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A CASE REPORT ON POLYNEURITIS CRANIALIS MULTIPLEX A RARE PRESENTATION OF GUILLIAN BARRE SYNDROME

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ABSTRACT

Polyneuritis cranialis (PNC), a sporadic 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 involving ophthalmoplegia, ptosis or papillary changes and bulbar signs like dysarthria or dysphagia are shown by the majority of patients. An uncommon disease that poses a lot of challenges in 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.

INTRODUCTION

Polyneuritis cranialis multiplex is a very 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 rare presentation that does not entail peripheral nerve damage or ataxia.[1,2,4] Polyneuritis cranialis typically affects the 4th, 5th, 6th and 7th cranial nerves.[2,4,5] It is frequently linked to idiopathic, immune-mediated or prior viral infections as the mechanism of 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[1,5,6], 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, lead, medications such as antibiotics including sulfonamide, chloramphenicol, metronidazole and isoniazid[10], chemotherapeutic agents like vincristine, cytarabine, vitamin deficiencies typically vitamin B6 or B12[12] and ophthalmoplegic migraine[13] 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.[14] 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[15] involving CSF findings and 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 which includes the use of 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, use of 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 4^{th} and 6^{th} nerve paralysis.

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 patch to avoid double vision. His illness started as double vision on 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 a consultation 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 4^{th} and 6^{th} 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.

TEST PERFORMED	RESULT
Peripheral smear	Normal
AChR	Negative
Anti-MuSK antibody	Negative
TFT	Normal
ds DNA	Negative
pANCA	Negative
cANCA	Negative
RNS	No decrement

Test results of patient performed during hospitalization

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.

Thus vasculitic panel was revealed to be normal. 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 1g in 100ml NS over 1 hour 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 complete blood count, SGOT, SGPT and blood sugar levels after 1 month. If total count <3,000 cells/Cmm, platelet count <1,00,000 lakhs/cumm, 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 till Methylprednisolone is continued and blood sugar becomes < 200 mg/dL. The patient was advised to do ocular exercises. He was discharged home with more than 50% improved clinical state.

DISCUSSION

Polyneuritis cranialis though considered a variant of Gullian-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 sets apart idiopathic cranial neuropathy from GBS is the preservation of tendon reflexes.[3,6] The deep tendon reflex in both upper and lower limbs were 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 left 4^{th} and 6^{th} nerve paralysis and relapsing headaches. *Lee et al* suggested that one other hallmark of PNC, in addition to several cranial neuropathies, should be a headache. A patient who presents with sudden symptoms of diplopia should be considered a clinical manifestation of Myasthenia gravis, anti-acetylcholine receptor antibody performed yielded negative result. *Dosunmu et al* revealed that the majority of isolated 4^{th} nerve palsies including those that manifest in adulthood are congenital.[16]

The effective treatment of multiple cranial neuropathy palsies necessitates a precise diagnosis and targeted therapy directed towards the cause.[3] Management challenges exist when the cause is unclear and precise suggestions 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 IV Methylprednisolone for 3 days which showed improvement in his clinical state. Perform routine laboratory tests as recommended 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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