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Primary localized amyloidosis of the ureter with osseous metaplasia presenting as a suspicious ureteral mass

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ABSTRACT

Primary amyloidosis of the ureter is a rare disease that is difficult to distinguish from urothelial carcinoma. Only 50 cases of primary ureter amyloidosis have been reported since it was first described in 1937. Of these, only five cases of ureter amyloidosis with osseous metaplasia were reported. In this study, we report the clinical presentation of ureter primary amyloidosis that presented as a mass with osseous metaplasia. The aim of this study is to provide clinicians with knowledge about the clinical/radiologic manifestation that raise the suspicion of amyloidosis, bearing in mind the importance of differentiating it from other "malignant" processes.

1. Introduction

Primary localized amyloidosis of the ureter is a rare disease that is clinically and radiographically difficult to distinguish from urothelial carcinoma.[1] It occurs predominantly in females.[1] The mean age at diagnosis is 58 years and most patients present with flank pain or hematuria. The majority of ureteric involvement (up to 60% of cases) was reported in the lower ureter, followed by the upper portion according to a report by Ding et al.[2] Only 50 cases of primary ureteric amyloidosis have been reported since it was first described in 1937.[2] Of these, only five cases of ureter amyloidosis with osseous metaplasia were reported. In this study, we report the clinical presentation and pathologic findings of primary amyloidosis in the ureter and demonstrate the unusual presentation of the primary amyloidosis of the ureter as a lobulated nodular mass and pertinent osseous metaplasia association. The aim of this case report is to provide clinicians with knowledge about the clinical/radiologic manifestation that raise the suspicion of amyloidosis, bearing in mind the importance of differentiating it from other "malignant" processes.

2. Case presentation

We report a case of 82-year-old female with past medical history of chronic diastolic heart failure, atrial fibrillation, and type 2 diabetes who presented with weakness and fatigue. A computed tomography (CT) scan of the abdomen/pelvis without contrast demonstrated an incidental finding of right ureteral mass with layering calculi concerning for neoplasm. The mass led to ureteral obstruction and right sided hydronephrosis. Urine culture grew *E. coli*. The patient underwent ureteroscopy with biopsy and stent placement.

Under light microscopy, the specimen revealed amorphous, homogenous, eosinophilic extracellular material expanded lamina propria and muscularis propria extending to the adventitia. Osseous metaplasia was also apparent. Fig. 1. Congo red staining was positive with unique apple-green birefringence on polarization. Fig. 2. Unfortunately, the patient did not pursue further follow up.

3. Discussion

Primary localized amyloidosis in the ureter is a rare entity caused by extracellular deposition of insoluble fibrillary protein that may result in

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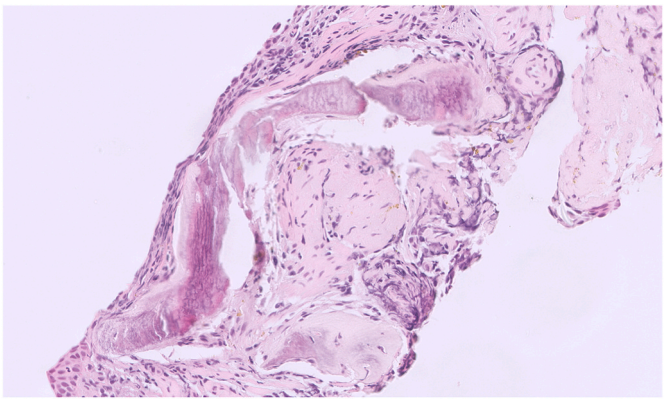

Fig. 1. H&E stains amorphic and eosinophilic acellular material in the ureter specimen.

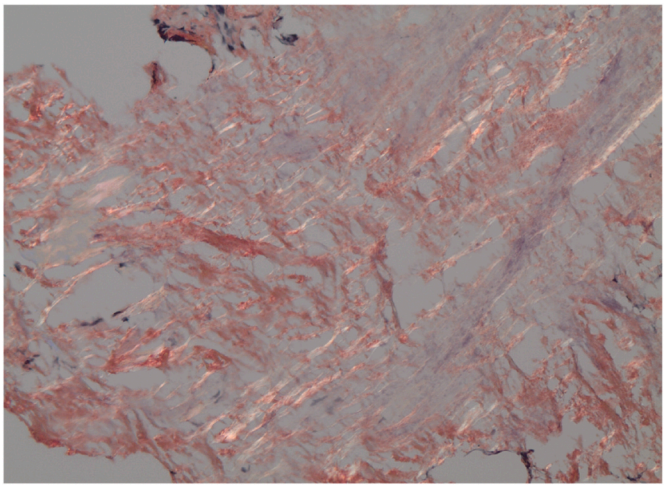

Fig. 2. Positive Congo Red staining consistent with the diagnosis of ureteric amyloidosis. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

severe organ dysfunction.[1] Amyloidosis is classified based on the type of the involved amyloidogenic protein or based on tissue distribution.[1] Primary amyloidosis is usually defined as amyloid immunoglobulin light chain disease that could involve different organs typically affecting the skin, soft tissues, genitourinary system, respiratory system, and gastrointestinal tract. Primary amyloidosis of the genitourinary tract is rare and most reported in the bladder and the prostate. The etiology of localized amyloidosis is unknown. Several studies suggested that primary amyloidosis could be related to chronic inflammation, monoclonal proliferation, and local secretion of light chains. Light chains accumulation may transform into amyloid by lysosomal degradation.

Secondary amyloidosis is often a result of a separate disease process, usually an underlying long term inflammatory disease such as rheumatoid arthritis, tuberculosis, or Crohn's disease. The kidney is the major involved organ in the genitourinary tract.[3] Alternatively, secondary amyloidosis could be an indication of a neoplastic disorder such as multiple myeloma. Because the clinical and radiologic findings are non-specific, histopathologic examination of the tissue remains the gold standard.

Amyloidosis, also named "The great imitator," because its nonspecific clinical presentation.[4] Although gross hematuria is a common manifestation, clinical signs and symptoms of ureteral obstruction such as hydronephrosis and hydroureter should raise the suspicion for amyloidosis in the genitourinary tract. This consideration could help in the avoidance of unnecessary surgery. Two conservative approaches can be used to preserve renal functioning after confirming the appropriate diagnosis by pathologic examination. One treatment option is segmental resection of the ureter with end-to-end anastomosis, ureteral reimplantation, and auto transplantation. The other treatment option is with dimethyl sulfoxide (DMSO).

Microscopically, the amyloid protein deposits in the tissue as an extracellular pale pink, homogenous, and hyaline acellular material on hematoxylin eosin staining, mostly deposited in vascular and perivascular tissue.[5] Congo-Red staining is generally recommended and reveals apple-green birefringence appearance under polarized light with high sensitivity and specificity.[5] Despite the relatively straightforward pathologic diagnosis of amyloidosis, additional subtyping is required for guiding the treatment. This subtyping is achieved by laser microdissection, then analysis by liquid chromatography/mass spectrometry technology. This methodology has emerged as the preferred method of amyloid typing and has its value for both confirmation of the diagnosis and subtyping and characterizing the peptide composition of the amyloidogenic protein with high sensitivity and specificity.

The differential diagnosis in this case included urothelial carcinoma, carcinoma in situ, and atypical reactive processes. Two interesting findings in our case were the presentation primary amyloid as a lobulated nodular mass formation that was suspicious for malignancy and the apparent osseous metaplasia formation. Local resection is the recommended clinical approach after ruling out a systemic disease process.

4. Conclusion

Primary localized ureteral amyloidosis is a rare and benign condition with only 50 cases reported in the literature. We report the sixth case of localized ureteral amyloidosis with associated osseous metaplasia. Due to the similarities in clinical/radiologic presentation with urothelial malignancy, a high level of awareness of this entity is required by the clinicians and pathologist to achieve an accurate diagnosis and appropriate patient management.

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