Case Report

Primary Renal Osteogenic Sarcoma: A Rare Entity

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Primary renal osteogenic sarcoma is a rare malignancy. Renal metastases from skeletal osteogenic sarcoma occur more frequently. Primary renal osteogenic sarcoma has an equal sex distribution and is a tumor of middle-aged and elderly individuals. We report a 60-year-old hypertensive woman who presented with left flank pain and who was found to have primary renal osteogenic sarcoma. We describe the clinical course of this unusual malignancy, with a short review of the literature. [Hong Kong J Nephrol 2006;8(2):75–7]

Key words: extraosseous, kidney, osteogenic, sarcoma

She underwent left radical nephrectomy with IVC thrombectomy. Microscopic examination of the tumor was suggestive of primary renal OS with presence of tumor thrombus in the IVC (Figure). Hilar and para-aortic lymph nodes were positive for metastasis. CT-guided fine needle aspiration cytology from the left lung nodule was positive for malignant cells suggestive of metastasis.

Figure. Microscopy of the kidney shows tumor composed of pleomorphic cells, with scattered giant cells, which are laying down osteoid (hematoxylin & eosin, original magnification ×4).

INTRODUCTION

Sarcomas account for only a small proportion of primary malignant renal neoplasms and include liposarcoma, leiomyosarcoma, fibrosarcoma and adult Wilms’ tumor [1]. Primary renal osteogenic sarcoma (OS) is an extremely rare entity [2–4] and was first described by Haining and Poole in 1936 [5]. Skeletal OS metastatic to the kidney occurs more frequently, with an incidence of nearly 12% [3]. The prognosis of primary renal OS is very poor as it is generally diagnosed at an advanced stage [3,4].

CASE REPORT

A 60-year-old hypertensive woman presented with the complaints of dyspnea on exertion and left flank pain of 15 days’ duration. Computed tomography (CT) of the chest revealed nodular lesions in the left lung; metastasis was suspected. CT of the whole abdomen showed a heterogeneous mass, measuring 9 × 8 cm, replacing the left kidney, with tumor thrombus seen in the left renal vein extending into the intrahepatic inferior vena cava (IVC) along with enlargement of left para-aortic lymph nodes and nodular lesions in both lungs. Bone scan study was normal.

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The patient was finally diagnosed with primary renal OS metastatic to the lung. She was put on a palliative chemotherapy protocol based on a combination of ifosfamide, doxorubicin and cisplatin. After three cycles of chemotherapy, chest CT demonstrated mild regression of the lung lesions. However, after the completion of five cycles of chemotherapy, she developed urination difficulty. CT of the abdomen and pelvis revealed irregular, non-mobile, heterogeneous lesions in the bladder along the left posterolateral wall, suggestive of metastases. She was started on palliative supportive care in view of disease progression and was discharged, with the palliative supportive care to be continued at home. However, after discharge, she was lost to follow-up.

**DISCUSSION**

Sarcomas account for approximately 1% of primary renal malignancies in adults, with leiomyosarcoma being the most common histologic subtype [6]. Extrasosseous OS is a known entity and was first recognized and reviewed in 1956 by Fine and Stout [7]. It is most commonly found in the breast, thyroid, uterus, bladder and soft tissues of the extremities. However, primary renal OS is an extremely rare entity, with only a few cases reported in the English language literature [1,2,8–10]. Primary skeletal OS, on the other hand, leads to renal metastasis in approximately 10–12% of patients [8,10,11]. The male to female ratio in primary renal OS is roughly equal and it usually occurs in middle-aged to elderly individuals [8,10]. Primary renal OS does not exhibit any propensity for laterality [9].

Calcified appearance on X-ray, especially with a sunburst pattern, strongly suggests the diagnosis of renal OS [10,12]. Other causes of calcified renal masses include vascular diseases, calculi, traumatic lesions, cystic lesions, and benign tumors [10,12]. Also, renal metaplasia associated with chronic inflammatory conditions and dysplasia can present as calcification. However, all these exemplify calcification rather than a true bone forming process within the tumor, as seen in primary renal OS [4]. Elevated serum alkaline phosphatase is a useful preoperative marker for calcified renal masses without demonstrable bone disease [10,12]. Ossification may also occur in various pathologic renal states, including hydronephrosis and chronic pyelonephritis [1,10]. Osteoid or bone production by tumor cells, lack of attachment to the bone, and absence of epithelial differentiation are the histologic criteria that need to be met for the diagnosis of extrasosseous OS [4].

Other differential diagnoses of primary renal OS include adult Wilms’ tumor, metastatic sarcoma and sarcomatoid variant of renal cell carcinoma (RCC), which accounts for 2% of RCC [2]. Pathologically, differentiation of sarcomatoid variant of RCC from various primary renal sarcomas, including OS, fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma and chondrosarcoma, is very important, and the presence of foci of epithelial differentiation is essential for the diagnosis of sarcomatoid variant of RCC [2,12].

The histogenesis of extrasosseous OS remains incompletely understood. Virchow’s theory proposed metaplastic reversion of connective tissue to primitive embryonic mesenchymal tissue with possible transformation to osteoblast [8]. This theory was first proposed in 1884 [1,10]. It is also hypothesized that, for primary renal OS, the origin of primary mesenchymal tumor in the renal cortex occurs by transfer of osteoblasts by the bloodstream. Also, the presence of mesenchymal cells from the developing renal blastema, occurrence of misplaced embryonic remnants of osteogenic tissue, and misplaced urothelium may induce bone formation [6].

Primary renal OS was first described at autopsy in 1936 [5]. Patients with primary renal OS tend to present late and have a poor prognosis. The clinical course is aggressive, with short survival, ranging from 6 weeks to 1 year. Like other primary renal sarcomas, about 85% of patients eventually die of disease [2,4,11]. These are usually fast growing and exhibit a tendency to local infiltration of contiguous structures and early metastasis [2]. Our patient had invasion of the IVC at the time of diagnosis. The tumors are characteristically of very hard consistency [12]. Clinical presentation of primary renal OS is like that of other renal tumors, with the presence of hematuria, flank pain and abdominal mass [2,10]. Metastatic spread of primary renal OS is usually to the lungs and liver, although spread to unusual sites such as the vulva and skin has also been reported [9].

Due to the rarity of this disease entity, there is scant documentation of the use of adjuvant systemic chemotherapy, but doxorubicin-based adjuvant chemotherapy has shown survival advantage [12].

To conclude, primary renal OS should be kept in mind in cases of renal masses, especially when there is calcification within the renal parenchyma, which do not offer the diagnosis of a benign renal process. Also, renal masses should be investigated thoroughly and treated aggressively with multiagent chemotherapy.

**REFERENCES**