

Idiopathic, Isolated, Recurrent, Ipsilateral Sixth Nerve Paresis in An Older Adult - A Rare Case Report

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Abstract

Recurrent, unilateral, isolated, idiopathic lateral rectus muscle paresis is an uncommon but well-recognized disorder in children but has not been recognized as well in other age groups. Here we are reporting a case of idiopathic, isolated, recurrent, ipsilateral sixth nerve paresis in an otherwise healthy older adult. A 46 years gentle man presented with double vision since 3 years. He had similar episodes of double vision 5 times all of which recovered completely, except the last time. On examination there was right esotropia, limitation of right abduction and a normal fundus. Diplopia and Hess charting verified partial 6th nerve paresis. The patient was investigated with a wide range of investigations to rule out tumors, demyelinating, infectious, restrictive and neuromuscular junctional causes all of which were normal. Patient underwent surgical correction of esotropia and he is asymptomatic till date and is under a close follow up.

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Introduction

Cranial nerve 6 palsy, also known as Abducens nerve palsy or Lateral rectus palsy is a relatively rare disorder in adults. There are numerous causes of cranial nerve 6 palsy, with most common being trauma, inflammation, tumour & vasculopathic disease. Recovery can occur spontaneously within weeks to several months & occurs most frequently in patients whose palsies have an unknown/vascular etiology. Recurrent, unilateral, isolated, idiopathic lateral rectus muscle paresis is an uncommon but well-recognized disorder in children but has not been recognized as well in other age group.¹ Here we are reporting a case of idiopathic, isolated, recurrent, ipsilateral sixth nerve paresis in an otherwise healthy older adult.

Case Report

A 46-years gentle man presented to our hospital with the complaint of double vision from past 3 years in primary position and more on looking towards the right lateral gaze. The diplopia disappeared on closing one eye. There were no associated other complaints like redness of eyes, proptosis, blurring of vision, fever, headache or vomiting. He did not have any other co morbidity. Previously, he had 5 similar episodes double vision all of which recovered completely except the last one which still persists since 2015 (Table 1).

Table 1: Details of each episode Time of presentation, duration, treatment given and recovered or not

Epi-sode	Year	Treatment & Duration	Recovery
1	2004	Tablet Diclofenac & Tablet Paracetamol Tablet multi vitamins - 1month.	Complete
2	2006	Oral prednisolone 40mg- gradually tapered.	Complete
3	2009	Tablet multi-vitamins- 1 month.	Complete
4	2011	No medication.	Complete
5	2015	Intra Venous methyl prednisolone 1 mg/kg followed by oral steroids for 8 months.	Partial

When the patient had 5th episode (last episode) in 2015, he was investigated thoroughly and all reports were almost normal. The following investigations were done- Complete haemogram showed a normal differential cell count and slightly elevated erythrocyte sedimentation rate (ESR; 15 mm/h), C-Reactive Protein was 8.2mg/dl. Angiotensin converting enzyme levels were normal. Rheumatoid Factor was negative. The renal, hepatic, and thyroid function tests were normal. Plasma glycated haemoglobin was normal. Lipid profile was normal. Anti-nuclear antibodies were negative. Serological testing for human immunodeficiency virus (HIV), Venereal disease laboratory research test and hepatitis B and C were negative.

Magnetic resonance images (MRI) of the brain with gadolinium enhancement did not reveal abducens nerve or meningeal enhancement. But MRI showed Empty -sella. Magnetic resonance Venography (MRV) normal. Electro Myography and Nerve conduction studies were normal.

In 2018 patient presented to our hospital with complaint of double vision. On examination, patients Best Corrected Visual Acuity (BCVA) was 6/6 & near vision N6 in both eyes. Extra ocular movements showed restriction of abduction of -3 in right eye (Figure 1) Stereo acuity was >400 sec of Arc. Worth's four dot test showed diplopia. On Prism Bar Cover Test (PBCT) there was 25 Prism Diopter (PD) & 20PD of esotropia for distance and near respectively. Secondary deviation (50PD) was greater than primary deviation (25PD). Neurological examination was normal. Anterior segment and Fundus examination was normal. Forced Duction Test (FDT) was positive for medial rectus of right eye suggestive of long standing right lateral rectus paresis. Diplopia charting and Hess charting verified partial right lateral rectus paresis. The patient was sent for CT angiogram of circle of willis and visual fields both of which were normal. From these, a clinical diagnosis of isolated, idiopathic, recurrent, partially recovered 6th nerve paresis of right eye was made.

As the patient is suffering from diplopia since 2015 surgical

correction of esotropia was planned and patient underwent medial Rectus Recession of 5mm of right eye. After 1week, postoperatively there is no symptomatic diplopia in primary position. Patient was orthotropic for both distance and near (Figure 2). But the right eye abduction deficit (-1) & diplopia in lateral gaze persisted. +1.50D glasses were prescribed for the patient. Till date patient is asymptomatic and satisfied even after one year of follow up as there is no diplopia in primary position (last follow up was in July 2019).



Figure 1: Pre operatively right eye esotropia in primary position and right abduction deficit due to paresis of right lateral rectus



Figure 2: Post operatively Orthotropia in primary position and improvement in right abduction deficit due to paresis of right lateral rectus

Discussion

Cranial 6th nerve palsy is a disorder that can be acquired in childhood and through-out life. The literature on recurrent isolated lateral rectus muscle palsy is mostly paediatric. However recurrent, isolated lateral rectus palsy has also been reported in adult patients where a variety of causes need to be considered like neuromuscular disorders like myasthenia, muscle restrictive disorders like thyroid eye disease, meningeal infiltrative and metastatic process like leukemia and brainstem gliosis and demyelination disorders like multiplesclerosis.¹

The following Table 2 gives previously reported studies on recurrent sixth nerve paresis in adults.

Our patient is a young healthy adult with 5 episodes of recurrent 6th nerve paresis, of which he recovered completely in 4 episodes except the last one.

Afifi et. al. and associates said, once the 6th nerve has been damaged by trauma/infection, the threshold to further damage is lowered which might be a reason for non recovery in last episode of 6th nerve palsy in our patient.¹³

Our patient was investigated thoroughly and completely to rule out infectious, neuromuscular junction, restrictive and demyelinating disorders and reports of all the investigations were normal. Hence we termed the patient as idiopathic.

The exact pathophysiologic mechanism of relapsing and remitting course of 6th nerve palsy remain unclear. Many hypothesis have been proposed none of which are conclusively proved, a few of which include –

- (a) Thickening and hyalinastion of nutrient vessels which lead to focal ischemic demyelination followed by remyelination and clinical recovery.¹⁴

Table 2

Authors Published Year [Reference #]	No. of patients	Duration of episodes of diplopia	Interval between episodes	No. of episodes	Etiology No. of Cases (%)
G. J. Hankey [1990] ²	1	Several days-4 Months	1-2 Years	5	Benign
Jane W Chan et.al.[2015] ³	7	2 Weeks - 3 Months	5 Months - 2 Years	-	4 (57%) - Tumor located in the parasellar or petrous apex cavernous sinus regions. 1 (14.29%) - Recurrent painful ophthalmoplegic neuropathy (Migraine) 1 (14.29%) - intracavernous carotid artery aneurysm 1 (14.29%) -ischemic mononeuropathy
DQ Nguyen et.al.[2006] ⁴	1	6 Weeks	3-18 Months	3	intracavernous carotid aneurysm
Mahale R et.al. [2016] ⁵	2	2 Days	2 Months 2-4 Years	2 3	1- Primary Systemic Sclerosis (SS) 1- Idiopathic Intra cranial Hypertension (IIH)
Blumenthal EZ et.al.[1997] ⁶	1	2-5 Weeks	-	7	Dolichoectasia of the Cavernous Internal Carotid Artery
Gupta N et.al.[2010] ⁷	1	2 Months	-	2	Sphenoiditis
Golnik KC et.al.[1992] ⁸	1	4 Months	-	2	Familial recurrent cranial nerve palsy
Vasconcelos LP et.al.[2008] ⁹	1	2 Weeks	1-2 Years	4	Ophthalmoplegic migraine
Volpe NJ et.al.[1993] ¹⁰	7	1-18 Months	-	-	skull base tumors
K A Sandvand et.al.[2008] ¹¹	1	3 Months	7 Months	11	neurovascular compression.
SR. Hamilton et.al.[1991] ¹²	5	Within 6 Months	Several Years	-	Benign

- (b) Intermittent compression by a structural lesion could create a cycle of demyelination followed by remyelination of 6th nerve.⁶
- (c) Direct compression of arterial supply of 6th nerve which can lead to ischemia followed by aberrant regeneration of 6th nerve to innervate 6th nerve.⁴

Our patient was investigated to rule out any structural lesion like tumors/aneurysm, but all the reports are normal.

In patients with unresolved 6th nerve palsy strabismus surgery with Medial rectus recession+/-transposition is a well established surgical procedure to correct residual esotropia.¹⁵ Our patient underwent Medial Rectus Recession, since only 25 PD of esotropia was present and patient is asymptomatic with good sensory fusion till date. In our patient till now no particular cause has been identified, but we are doing a close follow up.

Conclusion

Though isolated 6th nerve palsy is the most common oculomotor nerve palsy, if it recurs should be investigated thoroughly because the proposed causes carry high risk of morbidity and mortality. But even then sometimes cause may not be identified. In such case a close follow-up is deemed necessary.

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