Amyloidoma of the Bladder Mimicking a Bladder Tumor

Mesane Tümörünü Taklit Eden Mesanenin Primer Amiloidozu

Hikmat Jabrayilov[1], Murat Yavuz Koparal[2], Ipek Gonul Isik[3], Cengiz Kara[4]

[1]Medical Park Hospital, Urology, Ankara, Turkey
[2]Recep Tayyip Erdogan University, Faculty of Medicine, Urology, Rize, Turkey
[3]Gazi University School of Medicine, Department of Medical Pathology
[4]Izmir Katip Celebi University Ataturk Training and Research Hospital , Urology, Izmir, Turkey

ABSTRACT

Primary amyloidosis of the bladder is a rare disease reported in about 200 cases in the literature (1). Generally, this patients are admitted to the hospital with painless gross hematuria and lower urinary tract symptoms. Especially, symptomatic, radiological and cystoscopic findings of bladder amylodidosis and bladder cancer are similar. Neoplasia, hemorragic cystitis and lyphoma should be considered in differential diagnosis. Pathological diagnosis is imperative for the definitive diagnosis. Its pathophysiology is unknown, the prognosis is usually good and there is no specific treatment. Herein, we present the diagnosis and treatment of bladder amyloidosis, which is rare and mimics bladder cancer.

Key Words: Amyloidosis; bladder tumor; dimethyl sulfoxide; hematuria; transurethral resection.

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ÖZET

Mesanenin primer amiloidozu literatürde yaklaşık 200 vaka ile nadir görülen bir hastalıktır (1). Genellikle, bu hastaların hastaneye başvurma sebebi ağrısız gross hematüri ve alt üriner sistem semptomlarıdır. Özellikle semptomatk, radyolojik ve sistoskopik olarak mesane amilodidozu ve mesane kanseri bulguları benzerdir. Ayırıcı tanıda neoplazi, hemorjik sistit ve lenfoma düşünülmelidir. Kesin tanı için patolojik tanı şarttır. Patofizyolojisi bilinmemektedir, prognoz genellikle iyidir ve spesifik bir tedavisi yoktur. Burada, nadir rastlanan ve mesane kanserini taklit eden mesane amiloidozunu tanısı ve tedavisi ile sunuyoruz.

Anahtar Sözcükler: Amiloidoz; mesane tümörü; dimetil sülfoksit; hematüri; transüretral rezeksiyon.

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ORCID IDs. H.J. 0000-0001-6600-5769, M.Y.K. 0000-0002-8347-5727, I.G.I. 0000-0003-0058-5136, C.K. 0000-0002-6561-368X

Address for Correspondence / Yazışma Adresi: Hikmat Jabrayilov, MD Medical Park Hospital, Urology, Ankara, Turkey E-mail: dr.hikmetc@gmail.com

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INTRODUCTION

Amyloidosis is a disease characterized by extracellular deposits of a fibrillar protein called amyloid (1). It was first described by Virchow in 1853 (2). Primary amyloidosis of the urinary bladder is a rare disease, with approximately 200 cases reported in the literature.

Bladder amyloidosis is a rare disease and the first case was described by Solomin in 1897 (3). There is no reported sex predilection and the disease usually affects individuals between the fifth and seventh decades. Painless gross haematuria is the main presenting symptoms in most (>75%) cases (4). Localized urinary tract amyloidosis (UTA/ amyloidoma) is a rare disease that mimics neoplasia clinically, cystoscopically, and radiologically. Amyloidosis is categorized into two forms:

Primary amyloidosis - The process in which plasma cells overproduce protein rich portions of antibodies known as light chains (AL), these proteins are then deposited within the viscera. This is a primary condition requiring no secondary influencing condition.

Secondary amyloidosis (AA) - is most commonly associated with chronic inflammatory conditions such as rheumatoid arthritis, chronic osteomyelitis, or malignancies.

CASE REPORT

A 45-year -old, non-smoker female patient presented with a history of intermittent episodes of gross total painless haematuria of 2 months duration. Her family history was unremarkable. She had undergone an appendectomy 15 month ago with a diagnosis of acute appendicitis. Laboratory examination revealed no significant abnormality. Ultrasonography showed multiple solid lesions in the posterior wall of the bladder suggestive of urothelial carcinoma. Further evaluation with CT scan of abdomen and pelvis confirmed the presence of the lesion in the posterior bladder wall without bladder wall infiltration or adjacent structures (Fig 1). There was no ascites or any other abnormality in the regions that is observed in the available CT images. Flexible cystoscopy with local anastesia revealed a solid lesion in the posterior bladder wall mimicking an invasive carcinoma (Fig 2). Transurethral resection (TUR) was performed following of which macroscopic haematuria disappeared spontaneously. Histopathological examination of the TUR material revealed a localized deposition of an amorphous, pinkish extracellular material with focal chondroid metaplasia and dense fibrosis in the wall of the bladder. The urotelium was unremarkable. Congo red stain confirmed the deposition of amyloid with characteristic apple-green birefringence under polarized light. There was no evidence of malignancy (Fig 3). The patient consulted to nephrology and gastroenterology for systemic screening. No evidence of systemic amyloidosis was found. Serum protein electrophoresis showed non-specific abnormalities. Intravesical dimethyl sulfoxide (DMSO) for 10 weeks and oral colchicine therapy was started. Cystoscopy was repeated every 3 months and did not show any recurrence of amyloid within 17 months of follow-up period. The patient is currently free of symptoms.

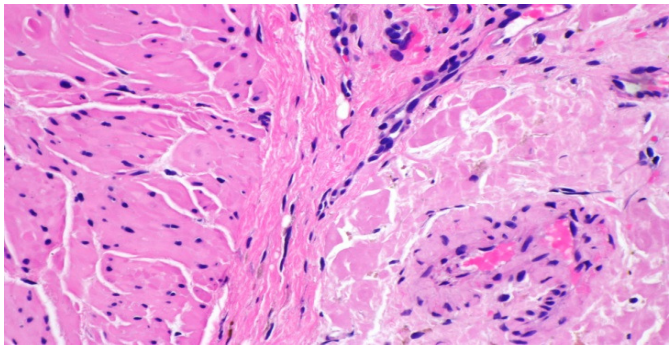

Figure 1. Apperance of nodular amyloid

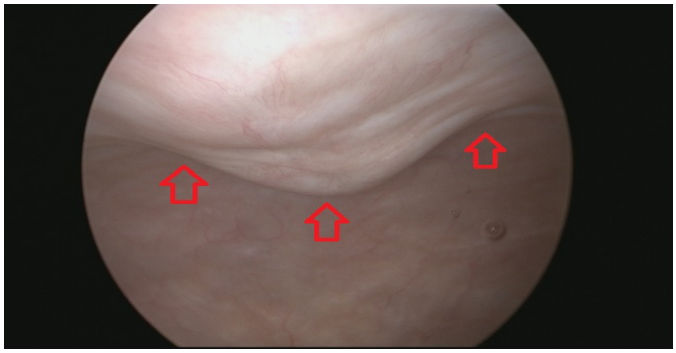

Figure 2. Cystoscopy image

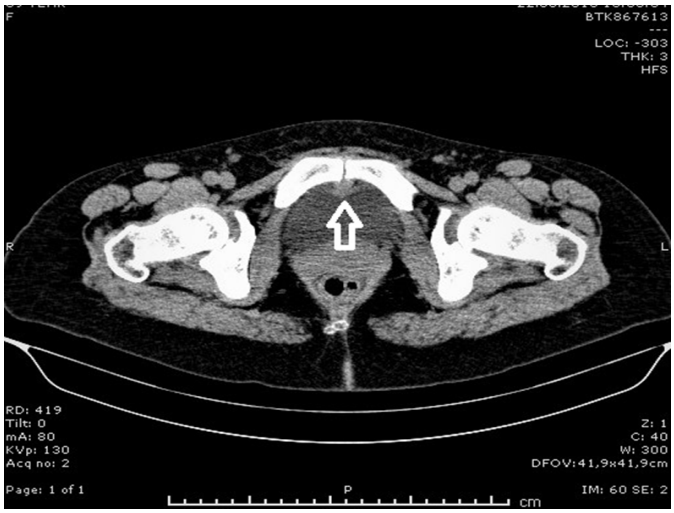

Figure 3. Preoperative CT image of the mass

DISCUSSION

Primary amyloidosis can occur anywhere along the urinary tract and has been reported in the kidney, renal pelvis, ureters, penis and even in the seminal vesicles (5-8). While the etiology of amyloidosis is unknown, several theories suggest a chronic monoclonal inflammatory response or an immunologic mechanism (9). Although bladder amyloidosis has been correctly diagnosed by cystoscopy in some reported cases, definitive diagnosis depends on histopathologic examination of the biopsy or resected specimen. Histologic examination shows proteinaceous amorphous eosinophilic deposits in the extracellular spaces. Diagnosis is confirmed by fluorescent apple-green birefringence after Congo red staining and visualization of the specimen under polarized light (5). In a study conducted by Biewend et al, none of the twenty patients with primary localized amyloidosis developed systemic disease during the follow-up of 7.6 years (10). This suggests that in primary bladder amyloidosis, there is a low risk of progression to additional sites.

Uracal cancer (UrC) is a very rare but highly malignant tumor with an incidence of <1% of all bladder cancers (11-13). The main differential diagnosis includes benign urachal tumors, nonurachal carcinomas of the bladder, and metastasis from different organs (prostate, colon, rectum, and female genital tract). Cystoscopy is important in the diagnosis of UrC. Imaging plays also an invaluable role at UrC workup. In ultrasound (US), it is generally recognized as a medium echo soft tissue mass or a fluid-filled cavity with mixed echogenicity and calcifications.CT scan is often used for local staging and evaluation of distant metastasis. It is usually depicted as a midline mass, superior to the bladder dome and adjacent to the abdominal wall. In the majority of cases, the tumor is mixed solid and cystic, the latter representing its mucin composition. Peripheral calcifications are often seen and are considered pathognomonic for urachal adenocarcinoma. On MRI, focal areas of high signal in T2 indicate the presence of mucinous component. CT and MRI are useful in demonstrating intra- or extravesical extension of the tumor (14). In our case, only localization similarity was observed.

CONCLUSION

Primary amyloidosis of the urinary tract is a rare condition that mimics malignancy in its clinical presentation and cystoscopic appearance and diagnostic imaging. The physiopathology is unknown, the prognosis is usually good and there is no specific treatment. Early eradication with fulguration or transurethral resection is indicated. Cystoscopic follow- up is necessary. Literature recommends a long term follow up. There is a need for large-scale investigation and new treatment modalities in the future of cases such as bladder amyloidosis that mimics bladder cancer.

Conflict of interest

No conflict of interest was declared by the authors.

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