Case Report

Mesenchymal chondrosarcoma of kidney

Ruchita Tyagi, Nandita Kakkar, Rakesh Kumar Vasishta, Mayank Mohan Aggarwal[1]

Departments of Histopathology, and [1]Urology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

ABSTRACT

Mesenchymal chondrosarcoma of the kidney is a very rare entity with no definite treatment protocol. Herein, we describe one such case with discussion of its diagnosis and management. The patient had a well circumscribed mass in right kidney extending into the inferior vena cava and metastasis to both the lungs. Right nephrectomy was performed and the histopathological examination confirmed the diagnosis to be renal mesenchymal chondrosarcoma. After surgical removal of the tumor, the patient was given chemotherapy with Cisplatin and Epirubicin, following which there was significant regression of lung nodules.

Key words: Chondrosarcoma, extraskeletal, kidney, mesenchymal

INTRODUCTION

Mesenchymal chondrosarcoma comprises less than 2% of all chondrosarcomas and is extremely rare in the extraskeletal sites.[1,2] There have been only a handful of cases of primitive mesenchymal chondrosarcoma arising in the kidney. Due to its rarity, there is no definite treatment protocol. Herein we present a case along with discussion of its diagnosis and management.

CASE REPORT

A 22-years-old female complained of hematuria, high grade fever, heaviness, and pain in right flank for 2 months. Contrast enhanced computed tomography (CECT) revealed a complex hypodense mass with variegated attenuation measuring 6 × 6 × 5.6 cm in mid-polar region of right kidney, involving the inferior vena cava. No calcification or fat was noted within the mass. No perirenal infiltration or lymphadenopathy was observed. Magnetic resonance imaging (MRI) revealed a large lobular mass measuring 6.5 × 6.2 × 5.6 cm in the right kidney with mixed hyperintense and hypointense signals on T2 weighted imaging. Positron emission tomography showed some subcentimetric non-FDG avid lung nodules which were suspected to be metastatic deposits. A clinical diagnosis of right sided renal cell carcinoma (T3bN0Mx) with invasion into inferior vena cava and metastasis to lungs was made. Right radical nephrectomy with IVC thrombectomy and lymph node dissection was performed. Per-operatively, the tumor involved the medullary region of right kidney. The IVC thrombus was infiltrating its wall and extended upto the level of caudate lobe of liver.

Pathology - Grossly, the right nephrectomy specimen measured 9 × 6 × 4 cm and showed a friable tumor measuring 4 × 4 × 3 cm in the medullary region, invading the renal parenchyma and compressing the pelvicalyceal system, but not involving the latter [Figure 1]. The tumor was fleshy in appearance and there were no glistening areas visible within the tumor grossly. The renal vein showed a tumor thrombus. IVC thrombus, sent separately, measured 5 × 2 × 2 cm. Attached ureter measured 2 cm and was grossly free of tumor. Histopathology showed a well circumscribed tumor comprising of an unusually large component of well differentiated cartilage alternating with stromal component which comprised of small sized, oval to spindle shaped cells with moderate nuclear pleomorphism, hyperchromatic nuclei, inconspicuous nucleoli, and moderate cytoplasm [Figure 2a and b]. The spindle shaped cells displayed a hemangiopericytoma like pattern of arrangement [Figure 2c]. Numerous atypical mitotic figures were noted along with focal areas of myxoid change and necrosis. The tumor was not involving the

For correspondence: Dr. Nandita Kakkar,
Department of Histopathology, Post Graduate Institute of Medical Education and Research,
Sector - 12, Chandigarh - 160 012, India.
E-mail: nandita_kakkar@yahoo.com

Access this article online

Quick Response Code:

Website: www.indianjurol.com

DOI: 10.4103/0970-1591.126914

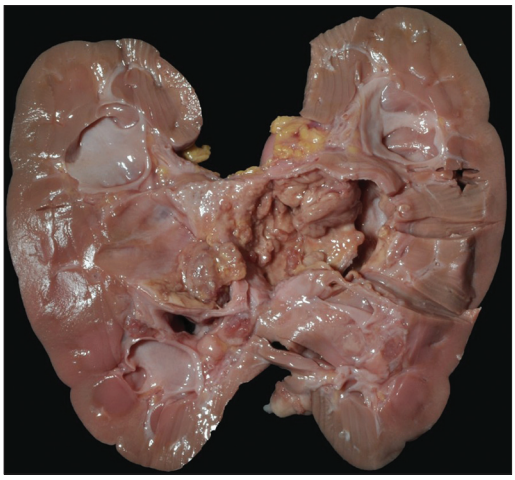

Figure 1: Gross photograph showing a friable tumor in the kidney, occupying the pelvicalyceal region

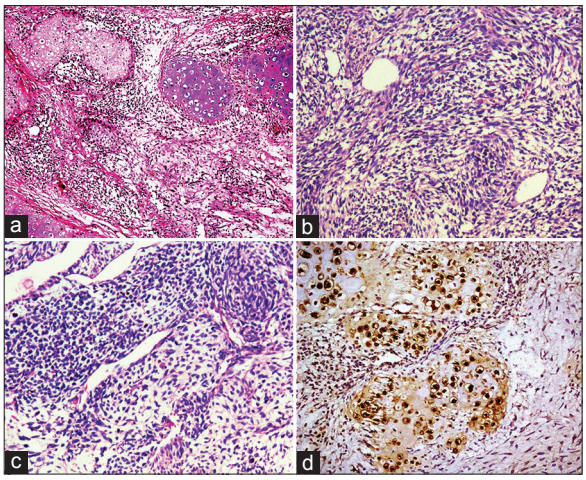

Figure 2: Photomicrograph of tumor showing (a) cartilaginous component admixed with stromal component (H and E, x100); (b) stromal component composed of oval to spindle shaped cells displaying moderate pleomorphism, with hyperchromatic nuclei and inconspicuous nucleoli (H and E, x400); (c) Stromal cells arranged in hemangiopericytoma-like pattern in the tumour. (H and E, x400); (d) IHC showing S-100 positivity in cartilaginous component

pelvicalyceal lining, which showed normal morphology and displayed no features of dysplasia/carcinoma *in situ*. On immunohistochemistry (IHC), the spindle cells were positive for vimentin and the cartilaginous component was strongly positive for S-100 [Figure 2d]. The tumor was involving the inferior vena cava. However, the ureter and the lymph nodes (29 in number) were free of tumor. Based on these findings, a diagnosis of extra skeletal mesenchymal chondrosarcoma of kidney was made.

Treatment - The patient was planned for close follow up at monthly intervals. Within 3 weeks of surgery, she presented with dyspnoea and chest pain of acute onset. CT pulmonary angiography was suggestive of pulmonary thromboembolism with partial IVC thrombosis. She was started on systemic chemotherapy with intravenous Cisplatin (90 mg/m^2) and Epirubicin (75 mg/m^2) on Day 1 at 3 weekly intervals. After six cycles of chemotherapy, the patient showed marked clinical improvement with significant regression in lung nodules [Figure 1c].

DISCUSSION

Mesenchymal chondrosarcoma was first described by Lichtenstein and Bernstein in 1959. This tumor is believed to arise from the undifferentiated cells derived from the primitive mesenchyme involved in chondroblastic differentiation.[3] Dowling reported the first case of extra skeletal mesenchymal chondrosarcoma.[4] It is commonly seen in second and third decade of life.

Microscopically, mesenchymal chondrosarcoma exhibits a characteristic bimorphic pattern composed of sheets of undifferentiated round, oval, or spindle shaped cells and abrupt presence of small, well defined nodules of well differentiated cartilaginous tissue which stains with S-100 on IHC. The undifferentiated cells have ovoid or elongated hyperchromatic nuclei and scanty, poorly outlined cytoplasm, are arranged in small aggregates or in a hemangiopericytoma-like pattern about sinusoidal vascular channels lined by a single layer of endothelium. The differential diagnosis includes hemangiopericytoma and synovial sarcoma. However, the presence of cartilage distinguishes mesenchymal chondrosarcoma from these entities. This case displayed unusually large areas of cartilaginous component. Mesenchymal chondrosarcoma arising in the kidney can also be misdiagnosed as Wilm's tumor as the oval shaped hyperchromatic stromal cells are mistaken as blastemal component and the cartilaginous component as the mesenchymal component.

Surgery is the mainstay of treatment. Radiotherapy and chemotherapy have been used but there is no definite treatment strategy. Mesenchymal chondrosarcoma is considered to be relatively sensitive to doxorubicin-based combination chemotherapeutic regimens.[5] A study showed the 5 year and 10 year survival rates to be 54.6 and 27.3%, respectively. Careful long-term surveillance of patients is essential as local recurrences and metastasis may occur even 10-20 years after surgical removal of the tumor.[6] The indexed patient responded to Cisplatin and Epirubicin. In view of complete surgical removal with adequate clearance of margins, radiotherapy was not given. The patient is on regular follow up and is doing well.

CONCLUSION

Herein we report a rare case of extra skeletal mesenchymal chondrosarcoma of the kidney with metastasis to lungs. Histologically, it is unusual to find such extensive chondroid component in this tumor. After surgical removal of the tumor, the patient responded well to chemotherapy, following which the metastatic lung nodules regressed significantly.

REFERENCES

1. Bertoni F, Picci P, Bacchini P, Capanna R, Innao V, Bacci G, *et al*. Mesenchymal chondrosarcoma of bone and soft tissues. Cancer 1983;52:533-41.
2. Kaneko T, Suzuky Y, Takata R, Takata K, Sakuma T, Fujioka T. Extraskeletal mesenchymal chondrosarcoma of the kidney. Int J Urol 2006;13:285-6.
3. Lichtenstein L, Bernstein D. Unusual benign and malignant chondroid tumours of bone: A survey of some mesenchymal cartilage tumors and malignant chondroblatic tumors, including a few multicentric ones as well as many atypical benign chondroblastomas and chondromyxoid fibromas. Cancer 1959;12:1142.
4. Dowling EA. Mesenchymal chondrosarcoma. J Bone Joint Surg 1964;46:747-54.
5. Nooij MA, Whelan J, Bramwell VH. Doxorubicin and cisplatin chemotherapy in high-grade spindle cell sarcomas of the bone, other than osteosarcoma or malignant fibrous histiocytoma: A European Osteosarcoma Intergroup Study. Eur J Cancer 2005;41:225-30.
6. Nakashima Y, Unni KK, Shives TC, Swee RG, Dahlin DC. Mesenchymal chondrosarcoma of bone and soft tissue: A review of 111 cases. Cancer 1986;57:2444-53.

How to cite this article: Tyagi R, Kakkar N, Vasishta RK, Aggarwal MM. Mesenchymal chondrosarcoma of kidney. Indian J Urol 2014;30:225-7.

Source of Support: Nil, **Conflict of Interest:** None declared.