Primary localized amyloidoma of the renal pelvis: A case report and literature review

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Abstract. Primary localized amyloidomas of the renal pelvis are challenging to diagnose, due to non-specific imaging results and the unusual location. The present study reports a rare case of primary localized amyloidoma of the renal pelvis and aims to illustrate the challenges in pre-operatively discriminating between this disease and transitional cell carcinomas. The present study identified that the mass was situated in the left renal pelvis using ultrasonography. A nephroureterectomy was performed following careful preparation. Finally, histopathological studies revealed that the tumor consisted of massive diffuse deposits of amyloid and microscopic amorphous eosinophilic material, which stained positively for Congo red, demonstrating potassium permanganate digestion. Consequently, a diagnosis of amyloid light chain-type amyloidoma was determined. Systematic examinations were performed following the unexpected diagnosis, which eliminated the possibility of amyloid associated-type amyloidoma. In total, 4 months post-surgery, the patient remained tumor-free.

Introduction

Localized amyloidoma is generally divided into two styles: AL-type and AA-type. AL-type occurs with an immunocyte dyscrasia while AA-type occurs with chronic infection, non-immunocyte neoplasia or inflammation. Localized amyloidoma occurs most often in the mediastinum or abdomen. Although amyloidomas may occur in almost all organ systems in the body; however, primary amyloidoma of the renal pelvis is rare. Using the keywords: ‘renal pelvis’, ‘amyloidoma’ and ‘amyloid tumor’ and search terms (renal pelvis) and (amyloidoma or amyloid tumor) in PubMed (National Center for Biotechnology Information, U.S. National Library of Medicine, Bethesda, MD, USA), a literature search was performed and only 26 cases of primary amyloidomas of the renal pelvis were identified (Table I) (1-23). Primary localized amyloidoma may present as hematuria and lumbago (14). A primary localized amyloidoma consists of amyloid deposits that may present as malignant tumors (15). The present study reports the rare case of a patient with amyloidoma that was confined to the renal pelvis, and the patient exhibited similar symptoms to those of upper urinary tract transitional cell carcinoma (TCC). In addition, 26 cases of patients with primary amyloidoma of the renal pelvis were identified from the literature were also reviewed. Nephrectomy was the most selected treatment in the reported cases. The majority of patients achieved long time disease free survival and good prognosis (7). The clinical and pathological features of the cases were discussed, in particular, the treatment methods and prognosis.

Case report

A 56-year-old man presented to the Department of Urology, Sun-Yat Sen University Cancer Center (Guangzhou, China) with left flank pain, symptoms of urinary irritation and intermittent gross hematuria. The patient had experienced the symptoms for 6 years; however, in the month prior to presentation, the
symptoms had become worse. Ultrasonography revealed a mass with a low-intensity heterogeneous pattern that measured 55x19 mm in size and obstructed the lumen of the renal pelvis. Dilatation of the renal pelvis was located in the left kidney (Fig. 1). A preliminary diagnosis of TCC of the renal pelvis was suggested based on sonography, and therefore, a preliminary diagnosis of TCC of the renal pelvis was suggested based on sonography, and therefore, a
<table>
<thead>
<tr>
<th>First author, year (Ref.)</th>
<th>Gender</th>
<th>Age, years</th>
<th>Symptoms or clinical finding</th>
<th>Medical history</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Akimoto, 1927 (22)</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Gilbert et al, 1952 (1)</td>
<td>F</td>
<td>52</td>
<td>Flank pain</td>
<td>None</td>
<td>Nephrectomy</td>
<td>NA</td>
</tr>
<tr>
<td>Sato, 1957 (23)</td>
<td>M</td>
<td>37</td>
<td>Hematuria and flank pain</td>
<td>None</td>
<td>Nephrectomy</td>
<td>NA</td>
</tr>
<tr>
<td>Chisholm et al, 1967 (2)</td>
<td>M</td>
<td>66</td>
<td>Painless hematuria</td>
<td>None</td>
<td>Nephrectomy</td>
<td>Succumbed to renal failure following a short period of time</td>
</tr>
<tr>
<td>Chisholm et al, 1967 (2)</td>
<td>F</td>
<td>58</td>
<td>Iron-deficiency anemia and mild azotemia</td>
<td>None</td>
<td>Left hemicolectomy for carcinoma of the sigmoid colon</td>
<td>Alive with no evidence of disease</td>
</tr>
<tr>
<td>Dias et al, 1979 (5)</td>
<td>F</td>
<td>67</td>
<td>Painless gross hematuria</td>
<td>None</td>
<td>Biopsy of renal pelvis</td>
<td>Alive with no evidence of disease</td>
</tr>
<tr>
<td>Ullmann, 1973 (3)</td>
<td>M</td>
<td>67</td>
<td>Left flank pain and rust-colored urine</td>
<td>None</td>
<td>Nephrectomy/partial ureterectomy</td>
<td>Succumbed to unknown causes 3 years later</td>
</tr>
<tr>
<td>Gardner et al, 1971 (3)</td>
<td>F</td>
<td>58</td>
<td>Hematuria and flank pain</td>
<td>None</td>
<td>Nephrectomy</td>
<td>NA</td>
</tr>
<tr>
<td>Dias et al, 1979 (5)</td>
<td>F</td>
<td>67</td>
<td>Painless gross hematuria</td>
<td>None</td>
<td>Radical nephrectomy</td>
<td>Alive with no evidence of disease</td>
</tr>
<tr>
<td>Gelbard et al, 1980 (6)</td>
<td>M</td>
<td>67</td>
<td>Left flank pain and gross hematuria</td>
<td>None</td>
<td>Nephrectomy</td>
<td>Alive with no evidence of disease 5 years later</td>
</tr>
<tr>
<td>Davis et al, 1987 (10)</td>
<td>F</td>
<td>51</td>
<td>Flank pain, fever and gross hematuria</td>
<td>None</td>
<td>Nephrectomy</td>
<td>Alive with no evidence of disease 2 years later</td>
</tr>
</tbody>
</table>

Table 1. Review of the reported cases of primary localized amyloidoma of the renal pelvis.
Table I. Continued.

<table>
<thead>
<tr>
<th>First author, year (Ref.)</th>
<th>Gender</th>
<th>Age, years</th>
<th>Symptoms or clinical finding</th>
<th>Medical history</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Borza et al, 2010 (15)</td>
<td>M</td>
<td>58</td>
<td>Gross, painless hematuria and right flank pain</td>
<td>None</td>
<td>Active surveillance</td>
<td>No clinical or radiographical signs of progressive disease 6 years later</td>
</tr>
<tr>
<td>Pan et al, 2011 (16)</td>
<td>M</td>
<td>70</td>
<td>None</td>
<td>Partial nephrectomy of the right kidney for an angiomyolipoma 4 years prior</td>
<td>Active surveillance</td>
<td>Recurrence in the unilateral bladder and ureter</td>
</tr>
<tr>
<td>Monge et al, 2011 (17)</td>
<td>M</td>
<td>68</td>
<td>Gross hematuria and renal colic</td>
<td>None</td>
<td>Nephrectomy, repeated, resections double J stent</td>
<td>No recurrence. Only mild renal insufficiency remained 5 years later</td>
</tr>
<tr>
<td>Paidy et al, 2012 (20)</td>
<td>F</td>
<td>72</td>
<td>Gross hematuria and right flank pain</td>
<td>Nephrolithiasis, hypertension, osteoarthritis stroke, transient ischemic attack, atrial fibrillation and hypercholesterolemia</td>
<td>Nephrectomy</td>
<td>NA</td>
</tr>
<tr>
<td>Zhou et al, 2014 (21)</td>
<td>F</td>
<td>77</td>
<td>Gross hematuria</td>
<td>None</td>
<td>Nephrectomy</td>
<td>Alive with no evidence of disease 12 years later</td>
</tr>
<tr>
<td>Zhou et al, 2014 (21)</td>
<td>M</td>
<td>71</td>
<td>Gross hematuria</td>
<td>None</td>
<td>Nephrectomy</td>
<td>Alive with no evidence of disease 6 years later</td>
</tr>
<tr>
<td>Grigor, 2015 (18)</td>
<td>F</td>
<td>60</td>
<td>Gross hematuria intraepithelial</td>
<td>Supraventricular tachycardia, neoplasia. Retinal detachment, macular hole and cervical lattice degeneration in right eye</td>
<td>Laparoscopic nephrectomy</td>
<td>Alive with no evidence of disease 30 months later, but diagnosed with breast cancer 21 months ago</td>
</tr>
<tr>
<td>Present study</td>
<td>M</td>
<td>56</td>
<td>Gross hematuria</td>
<td>Kidney calculi</td>
<td>Nephrectomy</td>
<td>Alive with no evidence of disease 4 months later</td>
</tr>
</tbody>
</table>

NA, not available; M, male; F, female.
Nephroureterectomy was advised. However, 5 urinary cytology tests were negative and pre-operative examinations revealed no abnormal signs, such as abdominal tenderness or rebound pain, during physical examination. Blood tests yielded the following results: Urine protein, 2+; urine red blood cell count, >3 cells/high power field; urine erythrocytes, 932 µl/l; left renal glomerular filtration rate, 18 ml/min; right renal glomerular filtration rate, 64 ml/min; and the serum creatinine levels, eosinophil numbers and basophil numbers were within the normal ranges. The patient had been a smoker for ~20 years and had suffered from nephrolithiasis for 6 years. The patient possessed no known drug allergies. Subsequently, the patient agreed to undergo a nephroureterectomy.

Following surgery, the excised mass underwent additional tests. Macroscopically, the surgical specimen revealed tumors located in the renal pelvis. No clear difference in the ureter was observed. A cut section of the tumor revealed a red-yellow surface with firm regions. Microscopically, histopathological studies revealed that the tumor consisted of massive diffuse deposits of amyloid and microscopic, eosinophilic, amorphous material (Fig. 2) and an absence of neoplastic cells. The tumor stained positive for Congo red (Fig. 3), which indicates the presence of material that is retained following potassium permanganate digestion. A final diagnosis of amyloid light chain-type amyloidoma of the renal pelvis was determined. The patient received regular surveillance and was alive with no evidence of disease 5 months later.

Written informed consent was obtained from the patient for the publication of the present study.

Discussion

Amyloidosis refers to a large heterogeneous group of diseases that is characterized by extracellular deposits of amyloid in individual organs or tissue. Amyloid is an amorphous, insoluble and proteinaceous material (17). Extracellular amyloidosis consists of specific protein fibrils (24). Amyloidosis can be classified into 4 groups, consisting of primary, secondary, heredofamilial and β2-microglobulin-associated amyloidosis. This disease may be additionally classified as localized amyloidosis, which involves a single organ, or systemic amyloidosis, which involves multiple organs and is the most common classification among reported cases (6). By contrast, localized amyloidosis occurs much less frequently. (15) Systemic amyloidosis may be primary, progressive and fatal. Primary amyloidosis is commonly associated with an underlying immune dyscrasia, such as multiple myeloma and Waldenstrom's macroglobulinemia (15).

The etiology of primary localized urinary amyloidosis remains unknown; however, it may be possible that amyloid deposits are produced locally, or the submucosa of the genitourinary tract may be targeted by light-chains that are produced elsewhere. Numerous studies support the first hypothesis, as there is an absence of a monoclonal plasma component. An additional hypothesis is that protein deposits in bladder non-amyloid associated (AA) localized amyloidosis consist of the immunoglobulin light chain subgroup I or IV (17).

Furthermore, non-AA localized amyloidosis has been described in the lung (25), nervous system (26), skin (24), larynx (27), intestinal tract (28) and genitourinary tract (17). Using PubMed, a literature search was performed and only 26 cases of primary amyloidomas of the renal pelvis were identified. The inclusion and exclusion criteria is whether primary localized amyloidoma. The majority of reported cases concerned with localized urinary tract amyloidosis are characterized as primary type, but secondary localized amyloidosis has been reported without a chronic systemic inflammatory state. Renal pelvis primary localized amyloidoma is an extremely rare condition, and is notable due to its clinical presentation and radiographic appearance, which mimics that of transitional cell carcinoma. In a review of the English and French literature by Monge et al (17), 169 cases of genitourinary tract localized amyloidoma were reported over the past 100 years. The renal pelvis was the most rare location identified, accounting for ~6% of the 169 cases, which was lower than the number located in the bladder, ureter and urethra. To the best of our knowledge, only 26 cases of renal pelvis primary localized amyloidoma have been reported (1-23).

Positive staining for Congo red is used to diagnose primary localized amyloidoma. In order to exclude AA-type amyloidosis, screening by pre-exposure of tissue slides to KMnO4, stain is performed, since false-negative results may be obtained using immunohistochemistry (negative for Congo red) (17). Clinical symptoms of primary localized amyloidoma of the renal pelvis typically present as gross hematuria, flank pain and urinary irritative symptoms, which mimic the symptoms of inflammation and neoplasm (21). It is challenging to distinguish between amyloidoma and upper urinary tract TCC solely from radiological findings, which may be non-specific (17). In addition, urine cytology does little to contribute to the diagnosis, since the majority of amyloid deposits are subepithelial. Consequently, to avoid misdiagnosis, upper urinary tract tumors should be evaluated microscopically by ureteroscopic biopsy when multiple urine cytology analyses are negative, or if possible, by surgical biopsy using frozen tissue sections examined prior to radical resection.

Primary localized amyloidoma possesses a relatively good prognosis in the genitourinary tract and other organ systems. Recurrence and implantation of the tumor has not been identified in reported cases. Nephrectomy was the chosen treatment in the present study, and is used in the majority of reported cases. However, by monitoring the progression of the primary localized amyloid tumor in the renal pelvis, using active surveillance with serial imageological examination, a similar outcome to nephrectomy may be observed (15). This should be considered by all urologists.

Primary localized amyloidoma is extremely rare in the renal pelvis (14,15). Consequently, there is a lack of standardized clinical symptoms and specific laboratory tests, and in addition, imageological examination mimics TCC. A pre-operative ureteroscopic biopsy or surgical biopsy is required when multiple urinary cytology analyses are not positive or a benign tumor, including primary localized amyloidoma, is suspected, to avoid an unnecessary nephroureterectomy. Early radical surgery is not required unless there is no renal function or a severe urinary tract obstruction. Instead, active surveillance with serial imageological examination may be used to monitor the progression of the lesion. In conclusion, primary localized amyloidoma of renal pelvis is a benign and rare tumor, which has a relatively good prognosis.
References