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BLADDER AMYLOIDODIS – A CASE REPORT

Urology

Revanasiddappa A Kanagali Department of Urology and Renal Transplant, St John's Medical College Hospital, Bangalore, India.

Raghavendra Kulkarni* Department of Urology and Renal Transplant, St John's Medical College Hospital, Bangalore, India. *Corresponding author

Suryakant Choubey Department of Urology and Renal Transplant, St John's Medical College Hospital, Bangalore, India.

ABSTRACT

A case of primary amyloidosis of urinary bladder was presented with painless heamaturia and irritative urinary symptoms. Radiological imaging showed multiple small masses within the urinary bladder with suspicion of multiple transitional cell carcinomas within the urinary bladder. Cystoscopy confirmed the multiple masses within the urinary bladder, some showing haemorrhagic papules and papillary projections over the masses. Histopathology of the biopsy material was negative for the malignancy. Immunostaining of the biopsy material with Congo red stain showed the presence of amyloid fibrils within the biopsy material. Further investigations of systemic illness exclude the secondary amyloidosis. Transurethral resection was done. Patient showed improvement after the resection and no recurrence was observed during follow up. The purpose of this case presentation is to create awareness among the urologists to think for the rare entity of urinary amyloidosis especially if the histopathology is negative for the malignant cells.

KEYWORDS

Amyloidosis, Transurethral resection, Hematuria

Introduction

Amyloidosis first described by Virchow in 1853 is characterised by deposition of homogeneous, eosinophilic, hyaline material in various tissues. Commonly affects urinary bladder, lung, larynx, skin, tongue and the periorbital region[1].

Amyloidosis can be classified as primary disease with no underlying cause and secondary disease which may result from chronic inflammatory diseases such as rheumatoid arthritis or tuberculosis. Amyloidosis is classified as systemic disease, where the material is deposited in a wide variety of organs or localized disease, in which the amyloid material is confined to a single organ. Males are affected at an earlier age than females with most frequent presentation being painless gross hematuria[2]. In the urinary tract, amyloidosis can affect kidney to renal pelvis, ureters, urinary bladder and urethra. The kidney being involved in nearly all the secondary amyloidosis and in approximately 50% of primary amyloidosis cases. However, urinary bladder is usually involved in primary amyloidosis which may closely resemble infiltrating tumor both on imaging and cystoscopy, hence the accurate diagnosis of amyloidosis becomes essential. Diagnosis is based on biopsy of the bladder lesion with Immunostaining with Congo red stain, showing absence of malignant cells on histopathology and positive amyloid fibrils on Congo red staining. Immunostaining of the biopsy material is the key for the diagnosis. The treatment ranges from conservative transurethral resection to total cystectomy with urinary diversion. Long term follow up with imaging and Cystoscopy is essential because of the higher incidence of multiple recurrences.

Case report

A Sixty one year old male patient presented with painless gross heamaturia and irritative voiding symptoms for few days. He was hypertensive and on antihypertensive medication. General physical examination was unremarkable. Hemoglobin was 9 mg /dl and his blood chemistry was normal. Urine was negative for Bence Jones protein. Urine cytology was normal. Initial Ultrasound examination of the urinary bladder showed small mass in the posterior wall of urinary bladder, suggestive of transitional cell carcinoma (Figure 1) No systemic deposits were seen. No ascites was defined. Cystoscopy confirmed two raised erythematous lesions less than 1 cm each in the left lateral wall and two in the posterior edematous wall of urinary bladder . Histopathology of the biopsied material showed no malignant cells (Figure 2). Immunostaining of the biopsied material with Congo red stain confirmed the presence of amyloid fibrils in the biopsy material (Figure.2) confirming the diagnosis of urinary bladder amyloidosis. Further investigation of the rest of the organs did not show the presence of amyloid. A diagnosis of primary amyloidosis of urinary bladder was established. Transurethral resection of the lesions was done. Patient showed improvement after resection and followed for one year with Ultrasound examination and CT scan with no evidence of recurrence.

Figure 1: Initial Ultrasound examination of the urinary bladder showed small mass in the posterior wall of urinary bladder

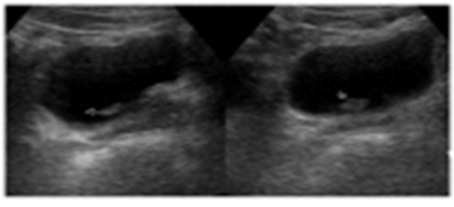
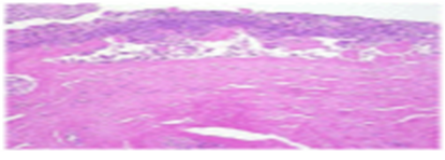


Figure 2 : Histopathology showing pink amorphous material in the lamina propria.



Discussion

Amyloidosis of the urinary bladder is a rare disease, with only approximately 200 cases reported in the literature[3]. Pathogenesis of amyloidosis of urinary tract is still unknown. Recent researchers consider abnormal metabolism of protein as the main reason of amyloidosis while another opinion is that chronic urinary tract infection and repeated inflammation of mucosa or submucosa leads to bladder amyloidosis disease[4].

Primary amyloidosis is more commonly distributed over the fifth, sixth and seventh decades of life in men, whereas it is diagnosed in women in sixth decade of life[5].

Other parts of the body are often involved in secondary amyloidosis. However the case which we are presenting denied medical history of chronic inflammation. And there was no evidence supporting any other organ involvement. Therefore we made the definitive diagnosis of primary bladder amyloidosis.

Primary bladder amyloidosis can masquerade as a malignancy, Radiological imaging and cystoscopy is usually misleading and may mimic malignancy. Surgical excision is the first line of therapy. Because of the high recurrence rate regular cystoscopy is recommended. Intravesical instillation of dimethyl sulfoxide and oral colchicine therapy have also given promising result[7].

As demonstrated in our case, transurethral resection is the treatment of choice for primary bladder amyloidosis. Medical treatments such as intravesical dimethyl sulfoxide instillation and oral colchicine have also been tried with limited success.

Recurrence rates post resection is estimated to be around 50%. Recurrence warrants full reassessment as cases of coexistent malignancy have been reported[8]. While no official guidelines exist for surveillance, most centers would advocate follow-up cystoscopy at 1–3 year interval.

Conclusion

Primary amyloidosis of urinary bladder is a rare disease, presenting as painless gross heamaturia. Primary amyloidosis can masquerade as a bladder malignancy. Radiological imaging and cystoscopy are usually misleading, mimicking malignancy. Histopathology is mandatory to exclude malignancy. Immunostaining the biopsy with Congo red stain is the only means to reach the correct diagnosis. Transurethral resection or coagulation and conservative treatment are best options for the treatment. Long term follow up is required[9].

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