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CASE REPORT

Metastatic Undifferentiated Pleomorphic Sarcoma of Kidney: Presenting as Hematuria - A Case Report

Sushama Gurwale, Banyameen Iqbal, Nilanjana Dhar Choudhury, Charusheela Rajesh Gore

Department of Pathology, Dr. D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pune, Maharashtra, India

Pleomorphic undifferentiated sarcoma (PUS) is an extremely aggressive soft-tissue sarcoma that occurs most commonly in the trunk and the extremities but is extremely rare in the kidney. This entity has no distinctive clinical or radiological signs for diagnosis. It has high local recurrence and distant metastasis rates along with poor prognosis. We describe a case of PUS of the left kidney with urinary bladder metastases. Radiological investigations suggested an initial diagnosis of multifocal urothelial carcinoma; however, a definitive and confirmatory diagnosis of PUS was made on the basis of histopathological and immunohistochemical examination. Subsequently, left nephroureterectomy along with bladder cuff excision was performed and postoperative histopathology revealed PUS. Follow-up investigations revealed metastatic deposits of tumor in the base of the urinary bladder for which an endoscopic surgery of transurethral resection of the bladder tumor was performed. The histopathological findings were consistent with metastatic high-grade PUS of the kidney with metastasis to the bladder.

Key words: Pleomorphic, renal, sarcoma, undifferentiated

INTRODUCTION

Sarcomas of different types can arise from the adult kidney, including the renal capsule.[1] A primary renal sarcoma constitutes about 1%–3% of all renal malignant masses and approximately 6% out of all primary renal sarcomas[2,3] Pleomorphic undifferentiated sarcoma (PUS), formerly known as malignant fibrous histiocytoma (MFH), accounts for around 20% of all soft-tissue sarcomas. The extremities and trunk are where PUS occurs most frequently, while the kidney is a very uncommon location.[4] The sixth and seventh decades of life are when it tends to happen most commonly with an equal gender ratio.[1,3] Its pathogenesis is still unclear.[5] Since there is no specific clinical representation or any pathognomonic radiological features, preoperative diagnosis of PUS becomes rarely possible.[4,6,7] Thereafter, the definitive and confirmatory diagnosis completely rests on postoperative histopathology.[7]

CASE REPORT

A 55-year-old female presented to our hospital with chief complaints of left flank pain and hematuria for the past 8 months. She was apparently asymptomatic 8 months ago when she developed pain in the left flank region which was insidious in onset, gradually progressive, and diffuse in nature. Low-grade fever with chills, fatigue, and loss of appetite was observed during the above course of the period. She also reported a weight loss of 10–12 kgs over the last 6 months. Abdominal examination revealed distension and a large nonmobile mass in the left flank region.

Computer-assisted tomography (CT) scan of the abdomen with CT urography revealed a large ill-defined mass (9.0 cm × 7.8 cm × 7.2 cm) in the left kidney involving the upper and mid poles and extending into the pelvis, pelviureteric junction, and proximal ureter for a length of approximately 3.4 cm [Figure 1a and b]. Two more similar lesions were noted-one in the mid-ureter at L4 vertebral body level (2.8 cm × 1.0 cm) and another in the left ureterovesical junction (2.8 cm × 1.5 cm). The final impression was that of multifocal transitional cell carcinoma involving the left ureterovesical junction, left

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Corresponding Author: Dr. Banyameen Iqbal, Department of Pathology, Dr D. Y. Patil Medical College, Hospital and Research Centre, Dr. D. Y. Patil Vidyapeeth, Pimpri, Pune - 411 018, Maharashtra, India. Tel: +91 9730388872; Fax: +91 20 27420010. E-mail: banzey@gmail.com

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mid-ureter, and left kidney causing obstruction of the collecting system. The left kidney was nonexcreting. Furthermore, a fluorine-18-fluorodeoxyglucose (FDG) positron emission tomography-CT scan of the whole body depicted FDG avid mass in the dilated pelvicalyceal system of the left kidney with extension to the upper ureter. The diagnosis was suggestive of left multifocal urothelial carcinoma. A biopsy specimen was obtained through cystoscopy and sent for histopathological examination which revealed tumor cells arranged in sheets and fascicles with high N:C ratio, hyperchromatic, and pleomorphic nuclei with scanty-to-moderate amount of eosinophilic cytoplasm. Brisk mitotic activity including atypical mitosis and extensive areas of necrosis were noted [Figure 2a and b]. immunohistochemical (IHC) studies revealed the tumor cells to be positive for desmin and focally positive for smooth muscle actin, and negative for epithelial membrane antigen, cytokeratin (CK 5/6), P-63, PAN-CK, and uroplakin III. Henceforth, the IHC findings favored the diagnosis of high-grade PUS of the kidney [Figure 3a-d].

Subsequently, left nephroureterectomy along with bladder cuff excision was performed on the patient and sent for histopathological analysis [Figure 4a and b]. The microscopic features were consistent with that of high-grade PUS. Furthermore, an IHC workup was done (tumor cells were negative for CD 34, calponin, and Myo-D1) that favored the above diagnosis [Figure 5a-d].

Follow-up investigations including CT scan abdomen and pelvis with CT renal angiography revealed a lobulated mass attached to the base of the urinary bladder, suggestive of a metastatic tumor deposit. Subsequently, a planned surgery of transurethral resection of the bladder tumor was performed. The histopathological findings were consistent with metastasis of PUS to the bladder.

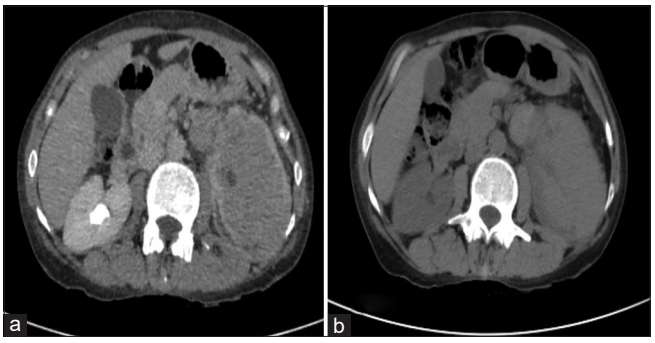

Figure 1: (a and b) A computed tomography (CT) scan with CT urography showing a mass lesion in the left kidney involving both the poles and the pelvis

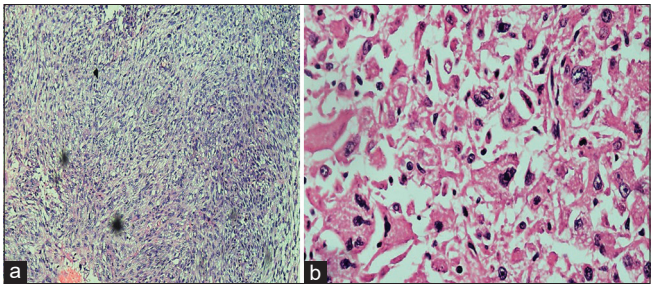

Figure 2: Photomicrograph shows tumor cells arranged in sheets and fascicles (a) (×200, H and E) Individual tumor cells show high N:C ratio, hyperchromatic pleomorphic nuclei, prominent nucleoli, and moderate amount of eosinophilic cytoplasm (b) (×400, H and E)

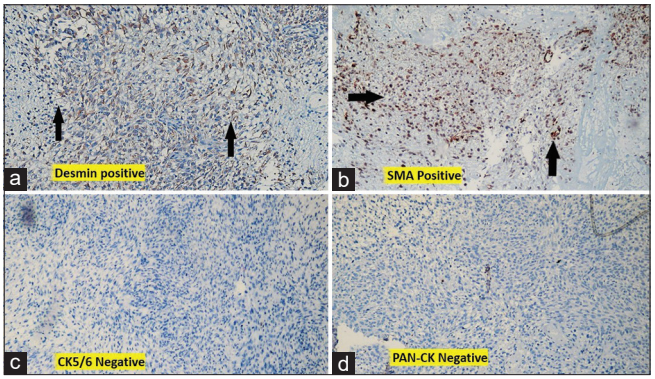

Figure 3: Photomicrograph shows immunohistochemical stains, (a) Desmin positive (black arrows), (b) Smooth Muscle Actin (SMA) Positive (black arrows), (c) CK 5/6 Negative. and (d) PAN CK Negative. [IHC X 200]

DISCUSSION

PUS was first described by O'bren and Stout in 1964, who termed them as MFH.[2,4,5] The tumor cells show mesenchymal as well as markers of mononuclear phagocytic system.[8] Due to their behavior being comparable to both sarcoma and lymphoma, Ghandur-Mnaymanch advocated changing its name to fibrous histiocytic sarcoma in 1987.[2,8] MFH was eliminated at the same time the term PUS was established in literature after the most recent World Health Organization modification.[2,8]

Till today, only around 60 cases of renal PUS have been published worldwide.[2] The most common site of occurrence of PUS is the extremities, followed by the body trunk and retroperitoneum. They have also been discovered in some rare sites such as the spermatic cord, prostate, bladder, and kidney.[3,6] The most commonly observed symptoms are abdominal or flank pain, the presence of any palpable mass, and/or weight loss.[3] Despite hematuria being known as a well-established common symptom of renal cell carcinoma (RCC), it can also rarely be found in renal PUS since the mass mostly arises from the capsule.[3] Similarly, our case presented with nonspecific complaints of left flank pain, hematuria, loss of appetite, and weight loss, which raised our suspicions toward RCC initially.

Moreover, since there are no specific clinical features or pathognomonic radiological signs to arrive at a provisional diagnosis of PUS, confirmatory diagnosis rests completely on histopathological examination.[3,9] The

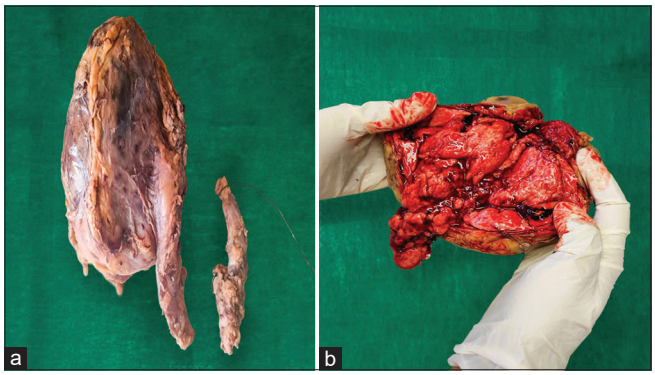

Figure 4: External surface (a) and cut section of the nephrectomy specimen (b)

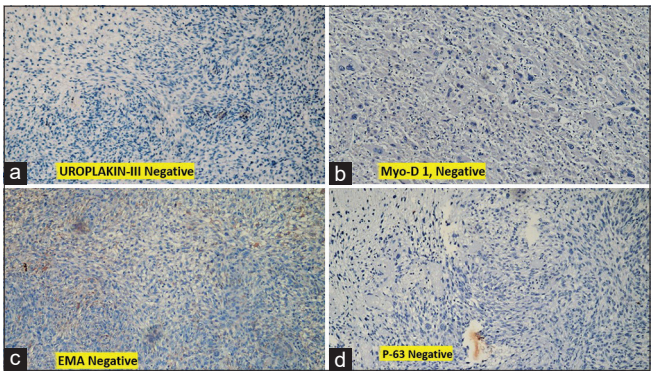

Figure 5: Photomicrograph shows immunohistochemical stains, (a) Uroplakin-III Negative, (b) Myo-D1 Negative, (c) Epithelial Membrane Antigen (EMA) Negative, and (d) P-63 Negative. [IHC ×200]

histological differentiation of the tumor can be done using immunohistochemical (IHC) staining.[5,10]

Since this tumor is so uncommon, there are no set management recommendations.[6] Most studies have found that total surgical excision of the tumor, followed by adjuvant chemotherapy, radiation, and/or immunotherapy, is the best course of action for PUS.[3] For our case, left nephroureterectomy along with bladder cuff excision surgery was performed, followed by adjuvant chemotherapy that has been ongoing for the past 4 months. On follow-up, there has not been any evidence of recurrence or distant metastases observed.

CONCLUSION

Renal PUS is an extremely aggressive tumor without any specific pathognomonic clinical or radiological findings. It is an aggressive form of tumor known for its high local recurrence and distant metastases. The primary challenge lies in the timely diagnosis of these tumors since their clinical and radiological findings are mostly nonspecific and could remain undiagnosed till a late stage. The preferred course of therapy is complete surgical resection, followed by adjuvant chemotherapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published, and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

Data availability statement

The data that support the findings of this study are available from the corresponding author, Iqbal. M, upon reasonable request.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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