

Visual rehabilitation following cerebellar stroke

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Abstract

Aim: To present a small case series of patients with visual complications following primary acute cerebellar stroke, document the types of visual deficit that occur, examine recovery patterns and identify rehabilitation strategies available.

Methods: A small, consecutive case series of patients referred to the Orthoptic Stroke Service at Sheffield Teaching Hospitals NHS Foundation Trust with primary acute cerebellar stroke in 2007–2008 is presented. The types of visual deficit that occurred and the orthoptist's contribution to patient rehabilitation are outlined.

Results: Seven patients were included in this series. All had manifest nystagmus with or without oscillopsia. The main complaint was of 'blurred vision'. Nystagmus varied in different positions of gaze, down gaze being the most troublesome for rehabilitation. Other deficits identified were skew deviation, internuclear ophthalmoplegia, and fixation abnormalities including saccadic intrusions and square wave jerks. Recovery was often incomplete, although in most cases some functional improvement did occur and most often in primary position.

Conclusions: Patients who have suffered cerebellar stroke, especially those with vague visual symptoms and those failing to respond to rehabilitation, should be referred for orthoptic assessment. This provides invaluable information that can explain difficulties with rehabilitation and give patients insight into their symptoms. Advice and therapy can help overcome problems in many cases.

Key words: Cerebellar stroke, Diplopia, Nystagmus, Square wave jerks

Introduction

In eye movement control, the cerebellum is responsible for image stabilisation and ensures smooth tracking of moving targets, maintains stable images on the retina during head motion and controls the amplitude of saccades or fast eye movements.¹

Isolated cerebellar stroke, from either infarct or haemorrhage, is comparatively rare, accounting for less

than 3% of strokes;² however, in a selected group of patients who were referred for orthoptic assessment this increased to 11.4%.³ Cerebellar stroke has a pronounced effect on visual function. Visual deficits are often missed as they can be subtle, often without obvious strabismus. There are generally more pronounced symptoms such as headache, ataxia, vertigo and vomiting⁴ that frequently occur following cerebellar stroke and in many cases patients have difficulty in defining or describing their symptoms.

Methods

All patients referred to the Orthoptic Stroke Service at Sheffield Teaching Hospitals NHS Foundation Trust with primary acute cerebellar stroke in 2007–2008 were included. Referral was made by the occupational therapy team on either the acute or rehabilitation stroke ward. All patients underwent full orthoptic assessment including visual fields to confrontation and assessment of visual inattention. Follow-up was for a minimum of 18 months in those patients whose symptoms persisted, or until signs and symptoms resolved. A small consecutive case series of patients is presented.

Results

Seven patients are included in this case series (Table 1). All patients had acute cerebellar infarcts of either the anterior or inferior cerebellar arteries, or cerebellar haematoma. In one case the lesion was not confined to the cerebellum but was more extensive and included a medullary infarct; in this case visual deficits were more complex. The average age of patient in this case series was 52 years (range 45–81 years) and average time from stroke onset to orthoptic assessment was 25 days (range 15–40 days) (see Table 1). The main complaint or reason for referral was 'blurred vision' rather than specific visual deficits.

All patients had some form of manifest nystagmus with oscillopsia. Nystagmus and/or fixation instability was present in primary position in all patients and nystagmus varied in different positions of gaze, down gaze being the most troublesome for rehabilitation, with activities such as reading and walking most affected. Recovery of nystagmus and eye movement deficits was often incomplete; however, in many cases some functional recovery did occur and this was most often in primary position leading to a significant improvement in symptoms. All patients with square wave jerks had some reduction in the amplitude of the eye movement

Table 1. Summary of cases

Case no.	Site of lesion	Age at onset of stroke (yr)	Days from stroke onset to assessment	Ocular diagnoses	Orthoptic management	Outcome
1	Cerebellar artery infarct	45	40	Nystagmus; square wave jerks	Advice and observation	Improved. Asymptomatic at discharge
2	Cerebellar artery infarct	49	15	Nystagmus; square wave jerks	Advice and observation	Improved. Asymptomatic at discharge
3	Cerebellar artery infarct	45	25	Nystagmus; square wave jerks	Advice; LVA	Improved. LVA returned. Discharged
4	Cerebellar artery infarct	50	30	Nystagmus; square wave jerks	Advice; LVA	Improved. LVA returned. Discharged
5	Cerebellar artery infarct	81	17	Nystagmus; square wave jerks	Line guides and LVA; optical advice (separate readers/distance)	Improved; LVA still required. Under review
6	Cerebellar haematoma	58	25	Skew deviation; nystagmus; square wave jerks	Occlusion on readers for nystagmus; line guides and LVA; prism for diplopia at 6 months	Diplopia resolved. Nystagmus improved but persisted. LVA returned. Infrequent review
7	Cerebellar and lateral medulla infarct	64	23	INO, bilateral VIth nerve palsies; nystagmus	Prism or occlusion for diplopia	Improved. Under review

LVA, low vision aid; INO, internuclear ophthalmoplegia.

with corresponding improvement in symptoms, but not all recovered fully. Patient outcomes are summarised in Table 1, and Table 2 shows the frequency of eye signs and symptoms experienced by these patients.

Case series

Cases were divided into two groups based on clinical findings. Group 1 ($n = 5$) consisted of patients with square wave jerks/nystagmus, while group 2 ($n = 2$) consisted of patients with diplopia, eye movement deficit and nystagmus.

Group 1: patients with square wave jerks/nystagmus ($n = 5$)

Five patients were referred with blurred vision and vertigo. Orthoptic examination revealed primary position, jerky, horizontal nystagmoid movements with loss of fixation/square wave jerks. Nystagmus was also present in tertiary positions and conformed to a pattern of right beating on right gaze, left beating on left gaze, with dissociated rotary vertical (downbeating) on depression.

In all 5 cases visual acuity was initially moderately reduced to at least 0.5 logMAR. No ocular motor palsy was present in any case. Follow-up in all 5 cases was for a minimum of 18 months.

Recovery occurred in all 5 cases but was incomplete. Primary position square wave jerks all improved significantly over a 6–12 month period with a corresponding improvement in visual acuity. In cases 1 and 2 these

were detectable only on extremely close scrutiny at their final 18 month follow-up. Tertiary position nystagmus remained but was described as very fine. In cases 3, 4 and 5 square wave jerks were still present but did not produce symptoms, and visual acuity continued to improve up to 15 months after the infarct. Nystagmus was still present on side gaze and in depression, causing some problems with reading and walking.

Temporary low vision aids were no longer required in 2 cases and the patients were discharged. One patient remains under review in the low vision clinic and manages well with her low vision devices.

Group 2: patients with diplopia, eye movement deficit and nystagmus ($n = 2$)

Two patients presented with a variety of visual symptoms, including blurred vision, diplopia and difficulty following objects. Both demonstrated nystagmus in primary position and tertiary positions of gaze, with defects of ocular motility.

Case 6 presented with ataxia and dizziness and was subsequently found to have a cerebellar haematoma. Ocular motility revealed a skew deviation causing vertical diplopia in all positions of gaze, jerky horizontal nystagmus on side gaze and dissociated downbeating nystagmus on depression. Macro square wave jerks/saccadic oscillations were present on attempted fixation with associated reduced vision (0.6 logMAR). She was fitted with a vertical Fresnel prism to alleviate diplopia at distance fixation and had occlusion of one lens of her reading glasses. She was issued with a temporary low vision aid to assist with reading. Nine months later the skew deviation had resolved, vision had improved (0.3 logMAR) and the square wave jerks, while still present, were significantly reduced. She no longer required low vision aids and remains under infrequent review.

Case 7 presented with diplopia and oscillopsia and was found to have a cerebellar infarct that also involved the lateral medulla. Skew deviation, internuclear ophthalmoplegia (INO) and bilateral asymmetrical sixth nerve palsies were present. Moderate, horizontal, manifest, right beating nystagmus was present in right

Table 2. Frequency and type of eye signs and symptoms

Signs	Symptoms	No. of patients
Nystagmus	Oscillopsia	7
VIth nerve palsy	Diplopia	1
Fixation deficits (saccadic intrusions and square wave jerks)	Blurred vision	6
Skew deviation	Diplopia	1
Internuclear ophthalmoplegia (INO)	Diplopia, oscillopsia	1

gaze, left beating in left gaze, with a significant rotary element with abducting nystagmus of the left eye. In primary position, nystagmus was documented as fine, rotary, manifest nystagmus. The INO recovered after 3 months but the nystagmus was essentially unchanged. There was some recovery of the sixth nerve palsies but the patient continued to have significant problems at 2 years follow-up due to horizontal diplopia and nystagmus. A Fresnel prism trial was successful in relieving the diplopia and prisms are now incorporated into his spectacles.

Rehabilitation strategies

Advice was given to the patients, their carers and the multi-disciplinary team (MDT) regarding the implications of the deficits on the patients' overall rehabilitation. This included:

- adopting a small, comfortable abnormal head posture to use a null zone where oscillations from nystagmus are least. This improves visual acuity, and reduces oscillopsia and vertigo;
- occlusion of one eye on a temporary basis if nystagmus dampened or if the nystagmus was asymmetrical. This was achieved using either Fresnel foils to degrade the vision sufficiently to eliminate symptoms or total occlusion;
- use of temporary Fresnel prisms to facilitate more comfortable binocular vision in cases of poorly controlled or manifest strabismus;
- explanation to the patients, carers and MDT regarding the optimum positioning of visual targets for both daily living and rehabilitation activities;
- advice on the most appropriate type of spectacles (bifocals/varifocals versus single vision lenses) depending on the position and type of nystagmus or eye movement defect;
- issue of low vision aids to assist with reading and other activities: patients with reduced visual acuity may benefit from a low vision assessment, and low-powered magnifying aids such as flat-field bars and domes can be useful for certain near tasks.

Discussion

In this case series isolated cerebellar strokes that resulted in visual deficits were all associated with nystagmus and fixation abnormalities such as micro or macro square wave jerks. Square wave jerks are described as involuntary saccades that move the eyes away from the point of fixation and are followed, after a latency of 100–200 ms, by a saccade that returns the eye to the fixation point.¹ They are often mistaken for nystagmus but do not conform to the definition of nystagmus. They cause significant visual disturbances, resulting in a moderate reduction in visual acuity, oscillopsia and vertigo.

Diplopia and eye movement abnormalities are also common and may be a result of involvement at supranuclear, nuclear and infranuclear levels. The ocular motor subsystems, pursuit, saccadic, vergence and vestibular systems are frequently affected and require

specific detailed assessment. Gaze palsies, INO, one and a half syndrome, and third, fourth or sixth cranial nerve palsies can also be present. Many of these defects have specific localising signs; however, the presence of nystagmus is less helpful in determining the exact site of the lesion.

In this case series there were two distinct groups based on the eye movement disorders, as all patients had some type of nystagmus or ocular motor subsystem deficit. Eye movement abnormality was associated with a more extensive lesion affecting both the cerebellum and medulla; however, it is difficult to attribute the specific signs to either the cerebellum or the medulla. Both patients demonstrated skew deviation; this is commonly due to lateral medullary infarct that often occurs from occlusion of the vertebral artery.⁵ In the patient with multiple eye movement defects (INO, skew, sixth nerve palsy) this is suggestive of pontine involvement as well as the cerebellum and medulla, which may have been a result of brain-stem compression by a large cerebellar infarct.

Patients with isolated cerebellar stroke tend to have a better outcome than those with extra-cerebellar lesions, as expected, which correlates with other findings.² This was also evident in this small case series, where ocular motility disturbances were also present in combination with ocular motor subsystems defects (group 2). In cases where nystagmus plus square wave jerks were the prime feature, without ocular motility disturbance (group 1), the infarct was isolated.

Conclusion

Visual deficit that results from cerebellar stroke frequently causes problems with rehabilitation which can result in an extended stay for the patient. Careful examination by orthoptists is recommended as visual defects are often complex and not well assessed.⁵ Patients' subjective accounts of their symptoms are often very variable in their reliability and accuracy, and many have difficulty in describing their problems. Objective signs such as obvious manifest strabismus, i.e. skew deviation or exotropia, are easy for most practitioners to elicit; however, subtle but debilitating nystagmus or fixation instability may be overlooked. The orthoptist can provide a thorough examination and detailed explanation to the patient, their carer and the MDT, and offer advice, strategies and treatments to help relieve symptoms, thereby aiding rehabilitation.

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