Positional oculogyric crises

Case report

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A young woman with a cystic glioma in the region of the posterior third ventricle is presented. Each time she turned to the supine position she developed immediate, forceful, conjugate upward gaze (oculogyric crises) and an inability to talk. If she turned to either side, she was alert and oriented, and could talk and lie comfortably. After stereotaxic removal of the cystic fluid, this phenomenon disappeared and did not return, even when the cyst refilled.

KEY WORDS: brain neoplasm • glioblastoma • oculogyric crisis • stereotaxic biopsy

In 1955, Freeman and Russell reported six cases in which ptosis, miosis, and anhidrosis changed from one side of the face to the other when the patients turned from one lateral lying position to the other. In each of these patients, they found an intramedullary cyst, resulting from trauma, at the C-6 level. The alternating Horner’s syndrome disappeared after the cysts were evacuated, but returned in two patients when their cysts refilled with fluid. The authors concluded that changes in the pressure gradients, produced by movement of the cystic fluid brought about by changes in the patient’s position, caused an alteration in irritation of the adjacent spinal cord neurons to produce the alternating neurological findings. Ottomo and Heimburger reported a similar case in 1980. They found severe scarring, causing adhesions between the spinal cord, dura mater, and spinal ligaments, but no intramedullary cyst. The alternating Horner’s syndrome disappeared after the scarring was freed, but returned in a year, presumably when the scar tissue re-formed.

Neurological deficits that manifest only in certain body positions are rare, usually indicating a minute shift in a neuronal structure due to a change in the direction of gravitational pull. The most common neurological deficit arising with positional change involves the vestibular system, and is due to change in labyrinthine fluid movement. No report of a previous case of oculogyric crises produced by positional change has been found.

Case Report

This 26-year-old married woman was hospitalized because of severe explosive headache, vomiting, blurred vision, neck stiffness, and urinary incontinence, starting on the morning of admission. During the previous 2 years she had suffered from intermittent headache and fatigue almost every evening. The headache had been more severe for the few months before admission, accompanied by irritability and night sweats.

Examination. The patient was a healthy-appearing woman with clear consciousness. She refused to lie supine for examination because she developed an immediate, forceful, painful, conjugate upward gaze of her eyes (oculogyric crises) in that position. During these episodes she was unable to talk, but seemed to understand. Only the sclerae could be seen under her eyelids. Her neck was retroflexed and stiff. As soon as she lay on either side, preferably the left, normal ocular position and extraocular movements returned, and her neck became supple. The remainder of her general physical and neurological examination was normal. She did not have papilledema or measurable loss of peripheral vision. Computerized tomography (CT) on the day
of admission showed a large low-density mass with peripheral ring enhancement in the right parietotemoral region filling the posterior portion on the right lateral ventricle and displacing the posterior portion of the third ventricle to the left. The mass was multiloculated and contained several cystic spaces (Fig. 1).

Operation. Application of the stereotaxic headholding device was difficult because the forceful, painful, conjugate upward gaze (oculogyric crisis) and retroflexion of the neck appeared as soon as the patient was placed in the supine position. Two milligrams of fentanyl citrate was given intravenously, making it possible to apply the head-holder under local anesthesia. The patient was placed in the stereotaxic frame lying on her left side. In that position she remained alert and comfortable, and was able to talk during the remainder of the procedure. A biopsy of the tumor was obtained and 15 ml of clear yellow fluid under considerable pressure was evacuated. Microscopic sections of the tumor showed a glioblastoma multiforme. When the patient was removed from the head-holding device she was able to lie supine without discomfort or any evidence of the previous oculogyric phenomenon or retrocollis.

Postoperative Course. A CT scan obtained 2 days after the cyst was evacuated showed it to be the same size as preoperatively, with probe tracts in the tumor and air bubbles in the cyst cavity (Fig. 2). Hyperfractionation radiotherapy produced some decrease in the tumor mass, but the cyst enlarged. A second attempt to evacuate the cyst stereotaxically yielded 5 ml of fluid, necrotic tissue, and pleomorphic glioblastoma cells. The oculogyric crises did not recur. The patient died at home 10 months after the glioblastoma multiforme was diagnosed. Autopsy was not obtained.

Discussion

Oculogyric crises (conjugate forceful upward eye movements) are most frequently associated with Parkinson's syndrome. A recent group of reports link the phenomenon with tardive dyskinesia, arising after prolonged use of phenothiazines and even L-dopa. This suggests that a change in the chemical substrate, as well as in a particular anatomical location, can be implicated. Nashold and Gills have observed changes in ocular motility during stereotaxically guided stimulation of the human mesencephalon. They wrote that "neural mechanisms for these ocular movements are not localized to discrete areas or centers." They also failed to observe vertical conjugate gaze in any of the stimulation or lesioning procedures performed in the human mesencephalon. Bender reported conjugate upward gaze in animals with bilateral simultaneous stimulation in the midbrain. Hess recorded head- and eye-turning in cats in response to stimulation of the ventral thalamus and centrum medianum. Nashold and Gills did not see these responses in humans, although posterior sensory thalamus stimulation produced ocular convergence and downward eye movements in one patient. Even though no specific nucleus or tract that controls vertical eye movements has been identified, the mesencephalon at the level of the quadrigeminal plate is implicated in the reports of changes in vertical eye positioning.

Compression in the area of the quadrigeminal plate must have been present in the patient reported here. Paralysis of upward gaze (Parinaud's syndrome), usually associated with lesions in this region, was not seen during this patient's illness. Even though this tumor was in the approximate anatomical location often occupied by pineal tumors, she had paralysis of downward gaze, rather than the expected paralysis of upward gaze. She gave no history of using medications which might produce tardive dyskinesia or oculogyric crises. Failure of the forceful upward eye movements to recur after
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fluid (and presumably pressure) returned to the cyst raises some doubt as to a purely anatomical cause for her unusual symptom. Its persistence until the cyst was evacuated, even after large doses of glycerol and steroids decreased the signs of elevated intracranial pressure, lends credibility to its occurrence as the result of irritation of the mesencephalic tegmental region. The apparent paralysis of downward gaze, rather than the expected paralysis of upward gaze (Parinaud's syndrome) produced by lesions in this region, plus the excitation of neurons producing upward gaze (oculogyric crises) only when the patient assumed a supine position seems to be worth reporting, and may be recognized in future patients.

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References


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