Upward Transtentorial Herniation
Seven Cases and a Literature Review

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- Seven cases of upward transtentorial herniation occurred. In each patient, coma with reactive, miotic pupils, asymmetrical or absent caloric responses, and decerebrate posture indicated brain-stem compression. In this setting, the development of unequal, then midposition, fixed pupils signaled midbrain failure from upward herniation. Vertebral angiography showed upward displacement of the superior cerebellar arteries. Results of autopsy confirmed the existence of growth of the vermis by the tentorial margins and, in one case, of anterior displacement and distortion of the midbrain. In five of 45 reported cases of upward herniation, the conditions were diagnosed antemortem. Instances of cerebellar hematoma and tumor predominated. In at least seven patients, performance of ventriculography may have precipitated herniation. Clinical details were provided in only nine patients and did not separate upward herniation from brain-stem compression. Cerebellar ischemic infarct found in one of our patients is a rarely reported cause of upward herniation.

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Upward herniation of posterior fossa structures through the tentorial notch is the least understood of the brain herniation syndromes. Most reports of expanding posterior fossa masses consider only compression and distortion of normal structures and downward herniation through the foramen magnum. Upward transtentorial herniation with midbrain compression seldom has been documented pathologically and rarely has been characterized clinically.

Two new cases of upward herniation of the cerebellum have been documented by serial examinations. We reviewed five additional pathologically confirmed cases in the files of the University of California Medical Center, San Francisco, but clinical details were insufficient for analysis. We also compiled 45 additional cases from a review of the literature. We reviewed the anatomy and pathophysiology of upward transtentorial herniation and analyzed the pathological findings in the seven new and 45 previously reported cases.

ANATOMY

The inner edges of the tentorium separate to form an oval opening, the tentorial incisura. As they descend anteriorly on either side of the midbrain, the inner, or free, edges of the tentorium pass below the superior colliculi and above or at the level of the inferior colliculi (Fig 1, top left). At the level of the tentorial notch, the superior vermis of the cerebellum lies closely opposed and just caudal to the dorsum of the midbrain. The midbrain is vulnerable to displacement by forces from below as well as from above the tentorium. A posterior fossa mass may expand dorsoventrally by compression of other posterior fossa structures, downward by displacement of the cerebellar tonsils through the foramen magnum (tonsillar herniation), or upward by displacement of the cerebellum through the tentorial notch (upward transtentorial herniation).

Direction Compression of the Brain Stem

When a mass lesion in the cerebellum increases in size, the pons and medulla are compressed against the clivus (Fig 1, top right). The fourth ventricle is distorted and obstructive hydrocephalus may occur.1,2 Infarction and hemorrhage may be a late and fatal result of compression of the vessels on the ventral surface of the brain stem.

Downward Tonsillar Herniation

As the mass effect increases, tonsillar herniation often occurs, which produces compression of the medulla and seals the inferior outlet of the posterior fossa (Fig 1, bottom left). Resultant obstructive hydrocephalus exerts a downward force at the tentorial incisura that opposes any upward movement of the cerebellum. Rapid descent of the cerebellar tonsils and impaction of the medulla causes sudden apnea and circulatory collapse. In almost half of a large series of patients with posterior fossa masses, tonsillar herniation was accompanied
by dorsoventral compression of the medulla, often with transverse grooving on its ventral surface caused by pressure against the anterior margin of the foramen magnum.

**Upward Transtentorial Herniation**

If tonsillar herniation is not fatal, increasing pressure in the posterior fossa causes upward herniation into and through the incisura (Fig 1, bottom right). Displacement of the cerebellum through the tentorial incisura is more likely to occur when the mass originates near the incisura, eg, in the cerebellar vermis, when drainage of the lateral ventricles relieves obstructive hydrocephalus and reduces pressure above, and when the opening in the tentorium is large.

The size of the tentorial opening is subject to wide variation that can influence the occurrence of upward herniation. Sunderland found that in the cadavers, the tentorial notch varied in length from 44 to 75 mm and in width from 26 to 30 mm. When the tentorial incisura is small, expansion upward is checked effectively and tonsillar herniation downward occurs first. When the tentorial opening is larger, upward herniation of the superior cerebellar vermis may occur more easily. As upward herniation develops, the displaced cerebellar vermis distorts the midbrain and cerebral aqueduct and buckles the quadrigeminal plate so that the inferior colliculi fold under the superior colliculi and together both structures shift upward beneath the splenium of the corpus callosum.

Galen's vein lies immediately above the posterior tentorial incisure. Herniation of the vermis through the notch displaces Galen's vein upward against the splenium and the unyielding free edge of the falx. Acute compression of Galen's vein may produce hemorrhagic infarction in the diencephalon and the adjacent white matter if venous collateral channels fail (case 3).

Hemispheric branches of the superior cerebellar arteries may be compressed by upward herniation of the cerebellum against the free edge of the tentorium, just as the posterior cerebral arteries that travel just above the tentorial edge are compressed in downward transtentorial herniation of the uncus. In some cases, this results in infarction of a portion of the...
cerebellar hemispheres below the tentorium.  

When a cerebellar mass expands, upward herniation is most prominent in the posterior aspect of the tentorial notch. By contrast, an expanding mass in the pons or the fourth ventricle causes more prominent anterior herniation, which displaces the pons into the interpeduncular cistern.  

The pons may be displaced as far as the infundibulum as the midbrain is carried anteriorly and superiorly. An extra-axial mass such as a cerebellopontine angle tumor causes asymmetrical upward shifts of the pons and the midbrain.  

REPORT OF CASES  

CASE 1.—Cerebellar infarction occurred in this case with upward herniation that was correlated with serial neurological examinations. A 23-year-old man with a history of psychiatric illness was admitted for observation after being found on a sidewalk amid broken glass below a shattered third-story window. He had numerous small lacerations on his extremities, but no evidence of head trauma. Results of neurological examination were entirely normal except for amnesia for the accident. Thirty-six hours after arrival, the patient became agitated and paranoid. He was given a total dose of 175 mg of chlorpromazine over a six-hour period.  

By next morning, the patient was unarousable but breathing spontaneously. Vital signs were as follows: blood pressure, 190/60 mm Hg; pulse rate, 90 beats per minute, and regular; respirations, 24/min, and regular; Pao₂, 51 vol%; Paco₂, 39 vol%; arterial pH, 7.40. After tracheal intubation, the results of a second determination of Pao₂ were normal. There was slight anisocoria, but both pupils reacted well to light. Corneal reflexes were present. Ice water caloric responses were absent. Noxious stimulation elicted decerebrate posture on the right and decorticate posture on the left. One hour later, the pupils were unchanged but ice water irrigation of the right tympanum caused partial conjugate deviation of the eyes to the right. Because of this change in caloric response and because the amount of chlorpromazine given was unclear at that time, the coma initially was thought to be drug induced.  

Lumbar puncture opening pressure was 240 mm H₂O and closing pressure was 130 mm H₂O. The CSF was clear and colorless. There was one mononuclear cell and 67 RBCs in the first fluid collected. The CSF glucose level was 97 mg/dL (simultaneous serum glucose level was 140 mg/dL). The CSF protein level was 81 mg/dL. Two hours after lumbar puncture was performed, the patient again lost all caloric responses, and the corneal reflexes as well. The degree of anisocoria increased such that the right pupil became 4 mm wide and the left pupil 2 mm, but both remained reactive to light.  

Carotid and vertebral angiograms showed severe enlargement of the lateral ventricles and a mass that expanded the vermis and left cerebellar hemisphere. Results of a right vertebral injection showed basilar artery compressed against the clivus, the right posterior inferior cerebellar artery pushed inferiorly, and the right superior vermian artery displaced upward. The left posterior inferior cerebellar artery did not opacify (Fig 2).  

Immediately after performance of angiography and before the patient could be taken to surgery, the pupils dilated and became fixed. Apnea required artificial ventilation. He died two days later.  

Autopsy results showed antemortem thrombosis of the left posterior inferior cerebellar artery with a large infarction of the left cerebellar hemisphere. There was no brain-stem infarction. The superior cerebellar vermis was forced upward.
through the tentorial notch, with distortion of the midbrain. There were no Duret’s hemorrhages of the midbrain.

An incomplete history misled the initial observer to suspect drug-induced coma; in retrospect, the early clinical deterioration was consistent with brain-stem compression due to the expansion of a cerebellar mass; that is, caloric responses were disconjugate at a time when the pupils still reacted to light. Thereafter, anisocoria developed with sluggish reaction to light, and finally the pupils became fixed and dilated. We suspect that distortion and buckling of the midbrain and the third cranial nerves due to upward herniation were responsible for the pupillary changes observed in this case.

The occurrence of a posterior inferior cerebellar artery occlusion in a young patient without preexisting cerebrovascular disease or bony abnormalities of the cervical spine is unusual. It seems likely that trauma was responsible. Sudden hypertensive neck movements have been followed by vertebral artery occlusion."

**Case 2.**—Upward herniation in this case followed rupture of an arteriovenous malformation in the cerebellum. In a previously healthy 40-year-old woman, a severe occipital headache developed. Two hours later, she vomited and rapidly lost consciousness. On arrival at San Francisco General Hospital, her blood pressure was 150/190 mm Hg, with a pulse rate of 100 beats per minute and regular respirations. The fundi were normal. The pupils were 2 mm wide and minimally reactive to light. Corneal reflexes were absent. There were no eye movements in response to ice water irrigation of the ear drums. All limbs were flaccid and without response to noxious stimulation. Tendon reflexes were normal on the left, hyporeactive on the right. The right plantar response was extensor, the left equivocal. Within 30 minutes of arrival, the pupils dilated to 5 mm wide on the right and 4 mm on the left, and were areflexic. Both plantar responses were extensor.

An angiogram showed that the right inferior vermian artery had aneurysms along its course and supplied a large arteriovenous malformation within the vermis. There was an associated mass. The superior vermian artery was displaced upward (Fig 3). A right cerebellar hemispheric hematoma was decompressed surgically, but the patient died four days later.

Autopsy results showed that the cerebral peduncles and midbrain tegmentum were compressed, and on their section, an upward herniation of the superior vermis and anterior brain stem were noted. There were no Duret’s hemorrhages of the midbrain, but there were punctate hemorrhages in the thalamus and in the splenium of the corpus callosum. The lateral ventricles were severely dilated. Examination of multiple sections through the cerebellum showed a large hematoma in the right cerebellar hemisphere that had replaced the vermis and filled the fourth ventricle. The surrounding tissue was edematous.

The initial presentation, with deep coma, small but reactive pupils, absent corneal reflexes, absent caloric responses, and bilateral signs of pyramidal tract dysfunction, suggested acute brain-stem compression. The later appearance of bilaterally fixed, dilated pupils indicated compression of the midbrain, probably from upward herniation.

Clinical details of the following cases were not available. They represent, however, five cases of autopsy-proved upward herniation.

**Case 3.**—A 62-year-old man became confused two months after the rupture and...
clipping of a right posterior communicating artery aneurysm. A ventriculoperitoneal shunt was placed after hydrocephalus was demonstrated by pneumoencephalography. Two weeks later, progressively severe nystagmus and gait ataxia developed, followed by rapid deterioration and death.

Autopsy results showed a large abscess of the left cerebellar hemisphere surrounded by edema. The superior vermis was herniated upward and grooved at the tentorial notch. Resultant distortion of the midbrain and pons was prominent. Bilateral tonsillar herniation with necrosis of the tonsillar tips had occurred. There was extensive hemorrhagic infarction of the hypothalamus, the medial thalamus, the midbrain, and the pons (Fig 4).

**Case 4**—A 34-year-old woman complained of headaches, tinnitus, and dizziness of three years' duration. Examination showed bilateral papilledema and right leg weakness with an atactic gait. A right vertebral angiogram showed hydrocephalus and a left cerebellopontine angle mass. A ventriculostomy was performed at the time of a left posterior fossa craniectomy. During the procedure, her pupils became fixed and dilated and she became hypotensive. She remained flaccid, unresponsive, and apneic until her death four days later.

Autopsy results showed a large left acoustic neuroma in which there was recent hemorrhage. An upward herniation of the cerebellum was found.

**Case 5**—A 5-year-old boy in 1966 was found to have a medulloblastoma and was treated with radiotherapy. He did well until 1971, when headaches and ataxia developed. In May 1971, a ventriculostomy was placed with relief of nausea, vomiting, and headaches. He became obtunded, and died in June 1971.

Autopsy results showed a cystic left cerebellar medulloblastoma, with a shift of posterior fossa structures to the right. Tonsillar herniation was prominent. The cerebellar vermis was herniated upward, overlying and compressing the colliculi (Fig 5).

**Case 6**—A 48-year-old man had confirmed metastatic malignant melanoma and headaches. Cerebral angiogram showed hydrocephalus and a posterior fossa mass. A ventriculogram confirmed the presence of obstructive hydrocephalus. A right ventriculoperitoneal shunt was placed with relief of his headache. He died one month later.

Autopsy results showed a massive tumor in the left cerebellar hemisphere with areas of old and new hemorrhage. Tonsillar herniation was present bilaterally, more noticeable on the left. There was upward transtentorial herniation of the cerebellum with compression and distortion of the midbrain, more on the left side. The aqueduct was pushed forward and the brain stem compressed.

**Case 7**—An 11-year-old boy underwent partial resection of an epedymoma of the fourth ventricle with postoperative radiotherapy in November 1970. In August 1971, headache and ataxia recurred and the patient died several weeks later.

Autopsy results showed a 5 x 6-cm tumor that replaced the entire vermis. There was moderate upward herniation of the superior vermis and severe bilateral tonsillar herniation.

**COMMENT**

**Pathologic Features**

In a review of the literature, we found 45 cases of upward transtentorial herniation confirmed at the time of autopsy.1-3,7,10-34 We excluded cases in which the pathologic changes suggested upward herniation but which were not so identified by the authors. Of these 45 cases considered with our seven cases, the lesion causing upward herniation was identified in 37 (Table 1).

A cerebellar lesion caused upward herniation most frequently (65%), often a hemorrhage. Medulloblastoma was the next most common lesion, a finding not unexpected since these tumors involve the cerebellar vermis and fourth ventricle and displace the vermis superiorly.

Upward herniation also was associated with lesions in the cerebellopontine angle (13%), in the pons (11%), and in the fourth ventricle (11%). It was impossible to determine for the entire series how frequently upward herniation coexisted with a direct compression of the lower brain stem and downward tonsillar herniation. We noted direct brain-stem compression in all seven of our cases and cerebellar tonsillar herniation in four. Direct brain-stem compression was referred to in only two of the 45 cases from the literature. In two other cases, tonsillar herniation was noted; it was not mentioned in the rest. In none was upward herniation specified to have occurred in isolation. Coexistent compression of the pons and medulla and tonsillar herniation may well have been present.

In nine of the 45 (20%) of the cases from the literature, ventricular drainage preceded death by an interval that ranged from minutes to days.5-12,14 Three of our seven patients in whom upward herniation developed had a ventricular shunt in place and the condition of a fourth patient deteriorated minutes after ventricular drainage was begun. Thus, in 18 of a total of 52 cases (25%), upward herniation occurred in the context of ventricular drainage. The possible danger of precipitating upward herniation by ventricular drainage should be emphasized; nevertheless, upward herniation occurred spontaneously in 75% of the cases reviewed.

**Angiographic Signs**

The angiographic signs of upward herniation have been documented previously by Perrett and Margolis.17 They are as follows: (1) upward displacement of the proximal portions of both the superior cerebellar and posterior cerebral arteries as they

<table>
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<th>Site of Lesion</th>
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<th>No. of Cases</th>
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<td>Cerebellum</td>
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**Total** | 37
course around the midbrain; (2) upward bowing of the superior vermian veins and the posterior mesencephalic vein; (3) anterior bowing of the precentral cerebellar veins; (4) these signs usually associated with anterior displacement of the basilar artery and anterior pontomesencephalic veins.

Clinical Features

In the cases of upward herniation that we found in the literature, clinical details were absent or did not suggest a consistent pattern. In a single case of upward herniation verified by cerebral angiography and ventriculography, Plum and Posner observed coma, hyperventilation, decorticate and decerebrate posturing, small fixed pupils, and absent vertical doll's eye movements with brisk horizontal responses. They concluded that the combination of miotic, fixed pupils (pontine compression) and absent vertical doll's eye movements (pretemporal compression) was diagnostic of upward herniation. The patient recovered after drainage of an abscess in the right cerebellar hemisphere. The presence of decorticate posturing, spontaneous respirations, and reflex horizontal eye movements, as well as the reversibility of coma, indicate that upward herniation was not far advanced in this patient. In our two patients in whom sufficient clinical data were available, anisocoria changing to fixed, midauditated, or large pupils occurred late in the course of the typical brain-stem compression syndrome. In both instances, anisocoria developed when oxygenation of blood and medullary respiratory and cardiovascular functions were well maintained. In case 1, fixed dilated pupils suggested dysfunction of the third cranial nerves. In case 2, the small, unequal pupils suggested midbrain instead of peripheral (infranuclear) third cranial nerve dysfunction.

Clinical-Pathologic Correlation

An expanding cerebellar mass produces a characteristic syndrome: coma, small reactive pupils, abnormal doll's eye reflex and ice water caloric responses, and signs of pyramidal tract dysfunction. Signs of midbrain dysfunction may occur in the course of brain-stem compression due to the presence of Duret's hemorrhages or venous infarction; however, neither direct pontomedullary compression nor downward tonsillar herniation will cause midbrain dysfunction before medullary compression has occurred. With pontine and medullary compression, the pupils remain equal and reactive until the end.

In our two patients, the progression from equal reactive pupils to anisocoria and then to midposition, fixed pupils indicated evolving upward herniation. In each case, pathologic evidence of upward transtentorial herniation with midbrain distortion and displacement was present. At the time of autopsy, the midbrain was free of infarction or hemorrhage. We postulate that upward herniation of the cerebellum through the tentorial notch caused these midbrain signs. Although no definite formulation of an upward herniation syndrome can be made from this limited number of observations, we suggest that the development of unequal fixed pupils in the setting of coma due to an expanding mass in the posterior fossa signifies upward herniation through the tentorial incisura.

Note added in proof: Two additional cases of upward herniation associated with infarction of portions of the cerebellum reported by Sypert and Alford were inadvertently omitted.

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Nonproprietary Name and Trademarks of Drug

Chlorpromazine—Chlor-PZ, Promacel, Thorazine.

References