Case report

Chronic inflammatory demyelinating polyradiculoneuropathy – A rare paraneoplastic syndrome of renal cell carcinoma

Abstract

Aims:

To identify the rare paraneoplastic presentation of renal cell carcinoma with chronic inflammatory demyelinating polyradiculoneuropathy

Presentation: A 73year old male presented with 6 months history of progressive weakness, was diagnosed as CIDP. His condition deteriorated despite usual immunosuppressive 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 and became ambulant

Discussion: Chronic inflammatory demyelinating polyradiculoneuropathy is commonly idiopathic in origin. However, it can have associations with malignancies- most common being non-Hodgkin's lymphoma and melanoma. Renal cell carcinoma is a very rare cause of paraneoplastic CIDP. 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.

Conclusion: Early recognition and management of the underlying malignancy can significantly improve neurological outcomes of patients with paraneoplastic aetiology.

Key-words

Chronic inflammatory demyelinating polyradiculoneuropathy, Renal cell carcinoma, Paraneoplastic syndrome, Quadriplegia

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 4year history of progressive weakness, which was initially diagnosed as chronic inflammatory demyelinating polyradiculoneuropathy.On further evaluation, a left sided enhancing renal mass was foundsuggesting 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.

Case History

A 73yearold male, known hypertensive, presented with a 4year history of progressive weaknessaccompanied byparaesthesiain the feetand difficultyambulating.The weaknessevolved into buckling of both knees resulting in multiple falls.Concurrently he developed paraesthesia in thehandswhich gradually evolved intoweakness 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 weaknessin 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). Patient was then taken up for left radical nephrectomy after getting an informed consent. 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 onemonth follow-up patient is walking without support. Physical examination showed grade 5 power in all four limbs.

Discussion

Incidence of CIDP in the backdrop ofmalignancy is a rarephenomenon.Haematological disordersare the most common association of CIDP ,particularly non-Hodgkin lymphoma, closely followed by malignant melanoma. While RCC is known for its diverse paraneoplastic manifestations, neurological involvement, especially CIDP, is exceedingly rare, constituting only a small fraction of cases.The presented case emphasizes the intricate relationship between RCC and paraneoplastic neurological syndromes, particularly CIDP.

This patient's significant improvement following surgery strongly suggests a paraneoplastic aetiology forCIDP, 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.[1,2,3]

The exact mechanisms underlying paraneoplastic neurological syndromes remainintangible. However, it is postulated that immune-mediated processes, possibly triggered by tumour antigens, play a crucial role.[4] In some cases, antibodies targeting neuronal proteins have been identified, but their absence in many reported casesincluding this one suggests a more complex pathogenesis.

In conclusion, even in the absence of overt cancer symptoms or identifiable antibodies, it is imperative to considerparaneoplastic CIDP when a patient presents with neurological

manifestationsas this can lead to early detection.This case further emphasizes the need to consider paraneoplastic aetiology in neurological cases even with classic presentation when they are not responding to standard treatment. A high index of suspicion is the crucial element in the right management of these patients .Prompt recognition and treatment of the underlying malignancyoffer 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.

References

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