kidney Transplantation, Khetarpal Hospital, India
[e] Department of Urology and Kidney Transplantation, Zydus Hospital, India

A B S T R A C T

Amyloidosis is defined as extracelluar deposition of amyloid, a fibrillary protein in one or more body sites. It can involve genito-urinary tract, primarily or secondarily, but isolated primary bladder amyloidosis is an extremely rare presentation. We herein report a rare case of 48-year-male patient presented with symptoms mimicking carcinoma urinary bladder especially painless haematuria. Transurethral resection of the mass was done in one sitting. The histopathological examination revealed to be a primary bladder amyloidosis. In the follow-up, patient had improvement in symptoms and no recurrence. We also briefly review the literature on primary bladder amyloidosis.

1. Introduction

Virchow first introduced the term “Amyloid” in medicine in 1953. Amyloidosis refers to abnormal extracellular deposition of amyloid, a fibrillary protein in one or more body sites. The commonly involved organs are skin, genitourinary tract (GUT), respiratory tract, gastrointestinal tract, heart, etc. In the GUT, amyloidosis involves the kidneys, renal pelvis, ureter, bladder, urethra, or corpora cavernosa, but isolated primary bladder amyloidosis is a rare phenomenon. The diagnosis of primary amyloidosis of the bladder is difficult as it resembles other bladder tumours on radiological imaging and similar clinical symptoms such as painless haematuria, irritative voiding symptoms or both. It can be accurately diagnosed only by histopathological examination comprising immunostaining with Congo red stain of the biopsied tissue.

2. Case report

A 48-years-old man presented in an emergency with painless gross haematuria for the past 2 days. As per the history, he also had intermittent haematuria and dysuria for the last 3–4 months but did not seek treatment. He was diabetic and on regular insulin. After the initial conservative management, the patient was further investigated. The blood investigation showed Hb(haemoglobin) of 9.5 gm% and serum creatinine of 1.2 mg/dl. Urine routine microscopy showed proteinuria, RBC (red blood cell) of 60–65/hpf. Urine culture was sterile. Computed tomography (CT) of the abdomen and pelvis showed diffuse irregular thickening of anterior, anterolateral and superior walls of the urinary bladder. It measured 1.4 cm in its maximum thickness with mild heterogenous enhancement. No evidence of perivesical fat infiltration or pelvic lymphadenopathy. [Fig. 1 (a-b)] Cystoscopy revealed an extensive broad-based tumour with papillary processes seen involving the bladder base, dome of bladder and both lateral walls. Transurethral resection of the bladder tumour (TURBT) was done in piecemeal. Histopathology of the biopsied material showed no malignant cells but showed pink amorphous material in the lamina propria in haematoxylin and eosin stain (H & E). [Fig. 1 (c)] It was further examined with Congo red stain which revealed a characteristic “apple-green” birefringence under polarized light suggestive of amyloid. [Fig. 1 (d)] We also noticed on cystoscopy that in between tumour tissues, there was a solid appearing area with yellowish appearance [Fig. 2]. Patient’s bone marrow examination was normal. Immunoglobulin A, Immunoglobulin G, free light chain (Kappa and Lambda) were within normal limits. On further evaluation, there were no evidence of amyloid deposition in other body sites. With this clinical picture, the patient was diagnosed to

* Corresponding author.
E-mail addresses: jaisukh2010@yahoo.com (J. Kalathia), girirajvala83@gmail.com (G. Vala), bhartittalreja@gmail.com (B. Talreja), drkslpatel@gmail.com (K. Patel), ayushkhetarpal@gmail.com (A. Khetarpal), dr.prathan@gmail.com (P. Joshi).

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have primary bladder amyloidosis. TURBT resolved the patients' symptoms of haematuria and dysuria. At 6 months, the patient was asymptomatic with no evidence of recurrence on cystoscopy. In the follow-up, the patient has been advised to have yearly cystoscopy.

3. Discussion

Amyloidosis is characterised by extracellular deposition of proteins. Amyloidosis may be primary or secondary. Primary bladder amyloidosis is caused by a disease with disordered immune cell function such as multiple myeloma and other immunocyte abnormalities. Secondary (reactive) amyloidosis is due to some chronic inflammatory disorders such as rheumatoid arthritis, tuberculosis, or any other chronic tissue destructive disease. In the urinary tract, amyloidosis can occur anywhere from kidney to renal pelvis, ureter, urinary bladder, urethra, and even penis. The kidney is nearly always involved in secondary amyloidosis and in approximately 50% of cases of primary amyloidosis.[1,2]

Primary urinary bladder amyloidosis is a rare clinical entity. Its importance is due to its similarity with urothelial carcinoma radiologically and cystoscopically. Chronic cystitis and inflammation of the bladder may lead to amyloidosis of bladder. It presents as painless haematuria, irritative voiding symptoms, or both. Amyloidosis may range in appearance from nodular to polypoid, single to multiple masses or bladder wall thickening with multiple yellowish plaques. Histologically, the diagnosis is confirmed by the presence of fluorescent apple green birefringence on congo-red stain under polarized light. The composition of amyloid AL and AA can be demonstrated with immunohistochemistry.[3]

Usually in primary bladder amyloidosis, deposits occur beneath the surface mucosa. In secondary amyloidosis, amyloid accumulates in the bladder vasculature which can cause massive haemorrhage. As a result, secondary amyloidosis of the bladder is reported to have a 30% mortality.[4]

Transurethral resection is the treatment of choice for primary bladder amyloidosis. Medical treatment has been described with intravesical Dimethyl sulfoxide instillation and oral colchicine. But this has a limited success. Cystectomy may be required in diffuse bladder involvement. Ligation of internal iliac arteries or cystectomy are occasionally necessary for control of massive haemorrhage in secondary bladder amyloidosis. Recurrence rate is around 50%. There are no definitive guidelines for surveillance, but most centres advocate follow-up cystoscopy every 1–3 years. When there is a recurrence, any co-existent malignancy should be ruled out.[5]

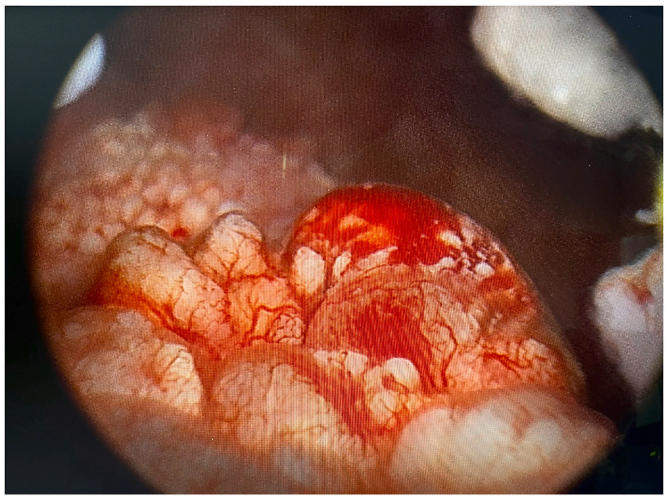

Fig. 2. Cystoscopy extensive broad based tumour with solid appearing areas.

4. Conclusion

Primary amyloidosis of the bladder is an unusual entity classically presenting with painless haematuria with irritative voiding lower urinary tract symptoms. Although it has recurrence but presently minimally invasive, transurethral resection is the treatment of choice with

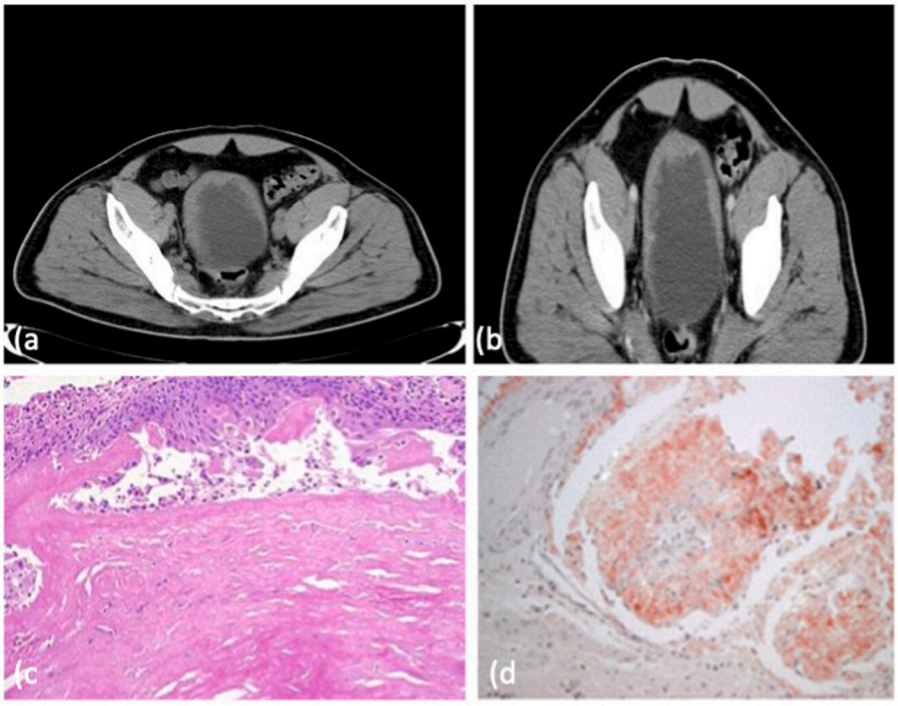

Fig. 1. A,b) CT-Scan reveals diffuse irregular thickening of walls of urinary bladder c) H & E stain shows pink amorphous material in lamina propria d) Congo red stain reveals a characteristic "apple-green" birefringence under polarized light suggestive of amyloid.

favourable outcome.

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Nil.

Declaration of competing interest

Nil.

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