Primary Renal Osteosarcoma Presenting as Hydronephrotic Kidney: A Case Report with Imaging Findings and a Literature Review

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Primary renal osteosarcoma is an extraordinarily rare primary tumor of the kidney. It occurs mostly in middle-aged and older individuals, and generally presents with non-specific symptoms such as abdominal pain, weight loss, palpable mass, and gross hematuria. Radiographically bizarre renal calcifications are often indicative of this uncommon neoplasm. Herein we report a case of primary osteosarcoma in a hydronephrotic kidney of a 76-year-old man, who developed lung metastasis soon after a radical nephrectomy. To the best of our knowledge, this is the first computed tomography based report of a patient with the hydronephrotic form of primary renal osteosarcoma.

Index terms
Primary Renal Osteosarcoma
Kidney
Multidetector Computed Tomography

INTRODUCTION

Primary renal osteosarcoma is extremely rare and only 27 cases have been reported (1). Most cases show bizarre calcifications within the renal contour (2, 3). To our knowledge, there have been no cases of primary osteosarcoma presenting as hydronephrotic kidney.

Hence, we report the first such case with CT findings and a literature review.

CASE REPORT

A 76-year-old man with a six-month history of left lower abdominal pain and weight loss was presented to the outpatient ward of our hospital. His medical history was unremarkable. Aside from a higher than normal serum alkaline phosphatase level of 262 IU/L (normal range: 40–129 IU/L), blood test results were within the normal range. The serum levels of tumor markers carcinoembryonic antigen, CA 19-9, and CA 72-4 were under the respective limits. A urinalysis showed microscopic hematuria, though the urine was sterile on culture and urine cytology yielded no malignant cells. Bone scan and skeletal survey results were normal, as was a chest radiograph.

On physical examination, a huge palpable mass was detected by the urology surgeon in the left lower quadrant of the abdomen.

Initial simple abdominal radiography indicated a soft tissue density mass with some calcifications, as well as lateral displacement of the adjacent bowel loops (Fig. 1A).

Three-dimensional kidney multidetector CT imaging (Light-Speed VCT; GE Healthcare, Milwaukee, WI, USA) was performed. On precontrast scans, there were dense amorphous or nodular calcifications and irregular solid components in the large mass (Fig. 1B, C). On postcontrast axial scans, irregular
enhancing solid areas were noted along the peripheral and central portion of the mass with focal fat obliteration between the mass and left descending colon (Fig. 1D). A reformatted coronal scan clearly revealed the marked hydronephrosis, calcifications and irregular enhancing solid areas in the lower portion of the left kidney (Fig. 1E). No evidence of hepatic metastasis or regional lymphadenopathy was found.

Based on the radiological findings, our initial diagnosis was transitional cell carcinoma, arising from longstanding hydronephrosis likely due to ureteropelvic junction (UPJ) stricture. Sarcomatoid renal cell carcinoma was included in the differential diagnosis.

We performed a left radical nephrectomy with left hemicolectomy due to adhesion. The resected specimen measured 20 × 16 × 4 cm. A pathologic specimen revealed marked dilation of the left kidney and irregular solid masses with multiple calcifications in the dilated lower portion. The left ureter could not be identified grossly or pathologically probably due to severe destruction and adhesion. The left colon that was adherent to the kidney showed severe adhesion but no evidence of invasion. Histopathologic and immunohistochemical analysis of specimens revealed the characteristic pattern of classic osteosarcoma with immature neoplastic osteoids and sarcomatoid stroma (Fig. 1F). The final diagnosis was primary renal osteosarcoma in the lower portion.

**Fig. 1.** A 76-year-old man with primary renal osteosarcoma.

A. Simple radiography shows soft tissue density mass on the left side of abdomen with calcifications (arrows).

B, C. Precontrast CT scans show amorphous or nodular calcifications and irregular soft tissue components (arrows) within the mass.

D. Postcontrast axial CT scan shows irregular enhancing solid areas along the peripheral and central portion of the mass with focal fat obliteration between the mass and left descending colon (arrows).

E. Postcontrast coronal reformatted CT scan shows irregular enhancing solid areas with calcifications in the lower portion of marked hydronephrotic left kidney (arrows).

F. Photomicrograph of specimen shows immature neoplastic osteoids and sarcomatous stroma (H&E, × 100).
of the kidney with marked hydronephrosis. Following surgery, the patient underwent no additional chemotherapy or radiation therapy. Just 7 weeks after surgery, multiple metastasis in the both lungs developed. The patient deteriorated over the course of several months and died 5 months after diagnosis of the primary renal osteosarcoma.

**DISCUSSION**

Sarcomas originating from the kidney, the majority of which are leiomyosarcoma, account for only 1% of all primary renal malignancies. Metastasis from the primary skeletal osteosarcoma to the kidney is found much more frequently than osteosarcoma originating from the kidney (4). According to the data, tumors occur more frequently in the sixth to seventh decades of life and the prevalence does not differ between males and females (5).

The patient might complain of various, but non-specific symptoms, including abdominal pain, flank pain, weakness, abdominal distension, weight loss, and, rarely, gross hematuria (4). Blood tests are often normal except for the serum level of alkaline phosphatase, which may be normal or slightly altered.

Radiologically, demonstration of renal mass containing dense or rim-like calcification suggests the diagnosis of renal osteosarcoma. The sunburst appearance of calcifications within the renal contour, which can be detected on CT, seem to be a characteristic finding (6), but several other configurations also have been noted. Signs of space occupying lesions, such as pyonephrosis with permeative mass or organ displacement, might be seen on ultrasonography (7).

The differential diagnosis includes adult Wilms tumor, metastatic sarcoma, and sarcomatoid renal cell carcinoma.

On angiography, primary renal osteosarcoma has lesser vascularity than normal renal parenchyma. Renal cell carcinoma, by contrast, usually shows greater vascularity (7). Frequently, primary renal osteosarcoma shows a regional growth pattern with early infiltration of the renal capsule and adjacent structure. It can metastasize to various organs such as the spleen, diaphragm, liver, small and large bowel, bone marrow, dura, skin, and most commonly, the lung (4).

The etiology is uncertain, but it is thought to involve metaplastic differentiation from connective tissue in the renal capsule to a pleuriopotent mesenchyme, that can differentiate into osteoblasts (5, 8). The typical microscopic findings are pleomorphic spindle cell and multinucleate giant tumor cell with diffuse calcified osteoids (5).

Various treatment approaches including radical nephrectomy, partial or total resection of contiguous organs, radiation therapy, and polychemotherapy have been pursued, but due to osteosarcoma's aggressiveness and usually advanced stage at diagnosis, the prognosis characteristically is poor (4, 5).

In our case, we initially thought that it had arisen from the hydronephrotic kidney due to the mass's aggressive imaging feature with hydronephrosis. Considering the prevalence of the renal mass, it could have been mistaken for a transitional cell carcinoma with UPJ stricture or a renal cell carcinoma with sarcomatoid change.

Unlike other previously reported primary renal osteosarcomas, the enhancing solid portions that were proven to be osteosarcomatous foci were located centrally rather than in the peripheral portion adjacent to the renal capsule. Additionally, the upper portion of the markedly hydronephrotic kidney showed no tumor infiltration. The left ureter could not be identified in the surgical field grossly or as a pathologic specimen.

In summary, we have presented a pathologically confirmed case of primary renal osteosarcoma presenting as hydronephrotic kidney depicted on CT imaging. Primary renal osteosarcoma could be differentially diagnosed in the case of a tumor arising from a hydronephrotic kidney associated with calcifications, although a primary renal osteosarcoma is a rare disease.

**REFERENCES**

수신증으로 발현된 원발성 신장 골육종: 증례 보고

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원발성 신장 골육종은 신장에 발생하는 아주 드문 종양이다. 일반적으로 중년 이후에 발생하는 것으로 알려져 있으며, 비특이적인 증상인 복부통증, 체중감소, 복부 종괴 및 육안적 혈뇨를 보인다. 영상의학적으로는 기이한 신장 석회화가 이 질환을 시사할 수 있는 소견으로 제시되고 있다. 우리는 76세 남성에서 수신증의 형태로 발현되었던 원발성 신장 골육종을 경험하였고, 이 환자는 근치적 신절제술을 시행 받았으나 그 후 폐 전이가 발생하였다. 원발성 신장 골육종이 전산화단층촬영에서 수신증의 형태로 나타난 경우는 아직까지 보고된 적이 없으며, 이에 증례 보고를 하고자 한다.

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