

Case Report

Primary renal plasmacytoma: Case report and literature review

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Extramedullary plasmacytoma (EMP) results from uncontrolled plasma cell proliferation and consists of monoclonal plasmacytic infiltration without bone marrow involvement. These tumors occur most commonly in the head and neck region, followed by gastrointestinal tract, central nervous system, thyroid, breast, parotid gland, testis and lymph nodes. Primary renal plasmacytoma is very rare. We report a primary renal plasmacytoma in a 76 year old female.

Key words: X-ray computed, plasmacytoma, renal neoplasm, tomography, positron emission tomography.

INTRODUCTION

Extramedullary plasmacytoma (EMP), accounting for approximately 3% of all plasma cell neoplasms, results from uncontrolled plasma cell proliferation and consists of monoclonal plasmacytic infiltration without bone marrow involvement (Weber, 2005; Dagan et al., 2009; Bachar et al., 2008). EMP occurs most commonly in the head and neck region, followed by gastrointestinal tract, central nervous system, thyroid, breast, parotid gland, testis and lymph nodes (Weber 2005; Dagan et al., 2009; Bachar et al., 2008; Park et al., 2007). Primary renal plasmacytoma is very rare; only less than 20 cases have been reported in the English medical literatures (Park et al., 2007; Igel et al., 1991; Rebelakos et al., 1995; Kanoh et al., 1992; Kandel et al., 1984; Morris et al., 1977; Silver et al., 1977; Catalona et al., 1974; Solomito et al., 1972; Fan et al., 2005; Monill et al., 2005; Sered et al., 2003), to the best of our knowledge. Herein, we report a primary renal plasmacytoma in a 76 year old female with emphasis on the relevance of imaging in the diagnosis and follow-up of plasmacytoma.

CASE REPORT

A 76 year old woman was referred to our hospital due to incidental finding of left renal mass diagnosed by abdominal ultrasonography at other hospital two months ago. Abdominal ultrasonography indicated a solid mass in the left kidney with the size of 3.8 × 3.9 cm. The patient complained of no specific symptom. The patient has back pain and hypertension history for many years and underwent coronary artery bypass graft surgery in our hospital in 2006. The patient was admitted into our hospital for further treatment of her left renal mass. Abdominal ultrasonography showed a hypo-echoic ill-defined mass with the size of 3.6 × 2.8 cm in the left lower renal pelvis (Figure 1). Abdominal contrast-enhanced CT scan was performed to define the nature of the renal mass. Unenhanced CT showed an iso-attenuation mass (CT number of 42.2 HU) with the size of 4.8 × 3.6 cm in the left kidney. The mass enhanced homogeneously at the cortical phase (80.7 HU) and medullary phase (90.9 HU) then washed out and became marked hypo-density mass at the delayed phase (68.5 HU, Figures 2A-D). Reformatted images showed the left renal artery penetrating into the mass but without signs of tumoral vessels (Figure 2E). No signs of venous filling

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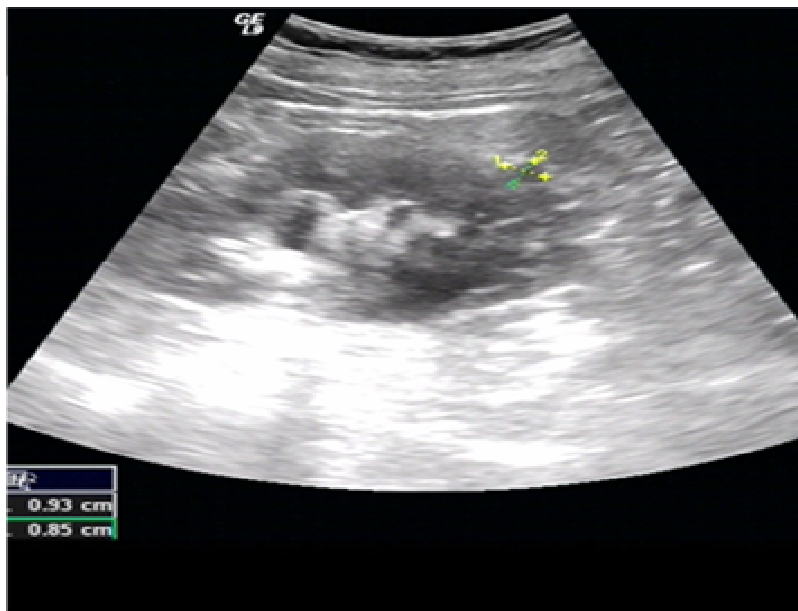


Figure 1. Abdominal ultrasonography showed the left renal hypoechoic mass with the size of 3.6 × 2.8 cm.

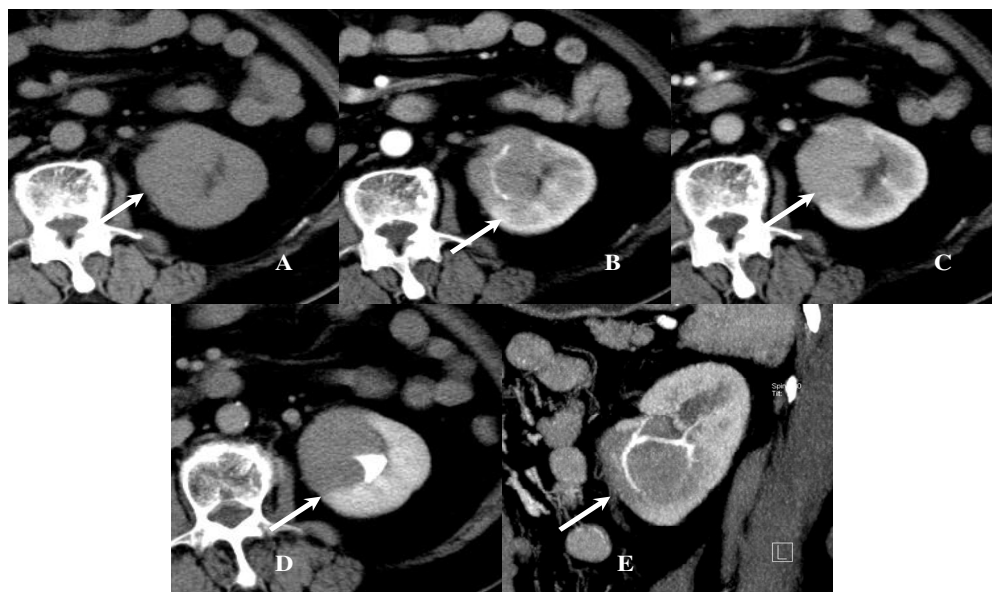


Figure 2. Abdominal CT findings of the renal mass. (A) Non-enhanced CT scan showed an iso-attenuation mass in the left kidney (arrow). (B) Contrast-enhanced cortical phase CT showed the homogeneously mild enhancement of the left renal mass (arrow). (C) Contrast-enhanced medullary and (D) delayed phase CT showed the contrast media washout from the mass (arrow). (E) Sagittal maximum intensity projection reformatted images of cortical phase showed the normal-appearing left renal artery penetrating into the mass (arrow).

defect in the renal veins and inferior vena cava were found. The left inferior renal pelvis was invaded by the mass shown at the delayed formatted images. The renal cell carcinoma was diagnosed based on CT findings.

The patient underwent laparoscopic radical nephrectomy

of left kidney. Gross pathological examination Revealed 5.0 × 4.0 cm well circumscribed grayish-white soft mass in the left kidney. On microscopic pathological examination, the tumor mass was composed of lymphoplasmacytic cells with diffuse plasmacytic

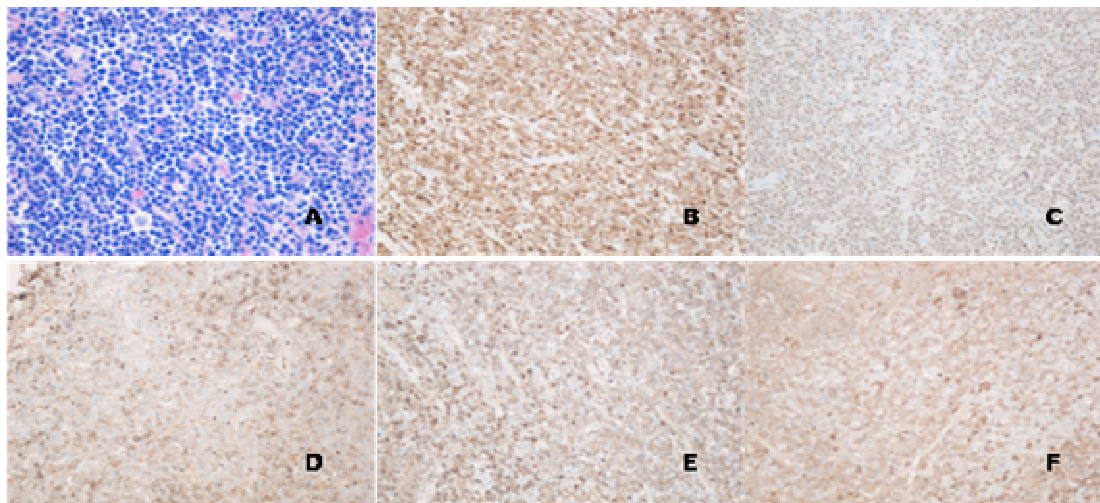


Figure 3. Pathological photographs. (A) Microscopic photograph showed the mass was composed of lymphoplasmacytic cells with diffuse plasmacytic differentiation (HE stain, original magnification $\times 400$). Immunohistochemical staining showed strongly positive for (B) CD79a, (C) Mum1, (D) CD138, (E) λ , and (F) κ (original magnification $\times 100$).



Figure 4. Whole body ^{18}F PET-CT showed multiple active retroperitoneal lymph nodes (arrows) with maximal standardized uptake value of 4.1; no other abnormal findings were observed.

differentiation showing eccentric nuclei, eosinophilic cytoplasm (Figure 3A). Immunohistochemical staining showed strongly positive for CD79a (Figure 3B), CD138 (Figure 3C), Mum1 (Figure 3D), λ (Figure 3E), κ (Figure 3F); focally positive for CD3, CD5, CD20; and negative

for CD10, Bcl-6. Thus, final diagnosis was extramedullary plasmacytoma of the left kidney. Postoperative complete blood test, hepatic and renal function tests showed normal findings. Bone marrow biopsy indicated normal finding. Serum free light chain (λ 40.46 mg/ml, and κ 50.6 mg/ml with ratio of κ and λ of 0.8) and immunofixation electrophoresis were normal. $^{99\text{m}}\text{Tc}$ -MDP bone scintigraphy was performed to define other lesions; only focal tracer uptake in the lumbar 4 and 5 was showed. Whole body ^{18}F fluorodeoxyglucose positron emission tomography (PET)-CT did not show any abnormal findings except for multiple small retroperitoneal lymph nodes with maximal standardized uptake value of 4.1 (Figure 4). Adjuvant chemotherapy was not recommended because physician believed no active lesions were found. Regular follow up was suggested for this patient.

DISCUSSION

EMP is a rare malignancy, composed of plasma cell dyscrasias localized to an extramedullary site in the soft tissue. The cause for plasmacytoma remains unknown, high radiation, virus infection, and autoimmune diseases can be associated with EMP. Over 50% of those with solitary plasmacytoma will develop multiple myeloma. Approximately 85% of lesions occur in the head and neck mucosa, and underlying bone involvement, particularly in the sinuses, may be noted (Weber, 2005; Dagan et al., 2009; Bachar et al., 2008). Gastrointestinal involvement, although, significantly less common, is the next most frequent site and other areas of involvement reported

infrequently include lung, bladder, thyroid, testis, ovary and tonsil. EMP arising in the kidney is a very rare malignancy (Park et al., 2007; Igel et al., 1991; Rebelakos et al., 1995; Kanoh et al., 1992; Kandel et al., 1984; Morris et al., 1977; Silver et al., 1977; Catalona et al., 1974; Solomito et al., 1972; Fan et al., 2005; Monill et al., 2005; Sered et al., 2003). Our patient had no evidence of multiple myeloma demonstrated by bone marrow biopsy, bone scintigraphy and PET-CT. Three fourths of EMP cases involve males. The median age of patients with EMP is 55 years. In 30 - 40% of cases, local lymph nodes are involved at presentation or upon relapse.

Primary renal plasmacytoma is not distinguished from a renal cell or transitional cell carcinoma using conventional imaging studies. Renal plasmacytoma may appear well defined or infiltrative, intrarenal (Park et al., 2007) or perirenal mass (Sered et al., 2003) with iso- or heterogeneous density and homogeneous or heterogeneous enhancement at CT. Our case showed well-defined mass with homogeneously mild enhancement, the left renal artery penetrating into the mass had normal-appearing morphology, which was different from CT findings of renal cell carcinoma. Though MR has an important role in the evaluation of spine involvement of plasma cell tumor, MR findings of renal plasmacytoma have been rarely reported due to its rarity. New imaging modalities for diagnosis and for monitoring of disease status (PET, in particular, combination of PET and whole-body MRI) can help further refine the diagnosis and identify high-risk groups for disease progression, occult disease and active disease can be more easily recognizable (Dimopoulos et al., 2009; Nanni et al., 2008). Renal plasmacytoma needs differentiate from other renal tumors. Renal cell carcinoma, the most common renal malignancy affecting adults, accounts for approximately 85% of renal tumors. Because of this high prevalence, our case was misdiagnosed as renal cell carcinoma preoperatively. But tumoral artery of renal cell carcinomas can be irregular and a large amount of tumoral vessels can be present. Lymphomas are typically hypovascular and homogeneous masses and demonstrate minimal homogeneous enhancement following administration of contrast media, which can help differentiate lymphoma from typically enhancing renal cell carcinoma. There are no clear guidelines for treatment of renal EMP due to its rarity. The treatment options are radical nephrectomy, radiation therapy, and chemo-therapy. It has been reported that EMP at other sites are highly radiosensitive with nearly all patients successfully achieving local control (80 - 100%) and approximately the 50 - 65% of patients remaining free of disease longer than 10 years. The recommendations of working group of the UK Myeloma Forum include a radiotherapy dose of 40 Gy in 20 fractions for tumors < 5 cm and up to 50 Gy in 25 fractions for tumors ≥ 5 cm with at least a 2 cm margin encompassing the primary tumor (Soutar et al., 2004). The risk of recurrence

was same for the three treatment approaches and no statistically significant differences in survival were seen.

In conclusion, renal plasmacytoma is a very rare renal malignancy. It should be listed as differential diagnosis when a solitary and solid renal mass with homogeneous mild enhancement was found. Normal appearing renal artery within the mass indicates the diagnosis of renal plasmacytoma. PET-CT can further evaluate the stage of renal plasmacytoma and make an appropriate management for the patient.

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