Pupillary Areactivity in Hydrocephalus of Recent Onset

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ABSTRACT: A patient who presented with bilateral loss of all pupillary reactions and normal ocular motor function is reported. Investigation revealed the presence of massive hydrocephalus. The syndrome developed shortly after transsphenoidal surgery for a suprasellar craniopharyngioma. Pupillary function returned to normal following the insertion of a ventriculo-peritoneal shunt. It is suggested that the syndrome was due to compression of the visceral oculomotor nuclei by a dilated sylvian aqueduct.

RÉSUMÉ: Absence de réactivité pupillaire dans l'hydrocéphalie d'apparition récente Nous rapportons le cas d'une patiente qui s'est présentée avec une absence des réflexes pupillaires sans atteinte oculomotrice. L'investigation a mis en évidence une hydrocéphalie massive. Le tableau s'est installé peu de temps après une intervention par voie transsphénoïdale d'un craniopharyngiome supra-sellaire. Il est proposé que le syndrome était dû à une compression de la portion parasympathique du noyau du nerf oculomoteur par un aqueduct dilaté.

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Isolated bilateral internal ophthalmoplegia is a rare syndrome except in the context of pharmacologic blockade or ciliary ganglion disease. We report a patient in whom isolated loss of pupillary reflexes occurred in relation to hydrocephalus of rapid onset.

Case Report

A 22-year-old woman presented in February of 1989 for bilaterally decreased vision of eight months duration as well as headaches, vomiting and secondary amenorrhea without galactorrhea.

On examination, she was found to have a pale complexion, short height (1.35 meters) and a low blood pressure. Her visual acuity was 3/200 in the right eye and 10/200 in the left eye. Her visual fields were limited to a nasal island of vision in each eye. There was bilateral optic atrophy. The pupils were isocoric and reacted normally to light and near. The remainder of the neurologic examination was normal.

Investigation showed the presence of partial panhypopituitarism characterized by diabetes insipidus, hypocorticism and hypothyroidism. Computed tomography (CT-scan) revealed a large cyst encroaching upon the floor of the third ventricle. Following transsphenoidal decompression of the cyst the visual field enlarged to a bitemporal hemianopsia and the visual acuity improved to 20/100 in the right eye and 20/200 in the left eye. Only bloody material was found in the aspirate. The patient was discharged on hormonal substitution therapy. Two months following discharge she was re-admitted because of recurring headaches. Her neuro-ophthalmologic examination was unchanged. CT-scan showed that the cyst which had regressed following surgery was again present.

Transsphenoidal excision of the lesion was performed. Histologic examination of the surgical specimen established the diagnosis of craniopharyngioma. Following surgery, the patient developed watery rhinorrhea secondary to a cerebrospinal fluid fistula, which was sealed through a transsphenoidal approach. She was discharged shortly after. Nineteen days following discharge, she presented with a three day history of slight confusion and urinary incontinence. She did not complain of headache. She was oriented, alert and cooperative but had some difficulty recalling the names of the doctors who normally looked after her and appeared less lively and less concerned about her illness than usual. Her visual acuity was unchanged at 20/100 in the right eye and 20/200 in the left eye for distance and near. The pupils measured 6 mm and did not react to light or near. Instillation of pilocarpine 2% in the right eye (the only one tested) resulted in normal pupillary constriction. The oculomotor examination including vertical saccadic and pursuit movements was normal.

CT-scan showed massive enlargement of the lateral and third ventricles and of the sylvian aqueduct (Figure 1) as well as a dilated fourth ventricle. Following a ventriculo-peritoneal shunt she quickly recovered a normal mental state and normally reactive pupils. CT-scan showed a gradual decrease in the size of the ventricular system with the exception of the fourth ventricle which remained unchanged. Analysis of the cerebrospinal fluid removed at the time of the shunt showed 7 red blood cells, 9 lymphocytes, 0 polymorphonuclear cells, normal protein and glucose values and absence of growth on cultures.

Six weeks after the last admission the patient was readmitted because of fever, vomiting and abdominal pain. She was again found to have unreactive pupils to light but the near response was not recorded. She also had paralysis of upward gaze. CT-scan showed recurrence of the hydrocephalus. She was found to have staphylococcus epidermidis meningitis secondary to an infected malfunctioning shunt. The shunt was removed and she was treated with external ventricular drainage and antibiotics. On this regimen, the pupillary and ocular motor signs gradually resolved and the hydrocephalus regressed.

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Discussion

There exists a previous report of a patient with a craniopharyngioma and poorly reacting pupils.[1] In that patient bilateral internal opthalmoplegia was attributed to midbrain compression by a large tumor. We believe that in our patient the absence of pupillary reactivity was the result of hydrocephalus and that as such it bore no direct relation to her craniopharyngioma. A cause and effect relationship between the hydrocephalus and the pupillary areactivity is suggested by the absence of other neurologic abnormalities which can affect the pupillary reaction and the regression of the pupillary syndrome following ventricular shunting. The reason for the hydrocephalus is unclear. The absence of change in the size of the fourth ventricle following shunting is evidence that the hydrocephalus was due to an obstruction of the aqueduct.

It is likely that the hydrocephalus affected the pupils by compressing the midbrain. An alternative explanation such as bilateral oculomotor nerve compression is unlikely in view of the normal ocular motor function and the preserved sensorium.

Damage to the dorsal midbrain in hydrocephalus is well recognized.[2-5] It is attributed to compression of the commissure area situated dorsal to the aqueduct by an enlarged suprapineal recess or by a dilated third ventricle and aqueduct. The resulting clinical picture is that of the dorsal midbrain syndrome which is characterized by pupillary light-near dissociation and ocular motor abnormalities of which the most common is paralysis of upward gaze. Rarely, in this syndrome, the ocular motor function is initially spared and the pupillary reaction is impaired to near as well as to light.[6] However, when, as in our patient, both these atypical features are present at the same time, the diagnosis of dorsal midbrain syndrome becomes difficult to maintain.

Other mechanisms by which hydrocephalus could abolish the pupillary reflexes include the interruption of input to the pupilloconstrictor motor neurons from both the light and near pathways and interference with the function of the pupilloconstrictor motor neurons themselves. The first of these mechanisms is unlikely in our case because the fibers carrying light and near information are not in close proximity to the third ventricle or the sylvian aqueduct except at the level of the posterior commissure. Their involvement at the latter site is ruled out by the absence of other elements of the dorsal midbrain syndrome.

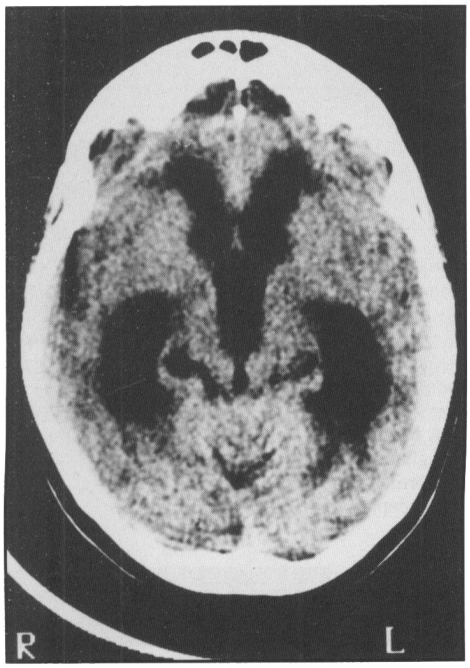

Figure 1 — CT-scan showing marked hydrocephalus.

We believe that in our patient the episode which was characterized by a lack of all pupillary reflexes is best explained by functional interference with the visceral motor nuclei of the third nerve from ventral compression by the dilated aqueduct. This interpretation is supported by the recent studies of Burde[7] which show that the visceral oculomotor nuclei, namely the anterior median nucleus and the dorsal visceral column, are closer to the third ventricle and aqueduct than is the somatic oculomotor nucleus. The reason for the pressure occurring mainly in a ventral direction, producing the pupillary syndrome described, rather than in a dorsal direction, giving rise to a dorsal midbrain syndrome as in the previously described cases, is not apparent.

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