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Paraneoplastic Chronic Inflammatory Demyelinating Polyradiculoneuropathy Associated with Renal Cell Carcinoma – A Rare but Critical Diagnosis

M K Manu [a], K Ram Mohan [b], Mohan Amal [a*], C Lingesh Sairam [a], Harikrishnan AB [a] and SJ Aquil Faris [a]

[a] Department of Urology, Govt. Medical College, Thiruvananthapuram, India.
[b] Department of Medical Neurology, Govt. Medical College, Thiruvananthapuram, India.

Authors' contributions

This work was carried out in collaboration among all authors. Authors MKM and KRM conceptualized the study and analyzed the intellectual content. Author MA managed the literature searches, data acquisition and data analysis. Author CLS prepared the manuscript. Author HAB edited the manuscript. Author SJAF reviewed the manuscript. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) in the context of renal cell carcinoma (RCC) is a rare occurrence. A 73year old male presented with 6 months history of progressive weakness, was diagnosed as CIDP. His condition deteriorated despite treatment, leading to quadriplegia. Further evaluation with imaging revealed a left sided RCC. Following radical nephrectomy, the patient experienced substantial sensory and motor function improvement.

**Corresponding author: Email: amalmohan006@gmail.com;*

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This case highlights a rare paraneoplastic link between CIDP and RCC, emphasizing the importance of considering paraneoplastic syndromes in patients with atypical or treatment-resistant neurological conditions. Early recognition and management of the underlying malignancy can significantly improve neurological outcomes.

Keywords: Chronic inflammatory demyelinating polyradiculoneuropathy; renal cell carcinoma; paraneoplastic syndrome; Quadriplegia.

1. INTRODUCTION

Renal cell carcinoma (RCC) is known for its association with various paraneoplastic syndromes, though neurological manifestations are uncommon, constituting only 0.5-1% of cases [1]. These neurological complications can affect both the central and peripheral nervous systems, often through poorly understood mechanisms. Some cases are theorised to involve antibodies targeting crucial nervous system proteins [2].

We present a case of 73year old male presented with 6 months history of progressive weakness, which was initially diagnosed as chronic inflammatory demyelinating polyradiculoneuropathy. On further evaluation, a left sided enhancing renal mass was found suggesting an underplay of paraneoplastic syndrome relating both.

This case highlights the importance of considering paraneoplastic syndromes in the differential diagnoses of patients presenting with atypical neurological symptoms or those demonstrating resistance to standard neurological treatments. Early identification and effective management of the underlying malignancy can significantly influence prognosis and neurological outcomes.

2. CASE HISTORY

A 73year old male, known hypertensive, presented with a 6 months history of progressive weakness accompanied by paraesthesia in the feet and difficulty ambulating. The weakness evolved into buckling of both knees resulting in multiple falls. Concurrently he developed paraesthesia in the hands which gradually evolved into weakness and loss of grip strength. Neurological evaluation, including nerve conduction studies and nerve biopsy, indicated a diffuse sensorimotor demyelinating polyneuropathy. Subsequent treatment with immunosuppressive agents, including cyclophosphamide, azathioprine, and mycophenolate mofetil, yielded limited improvement. One month prior to admission, he experienced a significant decline, developing quadriparesis and becoming bedridden. He received five cycles of plasmapheresis with limited benefit. Physical examination revealed left sided ptosis with esotropia, bilateral lateral rectus palsy and bilateral lower motor neuron type facial nerve palsy. Motor examination revealed weakness in both upper and lower limbs, with decreased reflexes and sensory deficits in the left lower limb. Further investigation with CECT abdomen revealed a 3.4 cm partially exophytic moderately enhancing left renal mass, suggestive of RCC. Metastatic disease was ruled out by Fluorodeoxyglucose Positron Emission Tomography (FDG PET). Intra-op and immediate post-operative period were uneventful. In the postoperative period, patient demonstrated gradual improvement of neurological deficit. Histopathology report was clear cell carcinoma with 70% sarcomatoid differentiation (ISUP grade 4). Within two weeks of surgery, cranial nerve involvement improved and he regained motor function in both upper and lower limbs, with sensory improvement as well. At one month follow-up patient is walking without support. Physical examination showed grade 5 power in all four limbs.

Table 1. Upper Limb motor power before radical nephrectomy [Medical Research council Scale (MRC Scale)]

	Right	Left
Shoulder Flexion	2	2
Shoulder Extension	2	2
Elbow Flexion	3	3
Elbow Extension	2	2
Wrist Extension	0	0
Wrist Flexion	1	0
Hand Grip	Absent	Absent

3. DISCUSSION

Chronic Inflammatory Demyelinating Polyneuropathy is a rare heterogenous immune mediated neuropathy [3]. Often it is characterised by immune mediated demyelination and

remyelination of segments of nerves [4]. Incidence of CIDP in the backdrop of malignancy is a rare phenomenon [5]. Haematological disorders are the most common association of CIDP, particularly non-Hodgkin lymphoma, closely followed by malignant melanoma [6]. While RCC is known for its diverse paraneoplastic manifestations, neurological involvement, especially CIDP, is exceedingly rare, constituting only a small fraction of cases [7]. The presented case emphasizes the intricate relationship between RCC and paraneoplastic neurological syndromes, particularly CIDP.

Table 2. Lower Limb motor power before radical nephrectomy [Medical Research council scale (MRC Scale)]

	Right	Left
HIP Flexion	2	1
HIP Extension	2	0
Knee Flexion	2	1
Knee Extension	2	1
Ankle Dorsiflexion	0	0
Ankle Plantar Flexion	0	0
Plantar Reflex	Absent	Absent

Table 3. Upper Limb motor power 4 weeks after radical nephrectomy [Medical Research Council Scale (MRC Scale)]

	Right	Left
Shoulder Flexion	5	5
Shoulder Extension	5	5
Elbow Flexion	5	5
Elbow Extension	5	5
Wrist Extension	5	5
Wrist Flexion	5	5
Hand Grip	Good	Good

Table 4. Lower Limb motor power 4 weeks after radical nephrectomy [Medical Research council scale (MRC Scale)]

	Right	Left
HIP Flexion	5	5
HIP Extension	5	5
Knee Flexion	5	5
Knee Extension	5	5
Ankle Dorsiflexion	5	5
Ankle Plantar Flexion	5	5
Plantar Reflex	Flexor Response	Flexor Response

This patient's significant improvement following surgery strongly suggests a paraneoplastic aetiology for CIDP, even in the absence of specific antibody identification. This aligns with previous reports highlighting the potential for neurological symptoms, including CIDP, to precede the detection of RCC, emphasizing the importance of considering paraneoplastic syndromes in patients with unexplained neurological deficits [1,2,8].

The exact mechanisms underlying paraneoplastic neurological syndromes remain intangible. However, it is postulated that immune-mediated processes, possibly triggered by tumour antigens, play a crucial role [9,10]. In some cases, antibodies targeting neuronal proteins have been identified, but their absence in many reported cases including this one suggests a more complex pathogenesis.

4. CONCLUSION

In patients presenting with neurological deficits, paraneoplastic aetiology needs to be considered as the potential cause of Chronic inflammatory demyelinating polyradiculoneuropathy even in the absence of overt cancer symptoms or identifiable antibodies. Prompt recognition and treatment of the underlying malignancy offer the best chance for neurological improvement and overall patient well-being. Further research is needed to elucidate the precise mechanisms underlying paraneoplastic neurological syndromes and develop more targeted therapeutic approaches.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Authors hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

CONSENT

Patient was taken up for left radical nephrectomy after getting an informed consent.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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