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Genitourinary

Amyloidosis of the renal pelvis presenting as flank pain

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ABSTRACT

Amyloidosis is a rare disease defined by accumulation of extracellular amyloid systemically or within a specific organ. Localized amyloidosis of the genitourinary system is extremely rare, with the predominate location being the bladder. The imaging findings are often non-specific and mimic urothelial carcinoma. We present a 49-year-old woman with a chief complaint of flank pain. A filling defect was discovered on radiological imaging. The defect was subsequently biopsied and proven to be a primary amyloidosis of the renal pelvis. We then review the radiological findings of amyloidosis of the genitourinary system.

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Introduction

Amyloidosis is a result of the accumulation of amyloid protein. It is a rare disease with approximately 6 cases per million. It can manifest as a systemic accumulation or localized, systemic being the more common form. Although the accumulation of amyloid can occur on any end organ, a localized amyloid is most common within the genitourinary, pulmonary, and cardiovascular systems. Localized amyloidosis of the renal pelvis is extremely rare. A literature review by Lu et al. demonstrated only 26 case reports of localized amyloidosis of the renal pelvis [1]. The present case report describes a rare case of a patient presenting with primary amyloidosis localized to the renal pelvis. The radiological findings are

reviewed and an overview of amyloidosis of the genitourinary system is presented.

Case description

History

A 49-year-old woman first presented to the emergency room for left flank pain. She described the pain as moderate to severe that has waxed and waned for the last several hours. She denied a fever or history of previous kidney stones. Her associated symptoms consisted of hematuria and nausea. There was no known pertinent family or previous surgical history. The urine

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analysis, the complete blood count, and the comprehensive metabolic panel were all within normal limits.

Imaging

Computed tomography (CT) imaging of the urinary tract was performed. The noncontrast CT images (Fig. 1A and B) showed fullness within the left renal pelvis demonstrating a focal area of hyperattenuation compared with the contralateral side. Subsequently, a CT urography was performed (Fig. 2A and B), which showed a filling defect within the left renal pelvis and moderate left-sided hydronephrosis. Additional images in the bone

window (Fig. 3A and B) enhanced the depicted filling defect. The most common process to present this way would be malignancy and was at the top of the differential at this time. Urology performed a cystoscopy and obtained a biopsy of the left renal pelvis mass. The pathology (Fig. 4A) demonstrated an urothelium overlying an expansile soft tissue proliferation demonstrating proteinaceous material. Additional staining with Congo red (Fig. 4B) showed the characteristic apple-green birefringence pathognomonic for amyloidosis.

In coordination with the findings and the pathologic result of amyloidosis, the patient underwent a nuclear medicine renal function test, which was normal. The patient will continue to

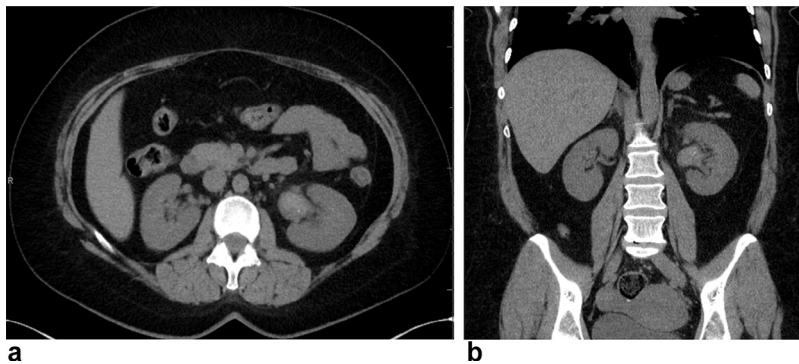


Fig. 1 – (A) Computed tomography noncontrast axial image at the level of the renal pelvis demonstrating a fullness of the left renal pelvis with areas of increased attenuation. **(B)** Computed tomography coronal noncontrast image through the level of the kidneys demonstrated the same fullness of the left renal pelvis with increased attenuation.

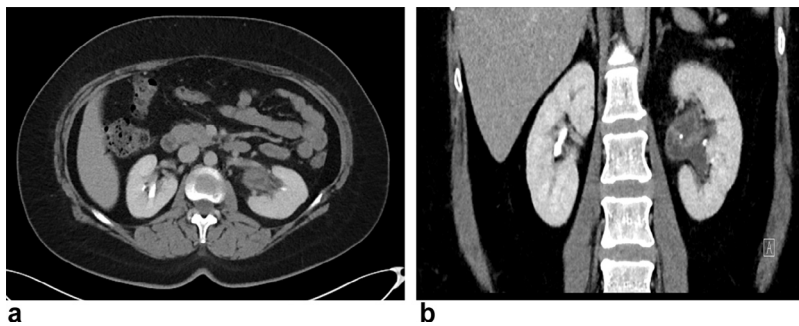


Fig. 2 – (A) Computed tomography urogram axial image demonstrates a filling defect in the left renal pelvis measuring 2.0×1.8 cm. **(B)** Computed tomography urogram coronal image demonstrating an associated moderate left-sided hydronephrosis. A tumor of the left renal pelvis could not be excluded.

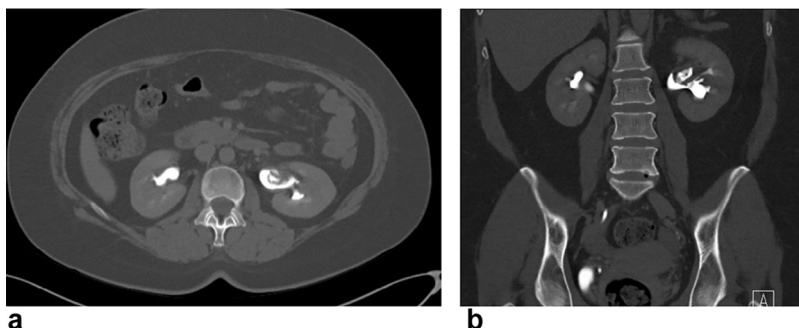


Fig. 3 – (A and B) Computed tomography urogram 10-minute delayed axial and coronal images on bone windows through the level of the kidneys demonstrating the filling defect within the left renal pelvis.

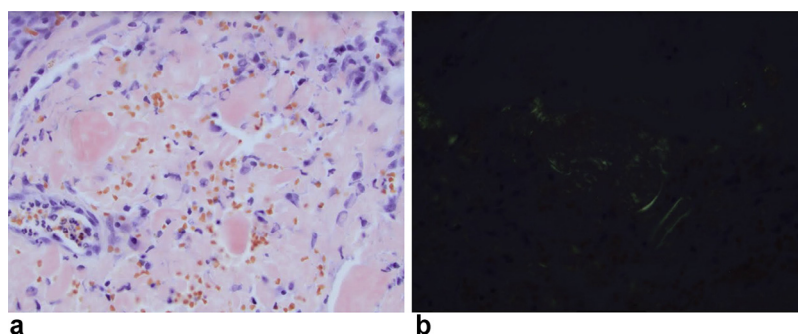


Fig. 4 – (A) The pathology slide demonstrates the urothelium overlying an expansile soft tissue proliferation showing proteinaceous material. (B) Congo red stain demonstrating the characteristic apple-green birefringence that is pathologic for amyloid.

be monitored closely with imaging and kidney function testing to watch for renal failure, which can occur as amyloidosis progresses.

Pathology: A surgical biopsy of the left renal pelvis was obtained. Sections demonstrated an urothelium overlying an expansile soft tissue proliferation containing proteinaceous material that, with Congo red stain, showed apple-green birefringence. The findings are consistent with amyloidosis.

Discussion

The human body readily makes various forms of amyloid by a process known as fibrillogenesis during protein synthesis. When errors occur in the folding of these proteins, the human body is unable to break down the amyloid, resulting in an accumulation systemically or within various end organs. This disease process is known as amyloidosis. There are several different subtypes of amyloidosis, the most common being amyloid light-chain (primary) results from the deposition of protein derived from immunoglobulin light-chain fragments [2]. A second type, amyloid A protein (AA), is more commonly associated with systemic disease resulting from a chronic inflammatory state such as rheumatoid arthritis, Crohn disease, and chronic osteomyelitis. Many other forms of amyloid can occur, such as β 2-microglobulin with long-term hemodialysis and A β 2 in Alzheimer disease. The clinical manifestations vary widely based on the type of protein precursor and the location of deposition. Systemic amyloidosis is far more common than localized amyloidosis, but the most common systems involved with localized amyloidosis are genitourinary, cardiovascular, and pulmonary.

Renal involvement by amyloid is often secondary to systemic disease by β 2 microglobulins, commonly as a result of chronic dialysis. In the case of systemic amyloidosis of the genitourinary system, the amyloid may infiltrate the retroperitoneal and pelvic soft tissues, encasing the urinary tract and resulting in progressive calcification [3]. The etiology of primary localized amyloidosis to the genitourinary system is unknown. Although a rare manifestation, the bladder is most commonly involved. The CT findings demonstrate a focal or a diffused bladder wall thickening or a focal filling defect. Less commonly, amyloid affects the kidneys or ureters, resulting in the

findings of kidney enlargement or focal filling defects within the calyx and the renal pelvis, or extending down the ureters. Chronic amyloidosis of the kidneys demonstrates the common findings of end-stage kidney disease showing atrophy of the kidneys with cortical thinning.

Isolated disease within the renal pelvis is extremely rare, and presenting symptoms are often vague and mimic systems of a kidney stone, infection, or neoplasm. Cytology offers little value with a majority of the deposition of amyloid within the subendothelium resulting in a very low quantity excreted in the urine. One imaging finding that may suggest amyloidosis is linear submucosal calcifications [4]. However, findings are often nonspecific and mimic urothelial carcinoma, and a histologic correlation is required for a definitive diagnosis. The pathognomonic apple-green birefringence on Congo red staining is diagnostic for a primary localized amyloidoma.

Conclusion

In summary, amyloidosis of the renal pelvis is rare with nonspecific radiological findings that mimic malignancy. Correlation with pathology is required for diagnosis, which will demonstrate the characteristic apple-green birefringence on Congo red stain. Although rare, it is imperative that a radiologist be familiar with the radiographic findings in the appropriate clinical setting to help guide proper diagnosis and early management.

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