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Case Report

A Case Report on Idiopathic Polyneuritis Cranialis Multiplex: A Rare Presentation of Guillian Barre Syndrome

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ABSTRACT

Polyneuritis Cranialis (PNC), a very infrequent disorder of multiple cranial nerve palsies without peripheral nerve involvement, is a variant of Guillain-Barré Syndrome (GBS). Facial weakness often combined with ocular signs such as ophthalmoplegia, ptosis, or papillary changes and bulbar signs like dysarthria or dysphagia, are shown by the majority of patients. It is a very uncommon disease that poses many challenges for early detection and management. Presented here is the case report of a 38-year-old male patient with recurrent headaches, a hallmark symptom of PNC, along with horizontal and vertical diplopia consistent with left 4th and 6th nerve paralysis.

Keywords: Polyneuritis cranialis, Guillian-Barre syndrome, Diplopia, Nerve palsies.

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INTRODUCTION

Polyneuritis cranialis multiplex is an uncommon disorder that progresses gradually and slowly involving multiple cranial nerve palsies. Even though it is a variant of Guillain-Barre syndrome, it is a very infrequent presentation that does not entail peripheral nerve damage or ataxia.[1-4] Polyneuritis cranialis typically affects the 4th, 5th, 6th and 7th cranial nerves.[2-5] It is frequently linked to idiopathic, immune-mediated or prior viral infections as the mechanism of the injury. The disease etiology is idiopathic although several conditions such as Guillian-Barre syndrome with its many variant forms found in adults and children may be associated with inflammatory causes,[1-6] infectious diseases such as diphtheria, Lyme disease, varicella zoster, botulism and Chagas disease, autoimmune disorders like systemic lupus erythematosus,[5-8] eosinophilic granulomatosis with polyangiitis,[3-7] myasthenia gravis, inflammatory bowel disease, Tolosa-Hunt syndrome[5-9] and sarcoidosis or toxins like arsenic, organophosphorus esters and lead, medications such as antibiotics including sulfonamide, chloramphenicol, metronidazole and isoniazid,[10] chemotherapeutic agents like vincristine, cytarabine, vitamin deficiencies typically vitamin B6 or B12[11] and ophthalmoplegic migraine[12] have been linked prior to polyneuritis cranialis. Due to the rarity of the disease, it's more difficult to characterise and the Incidence remains unknown.[1]

Facial weakness often combined with ocular signs involving ophthalmoplegia, ptosis or papillary changes and bulbar signs like dysarthria or dysphagia are shown by the majority of patients.[13] The pathological mechanism is believed to be due to occlusion of the vasa nervorum resulting in microinfarction of cranial nerves.[4] The differential diagnosis of PNC is broad and includes vascular, infectious, autoimmune and malignant etiologies[14] involving CSF findings, electrophysiologic features, serum levels and liver and renal function tests.[1] MRI with gadolinium may show postcontrast enhancement of cranial nerve roots. Management involves symptomatic relief, including corticosteroids to reduce swelling or inflammations and relieve pressure on the nerve (caused injury), using eye patches or glasses with prisms to reduce double vision and using painkillers and surgery to treat ptosis.

The present case report describes the case of an adult patient with this rare disorder of multiple palsies with horizontal and vertical diplopia consistent with left 4th and 6th cranial nerves.

CASE REPORT

A 38-year-old male patient was hospitalized in the neurology department with a complaint of vertical diplopia for one month. Currently not on any medications but uses an eye patches to avoid double vision. His illness started as a double vision in February last year. The first episode was preceded by a headache which improved quickly. It was followed by several relapses in March, April and May last year. The patient denies any fluctuation in double vision and improvement in sleep. The patient had a recurrence in July and September. The patient never had any ptosis, numbness, weakness of limbs or imbalance. After consultation

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with a local hospital, he was diagnosed with paralysis of the right lateral rectus. So as per old records, previous episodes involved the right rectus. In the present scenario left lateral rectus and left superior oblique muscles were involved. No other cranial nerves on either side were involved stating subacute cranial polyneuritis without any pachymeningitis. Family history and social history were unrevealing.

Neurological examination revealed convergent strabismus left eye, mild vertical disconjugation, left eye slightly elevated than the right. The patient has both horizontal and vertical diplopia consistent with left 4th and 6th nerve paralysis. No other signs, long tract involvement or systemic involvements were observed, neurologic findings were otherwise normal. His diagnostic work-up included Magnetic Resonance Imaging (MRI) to investigate the presence of hypertrophic pachymeningitis. However, the result yielded left nasal turbinate hypertrophy, DNS to the right, orbit and IOM were normal, partially empty sella and no meningeal thickening or parenchymal lesions, findings were otherwise normal.

The vasculitic panel was revealed to be normal (Table 1). Serology was negative, most probably due to several courses of immune modulators that the patient has received. The patient soon after admission started on Inj. Methylprednisolone at the dose of 1 g in 100 mL NS over 1 hr for 3 days, along with Tab. Azathioprine and Tab. Folic acid. The patient was told to consume tender coconut water once daily. Discussion with the patient regarding the condition and its management and emphasizing the need for oral medication to be continued for a minimum of 2 years. Further decisions will be made during routine follow-ups. Mild puffiness of the face, acne as well as altered blood sugar levels were anticipated, thus the patient was advised to avoid high-sugar foods while receiving Methylprednisolone.

Following discharge, methylprednisolone was intended to be administered for the following six weeks at a tapering dose until it was discontinued, along with Tab. Azathioprine and Tab. Folic acid. It was instructed to monitor a complete blood count, SGOT, SGPT and blood sugar levels after 1 month. If the Total count<3,000/cmm, platelet count <1,00,000 lakhs/cumm and SGOT>3 times elevated than normal range, then Tab. Azathioprine was advised to discontinue temporarily. Additionally, if blood sugar>200 mg/dL, Tab. Dapagliflozin at the dose of 10 mg can be administered once daily until Methylprednisolone is continued and blood sugar drops <200 mg/dL. The patient was advised to practice ocular exercises. He was discharged home with more than 50% improved clinical state.

Table 1: Test results of patient performed during hospitalization.

Test performed	Result
Peripheral smear test	Normal
AChR	Negative
Anti-MuSK antibody	Negative
TFT	Negative
ds DNA	Negative
pANC	Negative
cANC	Negative
RNS	No decrement

AChR: Acetylcholine receptor antibody; Anti-MuSK antibody: Anti-muscle-specific kinase antibody; TFT: Thyroid function test; dsDNA: Antidouble stranded deoxyribonucleic acid antibody; p-ANCA: Perinuclear Antineutrophil cytoplasmic antibody; c-ANCA: Cytoplasmic Antineutrophil cytoplasmic antibody; RNS: Repetitive nerve stimulation test.

DISCUSSION

Polyneuritis cranialis though considered a variant of Guillian-Barre syndrome is a very rare manifestation that does not cause ataxia or involvement of peripheral nerve damage. Torres *et al.* suggested that one of the main clinical characteristics that set apart idiopathic cranial neuropathy from GBS is the preservation of tendon reflexes.[3-6] The deep tendon reflex in both upper and lower limbs was normal in this patient. Despite the typical manifestations of PNC like facial weakness, ocular signs like ophthalmoplegia, ptosis or papillary changes and bulbar signs like dysarthria or dysphagia[4] the patient in this case report was presented with only horizontal and vertical diplopia consistent with 4th and 6th left nerve paralysis along with relapsing headaches. Lee *et al.* suggested that one of the hallmark symptoms of PNC, in addition to several cranial neuropathies, is a headache. A patient who presents with sudden symptoms of diplopia should be considered a clinical manifestation of Myasthenia gravis, an anti-acetylcholine receptor antibody test was performed and the result yielded negative. Dosunmu *et al.* revealed that the majority of isolated 4th nerve palsies including those that manifest in adulthood are congenital.[15]

Effective treatment for multiple cranial neuropathy palsies necessitates a precise diagnosis and targeted therapy directed towards the cause.[3] Management challenges exist when the cause is unclear, therefore precise recommendations cannot be made. Empiric therapy with corticosteroids appears to be a sensible management strategy.[1] The patient in this case report has begun the treatment with intravenous Methylprednisolone for 3 days which showed improvement in his clinical state. Performing routine laboratory tests as recommended would help to achieve better therapeutic outcomes and avoid any risks.

CONCLUSION

Early diagnosis and symptomatic treatments help in the effective management and prevention of further complications of polyneuritis cranialis. A multidisciplinary approach is usually considered essential for this rare condition to avoid unnecessary testing, provide a more effective diagnosis and reduce disease burden for the patient. Appropriate counselling about the

condition and its treatment contributes to the betterment of the patient.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ETHICAL APPROVAL

The study was approved by the institutional ethics committee.

ABBREVIATIONS

PNC: Polyneuritis cranialis; **GBS:** Guillian-Barre syndrome; **CSF:** Cerebrospinal fluid; **MRI:** Magnetic Resonance Imaging; **DNS:** Deviated Nasal Septum; **IOM:** Interosseous membrane **AChR:** Acetylcholine receptor antibody; **Anti-MuSK:** Anti-Muscle-Specific Kinase; **TFT:** Thyroid Function Test ds; **DNA:** Antidouble-stranded deoxyribonucleic acid; **pANCA:** perinuclear-Antineutrophil cytoplasmic antibody; **cANCA:** cytoplasmic-Antineutrophil cytoplasmic; **RNS:** Repetitive nerve stimulation test; **SGOT:** Serum Glutamic Oxaloacetic Transaminase; **SGPT:** Serum Glutamic Pyruvic Transaminase.

SUMMARY

A case report of a 38-year-old male with recurrent headaches and diplopia due to left 4th and 6th nerve paralysis is presented. The patient's diagnostic work-up included an MRI, which showed left nasal turbinate hypertrophy and partially empty sella but no meningeal thickening or parenchymal lesions. His test results for various antibodies and function tests were normal. His family history and social history were unrevealing.

The patient was finally diagnosed with idiopathic polyneuritis cranialis multiplex, which is a rare disorder, characterised by multiple cranial nerve palsies without peripheral nerve involvement or ataxia and is considered a variant of Guillain-Barré syndrome. It typically affects the 4th, 5th, 6th and 7th cranial nerves and is often associated with idiopathic, immune-mediated, or prior viral infections. The etiology remains idiopathic but can be linked to various inflammatory, infectious, autoimmune and toxic conditions and was treated with intravenous methylprednisolone, oral azathioprine and folic acid. Post-discharge, the treatment continued with a tapering dose of methylprednisolone and monitoring of blood counts and sugar levels to prevent any complications associated. The patient showed more than 50% improvement and was advised to do ocular exercises.

Early diagnosis and symptomatic treatment are crucial for managing polyneuritis cranialis. A multidisciplinary approach and appropriate counselling are essential to avoid unnecessary testing, provide effective diagnosis and reduce the disease burden on the patient.

REFERENCES

1. Torres AR, Salvador C, Mora M, Mirchandani S, Chavez W. Idiopathic Recurrent Polyneuritis Cranialis: A Rare Entity. Cureus. 2019.
2. Lee SS, Kwon S. Headache associated with Polyneuritis cranialis. Neurology Asia. 2018;23(2):189-91.
3. Carroll C, Campbell W. Multiple Cranial Neuropathies. Seminars in Neurology. 2009;29(1):53-65.
4. Tecellioglu M, Kamisli S, Erbay MF, Kamisli O, Ozcan C. A Rare Presentation of Cranial Polyneuropathy Without Rash Caused by *Varicella zoster* Virus. Medical Archives (Sarajevo, Bosnia and Herzegovina) [Internet]. 2017 Aug 1 [cited 2023 Oct 22];71(4): 293-5. Available from: https://pubmed.ncbi.nlm.nih.gov/28974853/
5. Keane JR. Multiple Cranial Nerve Palsies. Archives of Neurology. 2005;62(11):1714.
6. Ryan MM. Guillain-Barre syndrome in childhood. 2005;41(5-6):237-41.
7. Guillevin L, Cohen P, Gayraud M, Lhote F, Jarrousse B, Casassus P. Churg-Strauss Syndrome Clinical Study and Long-Term Follow-Up of 96 Patients. Medicine. 1999;78(1):26-37.
8. Omdal R, Løseth S, Torbergsen T, Koldingsnes W, Husby G, Mellgren SI. Peripheral neuropathy in systemic lupus erythematosus-a longitudinal study. Acta Neurologica Scandinavica. 2001;103(6):386-91.
9. Kline LB. NOSOLOGICAL ENTITIES?: The Tolosa-Hunt syndrome. Journal of Neurology, Neurosurgery and Psychiatry. 2001;71(5):577-82.
10. SNAVELY SR. The Neurotoxicity of Antibacterial Agents. Annals of Internal Medicine. 1984;101(1):92.
11. Graham SM, Arvela OM, Wise GA. Long-term neurologic consequences of nutritional vitamin B12 deficiency in infants. The Journal of Pediatrics. 1992;121(5):710-4.
12. Gelfand AA, Gelfand JM, Prabakhar P, Goadsby PJ. Ophthalmoplegic "Migraine" or Recurrent Ophthalmoplegic Cranial Neuropathy. Journal of Child Neurology [Internet]. 2012 Jan 12 [cited 2019 Apr 3];27(6): 759-66. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3562350/
13. Steele JC, Vasuvat A. Recurrent multiple cranial nerve palsies: a distinctive syndrome of cranial polyneuropathy. Journal of Neurology, Neurosurgery and Psychiatry. 1970;33(6):828-32.
14. Bailey Gleason Fitzgerald, Grant M, Kone GJ, Patwa HS, Rose M. Multiple Cranial Neuropathies as the Presenting Sign in a Patient with Metastatic BRAF-Mutated Lung Adenocarcinoma with Leptomeningeal Involvement. Case reports in oncology. 2020;13(3):1258-62.

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