

Primary Malignant Fibrous Histiocytoma Of The Kidney: Case Report And Review Of Literature

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Citation

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Abstract

Malignant fibrous histiocytoma as a primary renal tumor is rare. It is clinically and radiologically indistinguishable from a renal cell carcinoma and diagnosis is made by histopathology. We report a case of primary renal MFH of inflammatory type in a 45-year-old female whose CT scan revealed a well demarcated mass at the lower pole of left kidney. Preoperative diagnosis was a renal cell carcinoma and radical nephrectomy was performed. Histological examination revealed malignant fibrous histiocytoma of inflammatory variety. No adjuvant therapy was given. The patient died of metastasis. We present this case with review of literature.

INTRODUCTION

Malignant fibrous histiocytoma (MFH), formally called malignant fibroxanthoma, is the most common soft tissue sarcoma in adulthood.¹ It is a primitive mesenchymal tumour, occurs typically in extremities and retroperitoneum and seldom in genitourinary organs.² Primary involvement of the kidney is rare with less than 50 cases being reported.³ We report this rare case with review of literature.

CASE REPORT & MANAGEMENT

A 45-year-old female presented with left flank pain and intermittent gross hematuria with passage of clots for the past 4 months. She experienced occasional nausea and indigestion but no fever, anorexia, weight loss, burning micturition or frequency. Physical examination revealed pallor and no other abnormality was detected. Haemogram showed anaemia & leucocytosis. Blood chemistry showed normal kidney & liver function tests. Computerized tomogram (Fig-1) revealed a 7x4.7x6cm, well demarcated mass with low enhancement compressing the left pelvic calyceal system with no evidence of intramural calcification, haemorrhage, renal vein/IVC involvement or lymphadenopathy.

Figure 1

Figure 1: Abdominal CT scan showing a 7x4.7x6cm left renal mass.



Chest x-ray and bone scan were normal. With the pre-operative diagnosis of renal cell carcinoma, radical nephrectomy was done. Gross examination of specimen revealed a 7x6cm yellowish solid nodular tumor with multifocal necrotic areas in the lower pole of the kidney without perinephric, renal pelvic or renal vascular invasion. Histopathology showed a tumor arranged in nodular aggregates, separated by thin fibrous tissue, infiltrating the renal parenchyma. Cells were an admixture of heterogeneous populations of histocytes with foamy cytoplasm, lymphocytes, plasma cells, neutrophils, eosinophils, mononuclear cells and multinucleated tumor giant cells with

areas of active mitosis (5-10 hpf). There was no vascular involvement or regional lymph node metastasis. The tumor was stained negative for cytokeratin, epithelial membrane antigen (EMH) desmin, smooth muscle actin and S-1000 on immunostaining. In light of these findings the pathological diagnosis was inflammatory subtype of MFH arising from renal parenchyma. The patient was followed every 3 months. Eight months after surgery she had abdominal pain, chest pain and dysnoea. Investigation revealed local recurrence (ultrasonography) and lung metastasis (malignant pleural effusion on cytology). Her condition deteriorated and she died 11 months after surgery.

DISCUSSION

Malignant fibrous histiocytoma was first described by O'Brien and Stout in 1964.¹ It is a primitive mesenchymal tumor with some histiocytic and fibroblastic differentiation.² Typical locations are the extremities (65-75%) followed by retroperitoneum (6-16%).² Other sites reported are urinary bladder, prostate, spermatic cord, renal capsule, renal vein and renal parenchyma.²

The histological hallmark of MFH is a storiform arrangement of spindle cells with admixture of histocyte like cells, multinucleated giant cells and variable numbers of inflammatory cells.⁴ MFH can be divided into 4 subgroups, Storiform- pleomorphic (most common), myxoid, giant cells and inflammatory variant according to the predominant tumor component.²

Retroperitoneal MFH does not present clinical symptoms distinct from other retroperitoneal neoplasms.² The tumor is usually advanced when symptoms arise. Clinically and radiographically, MFH arising from the kidney or renal capsule cannot be differentiated from other renal or retroperitoneal neoplasms such as renal cell carcinoma, inflammatory pseudotumor or xanthogranulomatous pyelonephritis.⁴ However, selective renal arteriography usually reveals a hypovascular tumor as compared to renal cell carcinoma.⁵ Kitajima et al.⁶ reported that MR imaging was helpful in differentiating MFH of the renal capsule from other renal tumor. In particular, a hypointense area identified on T2-weighted images reflecting the fibrous component is an important characteristic. Round or ring like calcification around the tumor has been detected in 16% of abdominal MFH, especially in the storiform-pleomorphic subtype.⁴

Definitive diagnosis of MFH is made by the histological examination,² and immunohistochemistry.⁷ Diagnosis of

MFH is essentially a diagnosis of exclusion which is rendered when dealing with a malignant sarcoma with marked cellular pleomorphism, exhibiting no specific cell lineage of differentiation except that of a generic fibroblastic (vimentin positivity) and less importantly a possible histiocytic (CD68 positivity) one.³

Pre-operative diagnostic imaging usually includes ultrasonography and computerized tomography.² However, these patients undergo simple nephrectomy or radical nephrectomy on the suspicion of renal cell carcinoma because preoperative differentiation from renal cell carcinoma is not possible by currently available clinical or radiological modalities.²

Early and complete resection of the tumor is the most important treatment. If the tumor arises from the renal parenchyma, radical nephrectomy is the treatment of choice.³ However, kidney sparing surgery could be performed, if the tumor originates from the renal capsule with no parenchymal involvement.⁴

Failure of complete resection of the tumor results in poor prognosis. Despite radical surgery, MFH shows a strong tendency for local recurrence (>50%) and distant metastasis to lung, lymph nodes and bone.³ Even early detection of local therapy failure and promptly initiated aggressive salvage therapy may offer the chance of long term survival only in selected cases.⁸

The role of adjuvant chemotherapy and radiotherapy is questionable.² Eroglu et al.⁴ reported no recurrence for 15 months after surgical excision and adjuvant radiotherapy of 6600 rads. Papadopoulos and Rudolph,⁹ reported recurrence-free survival for one year after tumor nephrectomy, radiotherapy and chemotherapy with doxorubicin and ifosfamide. Chen et al.² reported that there was no recurrence during 12 months of follow-up after radical nephrectomy, 6 cycles of local irradiation and subsequent chemotherapy with doxorubicin and ifosfamide. Mureto et al.¹⁰, however, reported recurrence of metastasis in multiple sites 17 months after nephrectomy and adjuvant chemotherapy with adriamycin, vincristine, cyclophosphamide and decarbazine. On the other hand, Matsui et al.¹¹ reported a recurrence-free survival for 31 months with no adjuvant chemotherapy/radiotherapy for a MFH originating from the renal capsule.

The prognosis of MFH in retroperitoneal tissue is generally very poor with a recurrence rate of more than 50% and a 5-

year survival rate of only 14%.² Among reported cases of renal MFH, 25% of the patients died within one year after surgery.²

The treatment of renal MFH is not yet standardized and close, ongoing and lifelong follow-up is advisable.

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References

1. O'Brien JE, Stout AP. Malignant fibrous xanthomas. *Cancer*, 1964; 17: 1445-1455.
2. Kim SJ, Ahn BC, Kim SR, et al. Primary malignant fibrous histiocytoma of the kidney. *Yosei Med J* 2002; 43 (3): 399-402.
3. Chen CH, Lee PS, Han WJ, et al. Primary giant cell malignant fibrous histiocytoma of the kidney with staghorn calculi. *J Postgrad Med* 2003; 49 (3): 246-248.
4. Eroglu M, Bakirtas H, Cimentepe E, et al. Malignant fibrous histiocytoma arising from renal capsule. *Urol Int* 2005; 75 (4): 368-370.
5. Matsui Y, Kobayashi S, Sugino Y, et al. Malignant fibrous histiocytoma originating in a renal capsule: a case report. *Acta Urol Jpn* 2001; 47 (10): 727-9.
6. Kitajima K, Kaji Y, Monta M, et al. Malignant fibrous histiocytoma arising from renal capsule. *Mag Res in Med Sc* 2003; 2 (4): 199-202.
7. Wuk, Han Z, Ou T. Malignant fibrous histiocytoma of kidney. *Chin J Surg* 1996; 34 (9): 563-5.
8. AV May M, Marusch F, Heike C, et al. Locally recurrent malignant fibrous histiocytoma of the kidney and the spermatic cord. *Urologe* 2004; 43 (8): 976-81.
9. Papadopoulou I, Rudolph P. Primary renal malignant fibrous histiocytoma. *Urol Int* 1999; 63: 136-8.
10. Muretto P, Lemma E, Grianti C, et al. Inflammatory malignant fibrous histiocytoma of the kidney: an immunohistochemical and ultrastructural study. *Tumori* 1985; 71: 147-153.
11. Yonemoto Y, Oka N, Mizuno Y, et al. Spontaneous rupture of malignant fibrous histiocytoma arising from the renal capsule: a case report. *Acta Urol Jpn* 1998; 44(3): 159-62.

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