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Primary Renal Extra-Skeletal Mesenchymal Chondrosarcoma:A Case Report and Review of Theliterature

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1. Abstract

Primary Renal Extra-skeletal Mesenchymal Chondrosarcoma (PREMC) is a very rare, aggressiveand high-grade malignant tumor. Twelve cases have been reported in the English literature to date. We report the thirteenth case of PREMC in the right kidney of a 62-year-old male patient.

2. Case Presentation

Mesenchymal Chondrosarcoma (MC) is a rare and aggressive high-grade malignant tumor of bone and soft tissue which was first described by Lichtenstein and Bernstein in 1959 in a reviewing survey of a 25 unusual benign and malignant chondroid tumors of bone[1]. The incidence is approximately 1% of all chondrosarcoma neoplasms[2]. Extra-skeletal Mesenchymal Chondrosarcoma (EMC) is found in a third of the patient sin a one conducted case series [3]. Primary Renal Extra-skeletal Mesenchymal Chondrosarcoma (PREMC) is extremely rare and to date only twelve cases have been previously reported in the literature (Table 1).

Here in, we report the thirteenth case of PREMC in the right kidney of an 62-year-old gentleman who is known to have DM type II, Hypertension and was previously asymptomatic. He presented with an increase creatinine level on a regular checkup. The patient had no history of hematuria, traumaor fever and the physical examination was unremarkable. The patient was admitted to investigate his renal function. Ultrasound image and MRI of the right kidney (Figure 1) demonstrated a heterogeneous and hypoechoic lobulated mass (Fig 1a, white arrow) arising from the upper and mid pole of the right kidney. There was a central echogenic area within the mass with posterior shadowing corresponding to calcifications (Figure 1a, dashed arrow). Corresponding MRI T2 fat suppressed axial image (Figure 1b) demonstrated in homogeneous mixed signal of the lesion (white arrow) with dark central areas (dashed arrow) corresponding to bright signal on the dual echo T1 weighted images (Figure 1e&1f) and T1 spoiled gradient precontrast image (Figure 1g). The peripheral diffusion restriction of the mass appearing bright on high b value DWI image (Figure 1c) and dark on ADC map (Fig1d, white arrows). It is noticeable a heterogeneous enhancement ofthe mass on axial (Figure 1h, white arrow) and coronal (Figure 1i, white arrow) postcontrast T1weighted images. A primary renal malignancy was made and the patient underwent a right radical nephrectomy.

3. Macroscopic Examination:

We received a right kidney containinga tan/white and focally calcified mass measuring 8 x 6.5 x 5 cm that is infiltrating the renal capsule and extending to renal sinus fat(Figure 2). The tumour involves the vascular margin but he ureteric margin was free.

Section from the tumor (Figure 3) showed a biphasic (chondroid and blue cell component) with infiltrative growth pattern. The blue cell component consisted of undifferentiated spindle to oval shaped cells with hyperchromatic nuclei and scant clear cytoplasm arranged mainly in hemangiopericytoma-like pattern. The chondroid component comprised islands of atypical chondroid cells.

Table 1: Details of the previously published 12 PREMC.

	Author	Sex	Age (y)	Clinical presentation	Radiology features	Location	Tumor size in cm	Metastasis Or Recurrences	Follow-up
1	Pitfield et al, 1981 [4]	M	61	Severe loin pain	Rim of scattered amorphous calcifications	Lower pole of left kidney	23 x 18 x 14	Recurrent tumor in operation wound with underlying Mass	Died after two months of diagnosis.
2	Malhorta et al, 1984 [5]	M	27	Gross Hematuria and loin pain	Calcified mass	Lower pole of left kidney	9 x 8 x 7	Multiple Mets in the skeletal bones: Femur scapula sacroiliac joint vertebrae and ribs	Mets identified 6 years after the primary resection with no evidence of local recurrence
3	Gomez-Brouchet et al, 2001 [6]	F	52	Gross Hematuria	Mass with central calcification	Upper pole of right kidney	8	No	Uneventful course within 1 year
4	Kaneko et al, 2006 [7]	F	61	Incidental finding on US	Mass with calcification	Under renal pelvis	2.5 x 2	No	Uneventful course within 6 years
5	Buse et al, 2009 [8]	F	23	Gross hematuria and left loin pain	Inhomogeneous calcified mass	Upper pole of left kidney	7 x 5	Para vertebrae thoracic lesions	Stable disease within36 months
6	Xu et al, 2012 [9]	M	64	Gross hematuria and left loin pain	Low signal mass at MRI	Upper pole of left kidney	11 x 8 x 6	Ureter	Died after 2 months of diagnosis
7	Tyagi et al, 2014 [10]	F	22	Gross hematuria, high fever and loin pain	Complex hypodense mass	Right kidney pelvis	6 x 6 5.6	Lung	Stable disease after 6 cycle of chemotherapy
8	Chen and ZI YE et al, 2015 [11]	M	17	Sudden severe loin pain and high fever	Complex real mass with calcification	Left renal pelvis	10	No	Stable disease after 10 months of surgery
9	Salehipour et al, 2017 [12]	M	22	Progressive loin pain	Heterogeneous mass with coarse calcification	Mid part of Right kidney	9	No	Not mentioned
10	Wang and Guo et al, 2017 [13]	F	33	Gross hematuria	Well circumscribed calcified mass	Lower pole of left kidney	7 x 7	No	Uneventful course for 28 months
11	Pani et al, 2017 [14]	M	24	Right loin pain	Large cystic mass with foci of calcification	Mid and upper pole of right kidney	8 x 7 x 6	Not mentioned	Uneventful course after 6 months
12	Valente et al, 2018 [15]	M	35	Episodic gross hematuria and loin pain	Calcified mass	Most of renal parenchyma	20 cm	No	Uneventful course after 18 months

The transformation from the undifferentiated cells to the cartilage appeared abrupt in some areas. Foci of necrosis were seen and there were 15 mitosis per 10/hpf. No obvious clear cells or epithelial component was identified on morphological assessment.

Immunohisto chemical studies (Figure 4) showed that the chondroid component is positive with vimentin and focally positive with S100 and BCL2. The blue cell component waspositive with BCL2, CD99 and patchy positive with vimentin. The proliferation index (KI-67) varied from low in the chondroid component to very high (approximately 100%) in the blue cell component.

The following immunostains were negative in the blue cell component:

S100, MelanA, GFAP, EMA, MNF116, CK AE1/AE3, CK7, WT1, CD3, CD20, CD10, CD34, STAT6, SMA, Desmin, Myogenin, MyoD1, CD31, Caldesmon, Calponin,

Napsin-A and NSE.

The features were those of a high grade malignant extra skeletal mesenchymal chondrosarcoma and combatable with PREMC. The case was sent to experts in soft tissue tumours for confirmation of

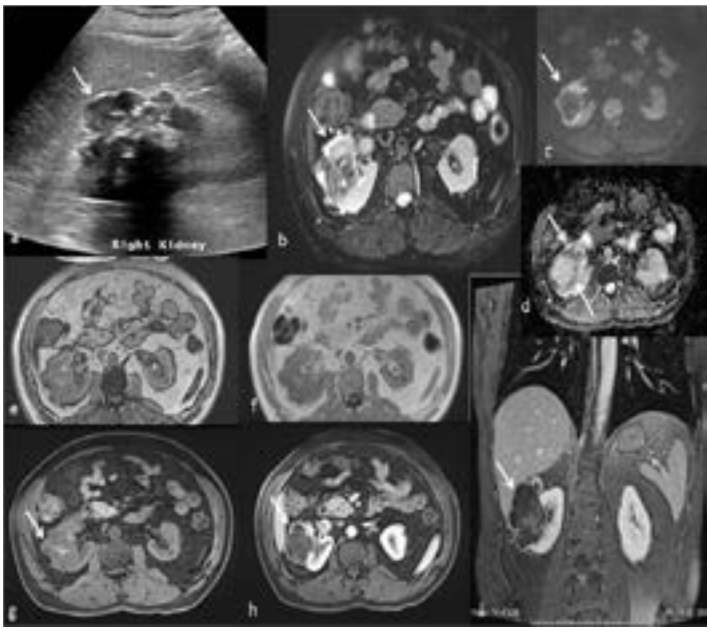

Figure 1a-i: US and MRI images of the right kidney (description in text)

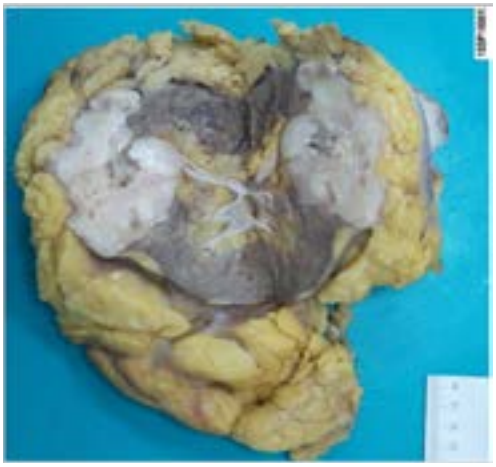

Figure 2. Gross Photograph showing PREMC in kidney invading the renal capsule and the kidney sinus fat

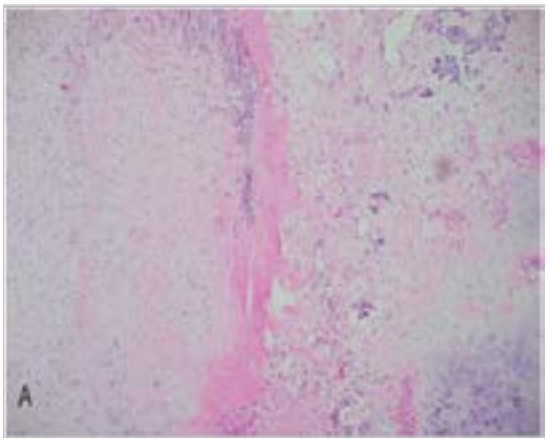

Figure 3a: chondroid component in PREMC (H&E, 100X)

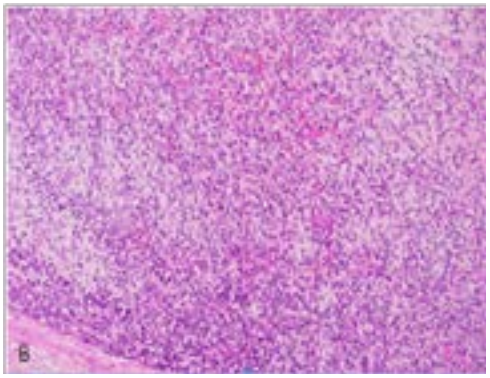

Figure 3b: Blue cell component (Mesenchymal cells) (H&E,200X)

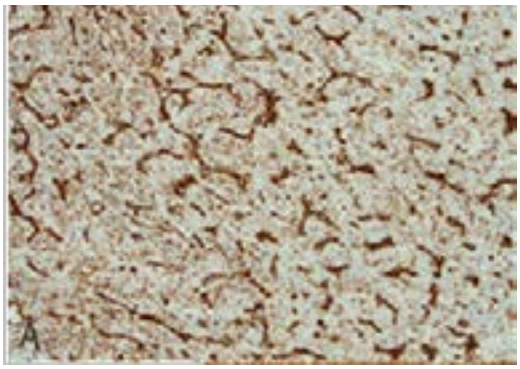

Figure 4a: Vimentin in Chondroid component, 100x

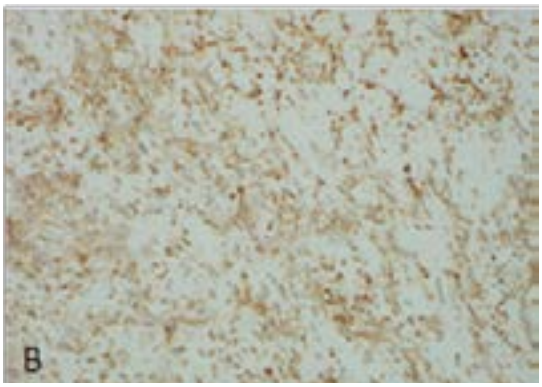

Figure 4b: CD99, Blue cell component, 200x

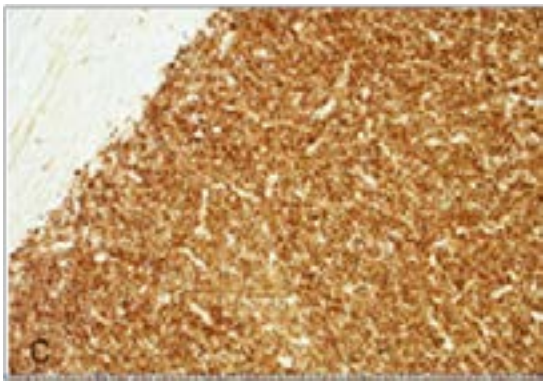

Figure 4c: BCL2, Blue cell component, 200x

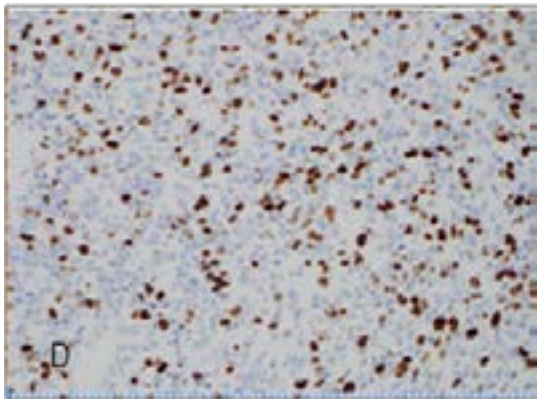

Figure 4d: KI-67 in Blue cell component, 200x

diagnosis and the experts agreed with our diagnosis.

4. Discussion

Mesenchymal Chondrosarcoma (MC) continues to present a difficult diagnostic, prognostic and management challenges due to the scarcity of the reported cases, despite being recognized as an entity for more than 60 years. A review of English literature was done through the PubMed database; twelve cases of PREMC were found and a summary of the key features of all these cases are displayed in Table 1.

In our review the tumor shows a male predominance with 8 out of 13 cases are male patients while the age of the presentation has been ranges from 17 to 64 years, mean age 34.5 years. Our patient is 62 years old. The clinical presentation is rather nonspecific where most of the cases identified by gross hematuria, loin pain and high-grade fever. The tumor ranged in size from 2 to 23cm and arose from the renal parenchyma in eleven cases while one case [10] the tumor originated from the renal pelvis and other case associated with a synchronous focus in the Ureter [9].

Radiologically the PREMC tumor usually presents as heterogeneous complex masses with coarse and rimmed calcification except two cases [9,10] which showed no calcifications. Most of the cases show calcifications on radiology imaging although nonspecific and the definitive diagnosis is still based on histopathological examination which illustrated as biphasic (chondroid and blue cell component) with infiltrative growth pattern whereas the blue cell component consists of undifferentiated mesenchymal cells. The course of the tumor exhibited distant metastasis and local recurrences in 5 out of 13 in different sites like thoracic paravertebral region, lung, skeletal bones, liver [16] ureter and span over a period from 6 month to 18 years.

The prognosis of the disease is poor where two patients died after 2 months of diagnosis[4,9]. Although there are no well-established treatment guidelines, the management for all these cases was radical nephrectomy with variable modalities of chemo-radiotherapy with no definitive evidence advocates their application. In our case the patient has been started on alternate VAC/IE Cycles (V; Vincristine-A; Adriamycin-C; Cyclohosphamide-I; Ifosfamide-E; Etopside) after the surgery. PET scan for the whole body was done after 3 months and showed no distant metastasis. However, the patient developed a neutropenic fever after the 1st cycle of chemotherapy and a derangement of his renal function(eGFR<30). Therefore, the second cycle was delayed and the Ifosfamide was omitted. PET scan was repeated 10 months after surgery and chemotherapy showed no sign of disease activity or distant metastasis. The patient is on a regular follow up and he is doing well so far.

5. Conclusion

We present another case of PREMC that is very rare tumor with very dismal prognosis. The handful reported cases, the lack of well-established treatment protocols and the variable tendency towards metastasizing and recurrences make regular follow up and close surveillance is highly recommended.

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