GIANT PRIMARY AMYLOIDOSIS OF URINARY BLADDER MANAGED BY SEGMENTAL CYSTECTOMY: A RARE CASE REPORT WITH REVIEW OF LITERATURE.

Urology

Mahesh Chandra*	Associate Professor, Department of Urology, GMCH, Chandigarh. *Corresponding Author
Tarun Mittal	Senior Resident, Department of Urology, GMCH, Chandigarh.
Aman Gupta	Senior Resident, Department of Urology, GMCH, Chandigarh.
Nikhil Bhachhes	Junior Resident, Department of Surgery, GMCH, Chandigarh.

ABSTRACT

Amyloidosis is a heterogeneous group of disorders with amyloid material deposit in the extracellular compartment of the body as fibrillary protein which may be generalized or localized. Primary amyloidosis of urinary bladder is rare and large lesion measuring more than 8x8cm² in size is not known. Majority, clinically presents with painless haematuria and irritative voiding symptoms which may confused with diagnosis of bladder carcinoma. Definitive diagnosis is made on histopathological examination of tissue removed through cystoscopy or biopsy and after special staining. It diagnosis should alert physician to arrange for referral or appropriate investigations. Medical treatment using colchicine or dimethyl sulphoxide (DMSO) has been reported in the literature with good results. However, the majority of cases are still managed by transurethral resections of lesion. To our knowledge, this is first case of giant amyloidosis of urinary bladder managed by segmental cystectomy.

KEYWORDS

Bladder, haematuria, amyloidosis, carcinoma, colchicine.

INTRODUCTION

Amyloidosis is a heterogeneous group of disorders with amyloid material deposit in the extracellular compartment of body as fibrillary protein which may be generalized or localized. The definitive diagnosis requires a special staining of tissue removed through cystoscopy or biopsy. Its diagnosis should alert the physician to arrange for referral or appropriate investigations. Primary urinary bladder amyloidosis is a rare and incidentally diagnosis on histopathological examination. To date, only about 200 cases of primary urinary bladder amyloidosis have been reported in the literature.¹ Primary amyloidosis of urinary bladder presenting clinically are usually of less than 3cm in size and an no case report of large size lesion measuring 8x8cm² is available. Patients present with painless haematuria and irritative voiding symptoms which may mimics urological malignancy. Transurethral resection urinary bladder amyloidosis is primary surgical modality of treatment.² To our knowledge, this first case of giant primary urinary bladder amyloidosis managed by segmental cystectomy.

Case report

A 46 year old man presented with recurrent painless gross hematuria for last two months in January 2018 with. no irritative voiding symptoms such as frequency, urgency and dysuria. He had no associated other clinical disorder. On clinical examination, his abdomen was normal and digital rectal examination revealed mild grade prostatomegaly with no other positive finding. Urine analysis was full of red blood cells (RBC). His haematological and biochemistry was normal except low haemoglobin (10gm/dl). Patient underwent ultrasound examination of kidney, ureter, and bladder and a suspicious growth of urinary bladder was noted. Patient underwent further evaluation using CECT scan (Contrast enhanced computerized tomography) of abdomen which showed a large size mass involving posterior-lateral wall and funds of urinary bladder suggesting a diagnosis of urinary bladder tumour (Fig.1). Subsequently, the patient underwent cystoscopy which revealed a fleshy, nodular and protuberant growth of posterior lateral wall extending to dome of bladder. Bilateral retrograde pyelogram done were negative for any ureteral obstruction or filling defects. A transurethral resection of tumour followed by taking of deep resection biopsies was done in April, 2018. Surprisingly, the histopathological examination of specimens suggested amyloidosis of urinary bladder with evidence of malignancy (Fig.2). Congo-red stain with typical apple-green birefringence upon polarization was positive for amyloid deposition in the lamina propria and a sub-epithelial connective tissue (Fig.3). In postoperative period, the patient became symptom free and was discharged on 3rd days with to come for regular follow up.

Following 3 months, he again started to have painless hematuria. His cystoscopy revealed recurrence, thus again underwent transurethral

resection of growth and evaluation of associated disorders. His histopathological examination again confirmed amyloidosis with no malignancy. On 4th day, patient was discharged oral Colchicine 0.6mg twice a day and advised to continue his follow up after two weeks.

However, patient remained asymptomatic for three month, when he again started to have same symptoms in December, 2018. The repeat imaging done using ultrasound confirmed the recurrence of lesion. Patient was again taken up for cystoscopy and transurethral resection of lesion. A complete resection of lesion was technically difficult, thus the open surgery was planned. The abdomen was opened through lower midline incision which revealed a smooth, hard and large growth measuring 8x8cm in size, arising from the poster lateral walls and fundus of urinary bladder which was managed by segmental cystectomy (Fig 4a & 4b). Postoperative period was uneventful and two weeks later, his per urethral catheter was removed. His voiding diary at four weeks, revealed a normal urine frequency with average voided urine volume of between 250-300ml during each voids. Biopsy report again reconfirmed amyloidosis. In the follow up for one & half years, presently the patient is asymptomatic with no evidence of recurrence.

DISCUSSION

Amyloidosis is defined by deposition of characteristic fibrillary proteins into extracellular tissue of body. It is classified as primary (AL), secondary (AA) and hereditary (ATTR) types. Aetiology for primary bladder amyloidosis is not known but secondary is due to chronic inflammations.

Primary giant bladder amyloidosis is a rare disease. It may mimics bladder malignancy and the diagnosis is made on histopathological examination of the resected tissue and confirmation made after special staining such as Congo-Red stains. It was first described by Solomin in year 1897 during an autopsy and since then about 200 cases of primary bladder amyloidosis has been reported in worldwide literature.³ To date, no author has reported a large size primary bladder amyloidosis measuring 8x8cm² in size. The most common site of involvement in primary bladder amyloidosis is posterior wall.³ Patients usually presents with painless hematuria or irritative voiding symptoms. In a case series of 31 patients by Tirzaman et al, they reported that 77% patients present with hematuria.⁷ In another study by Merrimen et al 62.5% incidence of hematuria was reported and irritative voiding reported in the literature were due to involvement of trigonal area.⁴ In our case report, the lesion measuring giant size lesion was present on the posterior wall and fundus of urinary bladder. The lesion had a fleshy, nodular and protuberant appearance on cystoscopic examination.

Currently, the transurethral resection of lesions is primary modality of

treatment and diagnosis is made after histopathological examination of the specimen despite high rate of recurrence. In a study by Tirzaman et al 31 patients were followed up to 36 years, only 2% remains disease free, 21% had stable disease, 54% had disease recurrence, 10% underwent partial cystectomy and 3% underwent total cystectomy.³ In another study by Merrimen et al, 9 patients underwent total cystectomy for symptom treatment.⁴ Our patient underwent transurethral resection of amyloidosis lesion twice but again had recurrence as a giant lesion and underwent segmental cystectomy. Various other treatment modality have been described in literature such as intravesical DMSO (dimethyl sulphoxide) and systemic colchicine with some good results.³ Although, there are no specific guidelines to follow-up these patients due to its rare incidence. Some reports have advised for annual cystoscopy.³ our patient also had recurrence after endoscopic surgery, thus we also recommend regular follow up with cystoscopy every 3 months for a year and after that annually.

CONCLUSION

Primary urinary bladder amyloidosis is rare disease and it resembles as bladder malignancy. The transurethral resection of urinary amyloidosis remains a primary modality of treatment. The medical treatment using Colchicine may be tried for symptomatic small amyloidosis lesion of urinary bladder. However, whenever recurrence is of large size and complete resection of amyloidosis is technical not feasible, the treatment using segmental cystectomy may be worth consideration. In view of high recurrence rate, a regular cystoscopic surveillance became mandatory.

IMAGES:

Fig. 1: CECT scan image of urinary bladder amyloidosis.

Fig. 2: Histopathology showing amyloid stroma

Fig. 3: Congo Red staining was positive for Amyloidosis.

Fig. 4(a & b): Specimen of giant primary amyloidosis of urinary bladder.

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