CLINICAL STUDY

Ocular manifestations of hindbrain-related syringomyelia and outcome following craniovertebral decompression

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Abstract

Purpose **To analyse and evaluate associated ocular symptoms and signs in hindbrain-related syringomyelia and their response to treatment.**

Methods **From a database of 275 patients treated in a single institution for hindbrain hernia and syringomyelia, 39 patients (14%) had ocular symptoms and signs. Only 31 patients were included in this study; the remainder were excluded due to inadequate follow-up information. All patients had confirmed evidence of hindbrain-related syringomyelia with MRI scan or CT myelogram. Treatment included craniovertebral decompression or ventriculo-peritoneal shunting. The mean follow-up was 23 months.**

Results **In addition to the well-recognised sign of downbeat nystagmus, classically associated with foramen magnum abnormalities, a number of other ophthalmic features were identified. Symptoms included diplopia, oscillopsia, tunnel vision and difficulty in lateral gaze. Signs included nystagmus (downbeat, horizontal, rotatory, and combinations), strabismus, disc pallor, anisocoria, ptosis and field defect. Patients were categorised into two groups depending on whether the ocular features were manifest at first presentation (group 1, $n = 14$) or developed later in the course of the disease (group 2, $n = 17$). The delay in diagnosis from first presentation was 5 and 6 years respectively. All patients underwent surgery. Craniovertebral decompression was performed in 13 patients in group 1 and in 15 patients in group 2. Ventriculo-peritoneal shunt was inserted in 1 patient in group 1 and in 3 patients in group 2, for the associated hydrocephalus. Following surgery, 100% of patients in group 1 and 82% of patients in group 2 had complete or partial resolution of their ocular symptoms and signs.**

Conclusions **The presence of unexplained ophthalmic features such as nystagmus or oscillopsia should alert one to the potential diagnosis of hindbrain-related syringomyelia. Delay in diagnosis is often associated with poorer outcome. Surgical treatment can offer excellent results for these patients.**

Key words Hindbrain hernia, Nystagmus, Oscillopsia, Syringomyelia

Syringomyelia is the condition characterised by longitudinally displaced cavities within the spinal cord, extending over more than one segment. It can be due to abnormalities in the craniovertebral junction or the spine. The commonest cause of syringomyelia is a structural abnormality at the region of the craniovertebral junction, referred to as Chiari type I malformation or hindbrain hernia.[1–10] This is characterised by descent of the cerebellar tonsils below the foramen magnum, which causes obstruction to cerebrospinal fluid circulation between the cranial and spinal compartment, and compression of neural tissue at the craniocervical junction.[4,6,7,9–12] Other causes of hindbrain-related syringomyelia include rare skull base bony abnormalities and development abnormalities (e.g. arachnoid cysts).

Neurological manifestations of syringomyelia include pain, motor symptoms, sensory disturbance and brainstem signs.[1–3,5,6,8–10,13,14] Ocular features such as downbeat nystagmus and oscillopsia are recognised as one of the many neurological symptoms or signs associated with this condition.[5,6,8–10,14–20] Less common and specific symptoms or signs such as blurred vision, diplopia, difficulty in focusing (particularly on lateral gaze), intermittent tunnel vision, rotatory and combinations of nystagmus, anisocoria and ptosis are often overlooked. Such signs, however, may be the sole presenting feature to alert one to the diagnosis. It has previously been shown that surgical intervention for this

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Received: 1 February 2000
Accepted in revised form: 14 July 2000

condition may improve or stabilise the neurological state.[3,9,10,14–17] There have also been reports of reversal of downbeat nystagmus following surgery, but there has been little mention of the effect of surgery on the other ophthalmological features associated with syringomyelia. This paper attempts to further evaluate all ocular symptoms and signs associated with hindbrain-related syringomyelia, both subjective and objective, and to analyse the effect of surgery.

Patients and methods

Patients were selected from a database of 275 patients with hindbrain herniation and syringomyelia treated at the Midland Centre for Neurosurgery and Neurology over a 10-year period (1985–1995) (the unit has since been relocated at the Queen Elizabeth Neuroscience Centre). The case notes were reviewed retrospectively. Of the whole group, 39 patients (14%) with hindbrain hernia and syringomyelia developed ocular signs; 31 of these were included in the study, the other 8 being excluded due to lack of complete follow-up information. There were 13 males (42%) and 18 females (58%). Mean age at diagnosis was 34.3 years (range 10–55 years). From diagnosis to presentation there was a mean delay of 5.5 years. The mean follow-up period was 23 months (range 2–108 months).

All patients underwent computed tomography of the head (CT), 5 patients underwent CT myelography and 26 had magnetiic resonance imaging (MRI) of the head and neck. All patients had radiological evidence of hindbrain hernia. In addition, 3 had associated hydrocephalus, 12 had syringomyelia and 1 had hydrocephalus and syringomyelia. All patients underwent surgery; 27 patients had craniovertrebral decompression only. This involved a small craniectomy over the region of the foramen magnum in the posterior fossa, decompression of the cerebellar tonsils and dissection of arachnoid adhesions. The dura mater was laid widely open. In 3 patients a ventriculo-peritoneal shunt was inserted only, to treat the associated hydrocephalus. In 1 patient both craniovertebral decompression and a ventriculo-peritoneal shunt were performed.

Results

Symptoms and signs

The patients were divided into two groups: group 1 included patients who presented with ocular symptoms or signs; group 2 included patients whose original presentation was with non-ocular features but who developed ocular features as a later manifestation of the disease process. There were 14 patients (45%) in group 1 (6 males) and 17 (55%) in group 2 (6 males). The mean ages at presentation were 34.9 years (range 10–54 years) and 33 years (range 12–55 years) respectively, the mean delay in diagnosis 5 and 6 years respectively and mean follow-up periods 17.6 months (2–60 months) and 23.3 months (3–108 months) respectively in groups 1 and 2.

The ocular symptoms identified are shown in Table 1. Diplopia and oscillopsia were the most common, followed by subjective visual field defect, tunnel vision and non-specific symptoms including difficulty in focusing, blurred vision on lateral gaze and intermittent loss of vision. The ocular signs are shown in Table 2 and include nystagmus, strabismus, visual field loss, optic nerve abnormalities, anisocoria and a deficit in accommodation. Nystagmus was the commonest finding and in Table 3 the different types are shown. Tables 4 and 5 show the various non-ocular symptoms and signs respectively seen at presentation.

Table 1. *Ocular symptoms*

Symptoms	Group 1	Group 2	Total
Diplopia	11 (79%)	10 (59%)	21
Oscillopsia	6 (43%)	6 (35%)	12
Tunnel vision	1 (7%)	2 (12%)	3
Difficulty on lateral gaze	5 (36%)	1 (6%)	6

Table 2. *Ocular signs*

Signs	Group 1	Group 2	Total
Nystagmus	10 (72%)	11 (65%)	21
Strabismus	2 (14%)	0	2
Disc pallor	1 (7%)	2 (12%)	3
Anisocoria, ptosis	0	1 (6%)	1
Field defect	0	1 (6%)	1

Outcome following surgery

Following surgery, 100% of patients in group 1 and 82% of patients (14) in group 2 had complete or partial resolution of their ocular symptoms and signs (Fig. 1). One patient in group 2 had deterioration of both the neurological and ocular features post-operatively necessitating further surgery 3 days later. Overall, the improvement of the ocular features compares favourably with that of the general neurological features. In comparison motor symptoms resolved completely in 35% of group 1 and 20% in group 2, and partially in 50% of the patients in group 1 and 60% in group 2. Sensory symptoms improved in 61% of the patients in group 1 and 50% in group 2. In all patients with syringomyelia the radiological appearance of the syrinx improved after surgery. There was no correlation between clinical outcome and the length of the syrinx, in either group.

Table 3. *Types of nystagmus*

Nystagmus	No. of patients
Downbeat	8
Horizontal	4
Rotatory	4
Combination	5

Table 4. *Non-ocular symptoms (both groups* n = *31)*

Pain (general)	26
Headache	22
Numbness	20
Gait disturbance	18
Impotence	13
Nausea/vomiting	11
Weakness in limbs	8
Dizziness	8
Clumsiness	8

Delay in diagnosis as a factor in the failure to improve after surgery

Of the 14 patients in group 1 whose initial presenting features included ocular symptoms or signs, 7 (50%) did not go on to develop any other neurological deficit. These 7 had a significantly shorter time from onset of symptoms to diagnosis and subsequent treatment compared with the remaining patients of the group. The mean delay for the former was 9 months compared with 84 months for the latter ($p < 0.001$, Student's t-test). Further evidence suggesting that the time to diagnosis may be responsible for the lack of improvement is also seen in group 2, where the mean delay in diagnosis was 108 months in those who improved compared with 120 months in those who failed to improve following surgical treatment ($p = 0.07$, Student's t-test).

Complications

There were few complications in this series of patients. One patient had superficial wound infection and one patient had a ventricular haemorrhage during shunt insertion. There was no mortality.

Case reports

The following cases have been selected to highlight some of the typical presentations.

Case 1

A 14-year-old female had been diagnosed as being 'a clumsy child' a few years earlier. At the time of presentation to the neurosurgical unit she complained of 'difficulty in seeing things', headaches and tinnitus. Her parents noticed that she would put her head down and look up just beneath her eyebrows in order to look at something. It was not known how long the problems had been present. On examination she had severe downbeat

Table 5. *Non-ocular signs (both groups,* n = *31)*

Gait abnormality	9
Abnormal reflexes	9
Weakness in limbs	8
Babinski reflex	8
Kyphosis	8
Disdiadochinesis/past pointing	5
Reduced range of movement	4
Ataxia	4
Trigeminal neuralgia	4

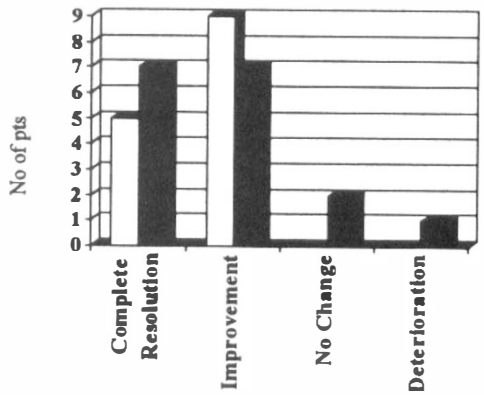

Fig. 1. *Change in ocular symptoms and signs following craniovertebral decompression.*

nystagmus which worsened on looking down, a slow beating nystagmus to the left and a rapid, fine beating nystagmus to the right, which became severe on looking to the right. There was no nystagmus on upgaze. No other ocular findings were noted and her visual fields were full. She was unsteady on her feet, suggestive of posterior column loss, but no actual spinal cord dysfunction was detected. MRI confirmed cerebellar tonsillar descent and syringomyelia from C2 to C7 (Fig. 2). Craniovertebral decompression was performed.

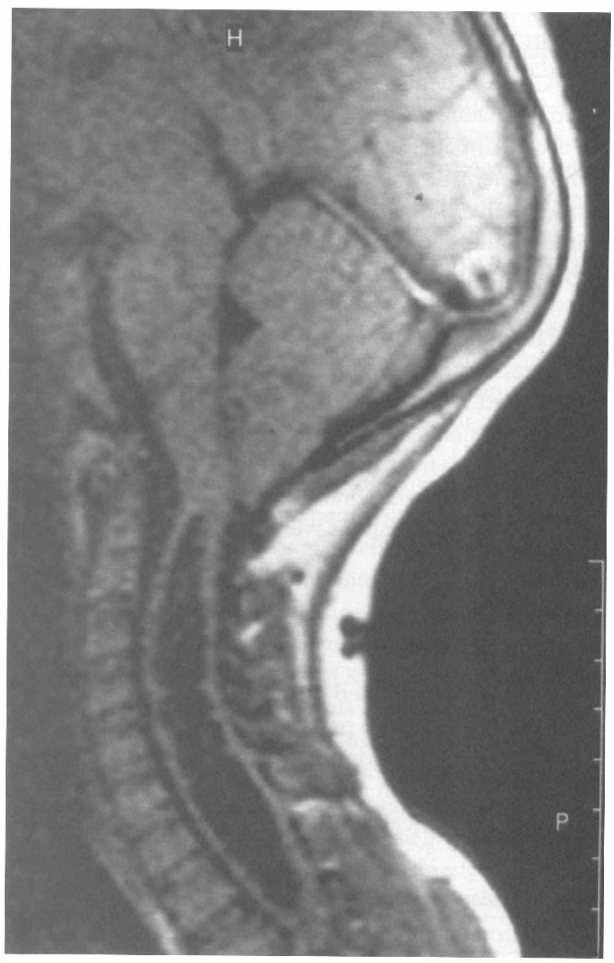

Fig. 2. *T1-weighted MRI scan of a 14-year-old female who presented with headaches, tinnitus and visual difficulties with severe nystagmus on lateral and downward gaze. A significant hindbrain hernia is seen. A syringomyelia cavity is occupying most of the cervical cord.*

Immediately post-operatively there was improvement in the nystagmus in all directions. On review 9 months later there had been further improvement, with complete resolution in all but one direction. The patient's symptoms had almost completely resolved at 1 year although she maintained a residual head posture, which the patient herself felt was due to habit rather than necessity.

Case 2

A 24-year-old man had been attending his general practitioner for 5 years with a history of headaches and sudden onset of diplopia. A diagnosis of multiple sclerosis had been made. On examination he had an alternating esotropia and mild, variable nystagmus. Ophthalmic and general examination were otherwise normal. On MRI there was a hindbrain hernia and syringomyelia in the cervical cord extending from C3 to C7. Appearances were very similar to those of Fig. 2. Craniovertebral decompression was performed but there was no immediate improvement in his ocular features. One year later there was total resolution of the nystagmus, the strabismus and the diplopia.

Discussion

A number of publications have already outlined the importance of downbeat nystagmus in localising foramen magnum abnormalities.[15–20] There are, however, few, if any, papers examining other ocular features which may be useful in aiding diagnosis.

The non-ocular features of hindbrain hernia-related syringomyelia are well documented and include sensory changes (the typical pain–temperature dissociation is seen in a minority of patients), cerebellar involvement, weakness, hypotonia, and a reduction or absence of reflexes.[1,3,5–10,13,14] Ocular features previously identified include diplopia, oscillopsia, nystagmus, anisocoria and ptosis.[5,6,8–10,14–21]

In cases of syringomyelia and syringobulbia associated with hindbrain hernia, direct pressure on the medulla, cerebellar tonsils and lower cranial nerves can cause damage both directly, leading to gliosis, as well as indirectly, compromising their corresponding blood supply. The effect of the tonsillar herniation is threefold: the tonsils become wedged and this promotes arachnoid adhesions thus worsening the disease process; the tissues become compressed, deformed and under tension; and lastly, if the descent of the hindbrain is unequal, then asymmetrical and rotational deformities may occur. The creation of a syringomyelic cavity inside the spinal cord is subsequently perpetuated by the deranged cerebrospinal fluid (CSF) circulation around the foramen magnum. The initial formation of the syringomyelia cavity is not well understood, but its further propagation has been outlined by Williams who described the mechanisms of 'suck' and 'slosh'.[9–12] The craniovertebral dissociation in CSF circulation keeps the syringomyelia cavity open by creating negative pressure outside the spinal cord in part of the respiration cycle ('suck'). When the thoraco-abdominal pressure rises the CSF is unable to return up to the head due to the subarachnoid block in the foramen magnum. This rise in spinal subarachnoid pressure causes forcible fluid movement upwards inside the syringomyelia cavity which dissects the cavity longitudinally even further ('slosh'). This theory is not universally accepted, was originally developed before the advent of MR scanning, and may well have to be modified in the light of newer information which has emerged in recent years following the introduction of cine phase contrast MRI.[4] The upward movement of fluid into the brainstem creates syringobulbia. This may explain why patients experience diplopia or other neurological features following a sneeze or cough.[7]

The abnormal eye movements may be in part explained by a lesion within the medial longitudinal fasciculus.[19] Involvement of the third and fourth nerves is thought to be rare, with involvement of the abducent nerve being more common. This has previously been described as being a result of destruction of the sixth nerve fibres in the pons. However, it appears that involvement of the nucleus itself seems a likely explanation in some of our cases and in addition it is possible that the downward movement of the brainstem has caused direct traction on the nerves.[19]

Nystagmus and oscillopsia are commonly associated with hindbrain herniation. The exact neural mechanism of downbeat nystagmus is unknown but is thought to be due to an inability to transfer downward visual velocity information to the neural network that generates smooth eye movement.[16,17,22,23] Abnormalities of the spinovestibular and spinocerebellar tracts are thought to be responsible.[16] Unlike congenital nystagmus, which is usually asymptomatic, patients who acquire nystagmus experience a sensation of the world jumping around. Hindbrain herniation is classically associated with a downbeating nystagmus but in this series of patients this was observed only in a third of the patients, the remainder having no particular pattern of nystagmus but a combination of vertical, horizontal and often rotational elements. Nystagmus in this series was more commonly reported than in other studies and improved more often post-operatively. No patient developed nystagmus post-operatively although one patient developed temporary deterioration of a pre-existing nystagmus.

Oscillopsia is very different from the visual disturbance experienced and described by some of our patients on lateral gaze. This is more difficult to explain. A possible explanation for this blurring of vision, of being unable to focus and 'fuzzy vision' on lateral eye movements is the phenomenon of saccadic suppression.[22,24] This is described as an increase in the visual threshold and a decrease in visual activity during the saccadic eye movements. Previous studies have indicated that the visual suppression lasted for 25 ms before the saccades began, continued throughout the saccades and for a further 25 ms after the saccades and finished. In patients with nystagmus on side gaze (as in

case 1), the eyes are involved in saccadic movement for a significant proportion of time and this may explain why the vision is impaired during this time.

The optic disc pallor observed in a small number of patients is most likely due to pre-existing long-standing intracranial hypertension secondary to the hindbrain hernia. Intracranial hypertension leads to optic disc swelling, which when of long standing results in optic atrophy and subsequent visual field defect. The association of intracranial hypertension and hindbrain hernia in the absence of hydrocephalus has only recently been appreciated, and patients have been encountered in clinical practice – outwith the context of this series – who had papilloedema at presentation that settled following craniovertebral decompression.

The introduction of MRI has improved the pick-up rate of the condition. The non-invasive nature of the examination has gradually lowered the threshold of request by the clinicians. Admittedly the pick-up rate of diagnosing hindbrain hernia among patients with only visual symptoms, in the absence of any other neurological signs, is likely to be small, but a high index of suspicion should always prevail, especially if symptoms persist with time or even deteriorate.

Craniovertebral decompression is associated with good success rate with respect to resolution of neurological symptoms and signs, and the results in this series compare favourably with previously published material.[3,5,9,10] The resolution of symptoms after craniovertebral decompression is probably due to release of pressure and traction on the cranial nerves and long tracts and blood vessels and a reduction in the compression of the brainstem in the foramen magnum. The observed difference in outcome in relation to the length of symptoms indicates that persistent neural compression may lead to irreversible damage, and points towards the need for early diagnosis and treatment.

In conclusion, ocular symptoms and signs are an important diagnostic feature of hindbrain-related syringomyelia. There is currently often a long delay in making the diagnosis and increased awareness of the associated ocular features may lead to an earlier diagnosis. A high rate of suspicion should prompt early investigation with MRI. Surgery in appropriately selected cases has particularly encouraging results with good chances of resolution of visual disturbance.

This manuscript in its original form was one of the last few scientific contributions that the late Bernard Williams had been working on prior to his untimely death. We remain indebted to his legacy.

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