Congenital ocular palsy

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Three patients with severe restriction of the ocular movements in one eye are described. They all preferred the poorly moving eye for fixation while the eye with full movements was amblyopic with nystagmus. This phenomenon has not, to my knowledge, been described before and is an exception to the general rule that congenital muscle palsies are not usually accompanied by amblyopia.

Case reports

CASE 1

A full-term male infant, weighing 8 lb 11 oz at birth, was delivered by forceps on July 15, 1960, because of occipito-posterior presentation. He had a large cephalhaematoma on the right side with considerable bruising and swelling around the right eye, the upper lid of which did not open for one month. When it did open, it was noted that the eye itself did not move, and that the pupil was small and did not react to light. Apart from this, the child was well.

At the age of 2 years the findings were as follows: the right palpebral fissure was smaller than the left due to ptosis; it narrowed slightly when the eye was abducted and widened slightly on adduction. The right upper lid became slightly retracted on attempting to look down but the lids closed normally in sleep. Bell's phenomenon was absent. The right eye had almost full movements in the horizontal plane but vertically there was only a very slight amount of elevation in adduction and a very slight amount of depression in abduction (Fig. 1*a-j*).

The movements of the left eye were full. The pupil of the right eye was smaller than the left (Fig. 1*b*) and it reacted sluggishly to light and accommodation. The cover test showed that the child maintained binocular vision some of the time, but he easily developed a left divergent squint with a rotatory nystagmus of the left eye. The right eye was occluded to encourage use of the left eye.

At the age of 5 years the findings were unaltered, except that the refraction then showed hypermetropic astigmatism, the prescription being R + 1 D sph., + 1 D cyl., axis 90°; L + 0.5 D sph., + 2 D cyl., axis 90°. The visual acuity with correction was R 6/6, L 6/24. At 11 years of age astigmatism had increased, the prescription being R + 2 D sph., + 3.5 D cyl., axis 80°; L + 0.5 D sph., + 3 D cyl., axis 75°; the visual acuity was still R 6/6, L 6/24.

Comment

This child had weakness of the levator palpebrae superioris and all the vertically acting muscles of the right eye; the pupil was small and reacted poorly to light and accommodation. He used the right eye for fixation in preference to the left which was amblyopic and had nystagmus.

CASE 2

A full-term male infant was born on June 15, 1951, by normal delivery, and weighed 7 lb 4 oz. He

FIG. 1 Case 1, aged 7 years In the right eye the vertical movements are very restricted, the pupil is small, and the upper lid moves poorly

was referred to the eye department at the age of 3 months because of ptosis of the right upper eyelid. It was also noticed that the right eye was divergent and that there appeared to be a complete right third nerve palsy, except for the pupil which reacted to light; no other abnormalities were found on general examination.

At the age of 1 year he was found to be using both eyes but preferred to fix with the right; the poor ocular movements and ptosis persisted.

At 3 years of age a Blascovitz operation was carried out on the right upper lid with some improvement in the ptosis. At 4 years of age he was found to have hypermetropic astigmatism, the refractive error being R + 2.5 D cyl., axis 120°, L + 2 D sph.; the visual acuity was R 6/24, L 6/60. The right upper lid was in a satisfactory position but there still appeared to be paralysis of the right superior, inferior, and medial rectus muscles. When he fixed with the right eye, the left eye deviated outwards and upwards with a marked nystagmus.

The condition remained substantially unaltered until, at the age of 14, he returned to see if anything could be done to improve his appearance. There was a moderate degree of right ptosis. The visual acuity was R 6/12, with + 1.25 D cyl., axis 135°; L 6/36, with + 2 D sph., + 1.75 D cyl., axis 15°. There was constant rotatory nystagmus of the left eye and an intermittent nystagmus of the right. In the primary position, when the patient fixed with the right eye, the left eye was elevated (Fig. 2c).

The right eye could be abducted from the midline but, apart from this, there was no movement in any other direction except very slight depression while the left eye had full movement in all directions (Fig. 2a-e). The pupils were equal in size and it was noted that the right one reacted poorly to light but not to accommodation, while the left reacted briskly to light but not to accommodation. The ocular media were clear and the fundi normal.

Operations

On July 29, 1965, when he was 14 years old, a left superior rectus recession of 5 mm. was carried out to reduce the hypertropia (this procedure sometimes reduces the size of the palpebral fissure as well and it was hoped it would lessen the disparity between the two fissures) and some improvement was obtained.

FIG. 2 Case 2, aged 14 years *Right ptosis with poor movements of the upper lid and severely restricted ocular movements except for abduction*

At the age of 16 years, recession of the left lateral rectus was carried out to restrict abduction of the eye, and re-resection of the right levator palpebrae superioris was performed by the anterior route; there was considerable improvement in his appearance after this (Fig. 3a-c).

FIG. 3 Case 2, aged 16 years, after left superior rectus and left lateral rectus recession, and re-resection of the right levator palpebrae superioris

Result

At the last visit the findings were unchanged except that the pupils were equal and both reacted briskly to light and accommodation. Bell's phenomenon was absent on the right side.

Comment

This patient appeared to have paralysis of all the extraocular muscles of the right eye except the lateral rectus. The right pupil reactions were reduced at one examination but normal at another. The patient used the eye with poor ocular movements for fixation while the other eye was amblyopic with constant nystagmus.

CASE 3

A boy born on October 10, 1964, weighing 4 lb. 11 oz. was one of fraternal twins. His condition at birth was good but he was found to have a harelip on the left side and ptosis of the right upper eyelid (Fig. 4a). The right eye was noted to be depressed and abducted and the pupil was small. No movements of the right eye could be elicited, but the left eye showed full movements and he appeared to look at objects with the left eye. Fixation could not be detected with either eye 2 months later. The left eye was showing rather jerky purposeless movements and the left upper lid became retracted when the eye was abducted. At the age of 6 months he was found to be looking at objects with the right eye (Fig. 4b) and turning his head to follow them in a birdlike manner. It was also noted that he had slight vertical nystagmus in the right eye and a rotatory nystagmus in the left. When he was looking at close objects the right eye fixed the object while the left eye became abducted and elevated with the upper lid retracted and a marked rotatory nystagmus; this gave the child a very bizarre appearance. He could fix objects quite well with the left eye when the right was occluded and the nystagmus in the left eye then became much less marked. His development was thought to be satisfactory at that stage and full general examination and x rays of the skull revealed no abnormality. Refraction under anaesthesia showed a moderate degree of hypermetropia—R + 3 D sph., L + 2 D sph. The ocular media were clear and the fundi normal. The mother was advised to occlude the right eye for one week and the left for one week alternately, and this was carried out with some difficulty.

FIG. 4 Case 3, aged 2 years Right ptosis, meiosis pupil, and no ocular movement

Operations

At the age of 13 months an operation was carried out to try to improve his appearance by restricting the movements of the left eye. A left superior rectus recession of 5 mm. and a medial rectus resection of 5 mm. were performed and 2 months later a left inferior rectus resection of 4 mm. and a lateral rectus tenotomy were carried out. The appearance was slightly improved especially when looking downwards.

At the age of 27 months resection of the right levator palpebrae superioris by the anterior route was performed with further slight improvement. At this time it was considered that the child was mentally retarded and there were severe behavioural problems, but he was then found to be autistic and has since been under treatment for this condition.

Result

At his last visit his condition was unchanged and no subjective tests were possible. The right pupil was small and did not react to light or accommodation but it did dilate with a mydriatic; the left pupil reacted well. He used the immobile right eye for looking at near objects and the left for looking around the room.

Comment

This child had practically no movement of the right eye except for very slight vertical nystagmoid movements. The pupil was small and not reacting. The left eye with full motility was probably amblyopic with nystagmus which was much less marked when the right eye was occluded (inverse latent nystagmus). Detailed examination of the child was made difficult by the autism.

Discussion

These three patients are unusual in that each has a severe ocular palsy of one eye but uses the poorly moving eye for fixation, while the eye with full movement is amblyopic with nystagmus. This nystagmus is also unusual because it becomes less marked when the fixing eye is occluded. Preferential fixation by the paretic eye does not seem to have been previously described in congenital palsy, but it does occur in some acquired ocular palsies in order to obtain wider separation of the double images (the secondary deviation being larger than the primary) and it is possible that this was the mechanism in these cases also. Other possible factors are that the right eye in all the patients may have been developmentally the master eye so that it was preferred for fixation although the refractive error was larger than in the left, or it may have been that the non-parietic eye was receiving such excessive neural impulses because of the attempt to move the paretic contralateral synergists (Lyle and Bridgeman, 1969) that the movements became excessive and uncontrolled so that the developing brain found it easier to utilize the visual messages from the paretic eye for correlation with spatial relationships and proprioceptive impulses. An analogous situation occurs in some cases of Duane's syndrome (Keith, 1961), in which the head turn, instead of being muscle-relieving as would be expected, places the eye in the direction of the maximal action of the most paretic muscle so that the eye is at the limit of movement in that direction and the other eye is brought into alignment with it. The mechanism may facilitate cerebral coordination of the severely disturbed proprioceptive impulses and the impaired movements. These adjustments are probably only possible when the cerebro-ocular organization is developing and could not be acquired later in life. It may particularly apply to affections of the muscles themselves rather than to nerve injuries. In many cases of congenital palsy, binocular vision is maintained by a head posture (Lyle and Bridgeman, 1959) and Case 1 does have some binocular vision while the other two have none.

The aetiology of the palsy in Case 1 was probably traumatic as he had an orbital haematoma at birth which could have caused fibrotic changes in the muscles and fascia so restricting the ocular movements. Unfortunately forced duction tests have not been carried out on any of the patients. Cases 2 and 3 had no obvious cause for the squint but the latter did have another congenital anomaly (harelip) and he was later found to be autistic. The Table shows that the lateral rectus muscle was only affected in Case 3, while the third and fourth nerve musculature was involved in all cases, but the pupil was small in Cases 1 and 3 instead of being enlarged as would be expected in a nerve lesion, so it is likely that the lesion causing the ophthalmoplegia was sited in the orbit rather than centrally and the muscles themselves were involved rather than the nerves to them. The

Table Findings in three patients

Case no.	1		2		3	
Eye	R	L	R	L	R	L
Lateral rectus	—	—	+	+	—	+
Superior oblique	—	+	—	+	—	+
Levator palpebrae superioris	—	+	—	+	—	+
Superior rectus	—	+	—	+	—	+
Medial rectus	+	+	—	+	—	+
Inferior rectus	+	+	—	+	—	+
Inferior oblique	—	+	—	+	—	+
Pupil size	Small	Normal	Equal	Equal	Small	Normal
Reaction to light	±	+	+	+	—	+
Reaction to accommodation	±	+	+	+	—	+
Amblyopia	—	—	±	+	±	+
Nystagmus	—	+	±	+	+	+
Visual acuity	6/6	6/24	6/12	6/36	—	—
Bell's phenomenon	—	+	—	+	—	—

Key + Good action or phenomenon present
 ± Some action or present to some degree
 — No action or not present

treatment was conventional in that the amblyopia was dealt with by occlusion and the prescription of glasses. Surgery was necessary in Cases 2 and 3 to reduce the amount of movement of the amblyopic eye and to try to align it with the paretic eye in the primary position. Levator resection was also needed to correct the ptosis of the paretic eye. With Case 2, in whom treatment has been completed, the appearance has been improved but is still only fair. Unfortunately a perfect cosmetic result in these severe cases of muscle palsy is very difficult to obtain.

Summary

Three patients are described, each of whom had severely restricted movements of one eye, but preferred the paretic eye for fixation while the eye with full movement was amblyopic with nystagmus. The causative lesion was probably sited in the orbit and in one case an orbital haematoma was present at birth. In two cases the pupil on the affected side was small with impaired reactions.

References

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