Acute isolated medial rectus palsy due to infarction as a result of hypercoagulable state: A case report and literature review

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Isolated medial rectus palsy in an otherwise healthy individual is a very rare entity. However, this may point towards underlying systemic pathology. This is a case report of an otherwise healthy young adult male who presented with sudden onset non-progressive blurring of vision in right eye. A series of investigations were performed and the patient was diagnosed to have a rheumatological disorder. Isolated muscle palsies in young patient may be masking a systemic disorder and needs to be evaluated thoroughly.

Keywords: Isolated medial rectus palsy, mid-brain infarct, pupil sparing third nerve palsy, strabismus

Any kind of extraocular muscle palsy can be a sign of neurological illness which in turn is commonly associated with systemic illness like diabetes mellitus, hypertension, hypercholesterolemia, cardiac illness etc.[1,2]

Isolated medial rectus palsy is a rare entity all the more in a younger patient which needs detailed systemic evaluation to point out the systemic etiology. To the best of our knowledge very few cases of isolated medial rectus palsy have been reported.[1,3-6] Most of the cases that were reported either had other signs of third nerve palsy like ptosis or mydriasis (anisocoria) or neurological signs suggestive of some central nervous system pathology. We report a case of a healthy young adult male who presented with sudden onset blurred vision in right eye and was diagnosed to have isolated medial rectus palsy. Detailed systemic evaluation lead to the diagnosis of hypercoagulable state due to rheumatological disorder. Our case was unique in terms of presenting symptoms, absence of other neurological signs and a documented normal previous neuroimaging. Rabadi *et al*.[6] reported a case of left medial rectus palsy with left partial ptosis as a consequence of midbrain infarction.

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Case Report

A 22-year-old otherwise healthy male presented in OPD with complaints of sudden painless loss of vision in right eye for distance since one day, which was non-progressive and experienced on waking up in the morning. On enquiry, it was reported to be associated with diplopia in left lateral gaze. There was no other relevant ocular history. He also complained of episodic headache mild to moderate grade since 8 days that was intermittent. For these complaints, he consulted elsewhere and was advised MRI brain – which was reported to be within normal limits. The case scenario was complicated due to presence of a previously reported normal MRI brain, but was found to have diagnostic features on repeat MRI scan.

On ocular evaluation, the findings were as follows: Unaided visual acuity was 20/30 in both eyes measured with Snellen's distance vision chart. Best corrected visual acuity was 20/20 in both eyes with -0.50 DS refractive correction in right eye and -0.75 DS refractive correction in left eye. Slit lamp evaluation revealed mild congestion in right eye. On extra-ocular movements examination, there was limitation of adduction in right eye [Fig. 1]. Orthoptic examination shows Right eye exotropia of 5 degree on Hirschberg corneal reflex test and 15 PD RXT on Krimsky test. Ocular motility showed limitation on adduction in right eye (-2), overaction in abduction in left eye (+1). Ocular motility in rest cardinal position appears full and is painless. There was no saccades on adduction in right eye. Forced duction test was negative. Diplopia charting with red-green goggles [Fig. 2] was done which showed maximum separation of crossed images on left lateral gaze suggestive of horizontal rectus muscle palsy (Fig. 1: Photograph of the patient; Fig. 2 Diplopia Charting of the patient).

Remainder of ocular exam was within normal limits. The rest of the cranial nerves examination revealed no deficit. No other signs of any neurological deficit was noted on systemic examination. A provisional diagnosis of medial rectus palsy in right eye (paralytic exotropia) was made. Differential diagnosis considered were viral Infections, orbital Myositis, orbital cysticercosis,[7] partial or complete third nerve palsy (pupil sparing): posterior communicating artery aneurysm, internal carotid artery aneurysm, or idiopathic, infarcts in the mid-brain involving lateral subnuclei of the midbrain. The patient was then referred to the Department of Neuromedicine, where a Radiology consultation was then sought.

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Neurology evaluation revealed no other abnormalities in central as well as peripheral nervous system. A repeat MRI brain was advised. Repeat MRI brain scan revealed small foci of diffusion restriction and FLAIR weighted hyper-intensities in the mid brain in the median and right paramedian region anterior to the aqueduct the largest of which measures 6.4 × 1 mm in the right paramedian location [Table 1 and Fig. 3].[8]

Diagnosis

Isolated medial rectus palsy of right eye due to mid-brain infarction involving the medial rectus nucleus of third cranial nerve nucleus as a result of hypercoagulable state due to autoimmune disorder. A differential diagnosis of Systemic lupus erythematosus, Anti Phospholipid Antibody syndrome, Scleroderma based on the laboratory investigations of ANA, LA, Antithrombin.

Management

Patient was initially managed with oral steroids in the form of tablet methylprednisolone 8 mg twice daily as a treatment for idiopathic or viral infection associated palsy[9,10] which was then tapered of as 4 mg twice daily for 1 week and 4 mg once daily for 1 week. Patient was managed for his hypercoagulable state with oral anti-platelet acetyl salicylic acid 75 mg once daily. Vitamin D and calcium supplements were prescribed to prevent untoward side effects of corticosteroids like vitamin D deficiency and bone damage.

Patient has best corrected visual acuity of 20/20 and extraocular movements full in all directions at 3 weeks' follow-up.

Table 1: Following are the results of investigations

Test	Results	
Hb	13.5 g/dl	
TLC	5740	
DLC	N 57 L 30 M 11 B 2	
PLT	3,56,000	
HsCRP	21.96	
RBS	75 mg/dL	
ESR	57 mm @ 1 h	
VDRL, HIV, HbsAg, HCV	Negative	
Mantoux	12 mm at 72 h	
KFT	BUN 27 Creat 0.42	
ANA	Positive	
LA		
LA1 screening	Patient: 53.2 sec	Control: 46.0 sec
LA2 confirmatory	Patient: 35.7 sec	Control: 36.8 sec
Antithrombin	138.7%	
Protein C and S	C: 133.7% S: 119.6%	
Activated protein C resistance	PCAT: 148.8 seconds PCAT/O: 73.2 sec Normalized ratio: 0.99	

Hb=Haemoglobin; TLC=Total leucocyte count; DLC=Differential leucocyte count; N=Neutrophils L=Lymphocytes M=Monocytes B=Basophills PLT=Platelet count; HsCRP=High Sensitivity C-Reactive Protein; RBS=Random Blood Sugar; ESR=Erythrocyte sedimentation rate; VDRL=Veneral disease research laboratory; HIV=Anti Human immunodeficiency virus antibody; HbsAg=Hepatitis B Antigen; HCV=Anti Hepatitis C Virus Antibody; KFT=Kidney Function Test; ANA=Anti-neutrophilic antibody; LA=Lupus Anticoagulant

Discussion

Isolated medial rectus palsy in an otherwise normal young patient is a rare entity. Pubmed search revealed only two[5,11] more cases being reported earlier in young adults. Although isolated medial rectus palsy has been reported in elderly patient with some or the other systemic illness, such a case in a young patient is very rare. Two important issues are associated with such cases. Firstly, a timely identification of the subtle finding, and secondly, a thorough knowledge of the systemic etiology.

Patients presenting complaint was blurring of vision with best corrected visual acuity of 20/20. On further enquiry, patient complained to have binocular diplopia during left gaze which subsided on occlusion of the affected eye that is the right eye. This proved that the diplopia was due to mal-alignment of the visual axis. In the absence of any known systemic illness, it was difficult to point out the etiology. To add into this dilemma, MRI brain done elsewhere was reported to be normal which

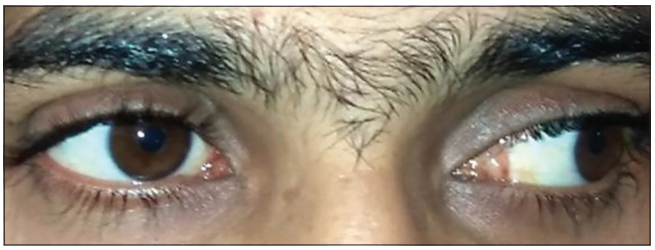

Figure 1: Photograph in levoversion

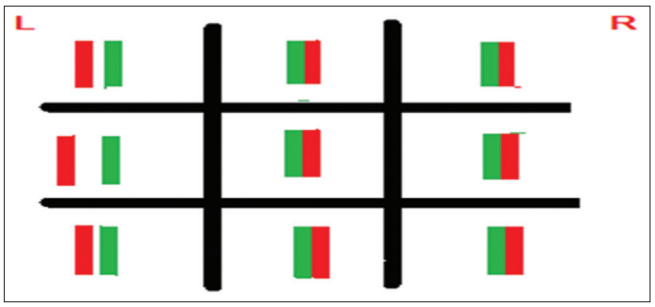

Figure 2: Diplopia charting of the patient

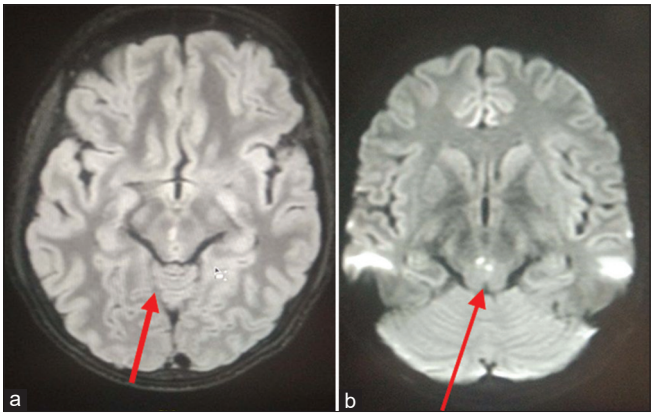

Figure 3: (a and b) MRI (Diffusion weighted imaging) images showing hyperintense foci in the midbrain suggestive of infarcts (Arrows pointing towards the pathology)

might be due to wider slice setting or a different machine. Repeat MRI revealed infarction in mid brain region. On the basis of neuroimaging following differential diagnosis could be made viz. isolated medial rectus palsy and internuclear ophthalmoplegia. Absence of nystagmus in other eye ruled out the diagnosis of internuclear ophthalmoplegia. Systemic evaluation was positive for ANA, Antithrombin and lupus anticoagulant which was pointing towards a hypercoagulable state. Clinically similar cases were reported by various authors[1,3,6,12] but in various age group and known systemic illness. Common thing was the neuroimaging in all these cases revealed infarction in mid-brain region.

Out of the cases reported, 2 were young adults one being male and the other female. Our case was different from Arif *et al*.[2] in a way that the presenting complaint was diplopia and ptosis in a young female. In the case reported previously, ocular examination revealed that there was complete ptosis, moderate hypotropia, large exotropia and no supraduction, infraduction or adduction which lead to the diagnosis of pupil sparing complete third nerve palsy. Neurological examination in that case revealed weakness of seventh cranial nerve. On investigation a patent foramen ovale was evident which was then treated surgically. Second case in a young adult male was reported by Simerpreet Bal *et al*.[5] in which the presenting complaint was giddiness followed by diplopia. This case was similar to ours in terms of ophthalmological evaluation which revealed medial rectus palsy. On investigation, it was found that patient suffered from dyslipidemia which was the reason for infarct.

Pupil sparing cranial nerve three palsy has been very well described[3] due to infarction in the mid-brain. Isolated muscle palsies are usually associated with the lesion in the orbit or the extra-ocular muscle itself. Reason for isolated involvement of medial rectus in our case is the strategic location of infarction in the ventral part of the cranial nerve three nucleus which comprises of fibres supplying ipsilateral medial rectus. Since there are a few more cases reported similar to ours, it prompts us to investigate regarding anatomical predilection for infarction in the ventral part of the cranial nerve three nucleus. This case is also emphasizes to the fact that diligence during evaluation of patients in OPD is of utmost importance.

Management of such cases is a question of debate. The role anti-platelet drugs in such cases is mainly for the prevention of further similar episodes. Recovery in extraocular movement occurs as a result of reactivation of the hibernating fibres. Prescription of calcium and vitamin D supplements in a young adult along with low dose oral corticosteroids might not be approved by some clinicians. However, some clinicians prefer to start patient on these supplements to as a preventive measure against the reported avascular necrosis of head of femur after low dose oral corticosteroids.[13]

Isolated palsy of extraocular muscle in a young patient should be evaluated thoroughly as these may point towards underlying systemic etiology commonly associated with central nervous system pathology.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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