Bilateral internuclear ophthalmoplegia associated with fourth ventricular epidermoid tumor

Case report

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A 49-year-old woman with multiple sclerosis diagnosed by her classical clinical history and bilateral internuclear ophthalmoplegia was found to have an epidermoid tumor of the fourth ventricle. Removal of the tumor resulted in resolution of her neurological symptoms and signs.

KEY WORDS • multiple sclerosis • bilateral internuclear ophthalmoplegia • epidermoid tumor • fourth ventricle

The patient who presents to a physician’s office with a long history of both intermittent and progressive central nervous system symptoms and a bilateral internuclear ophthalmoplegia usually has multiple sclerosis. The other diagnostic possibilities include various types of intrinsic brain-stem disease which, for the most part, carry such a poor prognosis that the diagnosis is primarily of academic interest. Yet, rarely, remediable lesions extrinsic to the brain stem have been said to cause bilateral internuclear ophthalmoplegia. This case is a reminder that patients with classical signs and symptoms of multiple sclerosis should have sufficient studies performed to exclude diseases amenable to surgical intervention.

Case Report

This 49-year-old woman had carried a diagnosis of multiple sclerosis for 5 years, and had been evaluated neurologically several times. Her symptoms began 7 years before the present admission, when she had episodes of vertigo that were aggravated by rapid movement of the head. She also developed episodic diplopia and unsteadiness in gait ranging in duration from a few hours to several days. She was admitted to a hospital at that time for evaluation and was told that an angiogram showed a possible lesion of an unknown type in the left posterior cerebral hemisphere. The patient declined further evaluation at that time. Over the next 4 years, she suffered episodes of diplopia, vertigo, and unsteady gait of varying duration and intensity. However, her functional status had changed only minimally in that she could still perform her usual household duties without assistance.

In 1976, she was again evaluated neurologically. Reports of this examination described horizontal and vertical nystagmus, a spastic-ataxic gait, diffusely hyperactive deep tendon reflexes, and bilateral extensor plantar responses. Because of the suspicion of a brain-stem tumor, a computerized tomographic (CT) scan was obtained, which was interpreted as probably normal but showing a large cisterna magna. Since 1976, she had been seen by several other physicians and had several courses of adrenocorticotropic hormone (ACTH) therapy. In the 3 to 4 months before being seen in our office, she had progressively increasing symptoms of ataxia, dysarthria, vertigo, diplopia, mental confusion, and a tendency to cry with the least provocation.

Examination revealed dysarthria, bilateral ptosis more severe on the left than the right, a moderately severe ataxic gait, vertical nystagmus on upward gaze, and a mild right central facial paresis. The most striking feature of her examination was the presence of a classical bilateral internuclear ophthalmoplegia (Fig. 1). There were no significant motor deficits in the extremities, but finger-to-nose testing showed mild dysynergia bilaterally, worse on the left than the right. Toe-to-object and heel-to-shin testing showed
mild dysfunction on the left. Deep tendon reflexes were normal in the arms and mildly hyperactive in the legs. A questionable left extensor plantar response was seen. There was a minimal diminution of pain over the left leg, trunk, and possibly the left arm, with no definite level. Visual acuity, visual fields, and funduscopic examination were normal. The patient was mildly disoriented to the date, and a little drowsy, but overall, her sensorium was intact.

The impression at the time of initial examination was that the patient had multiple sclerosis. The records of a recent hospitalization and the most recent CT scan performed 2 months previously were procured. These revealed the presence of a large radiolucent lesion in the midline of the posterior fossa with moderate hydrocephalus. The patient was admitted for further evaluation and treatment.

A CT scan on admission (Fig. 2) showed a large radiolucent midline posterior fossa mass with hydrocephalus. The patient underwent a suboccipital craniectomy with removal of a large cystic tumor that filled the fourth ventricle. The tumor contained oily material, and was completely removed except for that part of the capsule that was adherent to the floor of the fourth ventricle. Pathologically, it was an epidermoid tumor. Subsequently, the patient required a right ventriculoperitoneal shunt. After the first operation, the bilateral internuclear ophthalmoplegia showed moderate improvement, but was still present at discharge from the hospital. The shunting procedure did not produce any dramatic improvement in the eye signs. The ophthalmoplegia gradually improved to normal after several months (Fig. 3). Likewise, the patient's gait returned to normal and speech function improved considerably.

Discussion

Epidermoid tumors are rare, accounting for 0.3% to 1.8% of central nervous system tumors. These growths are not true neoplasms, and are the result of normal but ectopic ectodermal elements that accumulate keratin. Slowly progressive symptoms develop over a much longer time than one usually associates with intracranial tumors, with symptoms occurring at least 5 to 10 years before the tumor is diagnosed.

The location of this type of tumor is more variable than that of dermoid tumors, but 10% to 13% of intradural epidermoid tumors occur in the fourth ventricle. Although these tumors are almost invariably not infiltrating, brain-stem infiltration has occasionally been reported. The clinical signs and symptoms depend upon the location and size of the tumor. Organic mental symptoms are only rarely reported with epidermoid tumors, whatever the location. Extracranial movement disturbances are recorded very occasionally, although nystagmus is frequently present with intracranial epidermoid tumors, especially with lesions in the posterior fossa. However, we could find no mention of bilateral or unilateral internuclear ophthalmoplegia associated with this tumor.

Bilateral internuclear ophthalmoplegia is due to bilateral lesions of the medial longitudinal fasciculus, and for many years was considered “virtually diagnostic” of multiple sclerosis. Another neurological dictum stated that occlusive vascular disease usually produces unilateral internuclear ophthalmoplegia, but only exceptionally the bilateral type. Gonyea reported that while 26 of 41 patients with bilateral internuclear ophthalmoplegia had multiple sclerosis, 12 had occlusive cerebrovascular disease, and only three had other lesions. When tumors are a cause of
bilateral internuclear ophthalmoplegia, they are almost always intrinsic intra-axial lesions or else infiltrative to the brain stem. Only rarely have purely extrinsic tumors, including cerebellar astrocytoma, cerebellar tuberculoma, and supratentorial subdural hematoma, been implicated as the etiology. Other frequently described causes of bilateral internuclear ophthalmoplegia include syphilis, trauma, and Arnold-Chiari malformation.

The mechanism that explains the association of bilateral internuclear ophthalmoplegia with extrinsic lesions is at best speculative. Recently, Devereaux, et al. suggested that subdural hematoma could cause stretching of the basilar paramedian arteries, with resultant selective ischemia of the medial longitudinal fasciculus. They considered a vascular etiology more likely than actual brain-stem compression as the cause of medial longitudinal fasciculus dysfunction in patients with subdural hematomas. The end result, therefore, would be an intra-axial vascular lesion, a proposal that is consistent with the observed undramatic, very gradual improvement of bilateral internuclear ophthalmoplegia after removal of the subdural hematoma. In contrast, an argument for direct compression as a cause of this ophthalmoplegia could be made in cases in which the eye signs demonstrate rapid resolution. In one patient with Arnold-Chiari malformation and hydrocephalus, there was rapid resolution of the ophthalmoplegia following placement of a shunt, but it returned contemporaneous with the recurrence of the hydrocephalus. Our patient had a modest resolution of her bilateral internuclear ophthalmoplegia after removal of the tumor, but the eye signs continued to improve over the next several months. Shunting did not produce the dramatic improvement noted in the case of Arnold-Chiari malformation. The progressive improvement in our case is suggestive of a combination of direct compression and secondary vascular impairment as an etiology of the bilateral internuclear ophthalmoplegia, but the mechanism is far from clear.

In summary, although the combination of intermittent and progressive signs and symptoms in the presence of bilateral internuclear ophthalmoplegia is usually the result of multiple sclerosis, potentially remediable etiologies should be considered.

References


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