

A Rare Case of Malignant Fibrous Histiocytoma/ Pleomorphic Undifferentiated Sarcoma of the Kidney

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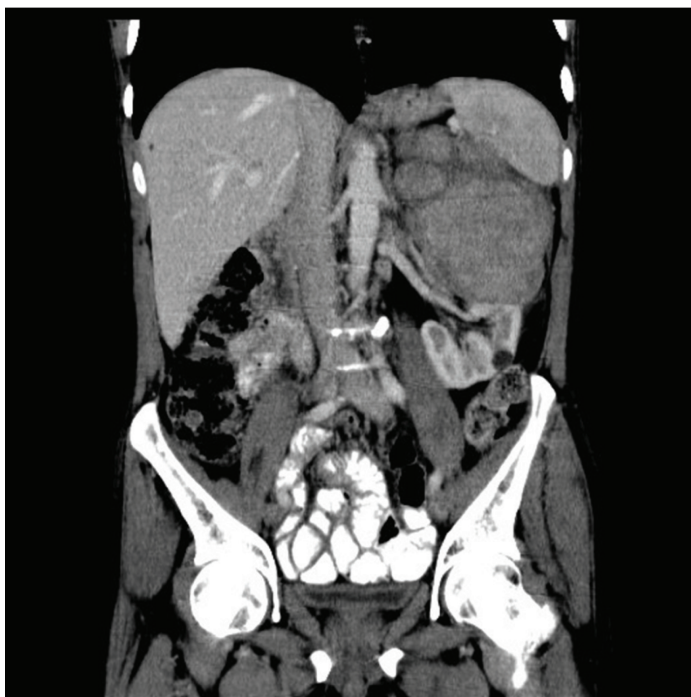
ABSTRACT

Malignant Fibrous Histiocytoma (MFH) usually arises from the extremities with the retroperitoneum being the second most common site. We present the case report of a 58-year-old man presenting with fever and was detected to have a renal mass on evaluation. He underwent a radical nephrectomy with a preoperative diagnosis of renal cell carcinoma. But the final histopathological report was MFH. Primary renal MFH is extremely rare and is usually diagnosed on histopathology with the aid of immunohistochemistry.

Keywords: CD 68, Kidney, Radical nephrectomy

CASE REPORT

A 58-year-old gentleman presented with history of fever and left loin pain for three months. On physical examination, he was found to have a renal mass. On laboratory evaluation he was found to be anaemic (PCV 23.8) with thrombocytosis (7,25,000/mm³) and had an elevated ESR (77). His CT abdomen revealed an 11 x 13 cm heterogeneously enhancing mass arising from the upper pole of left kidney [Table/Fig-1]. There was no involvement of the contiguous structures but superiorly, it was adherent to the spleen.



[Table/Fig-1]: CT scan showing a heterogeneously enhancing mass arising from the upper pole of left kidney

He underwent a radical nephrectomy and splenectomy as the tumour was adherent to the spleen. His post operative period was uneventful.

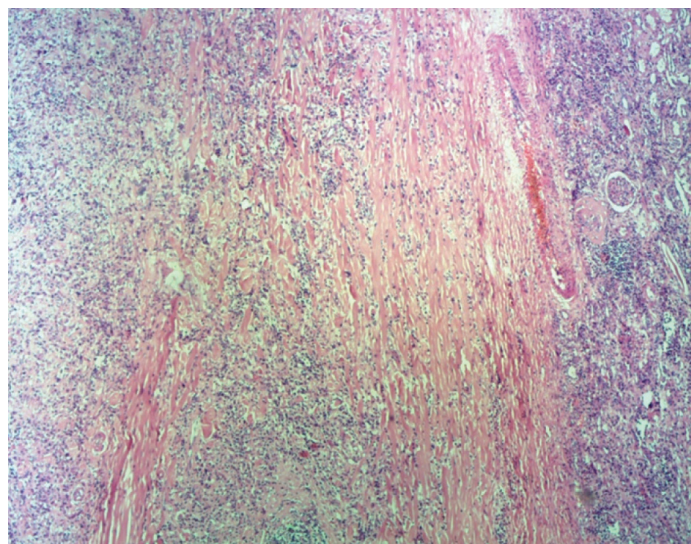
HISTOPATHOLOGY

Gross: A well-circumscribed tumour was seen arising from the upper and interpole of the kidney. The cut surface was grayish white, homogenous with focal areas of yellowish and myxoid change.

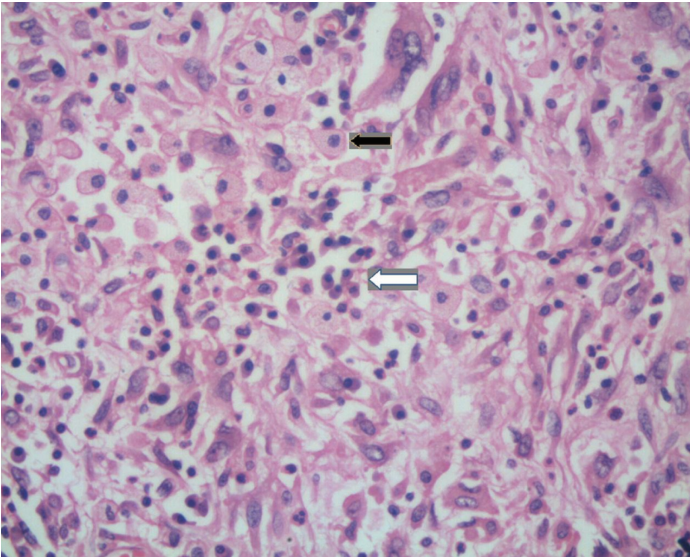
Microscopy: The renal parenchyma was infiltrated by a fairly circumscribed tumour [Table/Fig-2]; with clustered and scattered polygonal cells to spindle cells with markedly pleomorphic, vesicular nuclei and moderate amount of eosinophilic, cytoplasm. There were multinucleate giant cells, foamy macrophages and bizarre tumor cells. The intervening stroma showed many thick collagen bundle and dense infiltrates of lymphocytes, plasma cells, histiocytes, many neutrophils and eosinophils [Table/Fig-3]. There was focal hyalinization. There was no lymphovascular invasion or nodal involvement. The tumour cells were negative for desmin and myogenin but showed positive staining with CD68 [Table/Fig-4] on immunohistochemistry. These features were consistent with the diagnosis of inflammatory subtype of malignant fibrous histiocytoma. The tumour infiltrated the sinus fat with rest of the parenchyma being normal. Spleen showed features of congestive splenomegaly but was otherwise free of tumour [Table/Fig-5].

DISCUSSION

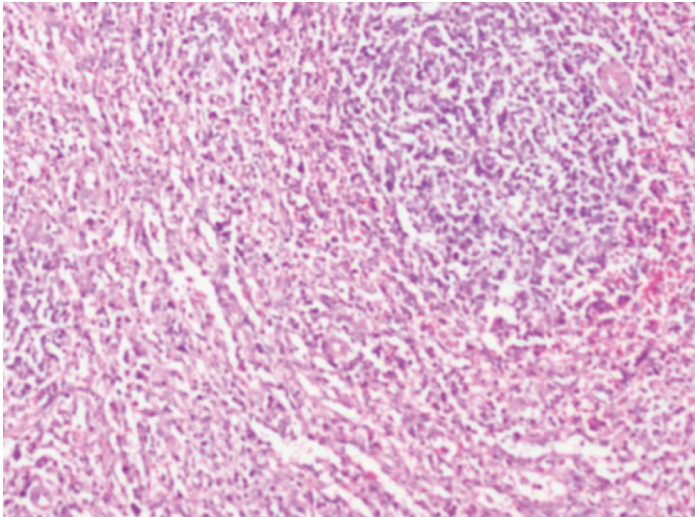
MFH is a pleomorphic sarcoma arising from fibroblasts and histiocytes and was first described by O'Brien and Stout in 1964 [1]. It is an extremely rare neoplasm and only 51 cases were reported till 2002 [2]. A few more cases have been reported since and have been highlighted [Table/Fig-6].



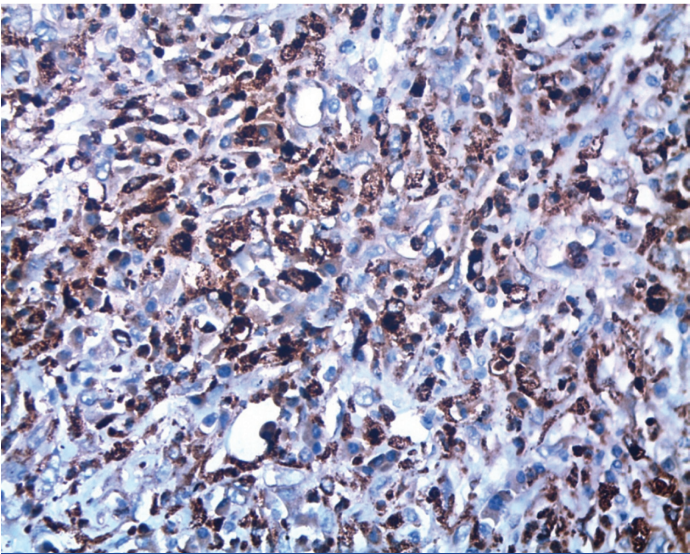
[Table/Fig-2]: H&E 5x, showing renal parenchyma infiltrated by tumour cells



[Table/Fig-3]: H&E 40x, showing clusters of pleomorphic spindle cells, interspersed with foamy histiocytes (black arrow) and mixed inflammatory cells (white arrow)



[Table/Fig-5]: H&E 40x, congested spleen



[Table/Fig-4]: CD68 400x, Cells showing positivity on immunohistochemistry with CD68

least common is the inflammatory subtype [13,14]. The histology of this patient has been reported as inflammatory type of MFH. It contains both fibrous and histiocytic elements. They stain positive for CD68 and anti $\alpha 1$ antichymotrysin but do not stain for cytokeratins [4].

According to the latest WHO update on renal sarcoma, MFH has been replaced with the term pleomorphic undifferentiated sarcoma after other forms of differentiation has been ruled out [15]. MFH is an aggressive tumour necessitating radical surgery with recurrence rates as high as 44%. It is associated metastases to lung (82%) and lymph nodes (32%) [13,16]. The major predictors of outcome are size, depth of involvement and inflammatory component of tumour [13]. They also require adjuvant chemotherapy usually with Ifosfamide and doxorubicin owing to the high rates of local recurrence and metastasis. Anecdotal reports of response to adjuvant radiotherapy to the tumour bed are also reported in literature [17]. Despite aggressive treatment, local recurrence was seen in the first two years and mortality rate was 66% within the first year of radical nephrectomy [6].

CONCLUSION

Malignant fibrous histiocytoma, now referred as undifferentiated pleomorphic sarcoma is a highly aggressive rare neoplasm with difficult preoperative diagnosis. Complete excision offers the only possible chance of cure with majority of patients succumbing to the illness in the first two years. Adjuvant therapy is of little or no benefit.

MFH presents in the 6-7th decades of life usually, with nonspecific symptoms like fever, weight loss and non specific abdominal pain [2]. The most frequent site is the extremities (49%) followed by the retroperitoneum (16%) [2]. The male to female preponderance

Author	Year	Pathology	Outcomes
Kitajima K et al., [3]	2003	MFH	Post radical nephrectomy, no adjuvant therapy. Follow up till 30 months, patient asymptomatic.
Chen CH et al., [4]	2003	Giant cell subtype MFH	Radical nephrectomy followed by 6 cycles of local irradiation and subsequent chemotherapy with doxorubicin and ifosfamide. Patient followed up for 12 months without evidence of tumour recurrence.
Erolu M et al., [5]	2005	MFH	Radical nephrectomy with adjuvant RT with 6,600 rads. No recurrence at 15 months.
Singh SK et al., [6]	2006	MFH	Patient died after 1 month of radical nephrectomy.
Ishibiki Y et al., [7]	2007	storiform-pleomorphic MFH arising from perirenal tissue	Refused adjuvant therapy after radical nephrectomy. Died due recurrence after 21 months.
Gupta R et al., [8]	2008	MFH	Patient dies after 2 cycles of chemotherapy.
Ghosh A et al., [9]	2008	Inflammatory subtype of MFH	No follow up.
Mellas S et al., [10]	2012	Pleomorphic undifferentiated sarcoma	Post nephroureterectomy, refused adjuvant therapy. Died after 10 months.
Matsushita M et al [11]	2013	MFH arising from renal capsule	Post radical nephrectomy, symptom free till 14 month post operatively.

[Table/Fig-6]: Case reports of MFH from 2001-2014

REFERENCES

[1] O'Brien JE, Stout AP. Malignant fibrous xanthomas. *Cancer*. 1964;17:1445-56.
[2] Kim SJ, Ahn BC, Kim SR, et al. Primary malignant fibrous histiocytoma of the kidney. *Yonsei Med J*. 2002;43(3):399-402.

- [3] Kitajima K, Kaji Y, Morita M, Okuda Y, Sugimura K. Malignant fibrous histiocytoma arising from the renal capsule. *Magn Reson Med Sci*. 2003;2:99-202.
- [4] Chen CH, Lee PS, Han WJ, Shen KH. Primary giant cell malignant fibrous histiocytoma of the kidney with staghorn calculi. *J Postgrad Med*. 2003;49(3):246-48.
- [5] Erolu M, Bakirta H, Cimentepi E, Unsal A. Malignant fibrous histiocytoma arising from the renal capsule. *Urol Int*. 2005;75(4):368-70.
- [6] Singh SK, Mandal AK, Agarwal MM, Das A. Primary renal inflammatory malignant fibrous histiocytoma: A diagnostic challenge. *Int J Urol*. 2006;13:1000-02.
- [7] Ishibiki Y, Matsumura T, Kawarai M, Hukushi Y. Malignant fibrous histiocytoma arising from perirenal tissue: a case report. *Nihon Hinyokika Gakkai Zasshi*. 2007;98(4):638-42.
- [8] Gupta R, Gupta S, Aggarwal D, Singh S. Primary pleomorphic undifferentiated sarcoma of the kidney: A rare renal tumor. *Indian J Pathol Microbiol*. 2008;51(4):573-76.
- [9] Ghosh A, Dwivedi US, Kumar A. Inflammatory malignant fibrous histiocytoma of kidney: a case report. *Pathol R Pract*. 2008;204(11):857-61.
- [10] Mellas S, Bouchikhi AA, Tazi MF, Khallouk A, Elammari JE, El Fassi MJ, et al. Primary Pleomorphic Undifferentiated Sarcoma—a Rare Renal Localization: A Case Report. *Case Rep Urol*. 2012;2012:862493.
- [11] Matsushita M, Okada T, Kawamura N, Ujike T, Nin M, Tsujihata M. A case report of malignant fibrous histiocytoma arising from the renal capsule. *Hinyokika Kiyo*. 2013;59(11):733-36.
- [12] Tarján M, Cserni G, Szabó Z. Malignant fibrous histiocytoma of the kidney. *Scand J Urol Nephrol*. 2001;35:518-20.
- [13] Weiss SW, Enzinger FM. Myxoid variant of histiocytoma. *Cancer*. 1977;39(4):1672-85.
- [14] Rosai J (ed.). Soft tissues. In: *Ackerman's Surgical Pathology*, 8th edn. Mosby, St Louis, 1996; 2021–138.
- [15] Fletcher CD. The evolving classification of soft tissue tumors: An update based on the new WHO classification. *Histopathology*. 2006;48:3-12.
- [16] Joseph TJ, Becker DI, Turton AF. Renal malignant fibrous histiocytoma. *Urology*. 1991;37:483-89.
- [17] Marchese R, Bufo P, Carrieri G, Bove G. Malignant fibrous histiocytoma of the kidney treated with nephrectomy and adjuvant radiotherapy: a case report. *Case Rep Med*. 2010;2010.

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