Case Report

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Pleomorphic Undifferentiated Sarcoma Located in the Right Kidney

Sağ Böbrek Yerleşimli Pleomorfik Andiferansiye Sarkom

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ABSTRACT

A 70-year-old male presented to Urology clinic with right flank pain. He underwent radical nephrectomy with a preoperative diagnosis of renal cell carcinoma. After the histopathological and immunohistochemical examination, the diagnosis of pleomorphic undifferentiated sarcoma was determined. Primary renal pleomorphic undifferentiated sarcoma is extremely rare and highly aggressive tumor. Differential diagnosis from other sarcomas should be made with a careful histopathological examination and appropriate immunohistochemistry. We present this case for its rarity.

Keywords: Pleomorphic undifferentiated sarcoma, Malignant fibrous histiocytoma, Renal sarcoma

ÖZET

70 yaşında erkek hasta Üroloji kliniğine sağ yan ağrısı şikayeti ile başvurdu. Preoperatif renal hücreli karsinom ön tanısı ile radikal nefrektomi yapıldı. Histopatolojik ve immünohistokimyasal incelemeden sonra pleomorfik andiferansiye sarkom tanısı konuldu. Primer renal pleomorfik andiferansiye sarkom son derece nadir görülen ve oldukça agresif bir tümördür. Diğer sarkomlardan ayırıcı tanısı dikkatli bir histopatolojik ve uygun immünohistokimyasal inceleme ile yapılmalıdır. Bu vakayı nadir olması nedeniyle sunuyoruz.

Anahtar Sözcükler: Pleomorfik andiferansiye sarkom, Malign fibröz histiyositom, Renal sarkom

INTRODUCTION

Histogenesis of pleomorphic undifferentiated sarcoma (PUS), formerly known as malignant fibrous histiocytoma (MFH), still remains as one of the controversial issues (1, 2). The lesion was first defined in 1964 by O'bren and Stout (2-4). Tumor cells carry the characteristics of both mesenchymal and mononuclear phagocytic system (1,2). Ghandur-Mnaymanch recommended to rename it as fibrous histiocytic sarcoma in 1987 due to the tumor's behavior similar to sarcoma and lymphoma (1,2). After the update by World Health Organization at 2002, the term PUS was preferred instead of MFH (1,5,6).

PUS is considered to originate from primitive mesenchymal cells that manifest both histiocytic and fibroblastic differentiation (7-9). It has been accepted as the most common soft tissue sarcoma in adults (4,9). Limb involvement is the most commonly seen characteristic (4,6,10).

Primary renal PUS is a rarely seen PUS subgroup, and the number of reported cases was approximately 60 by 2018 (1). We have aimed to report this rare case in a 70-year-old male patient.

CASE REPORT

A 70-year-old man presented with history of right flank pain at the Urology clinic. There was no characteristic finding on physical examination. In the laboratory examination, platelet count was found to be 426x10 µl (100-300x10 µl), BUN value was 34.3 mg/dl (8.4-25.7 mg/dl), creatinine value was 1.57 mg/dl (0.57-1.25 mg/dl). In addition, microscopic examination of the urine revealed 4 RBCs (0-3/hpf) at each field. Other biochemical parameters were within normal limits. Computerized tomography (CT) scan revealed a solid lobulated tumor with a diameter of 7 cm which have cystic and necrotic areas in the upper pole of the right kidney. Subsequently performed magnetic resonance imaging (MRI) scan supported the find-

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ings of the CT scan. RCC was considered radiologically. Radical nephrectomy was performed because of suspicious renal cell carcinoma.

Macroscopically; the right kidney and perirenal adipose tissue were observed with a dimension of 22x10x9 cm a weight of 1066 grams. On the cut surface of the kidney, a grey-white colored tumor with a size of 8x6x5 cm was seen. The perirenal adipose tissue was infiltrated by the tumor. The tumor was necrotic and located at the upper pole of kidney while its boundaries were irregular.

Histopathological examination revealed tumoral proliferation with a storiform structure in the kidney parenchyma (Figure 1A,B). The tumor was composed of spindled shaped cells, bizarre and giant cells (Figure 2A). Mitosis was over 10 in 10 high power field (>10/HPF). There were large necrotic areas (Figure2B). Immunohistochemically neoplastic cells were positive with Vimentin (cytoplasmic) and CD 68 (cytoplasmic) (Figure 3A,B). The tumor cells showed no immunoreactivity with Cytokeratin, EMA, Desmin, CD117, S-100, CD34, CK7, CK20, CD10, CD99 and BCL-2. Proliferation index was approximately 70-75% with Ki67. The tumor was determined to be grade 3 (tumor differentiation= score 3, Mitotic count= score 2, Tumor necrosis= score 2) according to the French Federation of Cancer Centers Sarcoma Group Grading System.

In the light of these histomorphological and immunohistochemical findings, the diagnosis of PUS was established by 2 pathologists (ED, DG). Chemotherapy was initiated after

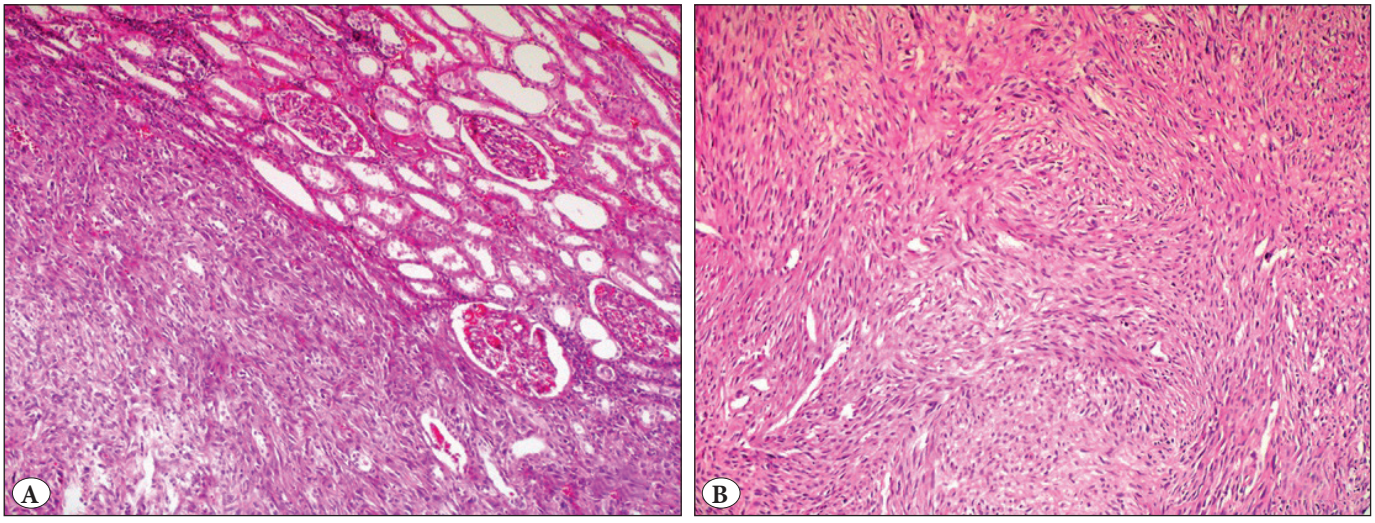

Figure 1: A) Tumoral proliferation in the kidney parenchyma (H&E, x100), **B)** Fibriohistiocytic cell proliferation with a storiform pattern (H&E, x100)

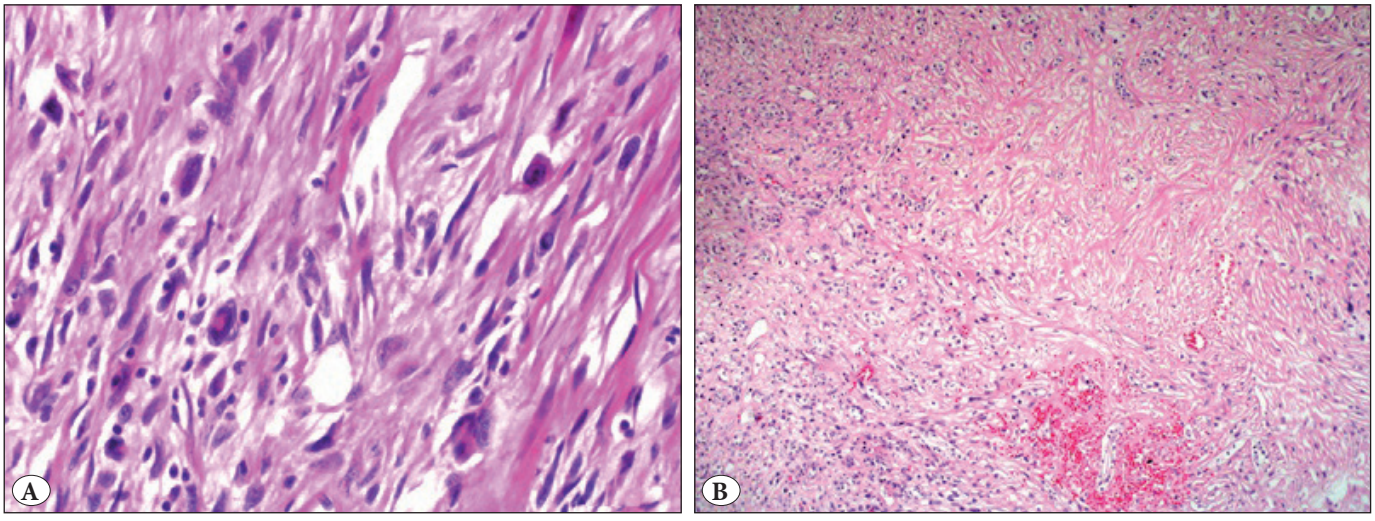

Figure 2: A) The tumor was composed of spindled shaped cells, bizarre and giant cells (H&E, x400), **B)** Large necrotic areas of necrosis (H&E, x100)

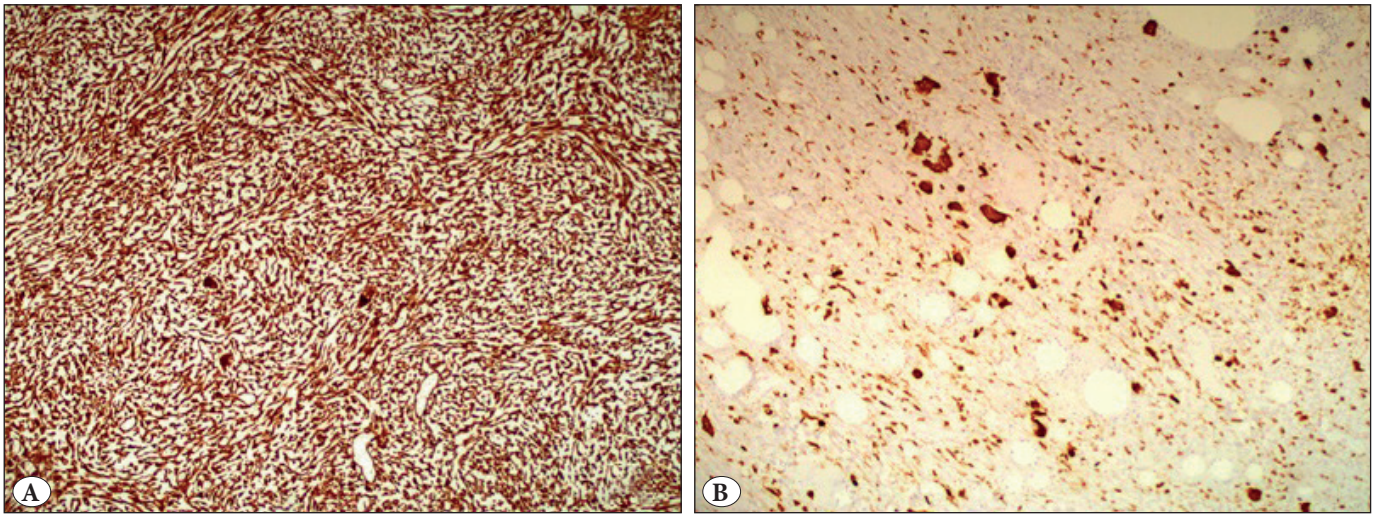

Figure 3: A) Immunoreactivity for Vimentin in tumoral cells (Vimentin, x100), **B)** Immunoreactivity with for histiocyte marker CD68 (CD68, x100)

treatment in the latter process. Four months after diagnosis, the patient had lung metastasis. The patient who developed lung metastasis and obstructive jaundice became exitus 9 months after surgery.

DISCUSSION

Renal sarcomas constitute a limited percentage of all malignant renal tumors, leiomyosarcoma is followed by liposarcoma, fibrosarcoma, and rhabdomyosarcoma (10,11). Renal PUS is an extremely rare tumoral formation that occurs with non-specific symptoms such as flank pain, fever, weight loss, a palpable mass and hematuria (3,6). Primary renal PUS occurs equally in both sexes and is most commonly seen between fifth and seventh decades of life (4,6,8). In diagnosis of PUS, no pathognomonic findings are detected by radiological imaging (4). PUS originating from the renal capsule is more common in the left kidney (4,11,12). In our case; age, sex, symptoms and radiological findings were consistent with most cases in the literature whereas right kidney localization was the characteristic that distinguished from other cases.

There are four main histological subtypes such as storiform-pleomorphic type, giant cell type, myxoid type and inflammatory type while storiform-pleomorphic type is the most common of those subtypes (3,5). Sarcomatoid carcinoma, renal cell carcinoma and other sarcomas should be eliminated before making diagnosis of renal PUS (11,12).

PUS is a diagnosis of exclusion when no other more specific entity is identified after extensive histological examination and use of ancillary diagnostic techniques. Microscopically, PUS is a heterogeneous malignancy with a variable cellular presentation and a predominantly fibrous stroma. All cases share a marked cytological and nuclear pleomorphism, with atypical mitosis and areas of necrosis, a predominant spindle-cell component, and scattered round histiocyte-like cells. Immunohistochemistry is of great importance in achieving a final diagnosis. PUS is distinguished from other sarcomas by the immunohistochemical positivity for CD68 (11,12). Although positivity for Vimentin and Alpha-1-antitrypsin are not specific to PUS, they are also expected (11,12).

PUS is easily differentiated from sarcomatoid carcinoma by the negative epithelial markers (11,12). The histopathological and immunohistochemical findings obtained in this case were consistent with the cases reported in the literature.

The significance of physical examination and radiology is limited in the differential diagnosis of PUS from other renal tumors (11,12). If Renal PUS is considered, the whole body should be examined considering the probability of another primary tumor localization.

The treatment of PUS is radical surgery (3,10-12). The role of adjuvant therapy is limited (3,10,12). PUS is an aggressive tumor that often recurs (4,7). PUS is the most commonly spread to the lungs and lymph nodes (4,6). Early surgical operation is critical with respect to the treatment of PUS. If there is local infiltration or remote metastases, the success rate is extremely low (7,11). The prognosis of PUS is usually disappointing with 5-year survival rate below 14% (2,7). Lung metastasis and obstructive jaundice developed in our case who received chemotherapy after surgery and became exitus 9 months after surgery.

CONCLUSION

Primary renal pleomorphic undifferentiated sarcoma is a rarely seen and highly aggressive tumor. The significance of physical examination and radiology is limited in the differential

diagnosis of PUS from other renal tumors. The treatment of PUS is radical surgery and the role of adjuvant therapy is limited. Differential diagnosis from other sarcomas should be made with a meticulous histopathological and appropriate immunohistochemical examination.

REFERENCES

1. Çilesiz NC, Erdoğan N, Özkan A, Kalkanlı A, Hazar Aİ, Balcı MBC, Aydın M, Özdemir E. Pleomorphic undifferentiated sarcoma: A case of a giant renal mass. Urol Case Rep. 2018;17:44-7.
2. Kumar S, Bansal P, Tiwari P, Kundu AK. Renal pleomorphic undifferentiated sarcoma: A rarity. Saudi J Kidney Dis Transpl. 2012;23(6):1241-2.
3. Kim SJ, Ahn BC, Kim SR, Kim YB, Joo HJ, Lee KB, Kim YS. Primary malignant fibrous histiocytoma of the kidney. Yonsei Med J. 2002;43(3):399-402.
4. Turk NS, Kelten C, Ozdemir NO, Duzcan E. Primary malignant fibrous histiocytoma of the kidney: Report of a case. Turk J Urol. 2010;26:165-7.
5. Adanur Ş, Keskin E, Demirci E, Ziypak T, Yapanoğlu T, Özbey İ. Primary renal pleomorphic undifferentiated sarcoma. Eur J Gen Med. 2014;1:66-8.
6. Pathrose G, John NT, Manojkumar R. A rare case of malignant fibrous histiocytoma/ pleomorphic undifferentiated sarcoma of the kidney. J Clin Diagn Res. 2015;9(1):PD27-9.
7. Singh S, Bhatia Y, Sharma J, Sen R. Malignant fibrous histiocytoma of the kidney. J Integr Nephrol Androl. 2017;4(2):67.
8. Ebrahimtabar F, Shafi H, Ranaee M, Darzi MM. A rare case of primary malignant fibrous histiocytoma: a sarcoma of the kidney. BMC Urol. 2019;19(1):45.
9. Kitajima K, Kaji Y, Morita M, Okuda Y, Sugimura K. Malignant fibrous histiocytoma arising from the renal capsule. Magn Reson Med Sci. 2003;2(4):199-202.
10. Ghosh A, Dwivedi US, Kumar A. Inflammatory malignant fibrous histiocytoma of kidney: A case report. Pathol Res Pract. 2008;204(11):857-61.
11. Göğüş Ç, Gökce Mİ, Süer E, Tulunay Ö, Şafak M. Primary malignant fibrous histiocytoma of the kidney: report of a case and literature review. Turk J Urol. 2013;39(3):194-7.
12. Tarján M, Cserni G, Szabó Z. Malignant fibrous histiocytoma of the kidney. Scand J Urol Nephrol. 2001;35(6):518-20.