Case Report
Extraskeletal mesenchymal chondrosarcoma arising from kidney: a case report and literature review

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Abstract: Mesenchymal chondrosarcoma (MC) is a malignant tumor that usually arises from cartilaginous structures. Primary renal MC is extremely rare. Here, we reported a 33-year-old woman with solitary left renal MC presented with asymptomatic gross hematuria. Abdominal Ultrasonography and CT revealed a 7 cm mass within the left kidney with obvious calcification. The tumor was removed by radical nephrectomy. Pathological specimen revealed the typical histology of mesenchymal chondrosarcoma. The patient received no chemotherapy or radiotherapy, there was no recurrence or metastasis occurred during a 28-months follow-up and the patient’s extended survival is attributed to the early diagnosis and treatment.

Keywords: Mesenchymal chondrosarcoma, asymptomatic gross hematuria, abdominal ultrasonography, CT, radical nephrectomy

Introduction

Extraskeletal mesenchymal chondrosarcoma (MC) is a rare malignant tumor with poor prognosis. MC frequently has a predilection for extraskeletal location and characterized microscopically by a dimorphic pattern. It rarely occurs in the kidney and only a few cases documented in the English literature [1-6]. No clear guideline for treatment are available due to its rarity. In this comprehensive review of the current literature, we presented radiographic and histopathologic features of this case; moreover, treatment and outcome were also discussed.

Case report

A 33-year-old woman, with no significant medical history, presented with asymptomatic gross hematuria for one day and a mass found in the left kidney by ultrasonography. Her family history and medical history were unremarkable, and there were no unusual findings on routine physical examination. Results of routine laboratory tests, including blood chemistry were within normal limits except for macroscopic hematuria. A conventional radiographic bone and chest survey also yielded no abnormal findings. An inhomogeneous and well-circumscribed mass with obvious calcification measuring about 7×7 cm was detected in the left kidney on CT scan (Figure 1A). Most of upper renal parenchyma structures were normal and no enlarged lymph nodes were detected. Most of the mass was no enhancement while peripheral mass enhanced slightly (Figure 1B). There was no evidence of metastasis of the neoplasm in any organ. Therefore, on the basis of these findings, preliminary clinical diagnosis of a renal cell carcinoma was determined; the patient underwent surgical exploration through a flank incision. On operation, the tumor was solid and rough mainly in the lower pole of the left kidney and invaded into perirenal fat tissues. The patient successfully underwent radical nephrectomy with an uneventful recovery.

Grossly, the removed kidney showed an inhomogeneous, calcifying tumor measuring about 7×7 cm in the lower pole of the kidney. Formalin-fixed, paraffin-embedded tissue was obtained for routine microscopic examination. Specimen was stained with hematoxylin and eosin. Microscopically, the tumor showed various-
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-sized lobules of neoplastic cartilage, which consisted of atypical chondrocyte-like cells and light basophilic matrix, divided by fibrous connective tissue. Nuclear pleomorphism was remarkable and occasional multinucleated cells were observed and mitotic figures were infrequent (Figure 2A), a bimorphic pattern comprising a sheath of undifferentiated oval- or spindle-shaped cells arranged in small, well-defined islets of cartilaginous tissue but no epithelial components. Pathologically, typical histologic features were identified consisting of small, round or spindled cells, interspersed with hyaline cartilage islands and undifferentiated mesenchymal cells. The differential diagnosis included the chondrosarcoma transformation of renal cell carcinoma (RCC), metastatic renal involvement from chondrosarcoma elsewhere in the body or the more frequent Wilms’ tumor with blastemal component and chondroid differentiation. Sox9 has

Discussion

MC is a specific variant comprising about 10% of all chondrosarcomas [7]. Microscopically, it is characterized by a dimorphic pattern. It occasionally occurs in the gastrointestinal tract, urogenital tract, thyroid, and head and pleura [8]. The etiology of extraskeletal MC is unknown, although it is not unexpected that extraskeletal mesenchymal chondrosarcoma would arise from the kidney on the basis of renal tissue normally does not contain cartilaginous components. It was hypothesized that mesenchymal cells from the developing renal blastoma that preserve their multi-potential capabilities give rise to mesenchymal tumors such as extraskeletal chondrosarcomas of the kidney or mesenchymal differentiation of a pluripotent stem cell [9]. Primary renal chondrosarcoma was first described in 1981 [10]. Although MC may present at any age, the median age at presentation is 50 years (21-70 years) and a slight male preponderance has been noted. After review of the current English literature, the clinical presentation is nonspecific and include abdominal flank pain or gross hematuria. Those tumors ranged in size from 3-23 cm. MC of the kidney usually remained asymptomatic till they are large enough to produce symptoms. Our case presented with gross hematuria which was similar to conventional renal cell carcinoma (RCC). The clinical characteristics of most MCs are indistinguishable from that of RCC.

Figure 1. CT shows an inhomogeneous and well-circumscribed mass with obvious calcification measuring about 7×7 cm in the left kidney.

Figure 2. A. Nuclear pleomorphism was remarkable and occasional multinucleated cells were observed and mitotic figures were infrequent. B. A bimorphic pattern comprising a sheath of undifferentiated oval- or spindle-shaped cells arranged in small, well-defined islets of cartilaginous tissue but no epithelial components.
been proposed as a unique marker for mesenchymal chondrosarcoma which may improve diagnostic specificity [11].

Radiologically, imaging findings of MC may be characteristic because osteogenic tissue or osteoblasts of embryonic mesenchyme sunburst appearance on X-ray and present as large soft tissue tumors with central ‘chondroid’ calcifications [12]. However, large nonspecific renal mass with both rim and amorphous calcification has also been described.

The natural history of the disease remains poorly understood, although with radical nephrectomy some cases had a recurrence or distant metastasis eventually. Some may be variable length of disease-free survival, a 6-year follow up with good prognosis case was attributed to the small size of the tumor (3 cm) and early treatment meanwhile recur ation or distant metastasis even after 18 years of nephrectomy has been described [13]. The most important prognostic features in chondrosarcoma are histology (low-grade, well-differentiated tumors have the best prognosis) and complete resection which has been shown to reduce the incidence of local recurrence. Because of its rarity, the optimum local and systemic treatment for this tumor remains controversial, initial radical excision of the tumor is certain indispensable. Data regarding the effect of chemo- or radiotherapy is still insufficient, most are resistant to chemotherapy and there is no indication for adjuvant chemotherapy in early stage. Palliative surgery may be more effective than chemotherapy or radiotherapy for metastasis tumor [13]. As a palliative approach, local metastatic lesion treated with intensity-modulated radiotherapy which may result in disease stable for a long time.

Chondrosarcomas are graded on a 1 to 3 scale corresponding to a histological classification from a well-differentiated to undifferentiated tumor, based on the rate of mitoses, cellularity, and nuclear size presented different behaviors. The patient with a low-grade tumor demonstrates the slow-growing nature, the grade of malignancy and the histological surgical margins are some of the most important prognostic factors just as our case. This emphasize the importance of histological tumor grade and appropriate treatment for the prognosis [3]. MC has a relatively potential of local relapse or distant metastases after surgery, long-term follow-up is recommended.

Disclosure of conflict of interest

None.

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