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Case Report

Bilateral Asymmetrical Cyclic Oculomotor Palsy with Spasm - A Case Report

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Abstract

This is a unique case of bilateral asymmetrical cyclic oculomotor palsy with spasm (COPS). This is an unusual presentation of COPS, which is usually unilateral. The patient initially presented with cyclic upper lid ptosis, exotropia, hypotropia, and mydriasis, lasting approximately 1.5 minutes. The affected eye would then recover, and the other eye would undergo the same pattern of symptoms in keeping with COPS. The right eye has shown spontaneous improvement of symptoms over the 7 years of follow-up. The use of ptosis surgery, together with patching, seems to have been able to prevent the development of amblyopia in the left, worse-affected eye.

Keywords: cyclic oculomotor palsy with spasm, oculomotor nerve regeneration, strabismus, neuro-ophthalmology

Abbreviations: COPS: cyclic oculomotor palsy with spasm

1. Introduction

Cyclic oculomotor palsy with spasm (COPS) is a rare disorder of aberrant 3rd nerve regeneration. It is characterized by episodic paresis of the muscles innervated by cranial nerve 3, resulting in mydriasis, ptosis, and exotropia with hypotropia. This alternates with a spastic phase whereby there is a return to midline, miosis, and elevation of the ptotic eyelid. This condition is normally unilateral and non-progressive. This case is unique in the bilateral but asymmetrical nature of the patient's presentation.

2. Case Report

An infant girl who had a normal prenatal history was born at term by an elective cesarean section. She had a tracheoesophageal fistula, which was repaired, and she was growing well and meeting her developmental milestones. The patient presents to the ophthalmologist for the first time at 7 months of age with a history of unequal pupil size and failure of left eyelid closure, which was noticed by her parents at 2 months of age.

On examination, she was able to follow objects with both eyes appropriately for her age. She was noted to have alternating episodes of ptosis (interpalpebral fissure 2–3 mm), exotropia and hypotropia, and 6 mm mydriasis, which lasted around 40 seconds. The affected eye would then recover, and the other eye would undergo the same changes of mydriasis, ptosis, and exotropia with hypotropia. The diagnosis of COPS was made.

She had no other neurological deficits. MRI of her brain, VEP, and ERG were normal, and she was seen by a geneticist to exclude any syndromic relationship between the COPS and her tracheoesophageal fistula.

An examination under anesthetic a year later at the age of 18 months was significant for right medial and inferior recti tightness as well as superior and inferior obliques on forced duction. All extra-ocular muscles save for the lateral rectus were tight in the left eye. She had normal discs and maculae but a granular appearance to her peripheral retina.

Despite the initially symmetrical presentation, her left eye became much more affected than her right eye. Gaze restriction was more severe in the left eye with full abduction but significantly reduced adduction (-2), depression (-4), and elevation (-3). Worsening of the left ptosis over the next two years resulted in occlusion of the left visual axis, necessitating a frontalis sling. Post-operatively, there was a clinical concern for left eye amblyopia. Patching of the right eye, as well as correction of refractive errors with spectacles, was undertaken.

At present, that patient is 7 years old and has a high astigmatism in both eyes and no evidence of stereopsis. Her best corrected visual acuity is OD 6/9 OS 9/12. The patient will continue to be monitored closely and aided in schooling as necessary.

3. Discussion

Cyclic oculomotor nerve paresis and spasms is a non-progressive condition defined by alternating spastic and paretic phases of the muscles supplied by the oculomotor nerve [1]. This condition was first reported by Rampoldi in 1884 [1]. While the origin of the disorder is still debatable, the most accepted theory is a combination of central and peripheral nerve damage, as suggested by Loewenfeld et al. [2]. They have postulated a primary lesion of the intracranial oculomotor nerve at birth and during early infancy, followed by secondary innervation changes in the oculomotor nucleus, resulting in cyclic spasm [2]. In effect, it seems to be a disorder of aberrant innervation [3].

The condition is noted in 92.5% of patients before 1 year of age and the etiology may be related to birth trauma or infections during infancy [1].

This disorder is characterized by a paretic phase – ptosis, mydriasis, exotropia and hypotropia, and lateral gaze - this alternates with a spastic phase – ptotic eyelid elevates, the eyeball is orthotropic, and miosis [1]. The paretic phase normally lasts 1–3 minutes, and the spastic phase 20 seconds but up to 1.5 minutes [1]. In the majority of these patients, neuroimaging is unremarkable.

The disorder is normally unilateral, with the right eye being affected more commonly [1]. This often results in amblyopia of the affected eye [1, 3, 4].

There is currently no proven treatment for COPS. Strabismus surgery with recession of the lateral and medial recti can assist with the exotropia [4]. Levator shortening and lid suspension can assist with the ptosis but must be carefully weighed with the risk of corneal exposure [1, 4]. Currently, no treatment has been proven to have an effect on cyclic spasms [5].

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