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Urology

OBSTRUCTIVE PRIMARY AMYLOIDOSIS OF RIGHT DISTAL URETER WITH MULTIFOCAL BLADDER INVOLVEMENT: DIAGNOSTIC DILEMMA.

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ABSTRACT Amyloidosis is disorder of protein metabolism with extracellular deposition of fibrillary protein(amyloid) in one or more body sites. It can involve genitourinary tract, primarily or secondarily. Primary amyloidosis of genitourinary tract is rare and isolated primary bladder with ureter involvement is even more rare. We herein report a rare case of 54-year-female patient presented with symptoms of right flank pain with haematuria and dysuria mimicking lower ureteric neoplasm with irregular thickening of bladder wall on CT imaging. Cystoscopic biopsy of bladder lesions with right ureteroscopic biopsy of ureteric mass done and right silicone DJ stent kept. The histopathological examination revealed to be a bladder and right ureteric amyloidosis and thus nephroureterectomy deferred. In the follow-up, patient had improvement in symptoms and no progression of disease.

KEYWORDS : Localised Amyloidosis, ureter and bladder, mimic malignancy.

INTRODUCTION:

Amyloidosis is disorder of protein metabolism with extracelluar deposition of fibrillary protein(amyloid) in solid organs or tissues. The commonly involved organs are skin, genitourinary tract, respiratory tract, gastrointestinal tract, heart. In genitourinary tract amyloidosis involves the kidneys, renal pelvis, ureter, bladder, urethra, prostate or corpora cavernosa. Isolated primary bladder and ureter amyloidosis is a rare phenomenon. The diagnosis is difficult as clinical and radiologic features mimic urinary tract cancer. It is accurately diagnosed only by histopathological examination comprising immunostaining with Congo red stain of the biopsied tissue.

CASE REPORT

A 54-year-old hypothyroid and diabetic female who presented with right flank pain, haematuria with dysuria since one week. She also complained of intermittent painless haematuria one to two episodes in a week and dysuria over past 3-4months. On investigating hemoglobin11.2gm/dl, creatinine of 1.1mg/dl, coagulation profile within normal limits. Urine routine microscopy showed RBCs of 60-65/hpf, proteinuria.

Ultrasound scanning showed obstruction of the right lower ureter with proximal hydroureteronephrosis. On computed tomography (CT), homogeneous soft tissue density mass of the right lower ureter (31–45 HU) was seen. The length of lesion was about 36 mm(Figure 1d). Enhanced images showed a strip soft tissue density showing significant enhancement (44 HU)(Fig 1e). Diffuse irregular thickening of anterior and anterolateral walls of bladder. No evidence of perivesical fat infiltration or pelvic lymphadenopathy.

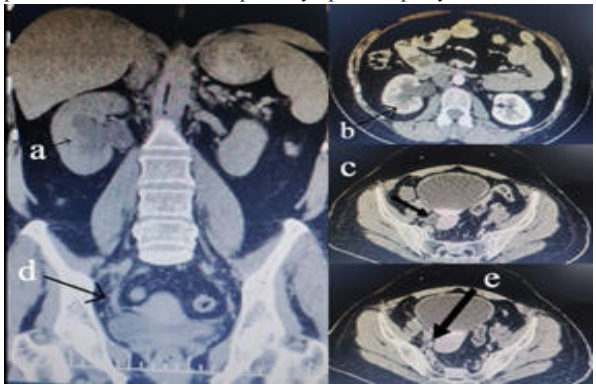

Figure 1: *a,b-Right hydronephrosis, c-Hydroureter till lower ureter, d-soft tissue density mass of right lower ureter with (e) significant enhancement.*

Cystoscopy showed broad-based lesions with multiple yellowish plaques seen involving the bladder base, dome of bladder and both lateral walls, further right ureteroscopy showed intraluminal irregular lower ureteric mass with significant narrowing.

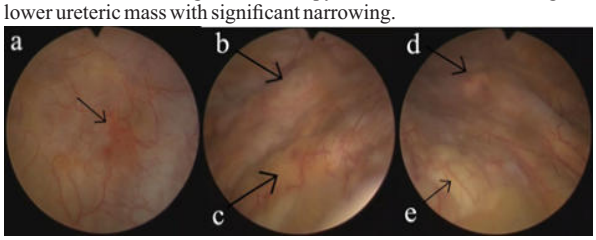

Figure 2 : *broad based lesions over dome(a),trigone(c), lateral wall(e). b– Left ureteric orifice, d- right ureteric orifice,*

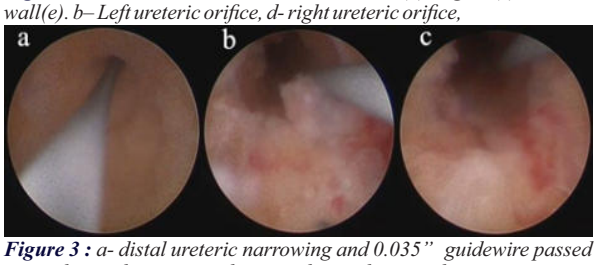

Figure 3 : *a- distal ureteric narrowing and 0.035" guidewire passed across luminal narrowing, b,c- intraluminal mass in lower ureter.*

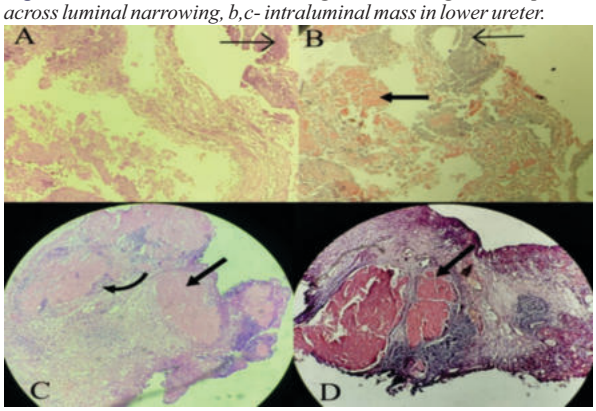

Figure 4 : *A) Ureteral mass Hematoxylin & Eosin stain. B) Ureteral*

mass congo red stain. C) Bladder mass Hematoxylin & Eosin stain. D) Bladder mass Congo red stain. Thin arrow- urothelium. Thick arrow – Amyloid. Curved thick arrow- giant cell reaction around amyloid.

Biopsy of lesions from both bladder and ureter taken. Urine from right ureter collected and sent for cytology and right silicone DJ stenting done. Histopathology of the biopsied material showed pink amorphous material in the lamina propria in haematoxylin and eosin stain. No evidence of malignant cells (H & E) [Fig.] Staining it with Congo red revealed a characteristic "apple-green" birefringence under polarized light suggestive of amyloid. [Fig.]. Urine cytology showed eosinophilic amorphous material amidst inflammatory cells which on Congo red stain shows apple green birefringence on polarising microscopy. No evidence of malignant cells.

On evaluation bone marrow examination was normal. Free serum light chain assay was done, which showed a normal kappa/ lambda ratio. Immunoglobulin A, Immunoglobulin G within normal limits.

With this clinical picture, patient was diagnosed to have primary right lower ureteric and bladder amyloidosis. Patient underwent right silicone DJ stenting. Over last year disease is non progressive.

DISCUSSION:

Amyloidosis has an incidence of 8 per million people each year[1]. Amyloidosis is characterised by pathological extracellular deposition of misfolded proteins(fibrils). It can be primary or secondary. Primary AL amyloidosis has clonal B-cell dyscrasia, including multiple myeloma, B-cell lymphoma, Waldenstrom macroglobulinemia, or another plasma cell neoplasia. Bone marrow biopsy required to exclude overt multiple myeloma. Secondary amyloidosis(reactive) arises as complication of chronic inflammatory disease (rheumatoid arthritis, chronic osteomyelitis, tuberculosis, Bronchiectasis).Kidney is nearly always involved in secondary amyloidosis and in approximately 50% of cases of primary amyloidosis.

Solonis first described bladder amyloidosis in 1897 at autopsy [2]. Gilbert and McDonald reported the first English literature about localized ureteric amyloidosis in 1952.[3] Primary urinary bladder and ureteric amyloidosis is rare. Chronic cystitis and inflammation of the bladder may lead to amyloidosis of bladder. It presents as painless haematuria, storage urinary symptoms or both. Although radiological examinations could detect hydronephrosis, bladder wall thickening in such cases, difficult to detect a specific etiology.[1] Its importance is due to its similarity with urothelial carcinoma radiologically and cystoscopically. Amyloidosis may appear as nodular to polypoid, single to multiple masses or wall thickening with multiple yellowish plaques on cystoscopy. Histologically, diagnosis of amyloid is currently based on specific Congo red staining on a biopsy of an involved organ by light microscopy under polarized light which shows apple green birefringence. The histopathologic diagnosis included AL localized amyloidosis, AA-type amyloidosis, TTR-type amyloidosis, AH type amyloidosis, Aβ type amyloidosis. It is histochemically possible to differentiate the specific protein that makes up the amyloid by treatment with potassium permanganate (KMnO4), if the deposits persist it is AL and if they disappear it is AA.[4] Deposits occur beneath the surface mucosa in primary bladder amyloidosis. Amyloid accumulates in the bladder vasculature in secondary amyloidosis which can cause massive haemorrhage. Hence, secondary amyloidosis of the bladder is reported to have a 30% mortality. Patients usually need early evaluation in order to rule out systemic amyloidosis, which requires different management and has poor prognosis.[5] Transurethral resection is the treatment of choice for primary focal bladder amyloidosis. Ligation of internal iliac arteries or cystectomy are occasionally necessary for control of massive haemorrhage in secondary bladder amyloidosis. Recurrence rate is around 50%. For surveillance there are no definitive guidelines. Follow-up cystoscopy is advocated every 1–3 years.

In cases of isolated ureteric amyloidosis surgery was the first choice, among which nephroureterectomy was predominant treatment. The reasons of a higher rate of nephroureterectomy were mainly because the ureteral tumor was difficult to exclude before and at surgery, and partly because the involved ureter was too extensive to salvage the ipsilateral kidney by conventional surgery. In this case ureteral stenting with conservative management and surveillance annually was chosen. There are no definitive treatment guidelines for management of localized urinary tract amyloidosis.

Surgical options for treatment: Double J stenting, ureteroneocystostomy, local excision and end anastomosis of the ureter, ileopyelostomy, autotransplantation, leal ureter, ureteric re-implantation with psoas hitch.

Tudor Borza and colleagues reported 2 cases with surveillance imaging and routine follow-up, without any medical or surgical interventions. With follow-up of 15 months, 6 years, and 8 years, none of the patients displayed clinical or radiographic signs of progressive disease.[6] Medical treatment has been described with intravesical dimethyl sulfoxide instillation and oral colchicine for diffuse amyloidosis.

Any co-existent malignancy should be ruled out when there is a recurrence. The association of amyloidosis with various malignancies, particularly multiple myeloma, medullary carcinoma of the thyroid, Hodgkin's disease, and renal cell carcinoma has been well documented.[7]

CONCLUSION:

Localised amyloidosis of urinary bladder and lower ureter is rare which can mimic malignancy on clinical and radiological evaluation. This case shows importance of obtaining histopathological conformation with high index of suspicion before undertaking radical surgical procedures. Congo red staining of biopsy is diagnostic. To prevent misdiagnosis and overtreatment awareness among urologist, radiologist and pathologists is important. Although nature of localised amyloidosis is benign, lifelong surveillance is recommended due to risk of local recurrence.

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REFERENCES

1. Kawashima A, Alleman WG, Takahashi N, et al.(2011). Imaging evaluation of amyloidosis of the urinary tract and retroperitoneum. Radiographics, 31(6), 1569-82. http://dx.doi.org/10.1148/rg.316115519
2. Jain M, Kumari N, Chhabra P, Gupta RK.(2008). Localized amyloidosis of urinary bladder: a diagnostic dilemma. Indian Journal of Pathology and Microbiology, 51(2), 247–249.
3. Mullin EM Jr, Trostle DR, Fetzer AE, & Stein A.J.(1984). Bilateral amyloidosis of the ureter associated with carcinoma. The Journal of Urology, 132(6),1181-1183.
4. García-Escudero López A, Padilla Nieva J, Infante Ria-o R, et al. (2011)Localized primary ureteral amyloidosis. Archivos Espanoles de Urologia, 64(4), 371-375.
5. Hinsch R, Thompson L, Conrad R. (1996). Secondary amyloidosis of the urinary bladder: a rare cause of massive haematuria. Australian and New Zealand Journal of Surgery, 66(2), 127-128.
6. Borza T, Shah RB, Faerber GJ, et al.(2010). Localized amyloidosis of the upper urinary tract: A case series of three patients managed with reconstructive surgery or surveillance. Journal of Endourology, 24(4), 641-644. http://doi.org/10.1089/end.2009.0383
7. Biyani CS, Fitzmaurice RJ, Upsdell SM.(1999). Localized amyloidosis of the urethra with transitional cell carcinoma of the bladder. BJU International, 83(6), 722–723.