MR Images of Primary Localized Amyloidosis of the Ureter and Bladder: A Case Report

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Primary localized amyloidosis of the ureter and bladder is a very rare condition. CT and cystoscopic findings are indistinguishable from malignant neoplasm or inflammatory lesions. We report a case of unilateral distal ureteral thickening caused by primary localized amyloidosis. MR image features of hypointensity on T2-weighted images and mural enhancing pattern on contrast enhanced T1-weighted images can be helpful for diagnosis of ureteral amyloidosis.

Index words : Amyloidosis
Ureter
Magnetic resonance (MR)

Introduction

Amyloidosis is characterized by extracellular deposition of an abnormal protein material. Localized primary amyloidosis of the urogenital tract is a rarely described cause of ureteral obstruction and renal failure. It is difficult to distinguish the lesions from malignant tumor and inflammatory lesion on cystoscopy or CT scan (1, 4). MR images of genitourinary tract have recently been reported to be helpful in diagnosis of bladder amyloidosis (1, 3, 4). However, MR image features of amyloidosis of the ureter have not been well documented in the radiologic literature. We report the MR images of a case presented with hematuria and found to have amyloidosis involving the unilateral distal ureter.

Case Report

A 49-year-old man presented with several episodes of gross hematuria for 15 months. He has a history of urinary amyloidosis treated 11 months previously by transurethral resection, and received biweekly bladder injection with DMSO (Dimethyl Sulfoxide) for 4 months. The physical examination was unremarkable. The initial investigation showed a normal complete blood cell count and serum electrolyte and creatinine level. Urine cytologic analysis demonstrated 6–10 red blood cells, 2–3 white blood cells on high power field, and negative for malignant cells.

Computed tomography (CT) demonstrated left hydronephrosis and segmental wall thickening with enhancement in left distal ureter (Fig. 1a, b). Cystoscopy

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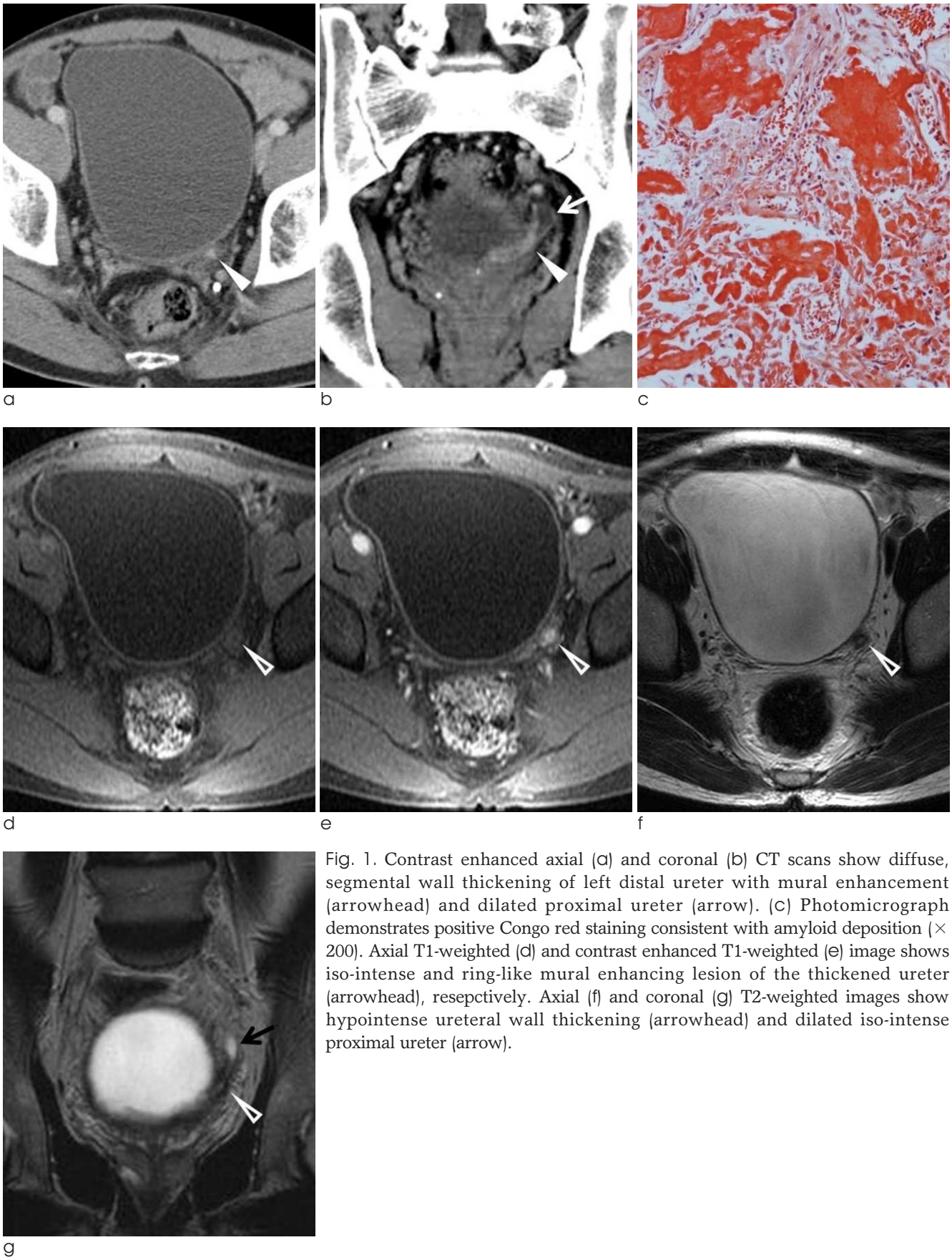

Fig. 1. Contrast enhanced axial (a) and coronal (b) CT scans show diffuse, segmental wall thickening of left distal ureter with mural enhancement (arrowhead) and dilated proximal ureter (arrow). (c) Photomicrograph demonstrates positive Congo red staining consistent with amyloid deposition (× 200). Axial T1-weighted (d) and contrast enhanced T1-weighted (e) image shows iso-intense and ring-like mural enhancing lesion of the thickened ureter (arrowhead), resepctively. Axial (f) and coronal (g) T2-weighted images show hypointense ureteral wall thickening (arrowhead) and dilated iso-intense proximal ureter (arrow).

showed irregular, edematous lesions in the lateral wall of urinary bladder and patchy submucosal yellowish plaque extended into the left ureteric orifice. No tumors or stones were seen in the bladder and left distal ureter. A urinary wash of the lesion revealed no malignant neoplasm with only reactive urothelial cells and fragments of amorphous material. Multiple biopsies were performed on bladder and distal ureter. Biopsy specimens showed submucosal proteinaceous material with positive Congo red staining consistent with amyloid with overlying benign urothelium (Fig. 1c).

After 4 months, MR images (Signa 1.5T GE. Milwaukee, WI, U.S.A.) demonstrated segmental wall thickening of left distal ureter that was isointense on T1-weighted images, hypointense on T2-weighted images, and ring-like mural enhancement on contrast enhanced T1-weighted images (Fig. 1d-g). There was no demonstrable mass lesion or regional lymphadenopathy. DMSO bladder instillation was repeated every 2 weeks at our institute and conservative management was performed. No systemic involvement of other organs was detected and after 8 months follow up, no symptomatic recurrence developed.

Discussion

Amyloidosis is characterized by extracellular tissue deposition of proteins, which can be classified as "primary amyloidosis", when no etiology is apparent with respect to amyloid deposition, or "secondary amyloidosis", when it occurs as a complication of an underlying chronic inflammatory disorder such as osteomyelitis, rheumatoid arthritis, leprosy, tuberculosis, or syphilis (5–7). Amyloid deposition may be confined to one organ (localized) or involve multiple organ systems (systemic). The correct diagnosis is confirmed by histopathological examination using Congo-red staining and the exclusion of other etiologies. When amyloid is detected in urinary tract, it is important to exclude systemic amyloidosis by performing serum electrophoresis, urine electrophoresis, chest radiography, and, possibly, rectal biopsy or fine needle aspiration of subcutaneous fat (7).

Primary localized amyloidosis of the urinary tract is a rare condition. In the urinary tract, the bladder is the most frequently affected site of isolated amyloid deposition with rare occurrences in the ureter or renal pelvis (5, 6). The primary form of disease is characterized by thick ureter walls due to deposition of light chains of immunoglobulin in the stroma and muscle, the distal third of ureter being the most common site involved (6). The etiology of primary localized amyloidosis is unknown but has been postulated to be related to recurrent, chronic inflammation, leading to chronic cystitis and migration of lymphoplasmacytic cells to the site (7). One of these proliferating cells becomes monoclonal, secreting light chains that are deposited in the tissue locally (8).

On radiologic examination, primary local amyloidosis showed focal narrowing of distal ureter, and should be differentiated from some benign or malignant lesions including severe edema or fibrosis, secondary to infection, inflammation, irradiation, complicated urolithiasis, malacoplakia, atypical pyelonephritis, tuberculosis, and urothelial carcinoma (4, 9). CT shows diffuse thickening of the urinary bladder wall in patients with amyloidosis, which is indistinguishable from bladder tumor and inflammation (3). Although some authors consider the presence of submucosal calcifications in the ureter as a pathognomonic sign for amyloidosis (6), these radiological findings did not indicate a specific etiology. In this case, CT showed relatively long segmental thickening of distal ureter without abrupt obstruction.

MR imaging is reported as useful modality for differential diagnosis of amyloidosis from other pathologic conditions: amyloid deposits is appeared as low intensity in T2-weighted images (1–4). In our case, T2-weighted images demonstrated hypointense wall thickening of the ureter. The deposition of amyloid and protein precursors could be pay a role in decreasing T2 relaxation times because they consist of β-plated sheet fibrils showing rapid phase dispersion and spin-spin interaction (3, 4). Transitional cell carcinoma and other inflammatory lesions show relative hyperintensity on T2-weighted images. The differential diagnosis of T2 shortening lesions includes amyloidosis, lymphoma, desmoplastic reaction to hemorrhage and calcifications (1).

Although amyloid material does not show enhancement (2, 3), few cases of localized amyloidosis involving urinay bladder have been described as homogenous or minimal enhancement on contrast enhanced T1-weighted images (1, 4). In our case, contrast enhanced T1-weighted images showed ring-like mural enhancement in thickened ureteral wall. Large amount of interstitial tissue and fluid, infiltration of inflammatory cells, and neovascularization were presented around the amyloid

material could play some important roles for the enhancement (4). Although urothelial carcinoma is a hypovascular tumor, moderate enhancement is seen with gadolinium contrast material, which could not be differentiated with amyloidosis.

Clinically, genitourinary amyloidosis usually manifests as symptoms of urinary obstruction. The main presenting symptoms of ureteral amyloidosis are abdominal pain and hematuria. Because of the difficulty in differentiating between localized amyloidosis and malignant conditions of the urinary tract, in some case even an unnecessary nephroureterectomy is performed. However, since localized amyloidosis is a benign condition, conservative surgical treatment is usually performed and allows renal preservation. After diagnosis of genitourinary amyloidosis, evaluation for systemic amyloidosis is recommended. Additionally, patients with primary amyloidosis should be evaluated for a plasma cell dyscrasia, while a systemic inflammatory condition should be sought in those with secondary amyloidosis (5). Since amyloidosis of the urinary tract frequently recurs, long term follow-up is recommended.

Conclusion

Primary localized amyloidosis of the ureter is a rare disease. Both the clinical and imaging presentations of amyloidosis are usually non-specific, simulate those of neoplasm or inflammation, and may cause delay in diagnosis and appropriate treatment. With this report we wish to alert radiologist to the diagnostic possibility of localized amyloidosis in patients with segmental wall thickening of distal ureter with low signal intensity on T2-weighted images and mural enhancement on contrast enhanced T1-weighted images in patients with persistent urinary symptoms that do not respond to the usual therapies.

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요관과 방광의 일차성 국소 유전분증의 자기공명영상: 증례 보고

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요관과 방광에 발생한 일차성 국소 유전분증은 매우 드문 질환이며, 전산화단층촬영과 요도방광경 소견상 악성 종양이나 염증성 병변들과 구분하기 어렵다. 저자들은 일차성 국소 유전분증으로 편측 원위부 요관이 비후된 증례를 보고하고자 하며, T2강조영상에서 저신호강도와 조영증강 T1강조 영상에서 벽의 조영증강형태가 요관 유전분증의 진단에 도움을 줄 수 있는 소견으로 생각된다.

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