

## “Malignant Peripheral Nerve Sheath Tumour Arising In Retroperitoneal Neurofibroma Infiltrating In Kidney ,Masquerading As Rcc On Cect-A Diagnostic Dilemma .”

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**Abstract:** Malignant peripheral nerve sheath tumors (MPNSTs) are very rare soft-tissue sarcomas with an overall incidence of 0.001% in general population . It may develop de novo or it may develop as a result of malignant transformation of previous plexiform neurofibroma. Retroperitoneal cases are fairly rare and clinically difficult to be detected. MPNSTs affect adults aged 20–50 years and are typically connected to the main trunks of nerves. Most MPNSTs are aggressive and have high rates of recurrence. Only 1% of MPNST are retroperitoneal. In most cases the diagnosis depends on pathological and immunohistochemical studies. We here report a case of 60 yr old male suspected with Renal cell carcinoma on CECT but came out to be retroperitoneal neurofibroma with malignant change – low grade MPNST on histopathological examination.

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### I. Introduction

MPNSTs are malignant tumours arising from a peripheral nerve or in extra neural neurofibromas in NF1 or rarely from neurogenic tumours (e.g. schwannoma, ganglioneuroblastoma, ganglioneuroma). MPNST commonly occur in the extremities and account for 5–10% of all soft-tissue sarcomas and up to 50% occur in patients with neurofibromatosis type 1 (NF-1), 10% are radiation-induced, and 40% are sporadic.<sup>1</sup> These tumors are usually found in patients between 20-50 years of age, in the head and neck region, trunk or in the soft tissue and showing nerve sheath differentiation.<sup>2</sup> These tumors originate from Schwann cells or pluripotent perineural cells of the neural crest .MPNSTS can either arise de novo or differentiate from a preexisting neurofibroma (most often plexiform limbs).<sup>3</sup> Renal or perirenal MPNST are exceptionally rare findings. MPNSTs are highly cellular tumors comprising of spindle cells which are reminiscent of schwann cells.<sup>4</sup>

### II. Case Report

A 60 -year-old male presented with complain of pain in left side of abdomen with mass felt in left side which was gradually increasing. Patient also complained of difficulty in micturition with loss of appetite ,loss of weight since six months . There was no history of hematuria or urgency of micturition. The clinical examination and CECT suspected a left renal tumour ( ? Renal cell carcinoma). Routine hematological and biochemical investigations revealed anemia with history of multiple blood transfusions during the past 6 months. RFTs were normal with only mild elevation of blood urea levels. (45 mg/dl). Nephrectomy was done and specimen sent to pathology department.

### III. Pathological Findings:

Gross findings : Renal tissue measuring 9x6x5cm along with an attached large bosselated soft tissue piece measuring 10x9x5 cm was received. (Fig 1). On cut section the soft tissue pieces showed creamish white surface with fish flesh consistency along with solid areas. Multiple pararenal masses measuring 9x7x3 cm altogether were also received which on cut section showed creamish white homogenous areas with fish flesh like areas and areas of multiple haemorrhages . (Fig 2,4).The kidney showed multiple glistening lobulated nodules involving the middle and lower pole with pushing of cortex at lower pole . The cortical thickness was 1.3 cm. (fig 3)



**Figure 1:** Image showing c/s of Kidney along with soft tissue mass cortical thickness is preserved with normal size of kidney



**Figure 2:** Image showing multiple glistening nodules of pararenal masses



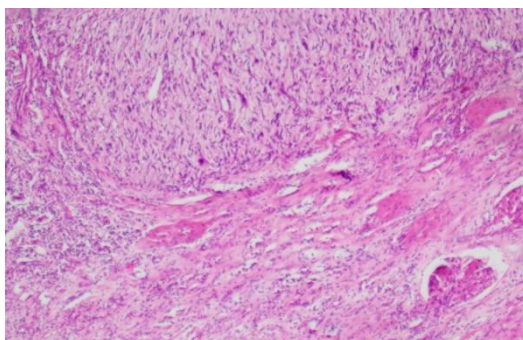
**Figure 3:** Image showing cut section of kidney with creamish white growth



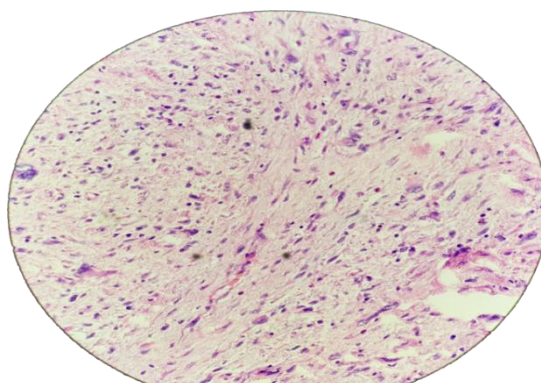
**Figure 4:** Image showing cut section of pararenal mass with fish flesh appearance

Microscopic findings: The renal mass showed tubules and glomeruli along with slender spindle shaped cells with serpentine nucleus with eosinophilic cytoplasm arranged as bundles and fascicles and lying in loose sheets along with fibroblasts and hyalinized collagenous stroma pushing the renal tissue at various foci.[Figure 5,6]. The tumor cells showed moderate to high nuclear pleomorphism, hyperchromasia with prominent macronucleoli.(figure 7)., Giant cells were also seen. In spite of high cellularity ,mitotic count was < 5/ 10 hpf. At places necrosis was evident but was less than 50 % of tumour tissue. At places shwannian stroma and non

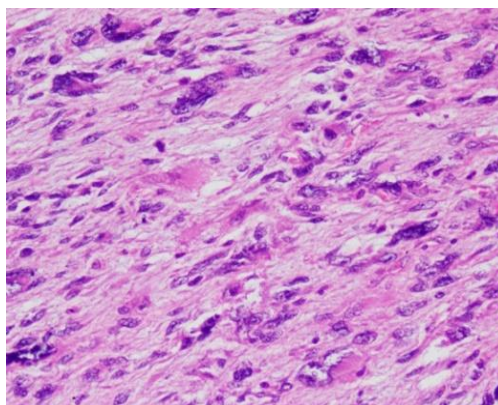
malignant neurofibroma tissue was also appreciated. Based on morphology, diagnosis of low grade malignant peripheral nerve sheath tumour was made seen arising in neurofibroma. IHC was done and tumour was found to be positive for S-100. The patient had no personal or familial history of neurofibromatosis.



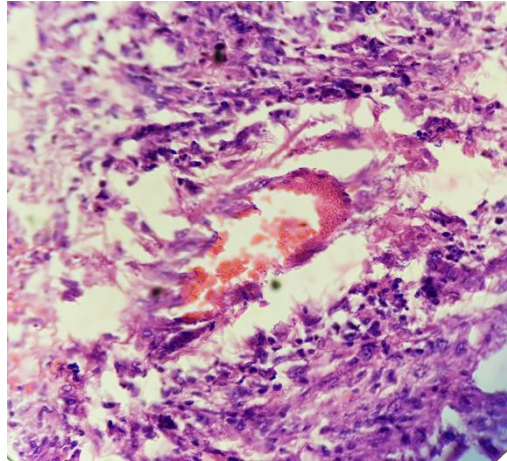
**Figure 5:** Image showing renal tissue in lower half and malignant neural cells in upper half(H&E,40 X)



**Figure 6:** Image Showing spindle shaped cells with serpentine nucleus arranged as sweeping fascicular pattern (H&E,40X)



**Figure 7:** Image showing tumor cells with moderate to high nuclear pleomorphism with vesicular nuclei. Bizarre cells are also seen



**Figure 8:** Image showing blood vessel lined by plump endothelial cells

#### IV. Discussion

MPNSTs are rare and highly aggressive soft-tissue sarcomas that are associated with neurofibromas in NF-1 patients but that also occur in association with radiation or sporadically. MPNSTs are sarcomas arising either *de novo* or in transition from pre-existing neurofibroma. Patient with NF-1 usually present with multiple rapidly enlarging masses or a new onset of pain associated with pre-existing plexiform neurofibromas, whereas the sporadic forms present with a new solitary, enlarging and painless mass.<sup>1</sup> The main recognisable benign precursor to MPNST is neurofibroma, in particular the plexiform type in the setting of NF1. The extremities are most common sites for MPNSTs, followed by the head and neck region and the trunk. Kidney involvement is uncommon, and it is very difficult to differentiate other kidney sarcomas from MPNSTs clinically or upon gross examination.<sup>5</sup> Thus, histopathological and immunohistochemical studies are needed for the diagnosis and grading of an MPNST.<sup>6</sup> Grossly, MPNSTs appear fusiform, have a large tumor size (>5 cm) and are gray-tan in color. Microscopically, these tumors exhibit features of spindle cells, with a fascicular pattern and varying degrees of mitosis, necrosis and tumor calcification. Associated benign neurofibroma or schwannian cells that have not undergone malignant transformation can be observed. If the histomorphological findings in a malignant soft tissue tumor consisting of spindle cells makes one think of MPNST, there should be at least one of the two following criteriorions in order to conclude a diagnosis of MPNST:-

1) Tumor should arise in a patient with NF1. 2) Tumour should appear to arise in a nerve or should develop in a previous neurofibroma<sup>8</sup>. Though several radiologic imaging methods are helpful for identifying some features of NF, histological examination and immunohistochemical staining provide definitive diagnosis.<sup>7</sup> MPNSTs stain positive for S-100 protein; neuron-specific enolase; and other proteins, such as actin, cytokeratin (CK), smooth muscle actin (SMA), desmin and vimentin, to differentiate from other spindle cell sarcomas.<sup>6</sup>

#### V. Conclusion

The MPNSTs can arise *de novo* from malignant transformation of benign nerve sheath tumors. The clinical course is usually short. Retroperitoneal involvement is uncommon, and it is very difficult to differentiate other kidney sarcomas from MPNSTs clinically or upon gross examination.<sup>6</sup> Thus, histopathological and immunohistochemical studies are needed for the diagnosis and grading of an MPNST.<sup>6</sup> Malignant nerve sheath tumors are very rare but aggressive neoplasms. As MPNSTs can arise from numerous cell types, its appearance can vary from one case to another. This makes diagnosis and classification quite difficult. The mainstay of treatment is surgical resection.

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