Short report

Cyclic oculomotor palsy: description of a case and hypothesis of the mechanism

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SUMMARY An unusual case of cyclic oculomotor palsy is described where the probable cause was a supraclinoid aneurysm. A mechanism is suggested by which the features of cyclic oculomotor palsy can be explained solely by a peripheral lesion.

Cyclic oculomotor palsy is a rare disorder in which a complete or partial third nerve palsy alternates with brief spasms of third nerve function. During a spasm the pupil constricts, the lid elevates and the globe turns down and in; partial forms of the disorder have been described in which only one or more of these changes occurs. The majority of cases are seen in infancy¹ and no patient has been described in whom the lesion responsible was identified. We report the case of an elderly female aged 69 years with a cyclic oculomotor palsy in which a structural cause was found.

Case history

The patient's illness began with the development of sudden severe headache, confusion and drowsiness. She was nursed at home and made a gradual full recovery over two months. Six months later she awoke at night with sudden severe pain in the left eye. This persisted for a few days and was associated with drooping of the left lid and periorbital swelling. The patient was referred to an ophthalmologist, who found a left partial third nerve palsy with severe restriction of vertical movement and a dilated poorly reacting pupil. Further assessment was declined until a year later. On this occasion the ocular changes of a cyclic oculomotor palsy were seen and pointed out to the patient, who was unaware of the cyclic changes.²

At that time, the only abnormality on neurological examination was of the left third nerve, the fourth and sixth nerves being intact. In the paralytic phase the patient had a partial third nerve palsy (fig 1A); there was partial

Fig 1A Paralytic phase showing left ptosis. Right pupil diameter 5.5 mm. Left pupil diameter 6 mm.

Fig 1B Spastic phase (photograph taken 36 s after fig 1A) showing over-elevation of left eyelid, mild depression and adduction of the left globe. Right pupil diameter 6.5 mm. Left pupil diameter 7 mm.

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left ptosis with a mild left exotropia and hypotropia. The pupil was dilated and reacted slowly to direct light and accommodation. Adduction was full but vertical move-

Fig 2 CT scan with contrast enhancement showing a round enhancing lesion in the left supraclinoid region.

ment was severely limited with no upward movement and only slight downward movement. There was marked lid lag on looking down. In the spastic phase (fig 1B) the lid became over-elevated and the globe slightly depressed and adducted, taking about 45 seconds for these changes to occur. The pupil was unchanged. The spastic phase lasted for a variable period of 45–90 seconds and then the lid slowly drooped as the globe returned to the position of the paralytic phase. Cycles occurred with varying intervals from three to seven minutes and were not affected by voluntary gaze.

A CT scan (fig 2) showed an enhancing mass in the left supraclinoid region reported as either an aneurysm or meningioma. A left carotid angiogram was not performed as the patient had a severe respiratory disability due to intrinsic asthma requiring steroid treatment.

Discussion

The cyclic changes described in this patient conform to the definition of cyclic oculomotor palsy since a partial third nerve palsy was associated with episodic brief spasms in which the lid became over-elevated and the globe turned down and in. The pupil was not constricted in the spastic phase which is unusual, but otherwise the cyclic changes are similar to the first case that Bielschowsky described.³ The clinical picture and the CT scan appearance leave little doubt that a supraclinoid aneurysm was the cause of the disorder.

Since there are no previously described cases in which the cause has been identified, there has been considerable dispute about the site of the lesion, some authors favouring a central brainstem lesion,^{4,5} others a peripheral one.^{6,7} More recently Loewen-

feld *et al*¹ proposed that a combination of a central and peripheral lesion was necessary to explain all the features of cyclic oculomotor palsy. In the case reported here the lesion was peripheral and the clinical history suggestive that the aneurysm caused a partial third nerve palsy with the cyclic changes developing sometime subsequently.

Following damage to the third nerve by an aneurysm, the misdirection syndrome, which has many similarities to cyclic oculomotor palsy, almost invariably develops if regeneration occurs.^{8–10} In the misdirection syndrome the lid elevates, the pupil constricts and the globe turns in on attempts to look up or down.⁹ There is good evidence that this is due to mass discharge in the misdirected regenerated fibres so that only unopposed ocular movements occur and vertical movement is absent since the actions of the superior and inferior recti cancel each other out.¹¹ The complete misdirection syndrome differs from cyclic oculomotor palsy only in that the ocular movements occur in the one voluntarily and in the other spontaneously; however patients with cyclic oculomotor palsy have been described in whom voluntary gaze influences the cycles,¹² and in some cases the two syndromes co-exist^{4,13} emphasising the close similarity between the two conditions.

In view of these considerations and the features of this case it is possible that cyclic oculomotor palsy could be due to aberrant regeneration of the third nerve together with an intermittent conduction block, in the regenerated fibres, caused by the scar. This hypothesis is able to account for the important features of cyclic oculomotor palsy. Regeneration of the third nerve and misdirection of these fibres explains the similarity of cyclic oculomotor palsy to the misdirection syndrome. The fact that the magnitude of the cyclic movement in one direction is inversely proportional to the physiological movements in the same direction is also explained by aberrant regeneration since physiological movements are caused by the undamaged fibres and the cycles by the regenerated fibres. In this case the pupil remained partially paralysed during the spastic phase and therefore it must be assumed that although it was partially denervated, no reinnervation had occurred. Vertical movement is rarely seen in cyclic oculomotor palsy, but if the inferior rectus was more strongly reinnervated, downward movement would occur during the spastic phase as illustrated by this patient.

The cyclic nature of the disorder can be explained by assuming that the regenerated fibres are only intermittently capable of conducting impulses or that the scar has caused an intermittent conduction block. Intermittent discharge of the regenerated fibres would lead to a cycle. This is supported by the

fact that in some cases voluntary gaze in a certain direction triggers or "holds" the cycles suggesting that an increased volley of impulses is capable of overcoming the temporary conduction block. It is therefore possible to account for the features of cyclic oculomotor palsy on the basis of a peripheral lesion and this case report supports this suggestion.

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