



Primary Renal Pleomorphic Undifferentiated Sarcoma

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ABSTRACT

Malignant fibrous histiocytoma (MFH) is the most common soft tissue sarcoma in adults occurring in the extremities and retroperitoneum. However, primary MFH of the kidney is extremely rare. Clinical, and histopathological diagnosis of MFH creates difficulties both for urologists, and pathologists. Herein, we aim to present this rarely seen lesion, and discuss its clinical, and radiological prognosis, as well as its histopathological and clinical diagnosis.

Key words: Malignant fibrous histiocytoma, kidney, treatment

Primer Renal Pleomorfik Undiferansiye Sarkom

ÖZET

Malign fibröz histiyostom, adultlarda retroperitonum ve ekstremitelerde oluşan en yaygın yumuşak doku sarkomudur. Ancak böbreğin primer malign fibröz histiyositomu oldukça nadirdir. Malign fibröz histiyositomun klinik ve histopatolojik tanısı ürologlar ve patoloğların her ikisi içinde zorluklar oluşturur. Burada histopatolojik ve klinik tanısının yanısıra, klinik ve radyolojik prognozunu tartışarak bu nadir görülen lezyonu sunmayı amaçladık.

Anahtar kelimeler: Malign fibröz histiyositom, böbrek, tedavi

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is the most frequently seen subgroup of soft tissue sarcomas. It is often seen in lower extremities in 65-75% of cases. The incidence of retroperitoneal MFH is 6-16 per cent. However, primary MFH of the kidney is relatively rare (1). Clinical, and histopathological diagnosis of MFH creates difficulties both for urologists, and pathologists. Herein, we aim to present this rarely seen lesion, and discuss its clinical, and radiological prognosis, as well as its histopathological and clinical diagnosis.

CASE

A 77-year-old male patient was admitted to our clinic with complaints of left flank pain and fatigue on going for the previous 3 months. His physical examination was

unremarkable. His serum creatinine level was 1mg/dl. On abdominal ultra-sonograms, a mass lesion measuring nearly 8 x9cm, which stemmed from the upper pole of the left kidney, and multiple simple cysts of differing sizes situated in the right kidney were detected. An abdominal MRI performed in another centre revealed a 96 x 99 x 94mm heterogeneous solid mass on the level of middle -upper pole with superiorly oriented epiphytic extension which contained necrotic areas with surrounding hyper-intensities marked with enhanced contrast uptake (Figure 1). The patient underwent left radical nephrectomy with the diagnosis of left renal mass at postoperative 15 month, a control abdominal MRI was obtained. Neither local recurrence or distant metastases were observed.

Gross examination of the specimen revealed a yellowish-white 9x9x7cm sized mass, with multifocal necrosis and cystic changes in the middle- superior pole of the

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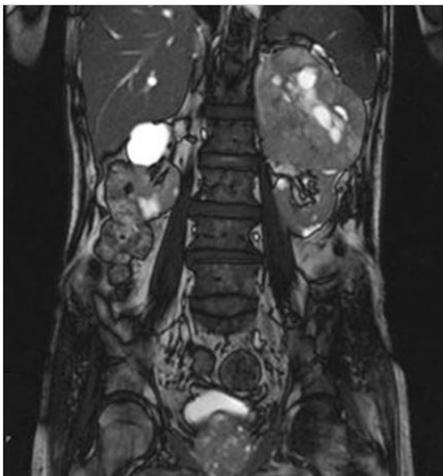


Figure 1. The lesion mass from the upper pole of the left kidney abdominal MRI

left kidney without renal vascular invasion. Multiple sections showed a tumour composed of predominantly spindle cells arranged in short fascicles with interspersed histiocytic cells including multinucleated giant cells. Marked nuclear pleomorphism and mitotic activity (5-7 per HPF) were seen. Areas of necrosis and focal inflammatory infiltrate were also present. The adjacent renal parenchyma and perinephric fat was tumour negative. On immunohistochemistry, the spindle cells stained for vimentin and focally for CD34 while histiocytic cells were positive for CD68. The tumour cells were negative for CK7, CK20, HMWCK, LMWCK, epithelial membrane antigen and S-100 protein (Figure 2a,b,c,d).

DISCUSSION

In the current World Health Organization classification of soft tissue tumors, MFH is considered synonymous with pleomorphic undifferentiated sarcoma. MFH was firstly described in 1963 and has currently become the predominantly reported histological diagnosis among soft tissue sarcomas (2). MFH, arises from renal capsule, and attacks both genders with comparable incidence. It is most frequently seen in the fifth and seventh decades of life. It tends to involve the left kidney (3). Flank pain, fever, weight loss, and a palpable renal mass are widely observed manifestations. Haematuria is rarely noted (4). Its nonspecific clinical signs correlate with the dimensions of the tumour. Before the onset of symptoms, le-

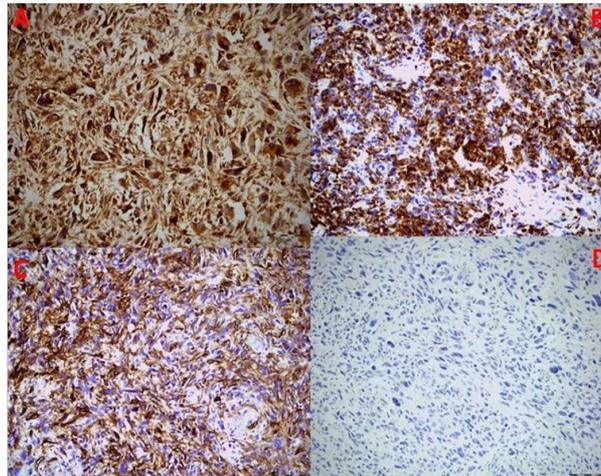


Figure 2. A: The tumoral area adjacent to the normal kidney tissue (CD68x400) B: Malignant fibrous histiocytoma stariform-pleomorphic type (Vimentinx200) C: Necrosis and hemorrhagic areas (CD34x400) D: Cells in malignant fibrous histiocytoma ara characterized by an extreme degree of pleomorphism and occassional multi-nucleation (EMAx400)

sions might become extremely bulky because of their retroperitoneal locations, and thus delay establishment of correct diagnosis (5). MFH has 4 histological subtypes including storiform-pleomorphic, giant cell, mixoid, and inflammatory forms (6).

Establishing clinical and radiological diagnosis of renal MFH and differentiating it from other renal tumours like renal cell carcinoma is a very challenging task in consideration of its relatively asymptomatic nature and nonspecific imaging characteristics (7). Definitive diagnosis can be only made by histopathological examination (3,4). Retroperitoneal location of MFH, physical examination findings, radiological scanning of extremities, proper macroscopic evaluation of the renal mass within the nephrectomy specimen provide important hints about the primary origin of the mass. Although its differentiation from other primary sarcomatous lesions of the kidney including leiomyosarcoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, and malignant schwannoma might pose difficulties, immunohistochemically CD68 positivity is specific for MFH (8). Also in our case, immunohistochemical analysis of histiocytic cells revealed the evidence of CD68 positivity, although CK7, CK20, epithelial membrane antigen (EMA) and S-100 protein

negativity. Initial treatment for all cases with MFH is radical nephrectomy because of overwhelming suspicion of renal cell carcinoma (3,4). Despite radical surgery, MFH tends to manifest local recurrences, and distant metastases to bones, and lungs in nearly 50% of cases (9). As most of the literature studies have indicated, local recurrences have reportedly occurred within postoperative 3-24 months, and 60% of cases died because of disease related complications within postoperative 12 months (3,9). Contrary to angiosarcomas and leiomyosarcomas, MFH is especially susceptible to doxorubicin and ifosfamide-based chemotherapeutic regimens. Still, generally it has an unfavourable prognosis. Tumoural size and histological grade are important prognostic parameters (9). We did not administer any chemotherapy protocol for our patient during postoperative period. Control MRI performed at postoperative 15 months did not reveal any evidence of local recurrences or distant metastasis. In conclusion, although primary renal MFH is a rarely seen entity, it can attain larger sizes without any symptomatic manifestation. Besides, because of its nonspecific radiological characteristics it cannot be readily discriminated from other renal malignancies. The absence of local recurrence and distant metastasis after surgery is the most important feature of this case. We think that the disease-specific survival rates of MFH can be increased if early diagnosis of this rarely seen aggressive disease can be made, and its systemic treatment can be instituted by collaborative efforts between clinicians and pathologists.

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