Radiologic Findings of Primary Localized Amyloidosis of Urinary Bladder

Primer Lokalize Mesane Amiloidozunun Radyolojik Bulguları

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ABSTRACT

Primary localized amyloidosis of the urinary bladder is a very rare disease with only 200 cases reported in the literature. However, it is an important condition because it can be easily confused with malignancy. It is most frequently observed during the fifth and sixth decade of life without sex preponderance. The classical presenting symptoms include painless gross hematuria and irritative lower urinary tract symptoms. The treatment of choice is transurethral resection, and histopathological evaluation is essential for the diagnosis and exclusion of malignancy. A close follow-up of the patient is required because of frequent recurrences. We present a case of primary localized amyloidosis of the urinary bladder with radiologic findings of a patient presenting with painless gross hematuria and mild dysuria.

Keywords: Primary bladder amyloidosis, painless gross hematuria, bladder tumor

ÖZ

Primer lokalize mesane amiloidozu literatürde sadece 200 vakanın bildirilmiş olduğu oldukça nadir bir hastalıktır. Fakat benzer klinik, görüntüleme ve sistoskopi bulgularına sahip olmasından dolayı kolaylıkla malignite ile karıştırılabileceği için oldukça önemlidir. Cinsiyet ayrımı gözetmeksizin en sık 5. ve 6. dekatlarda görülür. Klasik prezantasyonu ağrısız gros hematüri ve iritatif alt üriner sistem semptomlarıdır. Tanı için histopatolojik inceleme şarttır. Tedavisi transüretral rezeksiyondur. Rekürrens sıklığı nedeni ile hastanın yakın takibi gereklidir. Biz burada ağrısız gros hematüri ve hafif dizüri ile prezente olan primer lokalize mesane amiloidoz vakamızı radyolojik bulguları ile sunuyoruz.

Anahtar kelimeler: Primer lokalize mesane amiloidozu, ağrısız gros hematüri, mesane tümörü

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INTRODUCTION

Amyloidosis is a heterogeneous group of disorders associated with a systemic or localized deposition of insoluble eosinophilic fibrillar protein in the extracellular spaces of various tissues or organs. Both systemic and localized amyloidosis can be primary or secondary. If associated with monoclonal plasma cell dyscrasia, it is categorized as primary, and when it occurs as a complication of chronic inflammatory diseases, including rheumatoid arthritis, chronic osteomyelitis, or malignancies, it is categorized as secondary. Primary localized amyloidosis of the urinary bladder is a rare disorder; however, it has a clinical importance because of its similar presentation and work-up findings with malignancy that of the urinary bladder. Patients characteristically present with painless gross hematuria and irritative lower urinary tract symptoms. There is no pathognomonic imaging finding that distinguishes it from carcinoma of the bladder. Therefore, a histopathological examination is essential for diagnosis.

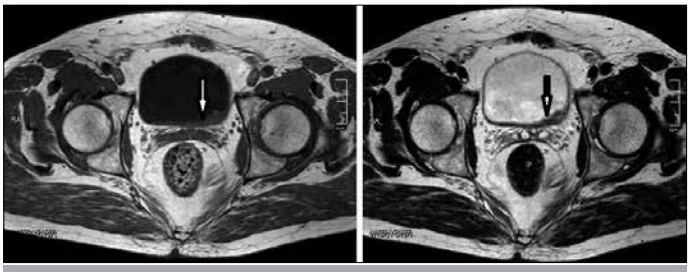

Figure 1. Hypointense mass on T1W and T2W images

CASE PRESENTATION

A 53-year-old man presented to the urology department with painless gross hematuria and mild dysuria with no associated constitutional symptoms. He was a nonsmoker. His past medical history and physical examination were unremarkable. Routine biochemical tests, including renal, liver, and bone, parameters were normal. A sonographic examination revealed a mass protruding into the lumen of the urinary bladder with an irregular surface and a diameter of 12 mm. Magnetic resonance imaging (MRI) showed that the mass was hypointense on T1-weighted T1W and T2-weighted (T2W) images (Figure 1) and had a marked enhancement on postcontrast images. A

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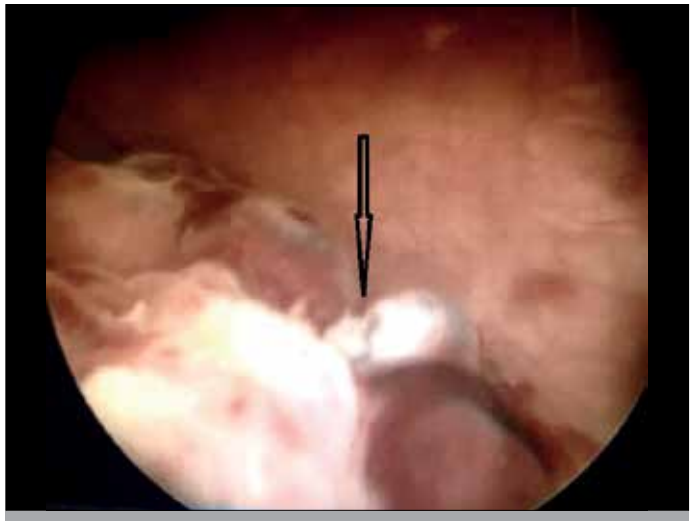

Figure 2. Cystoscopy showing urinary bladder nodules with edematous wall

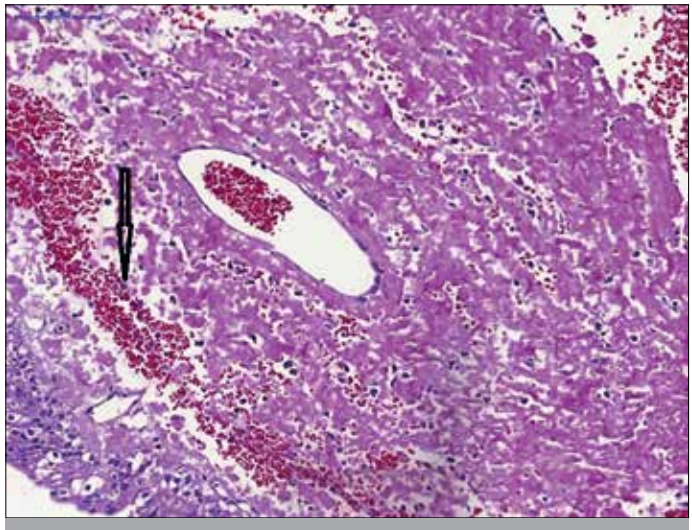

Figure 3. Histopathology (hematoxylin and eosin staining) showing pink amorphous material in the lamina propria

cystoscopy revealed nodular masses showing edematous wall (Figure 2). The patient underwent transurethral resection. A histopathological examination with routine hematoxylin and eosin staining demonstrated pink amorphous material in the lamina propria (Figure 3). To confirm the presence of amyloid, Congo red staining was performed, and it revealed the characteristic "apple green birefringence" under polarized light microscopy. Immuneprofile studies showed an AL (Lambda) subtype. All the findings were highly suggestive of primary bladder amyloidosis. To exclude systemic amyloidosis, a further evaluation was performed. Chest X-Ray and electrocardiogram revealed no pathologic findings. Urine analysis did not show proteinuria. There was no increased lymphocyte count and protein gap. Serum and urine protein electropheresis showed no abnormality with the normal level of kappa- and lambda-free light chains. After the exclusion of systemic amyloidosis, primary localized bladder amyloidosis diagnosis was confirmed, and a cystoscopic follow-up was suggested.Written informed consent was obtained from the patient who participated in this study.

DISCUSSION

Primary localized bladder amyloidosis is a very rare disorder. Only 200 cases have been reported in literature according to Wilkinson et al. (1). Localized amyloidosis, also known as amyloidoma, entails local amyloid accumulation in the extracellular compartments of tissues, resulting in nodular masses. It is observed at various sites in the body, including lungs, trachea, larynx, tongue, skin, nervous system, gastrointestinal tracts, and urinary tract (2). Although these amyloid deposits are derived from monoclonal light chains, they are not associated with systemic clonal plasma cell diseases. The usual presenting symptoms include painless gross hematuria and irritative lower urinary tract symptoms (3). While primary systemic amyloidosis is usually not associated with lower urinary tract symptoms, primary localized bladder amyloidosis is often accompanied by these clinical findings (4, 5). Bladder amyloidosis is radiologically seen as bladder wall thickening or mass lesion, and in most of the cases, it is difficult to differentiate from primary urothelial carcinoma. In bladder amyloidosis, a linear calcification of the bladder wall may be seen (6, 7). In the present case, there was no calcification. Another finding related with amyloidosis is hypointensity in the bladder wall on T2-weighted imaging. However, desmoplastic metastases and lymphomatous involvement may also demonstrate the same signal characteristics on T2-weighted imaging (8). Cystoscopically, amyloid deposits within the bladder wall mostly result in nodular or polypoid lesions. As most radiological findings and macroscopic appearances may mimic primary urothelial carcinoma, histopathological examination showing the presence of monoclonal light chains in the resection specimen is essential for diagnosis. After the exclusion of systemic disease, surgical excision is the choice of treatment. The clinical course is generally benign, but because the post-resection recurrence rate is as high as 50% (6, 9), long-term follow-up with a cystoscopic examination is recommended.

CONCLUSION

Primary localized amyloidosis of the urinary bladder is very rare, but because it presents with clinical, radiological, and cystoscopic findings that are similar to those of primary urothelial carcinoma of the bladder, it has a diagnostic importance. Although there are some imaging findings that are seen in amyloidosis, no pathognomic finding is present radiologically. It has a benign clinical course and transurethral resection is usually curative. Therefore, a histopathological examination is essential to make the definitive diagnosis and to avoid overtreatment. Another important point is the exclusion of the systemic disease when AL amyloidosis is diagnosed histopathologically. Because of the high recurrence rate, long-term follow-up is recommended.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

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