



Isolated prostate amyloidosis; a case report

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ABSTRACT

Identification of amyloidosis on a prostate biopsy specimen raises suspicion for systemic amyloidosis. For patients at risk of prostate adenocarcinoma, a transrectal MRI ultrasound fusion (MRI-TRUS) guided biopsy of the prostate is often needed for further investigation. Our patient is a 70 year old male presenting from his primary care provider with an elevated PSA. The patient underwent MRI-TRUS fusion biopsy of the prostate, which demonstrated AL (lambda)-type amyloidosis, confirmed by mass spectrometry in addition to prostatic adenocarcinoma. He was subsequently diagnosed with primary amyloidosis of the prostate without evidence of other amyloid deposition sites.

1. Introduction

Amyloidosis is characterized by extracellular deposition of insoluble fibrils following misfolding of various proteins. These pathologic fibrils are characterized by their β -pleated sheet conformation and when stained with Congo red dye have an apple-green birefringence, making them easily identifiable.¹ Amyloidosis can be either systemic or local, with local disease producing amyloidogenic protein at the site of deposition and systemic producing amyloidogenic protein distant from the original deposit site. Light chain (AL) amyloidosis is the most common form of systemic amyloidosis.²

Although amyloidosis is considered a rare disease, there is belief that its true prevalence is higher and is often underdiagnosed. However as the number of patients undergoing prostate biopsies for elevated PSA increases, the probability of identifying more systemic and local amyloidoses increases. We report a case of localized prostatic AL (lambda)-type amyloidosis and Gleason 6 (3 + 3) prostatic adenocarcinoma detected after MRI-TRUS fusion biopsy.

2. Case report

A 70-year-old male presented to the urology clinic following referral from a primary care provider with a chief complaint of “elevated PSA.” He has a past medical history of multiple stable subcentimeter bilateral

pulmonary nodules, as well a history of smoking and alcohol use. PSA value on presentation was 4.958 ng/mL and digital rectal exam revealed an enlarged prostate without any nodules. His prostate volume was calculated to be 22.7 cc, providing a PSA-density of 0.22. After discussion, he elected to undergo a transrectal prostatic needle biopsy which showed evidence of (1/12) cores positive for prostatic adenocarcinoma with 7% specimen involvement and a Gleason score of 6 (3 + 3). Thus, he was diagnosed with low risk prostate cancer per AUA/NCCN guidelines. After counseling on his treatment options, he elected to undergo active surveillance. Over the course of the next six and nine months, the patient’s PSA remained relatively stable at 5.249 and 4.642 ng/mL, respectively. After further discussion, he elected to undergo an MRI of his pelvis which showed a 1.4 × 1.4 cm T-2 hypointense lesion in the anterior transition zone of the left prostatic base with partially obscured margins, categorized as PI-RADS 3 (Fig. 1). He later underwent a subsequent MRI-TRUS fusion biopsy of the prostate. In addition to four cores of prostatic adenocarcinoma (two 3 + 4 and two 3 + 3), there was evidence of eosinophilic material in three separate cores (Fig. 2). This material was deemed by our local pathology department to be consistent with amyloid deposition via Congo red staining (Fig. 3). The congophilic material showed characteristic apple-green birefringence with polarized light. Further expert pathology review with mass spectrometry confirmed a protein profile consistent with AL (lambda)-type amyloid. Patient was referred to hematology to rule out systemic AL (lambda)-

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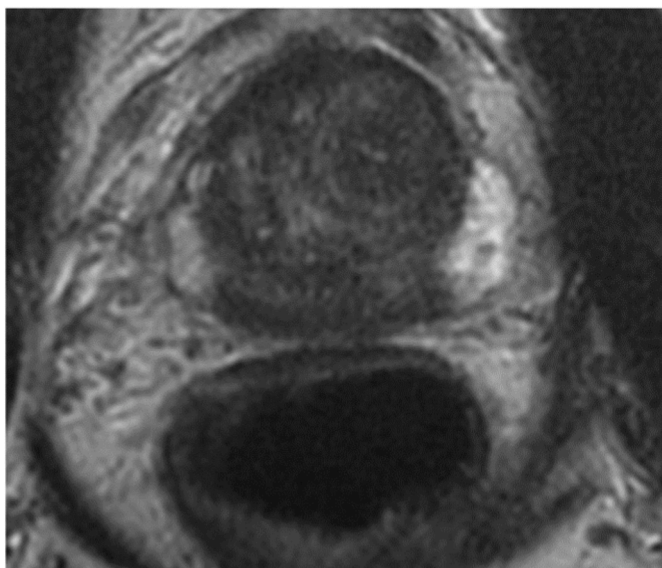


Fig. 1. T2-weighted axial MRI of the prostate (with rectum below) showing a rounded 1.4×1.4 cm hypointense lesion in the anterior transitional zone with partially obscured margins, considered a PI-RADS 3 lesion.

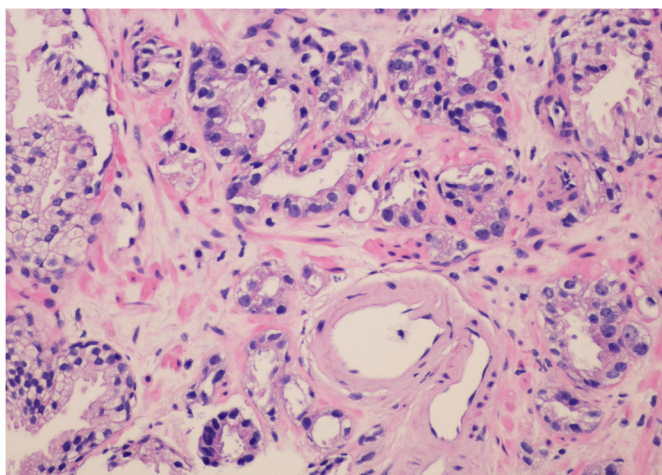


Fig. 2. Small artery with amyloid surrounded by prostatic adenocarcinoma.

type amyloidosis and cardiac evaluation. Extensive further exploration for systemic amyloidosis took place including: serum and urine protein electrophoresis, immunofixation, and free light chains; all without further evidence of additional amyloid deposition. PET CT was performed that showed no aberrant uptake and was deemed negative.

3. Discussion

Amyloidosis can be characterized as having systemic or local involvement, with single organ system involvement making up 6–9% of all cases.³ With incidental discovery of genitourinary amyloidosis, an evaluation for systemic and cardiac involvement should be undertaken.¹ After workup for systemic amyloidosis in our patient, amyloid deposition was solely found within the prostate. At the time of workup, our patient had no symptomatology related to his prostatic amyloid or Gleason 7 (3 + 4) prostatic adenocarcinoma and elected to continue active surveillance.

Amyloidosis of the prostate is one of the rarest forms of genitourinary amyloidosis with only a few cases being reported. A recent extensive review revealed quite a low prevalence, identifying only 7 out of many

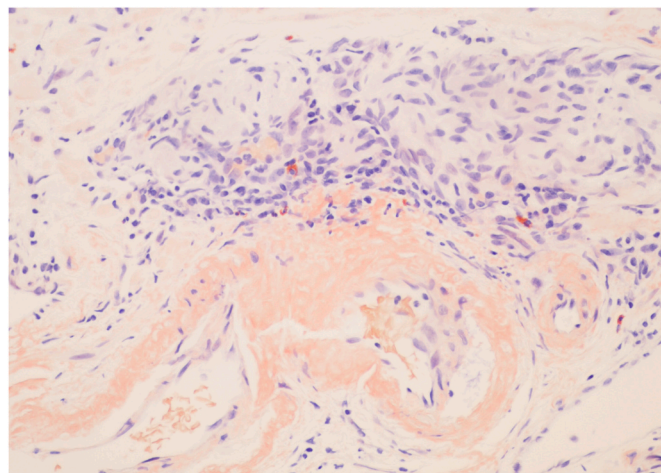


Fig. 3. Congo red stain with characteristic salmon amyloid deposits. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

thousands of patients from 2008 to 2018 were positive for amyloid deposits.⁴ In addition to the seven patients found to have amyloid deposits, three were diagnosed with concomitant prostate adenocarcinoma and five were diagnosed with cardiac amyloidosis. A different study examined prospectively 75 patient samples without a history of systemic amyloidosis who underwent transrectal biopsy of the prostate, of which eight (10%) had amyloid deposits.⁴

In systemic AL amyloidosis, early identification can have great implications as it is the most severe of the systemic varieties with a median overall survival of 59 months.⁵ Aggressive treatment aimed at targeting the underlying plasma cell clone can lead to complete remission, although prognosis is still poor. Through selective targeting of plasma cell dyscrasia, amyloid fibrils and intermediate soluble fibrils are eliminated, allowing for organ recovery. Currently, treatment options for systemic AL amyloidosis include multiple myeloma therapeutics, while for low risk patients high dose melphalan supported by autologous stem cell transplantation is the standard treatment.⁵ Although current treatment options have increased mean survival from months to years for patients at all stages of disease, further research into early diagnostic testing and treatment options is necessary.⁵ If urologists should find evidence of amyloidosis, additional testing by the appropriate specialty would be prudent.

4. Conclusion

Incidental discovery of amyloidosis of the prostate is a rare entity but important clinically as it may be evidence of a systemic amyloidosis. In our patient, an elevated PSA led to a prostate cancer diagnosis. Subsequent active surveillance including subsequent MRI investigation of the pelvis along with an additional biopsy led to an unusual isolated finding of prostatic amyloidosis. Further investigation and surveillance for sequelae of a systemic amyloidosis process should be undertaken. Our patient fortunately is currently without cardiac or suspicion of alternative organ involvement, however continues to undergo active surveillance.

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