Delayed Oculogyric Crises Associated with Striatocapsular Infarction

Grant T. Liu, M.D., Enrique J. Carrazana, M.D., Jeffrey D. Macklis, M.D., and Mohamad A. Mikati, M.D., F.C.P.

Oculogyric crises are dystonic, usually upward, conjugate eye deviations. We describe an 11-year-old girl who developed oculogyric crises 3½ years after infarction of the right caudate, putamen, and internal capsule. Her abnormal eye movements responded to anticholinergic agents. This is the first reported association between oculogyric crises and striatocapsular infarction.

**Key Words:** Basal ganglia—Dystonia—Oculogyric crises—Stroke—Varicella.

Oculogyric crises are a dystonia of ocular muscles (1) characterized by dramatic involuntary conjugate deviation of the eyes, usually upward, lasting seconds to hours (2,3). Frequently they are accompanied by other dystonic or dyskinetic movements such as tongue protrusion, lip smacking, blepharospasm, choreoathetosis and anterocollis (4–7). In the 1920s the French first described crises (crises oculogyres) as complications of postencephalitic parkinsonism (2,8), but today such crises are thought to be usually acute (3) or tardive (4–7) extrapyramidal reactions to neuroleptics. In contrast, we describe a patient who developed a striatocapsular infarction and delayed oculogyric crises, a heretofore unreported association.

**CASE REPORT**

An 11-year-old right-handed girl presented to the neurology service complaining of uncontrollable annoying eye movements. She was born with a double outlet right ventricle and pulmonary stenosis. At age 5 she underwent a right atrium to pulmonary artery anastomosis and intra-atrial baffling of the pulmonary venous return (modified Fontan procedure). Her postoperative course was complicated by persistent pericardial and pleural effusions, requiring chronic steroid treatment and Denver pleuropertoneal shunting.

At age 7 years, 5 months, while still on steroids, she contracted a primary varicella infection treated with acyclovir. Two months later she developed a right frontal headache, then acute weakness of the left face, arm, and leg. Initially, computerized tomography of the brain was normal; however, a repeated study 11 days later documented an infarction in the right basal ganglia and internal capsule. A work-up for the etiology of her stroke was unrevealing.
Within weeks her hemiparesis improved, although she developed dystonic posturing of the left hand, marked by finger hyperextension that was worsened by hand or finger activity. There was no family history of dystonia or tics. Eight months after the stroke, the posturing spontaneously resolved, and the hemiparesis was mild.

At age 11 she began experiencing rolling eye movements which gradually increased in frequency and intensity over 4 days and interfered with her schoolwork and play. She had no headache, diplopia, or abnormal thoughts. The eye movements were not preceded by any irresistible urge, and she did not exhibit coprolalia, echolalia, stuttering, or spasmodic dysphonia. Her medications included aspirin and Inderal for migraine headaches, and Lasix, Aldactone, and digoxin.

Her neurological exam was remarkable for left hemihyponasia, hemiparesis, and hyperreflexia. There was neither dysmetria nor gait ataxia. Visual acuity and fields and funduscopic exam were normal. Her pupils were equal and normally reactive.

Eye movements. Primary gaze was conjugate, and vergence movements were normal. Randomly and independent of position, her eyes deviated conjugately upward and obliquely to the right or left. The episodes lasted 1-3 seconds, and then her eyes returned to primary position. During a timed typical 1-minute period, 5 events were oblique to the right, and 6 were oblique to the left. She was unable to suppress them, and they persisted during venipuncture while she cried. During observation without her awareness, the eye movements continued. Frequently she manifested brief left and rightward spasmodic, dystonic head turning which was temporally and directionally unrelated to her abnormal eye deviations.

Electroencephalography (EEG) during the eye movements showed no epileptiform activity. Sedimentation rate, antinuclear and antistreptolysin-O antibodies, copper and ceruloplasmin levels, and cerebrospinal fluid and toxicology screen were all normal. Magnetic resonance imaging of the head demonstrated the old infarction within the right caudate, putamen, and internal capsule (Fig. 1).

The patient was given intramuscular placebo (3 cc 0.9% NaCl) and then Benadryl (50 mg/3cc 0.9% NaCl). The eye movements did not respond to placebo despite suggestion. However they responded to Benadryl gradually over 10-15 minutes. Their rate decreased from 8-10 movements per minute to 0–1 per minute.Cogentin (0.5 mg bid) was prescribed. Two months later eye movements continued to occur at a rate of 2 or 3 per hour, and the head turning resolved. Five months after their onset, the eye deviations were completely controlled. After a subsequent taper-off of Cogentin, the abnormal eye movements recurred; therefore, the Cogentin was restarted.

FIG. 1. Cranial magnetic resonance imaging (TR = 2000 msec, TE = 80 msec) revealed previous infarction of the right internal capsule, caudate, and putamen (arrow).

DISCUSSION

Our patient developed a right striatocapsular infarction either from a cardiac-source embolus in the setting of congenital heart disease (9) or a varicella-related delayed vasculopathy, which typically involves the lenticulostrate arteries 1–3 months after the initial infection (10). However, neither etiology could be verified with certainty.

Her spasmodic involuntary upward eye movements 3½ years later are reminiscent of oculogyric crises (2,3), and the head turning (which we feel was torticollis), as well as the response to anticholinergic agents, suggest such eye movements were indeed dystonic ocular deviations. They probably are not hysterical, because the eye movements persisted while she was unaware of observation and while she cried during venipuncture, and they did not respond to administration of placebo. The absence of a repetitive to-and-fro quality makes it unlikely that they are nystagmus, and they are too slow to be considered ocular flutter, opsochonius, or ocular myoclonus. Reverse ocular bobbing is primarily vertical rather than oblique.
and occurs in comatose patients (11). Dysmetric overshoot is related to pursuit. The normal EEG during the eye deviations rules out epilepsy.

Dystonic eye rolling and torticollis have been described in tic disorders (12-14), but for several reasons we feel that this is an unlikely possibility in our patient. Tourette’s syndrome is inherited, and children with this disorder have vocal tics and behavioral disturbances in addition to motor tics (14). Ninety percent of Tourette’s patients develop their first motor tic by age 10 (15). Isolated motor tics can be idiopathic or secondary to drugs or basal ganglia injury (‘acquired tourettism’) (14). Unlike most individuals with tics, however, the patient lacked a typical preceding irresistible urge and an ability to suppress the eye movements (14). Furthermore, tics respond to dopamine antagonists, whereas our patient’s eye movements improved withCogentin, which worsens tics (14).

Although oculogyric crises are normally associated with a thought or emotional disturbance, are preceded by a brief stare, and last for hours (2,3,5,8), they can occur without these features and last only a few seconds (2,3,6). The eyes usually deviate straight upwards or up and to the left or right, and the position can change from crisis to crisis (2). Oculogyric crisis are features of symptomatic rather than idiopathic dystonias (1). In addition to postencephalitic parkinsonism, oculogyric crisis have been associated with Parkinson’s disease (16), familial Parkinson-dementia syndrome (17), neurosyphilis, multiple sclerosis (18), ataxia-telangiectasia (19), Rett’s syndrome (20), cerebellar disease, trauma (21), acute herpetic brain stem encephalitis (22), and a third ventricular cystic glioma (23). Our patient had none of these disorders. Besides neuroleptics (3–7,24), Tegetrol (25), tetrabenazine (26), and several other drugs (21) can cause oculogyric crisis, but our patient did not use any of these. Hereditary risk factors for dystonia such as Wilson’s disease, Huntington’s disease, Hallervorden-Spatz disease, and metabolic disorders (1) were notably absent.

Rather, we attribute our patient’s oculogyric crises and torticollis to delayed effects of her original striatocapsular injury. Acutely she acquired a left hemiparesis and subacutely a dystonic posturing of the left arm, both of which eventually resolved. Contralateral focal and hemidystonias are known sequelae of striatal infarction (27). As in our patient, dystonias can manifest themselves several years after a nonprogressive cerebral insult, perhaps as a result of aberrant neuronal sprouting (28). In addition, caudate lesions can cause torticollis (27). To our knowledge oculogyric crises have not previously been associated with basal ganglia infarction.

Injury to the striatum can cause dystonias by interrupting extrapyramidal pathways from frontal association, premotor, and sensorimotor cortices to globus pallidus and substantia nigra, both of which then send fibers to the ventrolateral thalamus (29). Unfortunately we are unable to explain how a unilateral basal ganglia lesion can lead to upward and leftward as well as rightward deviation or to torticollis with bilateral head turning. The direction of the torticollis is usually only contralateral to the lesion (27).

The etiology of oculogyric crises is not certain. In postencephalitic patients, some authors believe they result from a release of supranuclear control of oculomotor centers due to injury to the corpus striatum or subthalamic nucleus (2). Onuaguluchi (8) hypothesized that these crises were due to an abnormal vestibulo-ocular reflex in the setting of brain stem lesions involving vestibular pathways. Leigh et al. (5) attributed the deviations to an incorrect efference copy of eye position. Based on the response to anticholinergic agents in neuroleptic-induced crises, they alternatively invoked a defect in mesencephalonic vertical gaze-holding mechanisms normally dependent on balanced cholinergic and dopaminergic systems (5). In our patient the association between oculogyric crises and a lesion in the striatum implies that this balance may be mediated within the cortico-striato-pallidol/nigral-thalamic loop (27,29).

The findings in our patient suggest that striatocapsular infarction should be added to the list of disorders associated with oculogyric crises.

REFERENCES


