Synergistic Convergence in Congenital Extraocular Muscle Misinnervation

Synergistic divergence is a well-established clinical condition in which abduction arises on attempted adduction, leading to simultaneous abduction of both eyes on lateral gaze. Synergistic divergence can occur as an extreme form of Duane syndrome, where most if not all oculomotor nerve branch fibers originally directed to the medial rectus muscle innervate the lateral rectus muscle.1 Synergistic convergence is an extremely rare form of ocular motor synkinesis characterized by simultaneous adduction on lateral gaze. It has only been described once in a case of congenital fibrosis of the extraocular muscles.2 Here we describe a girl with isolated congenital synergistic convergence and discuss its probable pathophysiology.

Report of a Case. A 5-year-old girl had orthophoria and reduced stereovision in primary position. On right gaze, she fixated with the left eye in adduction, and a large esotropia of about 52 prism diopters (Δ) appeared. On left gaze, she fixated with the right eye and, very similarly, a large convergent angle of greater than 50° appeared (Figure 1). Thus, the right and left gazes evoked a similar convergent misalignment of the eyes. However, on changing the gaze direction, we observed a marked modification of the eyelid fissure. Due to globe retraction of the nonfixating eye, the right eyelid fissure narrowed during right gaze, whereas the left eyelid fissure narrowed on left gaze (eFigure1 [available at http://www.archophthalmol.com]). No ptosis was apparent on primary gaze. Upgaze and downgaze were normal.

There was no change in pupil diameter with change of gaze. Convergence could be elicited up to 6 cm with no difference in the eyelid fissures. Magnetic resonance imaging examination was carried out at a 3-T magnetic resonance scanner (Trio Tim Siemens, Erlangen, Germany). The examination protocol requested axial and sagittal T2-weighted images with 5- and 6-mm slice thickness, axial T1-weighted images of the orbit with 2 mm, coronal and sagittal T2–fat-suppressed slices with 2 mm, coronal T1–fat-suppressed slices with 3 mm, and a T2-weighted 3-dimensional Constructive Interference in Steady State sequence with 0.5-mm slice thickness enabling secondary reconstructions along the cranial nerves. The oculomotor nerves showed a slight difference in thickness (1.80 mm for the right and 1.50 mm for the left) (Figure 2A), with the left one being slightly thinner than the mean (SD) of 2.01 (0.36) mm in healthy subjects.3 Abducens nerves (eFigure 2A) had a normal configuration at the subarachnoid space course. The orbits and the nucleus regions of the brainstem revealed no abnormality. Extraocular muscles were normal in size, showing no fibrotic changes (Figure 2B). There was no family history of ocular misalignment, ophthalmoplegia, or abnormal eye movements.

Comment. Synergistic convergence has been described only once in a patient who had congenital fibrosis syndrome and whose eye motility was limited in all directions in both eyes.2 Magnetic resonance imaging in this patient disclosed absence of the abducens nerves as well as hypoplasia of the oculomotor nerves with atrophy of the superior and medial rectus muscles of both eyes. Pronounced globe retraction depending on gaze direction was not present. By contrast, our patient had no ptosis, no extraocular muscle hypoplasia, and no family history to suggest the diagnosis of congenital fibrosis syndrome. Therefore, our patient’s pathomechanism of synergistic convergence is most likely different. Her congenital, isolated synergistic convergence with globe retraction suggests aberrant nerve sprouting during embryogenesis as the underlying cause. It might be due to a pattern of aberrant innervation similar to that in Duane syndrome, which leads to synergistic divergence (type IV). However, instead of aberrant oculomotor nerve fibers, here we suspect a miswiring of abducens motor neurons to the medial rectus muscle. If most of the abducens nerve fibers were misdirected to the medial rectus muscle, intended abduction would lead to adduction of the eye, and the co-contraction of the lateral and medial rectus muscles would result in globe retraction and eyelid fissure narrowing (Figure 3). In contrast to Duane syndrome, in which there is evidence of primary abducens nerve hypoplasia4 that predisposes to the miswiring, our patient showed no clinical or neuroimaging signs of oculomotor nerve hypoplasia (Figure 2A and B). Contraction of the medial rectus muscles during convergence was normal; thus, we observed no obvious explanation of aberrant nerve sprouting. An unusual congenital sixth-nerve palsy similar to congenital third-nerve palsies5 should also be considered a cause of miswiring. Congenital third-nerve palsies often occur with anomalous reinnervation and no other neurologic or systemic ab-

Figure 1. In primary gaze, the eyes are mostly parallel and no difference in eyelid fissure is visible. In right and left gaze, a tremendous esotropia becomes obvious. Depending on the gaze direction, one observes a retraction and narrowing of the eyelid fissure of the nonfixating eye.
normalities. Most regenerated abducens fibers would have been directed to the medial rectus muscle in our patient. However, a misdirection of regenerated nerve fibers to a muscle originally innervated by a different cranial nerve is less probable than a primary aberrant abducens nerve innervation.

Figure 2. Magnetic resonance images. A, T2-weighted images (3-dimensional Constructive Interference in Steady State sequence, 0.5-mm slice thickness) with axial and sagittal reconstruction along the oculomotor nerves (black bars). Cerebrospinal fluid is depicted in white. There is a slight difference in nerve thickness, showing a 1.50-mm diameter of the left nerve and a 1.80-mm diameter of the right nerve. A indicates anterior; R, right; L, left; 1, pons; 2, medulla oblongata; 3, posterior communicating artery; and 4, infundibular stalk. B, Axial T1-weighted images of the midbrain and pons demonstrate the integrity of the oculomotor and abducens nerve nuclei. A indicates anterior; 0, midbrain; and 1, pons.
Synergistic convergence is a rare variant of congenital extraocular misinnervation syndromes. Our case demonstrates that it can develop bilaterally in the absence of deficient ocular motor innervation to the extraocular muscles.

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Pituitary Apoplexy Causing Isolated Blindness After Cardiac Bypass Surgery

Pituitary apoplexy is a clinical syndrome that usually results from infarction of, or hemorrhage into, a pituitary macroadenoma. Typically, there is a rapid increase in tumor volume resulting in the abrupt onset of a variable combination of symptoms and signs that may include headache, meningismus, vomiting, visual loss, ophthalmoplegia, and stupor. Many factors have been implicated as precipitants, including major surgery. We describe 2 patients who developed blindness, without any other symptoms or signs to suggest pituitary apoplexy, due to infarction of undiagnosed pituitary macroadenomas during coronary artery bypass grafting (CABG).

Report of Cases. Case 1. A 79-year-old man with no prior symptoms to suggest neuro-ophthalmologic or endocrine disease underwent CABG. Following extubation, he reported blindness but denied headache. On examination, he was alert and oriented and there was no meningismus. He had no light perception OU. His pupils were 2 mm in diameter and unreactive to light. Examination findings were otherwise unremarkable.

A large pituitary mass was evident on computed tomography (CT) (Figure, A). On magnetic resonance imaging (MRI), the mass was seen to be extending into the suprasellar cistern to compress the optic chiasm (Figure, B). Pituitary apoplexy was diagnosed and dexamethasone sodium phosphate (8 mg) was administered intravenously. Transsphenoidal decompression was performed 24 hours after CABG. Histopathologic examination of the tissue revealed a necrotic pituitary adenoma. Postoperatively, the patient’s pupils became dilated but remained unreactive, and optic atrophy developed. On follow-up, he remained blind with no light perception OU.

Case 2. A 64-year-old man with no prior symptoms to suggest neuro-ophthalmologic or endocrine disease underwent CABG. Following extubation, he reported blindness but denied headache. On examination, he was alert and oriented and there was no meningismus. He had no light perception OU. His pupils were 2 mm in diameter and unreactive to light. Examination findings were otherwise unremarkable. Because it was thought that a stroke had produced his blindness, treatment with intravenous heparin sodium was commenced.

A large pituitary mass was visible on CT. On MRI, the mass was noted to be extending into the suprasellar cistern to compress the optic chiasm. There was no stroke. Pituitary apoplexy was diagnosed, heparin was stopped, and dexamethasone sodium phosphate (12 mg) was administered intravenously. Transsphenoidal decompression was performed 64 hours after CABG, following the correction of thrombocytopenia (platelet count, 72 × 10^9/L) and coagulopathy (international normalized ratio, 2.3; activated partial thromboplastin time, >180 seconds). Histopathologic examination of the tissue revealed a necrotic pituitary adenoma with secondary hemorrhage. Postoperatively, the patient’s pupils became dilated but remained unreactive, and optic atrophy developed. On follow-up, he remained blind with no light perception OU.

Comment. Cardiac bypass surgery is rarely implicated as a precipitant for pituitary apoplexy. In one series reviewing 11 cases, 1 patient was known to have a macroadenoma and 3 had symptoms or signs of endocrine or neuro-ophthalmologic disease prior to surgery. Postoperatively, all developed multiple symptoms and signs to suggest a diagnosis of pituitary apoplexy. In contrast, our patients developed blindness alone. To our knowledge, these are the first reported cases of pituitary apoplexy causing isolated blindness following cardiac bypass surgery. The blindness resulted from chiasmal compression, and loss of the pupillary reaction to light was