



Taibah University

Journal of Taibah University Medical Sciences

www.sciencedirect.com



Case Report

Primary localised amyloidosis of the urinary bladder: A recurrent and progressive disease

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Received 18 December 2017; revised 12 February 2018; accepted 19 February 2018; Available online 10 March 2018



المخلص

ينتج الداء النشواني الأولي من ترسب ليفيات بروتين الأميلويد في المساحة خارج الخلية، وفي كثير من الأحيان يشمل المثانة البولية. نعرض حالة مريض ذكر عمره ٤١ عاماً تم تشخيصه بالداء النشواني الأولي للمثانة البولية، وتم استئصال ورم في المثانة عن طريق الإحليل مرتين قبل ٤ أعوام. دخل المريض المستشفى مؤخراً، عن طريق وحدة الطوارئ يعاني من بول دموي واضح غير مؤلم. وأظهرت الأشعة المقطعية للبطن والحوض ورماً في المثانة أكبر من الذي تم ذكره سابقاً. وتم إعادة تنظير للمثانة واستئصال الورم وأظهر التشريح المرضي للنسيج الداء النشواني الأولي للمثانة البولية. كما تم إجراء فحص شامل لاستبعاد الداء النشواني الشامل.

الكلمات المفتاحية: المثانة البولية؛ ورم حميد؛ الداء النشواني؛ متكرر؛ مترقق؛ بول دموي

Abstract

Primary amyloidosis results from the deposition of amyloid protein fibrils in the extracellular space and rarely involves the urinary bladder. We present a 41-year-old man who was diagnosed with primary amyloidosis of the urinary bladder and underwent two sessions of transurethral resection of the bladder mass 4 years prior. Recently, the patient was admitted through the emergency with painless frank haematuria. Computed

tomography of the abdomen and pelvis revealed a bladder mass that was larger than the previously reported mass. A repeat cystoscopy and resection of the mass was performed. Histopathological examination of the resected tissue revealed primary amyloidosis of the urinary bladder. A comprehensive examination was performed to exclude systemic amyloidosis.

Keywords: Amyloidosis; Benign tumour; Haematuria; Progressive; Recurrent; Urinary bladder

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Introduction

Amyloidosis is a disorder of protein metabolism characterised by the deposition of amyloid protein fibrils in the extracellular spaces of different body tissues. It may remain localised to a single organ or present as a systemic disorder involving multiple organs.^{1,5} Primary localised amyloidosis of the urinary bladder is a rare condition, first described by Solomin in 1897,¹ with less than 200 cases reported worldwide.² The main presentations of primary amyloidosis of the urinary bladder are intermittent, painless frank haematuria with irritative voiding symptoms.^{1–3} The condition can be easily mistaken for a malignancy on the basis of imaging and cystoscopy findings.^{6,7} Therefore, transurethral resection of the lesion and histopathological

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Peer review under responsibility of Taibah University.



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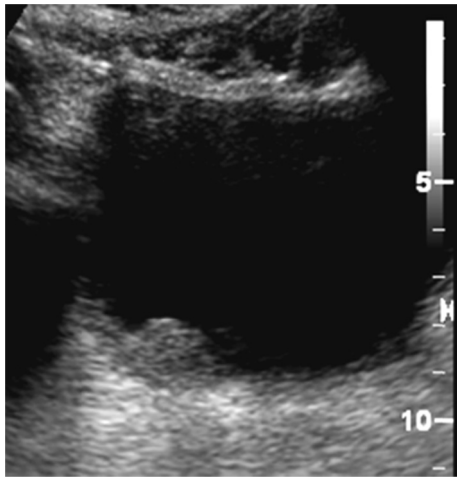


Figure 1: Ultrasonographic image showing a bladder mass.

examination are considered the standard strategy for diagnosis and treatment and to exclude malignancy.^{6,7}

Case report

A 41-year-old man presented for the first time to the Accident and Emergency department in 2012 complaining of haematuria that lasted for 10 days duration that was intermittent, painless, terminal and frank, with passage of clots associated with mild dysuria. The patient was a non-smoker with no comorbidities.

The results of a urinalysis and a urine culture were negative. His urine cytology showed atypical urothelial cells and was negative for malignancy. Ultrasound of the kidney, ureters and bladder revealed a normal upper urinary tract and two echogenic lesions measuring 0.87×0.65 cm and

1.7×1.4 cm, respectively, in the posterior wall of the urinary bladder (Figure 1).

He underwent a cystoscopy that revealed a polypoid nodular lesion measuring approximately 2 cm in size, with hyperaemia and ulceration involving the posterior bladder wall; this was highly suggestive of malignancy. Both ureteric orifices were not affected by the lesion. Transurethral resection of the lesion was performed. His histopathological findings were consistent with amyloidosis and were confirmed using special stains such as CD 138, Congo red, and haematoxylin & eosin (H&E) (Figure 2a,b,c).

The patient was fine and appeared healthy at follow-up. Systemic investigations were performed to exclude systemic amyloidosis. His 24-h urine protein electrophoresis and urinary Bence Jones protein test results were negative. Additionally, the sigmoid colon biopsy result was also negative. His electrocardiogram was normal.

One year later he developed similar symptoms and underwent computed tomography (CT) which revealed a plaque-like, focal mural thickening along the left posterolateral aspect of his urinary bladder with a maximal thickness of 4–5 mm (Figure 3).

Urine cytology was negative for malignancy. He was treated conservatively.

One year later, intermittent, gross painless haematuria with passage of clots recurred. CT was repeated and revealed that the urinary bladder was moderately distended, with focal wall thickening along the left posterolateral aspect with maximum thickness of 9 mm. The vesicoureteral junction was also affected and showed wall thickening (Figure 4).

The patient was offered a check cystoscopy and transurethral resection of bladder tumour; but he refused and was lost to follow-up for approximately a year. He visited the Accident and Emergency again in November 2016 with painless, intermittent frank haematuria associated with the

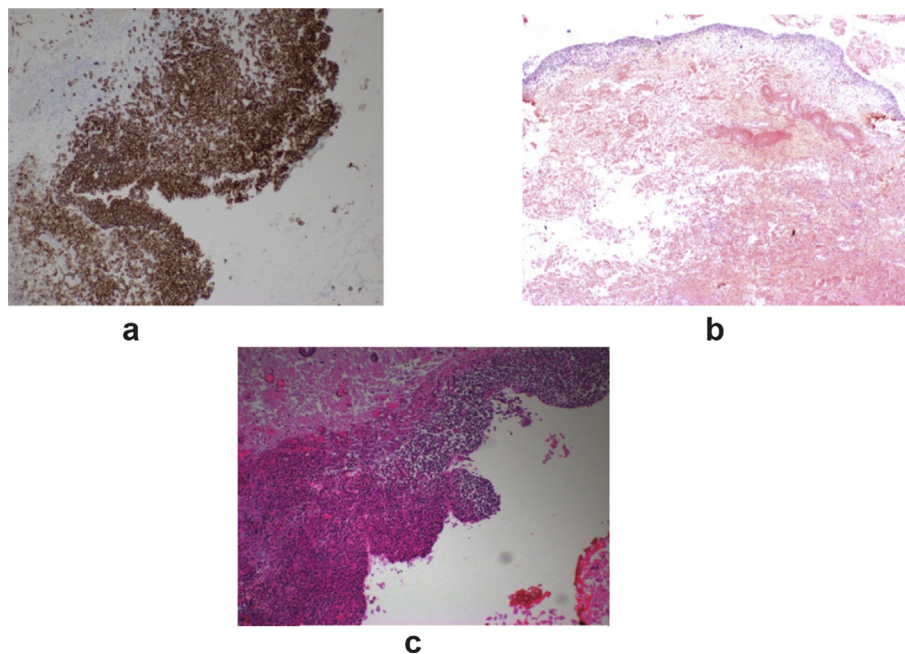


Figure 2: a: Histopathological examination of a specimen using CD138 stain. b: Histopathological examination of a specimen using Congo red stain. c: Histopathological examination of a specimen using haematoxylin & eosin (H&E) stain.

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