A Rare Case Report: The Cause of Painless and Gross Hematuria Is Primary Amyloidosis of the Bladder

H. Jabrayilov*, C. Kara

*Medical Park Hospital, Ankara, Turkey; *E-mail: dr.hikmetc@gmail.com*

Primary amyloidosis of the urinary bladder is a rare disease, with only approximately 200 cases reported in the literature. Localized bladder amyloidosis is a rare disease that mimics neoplasia clinically, cystoscopically, and radiologically. The physiopathology is unknown, the prognosis is usually good and there is no specific treatment. Here we present a case report of the bladder amyloidosis, diagnosis and management of this rare condition.

Keywords: *Amyloidosis, transurethral resection, hematuria*

INTRODUCTION

Amyloidosis is characterized by extracellular deposits of the fibrillar protein, amyloid (Huang et al., 2006). It was first described by Virchow in 1853 (Virchow, 1854). Primary amyloidosis of the urinary bladder is a rare disease, with only approximately 200 cases reported in the literature. Bladder amyloidosis is considered to be a rare occurrence with the first case of primary bladder amyloidosis being described by Solomin in 1897 (Auge and Haluszka, 2000). Both sexes are equally affected between the fifth and seventh decade. Painless gross haematuria is the main presenting symptoms in most (>75%) cases (Johansson and Cohen, 1996). Localized urinary tract amyloidosis (UTA) 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. Here, we see widespread systemic deposition of amyloid proteins.

CASE REPORT

A 45 year old female patient in July 2016, presented with history of intermittent episodes of gross total painless haematuria of 2 months duration. She had no known drug allergies. Surgical history included appendicectomy. There was no significant family history. She was a non-smoker. Laboratory examination revealed no significant abnormality. Sonography showed multiple solid lesion posterior wall of bladdersuggestive of transitional cell carcinoma. Further evaluation with CT scan of abdomen and pelvis was done showing multiple small lesions in the posterior bladder wall without bladder wall infiltration to the adjacent structures. No systemic deposits were seen. No ascites was defined. At flexible cystoscopy with local anastesiawas seena solid lesion on the posterior bladder wall resembling invasive bladder (Fig 1).

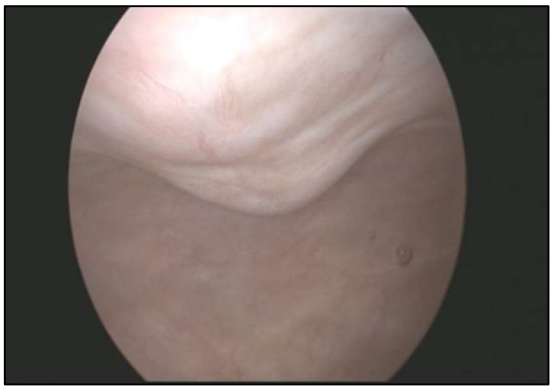

Figure 1. Cystoscopy image.

Transurethral resection was performed and submitted for histological examination. Macroscopic haematuria disappeared spontaneously after the treatment. Histopathology of the biopsied material showed no evidence of cancer.. Immunostaining of the biopsied material with Congo red stain confirmed the presence of amyloid fibrils in the biopsy material confirming the diagnosis of urinary bladder amyloidosis (Fig 2). 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 the 10 weeks and oral colchicine therapy was started. The patient has been followed up for 17 months and is currently free of symptoms. Cystoscopy was repeated every 3 months and did not show any recurrence of amyloid within a 17 month follow-up period.

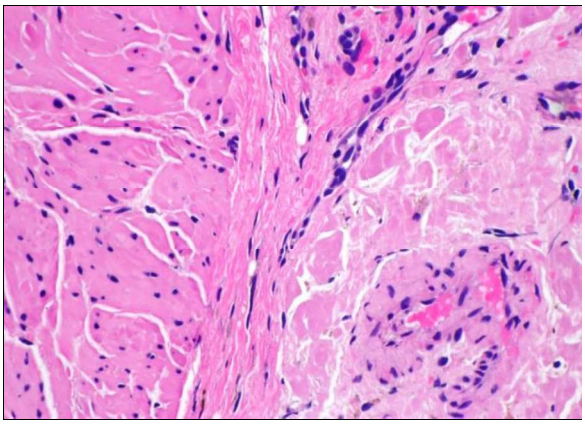

Figure 2. Apperance of nodular amyloid.

DISCUSSION

Primary amyloidosis can occuranywhere along the urinary tract and has been reported in the kidney, renal pelvis, ureters, penis and even in the seminal vesicles (Gardner et al., 1971; Krane et al., 1973; Caldamone et al., 1980; Gulmi et al., 1988).While the etiology of amyloidosis is unknown, several theories suggest a chronic monoclonal inflammatory response or an immunologic mechanism (Cohen, 1967). 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 (Biewend et al., 2006). This suggests that in primary bladder amyloidosis, there is a low risk of progression to additional sites.

CONSLUSIONS

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.

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Nadir Rastlanan Xəstəlik: Ağrısız Hematuriyanın Səbəbi Sidik Kisəsinin İlkin Amiloidozudur

H.Cəbrayılov*, C.Kara

Medical Park Hospitalı, Ankara, Türkiyə

Sidik kisəsinin primer amiloidozu nadir bir xəstəlikdir və ədəbiyyatda təxminən 200 belə xəstəlik bildirilmişdir. Klinik, sistoskopik və radiolojik cəhətdən neoplatik xəstəlikləri təqlid edir. Bu xəstəliyin fiziopatologiyası bilinmir, proqnozu adətən yaxşıdır və bilinən bir spesifik müalicəsi yoxdur. Burada da biz nadir görülən sidik kisəsi amiloidozu olan xəstəmizi, onun diaqnozunu və xəstəmizin müalicəsini təqdim etdik.

Açar sözlər: *Amiloidoz, transuretral rezeksiya, hematuriya*

Редко Встречающаяся Болезнь: Причина Безболезненной Гематурии – Первичный Амилоидоз Мочевого Пузыря

Х.Джабраилов*, Ч.Кара

Госпиталь Медикал Парк, Анкара, Турция

Первичный амилоидоз мочевого пузыря -это редкое заболевание, и в литературе отмечается примерно 200 таких случаев. Локализованный амилоидоз мочевого пузыря имитирует неоплазию клинически, цистоскопически и радиологически. Физиопатология этого заболевания неизвестна, прогноз, как правило, благоприятный, конкретного специфического лечения нет. В статье представлена информация о пациенте с первичным амилоидозом мочевого пузыря, а также данные о диагностике и лечении этого заболевания.

Ключевые слова: *Амилоидоз, трансуретральная резекция, гематурия*