Multiple cranial nerve signs from supratentorial tumors

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A case of sphenoid meningioma with multiple cranial nerve signs is described in which the olfactory and optic brain tracts were involved unilaterally by the local effects of the tumor. All true cranial nerves except the eleventh were bilaterally, symmetrically, and selectively involved as a result of the general pressure effects of the tumor. The mechanisms that can produce this syndrome of multiple cranial nerve failure by a sequence of various brain shifts and hernias, hydrocephalus, and foraminal impaction, are discussed and demonstrated.

This case of a large sphenoid wing meningioma presented hydrocephalus, foraminal impaction with tonsillar herniation, and cingulate, central-transentorial, and uncal herniation, combined in such a sequence as to produce the unusual syndrome of multiple false localizing cranial nerve signs.

Case Report
This 42-year-old right-handed woman had had tinnitus, dizziness, and unsteady gait for 1 month, and occipital and posterior neck pain with intermittent, severe bifrontal headache, for 3 weeks. A week before admission she complained of progressive blurring of vision. Generalized weakness confined her to bed. Three days before admission she developed difficulty in swallowing and hoarseness.

Examination. The woman appeared extremely ill upon her arrival at the Montreal Neurological Hospital. She was obtunded but readily arousable. Her respirations were shallow and slow, and there were excessive oropharyngeal secretions due to inability to swallow. The pulse was 60 and blood pressure 160/80. She was able to answer questions when stimulated, and spoke intelligibly but in a soft whisper. There was moderate bilateral papilledema. The pupils were equal and midposition in size, but they reacted sluggishly to light and accommodation. There was restriction of all extraocular movement bilaterally. Corneal sensation and the corneal reflex were diminished on both sides. Bilateral facial weakness of peripheral type was present. There was absence of the gag reflex and an apparently complete paralysis of the palate, pharynx, and larynx. There was difficulty with tongue movement and inability to protrude the tongue. At intubation there was paralysis of the vocal cords, which were situated in the median plane. There was no motor or sensory deficit in the extremities but there were bilateral Babinski signs. There was no ataxia, the patient being tested only in the supine position.

Angiography. Because of the urgent nature of the patient's illness and the many
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Fig. 1. Left vertebral angiogram showing downward displacement of the brain stem, posterior inferior cerebellar, superior cerebellar, and posterior cerebral arteries. Note the elevation of the right middle cerebral artery which fills via the right posterior communicating artery.

brain stem signs, an emergency left vertebral angiogram was carried out (Fig. 1). The left posterior inferior cerebellar artery was displaced below the foramen magnum to the level of the lamina of C-1. Both superior cerebellar and both posterior cerebral arteries were displaced inferiorly, the right side more than the left. The brain stem was shifted downward to the left. There was filling of the right middle cerebral artery via the right posterior communicating artery. The Sylvian vessels were elevated and displaced mesially, indicating the presence of a supratentorial mass.

To investigate this displacement, a right carotid angiogram was performed (Fig. 2). The right posterior communicating and posterior cerebral arteries were displaced inferiorly and stretched proximally. There was subfalcial herniation of both the pericallosal and cingulate vessels 11 mm to the left. There was moderately severe ventricular dilatation. The Sylvian group was dislocated mesially, and multiple pathological vessels were present in the region of the sphenoid ridge. The middle meningeal artery was enlarged but there was also evidence of Sylvian middle cerebral supply to the tumor. There was a 14 mm shift of the internal cerebral vein to the left, and prolonged circulation time was noted. A diagnosis of right sphenoid ridge meningioma with hydrocephalus and associated subfalcial, transtentorial, and tonsillar herniations was made.

Preoperative Course. After angiography, complicated by a difficult intubation for the anesthetic attended by vomiting and aspiration, the patient was slow in waking, and a left hemiplegia was noted. Tracheostomy was performed. Mannitol and intravenous steroids were administered. She became unresponsive to all painful stimuli. The right
pupil was now larger than the left, and neither reacted to light. Hypotension and tachycardia supervened, along with a brief respiratory arrest. The left lateral ventricle was immediately punctured through a twist drill hole, and cerebrospinal fluid released under markedly increased pressure. Ventricular drainage was rapidly followed by marked improvement in her state of consciousness and by some diminution of facial and bulbar palsy and left hemiplegia. Pupillary reactions were restored, although the right pupil remained larger than the left. There was a tendency for head and conjugate eye deviation to the right. Seven hours later there was a transient episode of total blindness. However, maintenance of external ventricular drainage was accompanied by continuing improvement in the level of consciousness.

Operation. Thirty-six hours after admission the patient was alert and was taken to the operating room. A right external carotid ligation in the neck was followed by craniotomy with removal of a globular right sphenoid ridge meningioma 7 cm in diameter. Despite external carotid ligation, the bone and dura were extremely vascular. Only a small portion of the tumor was visible at the surface Sylvian point adjacent to the pterion. Both the frontal and temporal opercula were undermined by the extremely large meningioma. The tumor extended from the outer to the inner sphenoid ridge, along both lesser and greater wings of the sphenoid. Its blood supply originated from the middle meningeal artery, from pathological vessels along the entire sphenoid ridge, and from middle cerebral vessels in the depths of the Sylvian fissure.

Postoperative Course. The multiple bilateral cranial nerve signs regressed rapidly. The patient was alert, and her intermittent agitation and confusion persisted for only a few days. She had no evidence of any motor deficit 24 hours after surgery. The only persistent sensory deficits were anosmia on the side of the tumor, an incongruous left homonymous hemianopia with tunnel vision in the left eye (Fig. 3), and resolving papilledema with return of venous pulsations. At the time of discharge she was clinically asymptomatic, fully ambulatory, and able to care for herself.

Discussion

In 1926, Collier and Adie noted the relationship of false cranial nerve paralysis to elevated intracranial pressure and the dynamics of the shifting brain:

"Paralyses of cranial nerves are serious pitfalls. They are of value in localization when occurring early and in association
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with alternate hemiplegias, and paralyses of the eighth, ninth, tenth, eleventh and twelfth are always of sure localizing value. Paralysis of the sixth cranial nerve, perhaps, should always be disregarded as a localizing sign. . . With further increasing pressure . . . shifting backwards of the medulla and cerebellum (into the foramen magnum) will cause stretching of those cranial nerves attached to the medulla, in proportion as they are directed anteriorly, posteriorly, and take a straight course between their attachments to the dura mater and their origin from the medulla, and of these the sixth nerves will be most affected, and afterwards the third, seventh and fifth in that order. These nerves will not only be stretched, but are subject to the increased pressure also, and they may accordingly cease to function simply as the result of increased pressure.  

Cushing and Eisenhardt\(^6\) in their classical description of meningiomas divided the sphenoid ridge into three parts, since tumors arising at each site present a characteristic syndrome. The inner, deep ridge or "clinoid" meningiomas produce relatively early symptoms and signs (unilateral optic atrophy, exophthalmos, early oculomotor palsy). Middle ridge or "alar" meningiomas, on the other hand, usually attain enormous size before revealing themselves, at which time elevated pressure, papilledema, and brain shift appear. Global tumors of the outer ridge, "pterion" meningiomas or "Sylvian point" tumors, similarly produce symptoms and signs late in their growth (focal cerebral seizures, contralateral facial weakness, and choked discs). "Meningiomas en plaque" of the pterion produce a fourth and highly characteristic syndrome of hyperostosis of the greater wing, which because of its non-space-occupying nature, is not pertinent to the present discussion.

The meningioma in our case clearly had involved the entire ridge at the time of its clinical emergence. Medially, the anterior clinoid was eroded, while laterally at the pterion, the frontal and temporal lobes were separated. The middle ridge origin of this tumor is, as Cushing noted, compatible with clinical silence for many years, particularly on the non-dominant side.

Slowly progressive homonymous hemianopia may occur with middle ridge tumors on the basis of local pressure on the optic tract. The visual fields in this case suggest that, in addition to the constriction of the peripheral field related to papilledema, there was an underlying incongruous homonymous hemianopia. This is compatible with direct pressure on the adjacent optic tract, and contrasts with the congruous hemianopic field defect sometimes seen with a tentorial pressure cone resulting from stretching of the posterior cerebral artery and calcarine ischemia.

A unilateral anosmia due to rostral extension of the middle ridge tumor, and an incongruous field cut due to mesialcaudal tumor growth, could easily have passed unnoticed by the patient for a considerable period of time. The asymmetrical homonymous hemianopia of optic tract involvement is associated with normal central visual acuity as long as the chiasm is not simultaneously involved.\(^3\) This allows for a relative neglect of this symptom.

True cranial nerve involvement, from oculomotor to hypoglossal, awaited the development of hydrocephalus, brain shift, and herniation.

Plum and Posner\(^13\) described three major patterns of supratentorial brain shift: cingulate herniation, central-transtentorial herniation, and uncal herniation. Unilateral expanding lesions tend to result in midline shift with cingulate herniation before downward shift through the tentorial incisura occurs. The slow growth of the meningioma also favors this initial midline shift; downward herniations through the incisura are more com-
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mon in rapidly expanding supratentorial lesions. However, as pointed out by Johnson: 10 "Downward displacement may also be extreme in a slowly-growing tumor, especially if associated with a gradually developing hydrocephalus. It is important to recognize this when searching at autopsy for the cause of midbrain hemorrhages and for the cause of death; much of the impressive displacement may have occurred long before death and may have been compatible with a relatively healthy existence." Plum and Posner 13 also state that supratentorial mass lesions almost invariably produce a gradual, orderly failure of diencephalic, midbrain, pontine, and medullary functions, in that order. Our case represents an exception to this dictum, in that marked bulbar symptoms (dysphagia and hoarseness) and bulbar signs (complete palatal, pharyngeal, and laryngeal paralysis and paresis of tongue movement) were present during the period in which the patient was readily arousable. Bulbar paralysis is quite compatible with the conscious state. Transmission of pressure from supratentorial to bulbar levels can conceivably be accomplished via the ventricular system, provided that the aqueduct is not already blocked by midbrain compression at the incisura.

Our patient's history of severe and intermittent bifrontal headache associated with occipital pain is very suggestive of the "hydrocephalic attacks" described by Howell 8 and associated with marked dilatation of the ventricles including the fourth. "Foraminal impaction" of the cerebellum and medulla at the foramen magnum, described initially by Cushing, 5 prevents escape of ventricular fluid into the basilar cisterns during the hydrocephalic attack. Transient deafness and blindness may occur during these episodes. Howell reported several cases of bilateral cranial nerve palsy from the seventh to the tenth nerves resulting from foraminal impaction secondary to hydrocephalic attacks. Transitory bilateral deafness was the most common complaint. Dysphagia was noted in three patients. The seventh and eighth nerves, emerging rostral to the flocculus, and the ninth and tenth, caudal to the flocculus, were particularly liable to compression between the flocculus and the adjacent cerebellum in cases of foraminal impaction. Paralysis of the eleventh and twelfth cranial nerves as a result of impaction due to hydrocephalus was not reported.

Johnson and Yates 9 emphasized the importance of downward movement of the midbrain through the tentorial notch in the so-called "central-transtentorial syndrome." Both oculomotor nerves may be stretched and sharply indented by the petroclinoid ligaments as demonstrated by Blackwood, et al. 3 The resultant oculomotor palsy may occur in the absence of uncal or hippocampal herniation. Downward displacement of the brain stem also accounts for stretching of the abducens nerves between the pons and clivus during their long course to the point of dural exit. The basilar and posterior cerebral arteries are relatively fixed by their attachment to the circle of Willis, and stretching of their small branches by downward movement of the brain stem was postulated by Dott and Blackwood. 7 However, in the absence of coma and multiple long tract signs, stretching of midline arteries seems an unlikely cause of selective involvement of the cranial nerve nuclei.

The onset of left hemiplegia and anisocoria with right pupillary enlargement and deepening coma following angiography indicated the full development of uncal herniation with midbrain compression. This was first described by Meyer. 12 The concept was given a firm pathological basis by Kernohan and Woltman. 11 Vincent, et al., 15 coined the term "tentorial pressure cone" and later 15 spoke of both hippocampal and uncal hernias. Reid and Cone 14 experimentally demonstrated the relationship between fixed dilatation of the pupil and hippocampal herniation. Johnson 10 emphasized the importance of the more posterior incisural herniation of the hippocampus as a cause of "tentorial packing," terminal deterioration, and loss of upward movement of the eyes.

Thus, a combination of local and general factors accounted for the multiple cranial nerve signs in our case. Involvement of true cranial nerves was falsely localizing, in contrast to the unilateral local effects of the meningeoma on the olfactory and optic tracts. An internuclear ophthalmoplegia was closely mimicked by severe stretching of oculomotor, trochlear, and abducens nerves bilaterally between the dislocated brain stem and their dural exits. "Tentorial packing"
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with compression of tectal and preptectal areas may have contributed to the ophthalmoplegia. Diminished corneal sensation and the sluggish corneal reflex suggested bilateral trigeminal palsy compounded by symmetrical peripheral facial weakness.

As Collier and Adie noted, cranial nerves 3, 5, 6, and 7 produce false signs in the face of elevated intracranial pressure. Hydrocephalic attacks with foraminial impaction and tonsillar herniation resulted in bilaterally symmetrical paralyses of cranial nerves 7, 9, and 10, and possibly were responsible for the patient’s tinnitus, dizziness, and transient blindness. Pressure effects upon the hypoglossal trigone in the floor of the fourth ventricle, or at the hypoglossal canal adjacent to the foramen magnum, might be postulated to account for impaired tongue movement.

References


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