Simple cysts of the cerebellum

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Four cases of simple cyst of the cerebellum treated at Stanford University Medical Center are presented. Review of the literature and past experience at Stanford suggest that they are uncommon. They seem to occur most often in middle age, presenting the signs and symptoms of an expanding cerebellar mass, and appear to be adequately treated by unroofing and draining the cyst cavity. It is suggested that these cysts do not have a congenital origin but rather represent a degenerated form of cerebellar astrocytoma.

KEY WORDS • brain tumor • cerebellar cyst • astrocytoma

In the past 10 years we have seen four case of so-called "simple cyst" of the cerebellum, presenting signs, symptoms, and radiographic evidence of an expanding cerebellar mass. At surgery or autopsy an intracerebellar cyst was found, filled with protein-rich, yellow fluid. No mural nodule was present, and multiple biopsies of the cyst wall failed to disclose the suspected astrocytoma. No connection with the fourth ventricle was seen. Microscopic examination showed an intraparenchymatous cyst, the wall composed of a dense astroglial proliferation, devoid of lining cells. Scattered throughout the glial wall were many thickened hyaline, apparently degenerated, astrocytic processes (Rosenthal fibers).

Although the literature contains many cases of cerebellar cyst, few well-documented cases of "simple cyst" have been reported. Because of the sketchy nature of many of the earlier recordings, it is not always possible to distinguish these cysts from presumably congenital cysts lined by ependyma or arachnoid, or cysts associated with tumor or parasites.

These simple cysts are uncommon, and their pathogenesis remains conjectural. Although in three of the following cases, there is not autopsy proof that a microscopic tumor was not present, the cysts were thoroughly explored and generously sampled at surgery without the discovery of any tumor mass.

Case Reports

Case 1

A 50-year-old physicist had increasing dizziness, headache, dysequilibrium, nausea, and vomiting for 1 month prior to seeking medical attention.

Examination. There was bilateral horizontal nystagmus, more marked on right lateral gaze, and vertical nystagmus on upward gaze. Romberg's sign was present, and the patient did poorly on tandem walking. There was a noticeable tremor on finger-to-nose testing on the left and ataxia on left heel-to-shin testing. No papilledema was seen and the reflexes were equal and active. The plantar responses were flexor. Lumbar puncture showed an opening pressure of 180 mm H₂O with no cells and a protein content of 68 mg%. Brain scan was felt to be diffusely abnormal, consistent with an "encephalitis," but carotid arteriograms showed significant hydrocephalus. A vertebral angiogram outlined an avascular mass in the right cerebel-
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lar hemisphere, later confirmed by ventriculography.

Operation. At suboccipital craniectomy, the right cerebellar hemisphere bulged outward, with thinning and flattening of the cortex posteriorly. Needle exploration disclosed a large cyst filled with yellow fluid (protein content 3.8 gm\%\). The cyst was surgically exposed by removing a portion of the posterior wall. It was seen that the cyst extended across the entire width of the right cerebellar hemisphere and through the vermis to encroach upon the left cerebellar hemisphere. Thorough exploration failed to uncover a mural nodule, and multiple biopsies showed no tumor on frozen section. Permanent sections showed a simple cyst. The patient made an uneventful recovery from surgery. When last seen 2 months postoperatively he seemed entirely well and no neurological abnormalities were noted.

Case 2

A 62-year-old woman for 7 months had had progressive ataxia and falling backward and to the left. Examination confirmed the ataxia with a tendency to fall to the left. Carotid and vertebral angiography were interpreted as normal. The patient refused a pneumoencephalogram. Five months later she returned with severe truncal ataxia with an intention tremor and past pointing on the left. No papilledema or nystagmus was noted. Reflexes were symmetrical, and the plantar responses were flexor. A pneumoencephalogram was attempted, but initial films showed bilateral cerebellar tonsil herniation. A ventriculogram then showed marked obstructive hydrocephalus, and a midline cerebellar mass was seen compressing the fourth ventricle.

Operation. Suboccipital craniectomy exposed a large cyst of the vermis, projecting into the right cerebellar hemisphere and filled with yellow fluid. No mural nodule was seen. The cyst lining was removed for microscopic examination and showed a simple cyst. The patient died of respiratory complications on the second postoperative day. Permission for postmortem examination was not obtained.

Case 3

A 52-year-old contractor was first seen because of progressively incapacitating headache occurring over the previous 6 months. No neurological abnormalities were noted at that time. One month later he returned with continued headache, vomiting, and unsteadiness of gait. A pneumoencephalogram was attempted but no air was seen to enter the ventricles. He was discharged only to return 2 weeks later with more severe headache, nausea, vomiting, double vision, and more incapacitating ataxia.

Examination. The patient was found to have bilateral early papilledema, bilateral horizontal nystagmus, more marked on gaze to the left, and vertical nystagmus on upward gaze. There was marked truncal ataxia with bilateral dysmetria. The deep tendon reflexes were symmetrical. Plantar responses were flexor, and no sensory impairment was found. A ventriculogram showed increased intracranial pressure with obstructive hydrocephalus, and a midline cerebellar mass was seen compressing the fourth ventricle.

Operation. Suboccipital craniectomy disclosed a cyst of the vermis the size of a hen’s egg filled with clear yellow fluid and having a protein content of 1.7 gm\%. Operative exposure showed no mural nodule and multiple biopsy specimens disclosed only a simple cyst. Except for mild residual truncal ataxia, the patient made a complete recovery and has been well for the past 3 years.

Case 4

This frail 61-year-old woman was first seen because of a 5 months’ history of dysphonia. On examination she was found to have paralysis of the ninth, tenth, eleventh, and twelfth cranial nerves on the left side. Plain x-ray films of the skull showed erosion of the left jugular foramen. She was followed as an outpatient for 9 years while her neurological signs and symptoms remained stationary. At the age of 70 she was readmitted to the hospital after 3 weeks of nausea, vomiting, headache, and unsteadiness of gait.

Examination. The patient was found to have, in addition to the previously noted cra-
FIG. 1. Case 4. The coronally sectioned cerebellum at autopsy. The cyst lies completely within the substance of the right cerebellar hemisphere, surrounded by white matter. It is displacing the fourth ventricle and vermis across the midline.

Cerebellar nerve palsies, hypotonia, ataxia, and an intention tremor of the left arm and leg. Routine laboratory investigations were normal. Plain skull films and tomograms showed the previously noted erosion of the left jugular foramen not to be significantly larger than when she was first studied. Vertebral angiography showed an avascular mass on the left in the posterior fossa. Because it was felt that she would not survive a posterior fossa exploration she was treated with 5000 R to the posterior fossa. She continued to deteriorate and died 1 month after completing the course of treatment.

**Autopsy Examination.** A 1.5 cm neurofibroma arose from the ninth cranial nerve on the left, excavating the jugular foramen, and a 3.5 cm simple cyst was found in the left cerebellar hemisphere (Fig. 1). There was a marked obstructive hydrocephalus secondary to compression and displacement of the aqueduct and fourth ventricle. Serial sections of the cyst failed to reveal any tumor.

**Neuropathology**

Multiple sections were examined microscopically and found to be similar in all cases. The cysts were situated within the cerebellar white matter: there was always a margin of granular neurons and/or a portion of the molecular layer between the cyst wall and the meninges (Fig. 2). Several Purkinje cells could still be seen near the cyst wall, but there was a marked loss of both Purkinje cells and granular neurons in the vicinity of the cyst.

The cyst walls were composed of a variable thickness of glial fibers (Fig. 3), in some areas nearly as thick as the normal molecular layer, in other areas thin and ribbonlike. Very obvious in almost all areas was a profusion of predominantly oval or club-shaped eosinophilic hyaline bodies (Fig. 4). These Rosenthal fibers appeared to be greatest in number near the inner margins of the cyst wall. No lining cells were identified. A flattened astrocyte nucleus was occasionally noted on the inner margin of the cyst wall. There were a few astrocytic nuclei within the wall of the cyst, but no evidence of tumor. Because of the crowding together of many cerebellar folia, the granular and molecular layers seemed almost fused in places, with intervening meningeal vessels giving some areas a very vascular appearance. However, there was no evidence of new vessel formation.

In Cases 1 and 3, several pieces of cerebellum were taken deep beneath the cyst wall. In Case 4 the entire adjacent cerebellum was examined. Several areas of the biopsy and autopsy material showed a large...
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number of interlacing, loose glial fibers in an area resembling micro-cystic degeneration. A number of large stellate astrocytes were seen within this glial stroma (Fig. 5) but insufficient cells were present to make a diagnosis of astrocytoma.

Discussion

In 1907 Henschen reported the autopsy findings in a case of ependymal cyst of the cerebellum and reviewed the literature on cysts of the cerebellum up to that time. He collected 17 cases of supposed simple cyst, but histological descriptions of the cyst linings were scanty. Williamson noted in 1910 that “in cases presenting the symptoms of cerebellar tumor, the lesion is sometimes found, at autopsy or operation, not to be a tumor but a cyst.” He reviewed 19 such cases but histology was lacking. Bartel and Landau reported eight cases of cerebellar cysts; however, the majority of their cases were associated with either macroscopic or microscopic tumor.

Antoni published three cases of cerebellar cyst, labeling them “syringomyelia of the cerebellum.” Two cases were in children who survived surgery, but one was that of a 45-year-old woman who succumbed after an unsuccessful exploration. At autopsy a 4 × 5 cm cyst was found in the right cerebellar hemisphere. No communication was noted...
with the fourth ventricle. On microscopic examination the cyst wall was made up of neuroglial fibers, rich in Rosenthal fibers. No lining cells were seen. The cyst wall was carefully studied with serial sections, and no tumor was found.

Lindau reviewed a large autopsy series of cerebellar cysts. He discussed many cysts associated with hemangioblastomas and gliomas, and also noted two cases of simple cysts without gross or microscopic evidence of tumor. As in Antoni's case, these cysts were within the parenchyma of the cerebellum and contained clear yellow fluid. They were composed of loosely interwoven, cell-poor glial fibers, many thickened and degenerated. No tumor was found on serial sections of the cysts.

Szigethy recorded another well-documented autopsy case of simple cyst of the cerebellum having only a glial fiber wall with no lining and no tumor. Finally, Cushing, in 1931, listed two cases of simple cyst within his series of cerebellar astrocytomas. Only one had autopsy documentation, discovered as a chance finding in a man who had a long history of cerebellar symptoms. The second case was apparently cured by surgery, but the lesion was carefully documented by Dr. Cushing's operative sketches. Both cysts were noted to have linings microscopically identical to the non-neoplastic portions of the cysts associated with cerebellar astrocytomas.

From the cases documented at autopsy (Table 1), an entity emerges that appears separate from other cystic lesions of the cerebellum.

Review of the cases gleaned from the literature (Table 1) plus our four suggests that simple cysts of the cerebellum occur in later middle age (average 52 years), and are as frequently lateral as medial in the hemisphere. They usually present a progressive cerebellar syndrome of headache, ataxia, nystagmus and, if they involve one hemisphere exclusively or predominantly, lateralizing cerebellar defects in the appropriate arm or leg. The slow progression of symptoms becomes more precipitous when hydrocephalus supervenes. Nothing in the history or presenting signs appears to separate simple cysts from other slowly expanding masses within the cerebellum. If one assumes that Cushing's surviving case and our two survivors were true simple cysts, then the prognosis is probably quite favorable if the cyst is partially excised and drained.

As there is no evidence for infection, trauma, vascular, or degenerative disease playing a role in the evolution of simple cysts, one is left with two possibilities: a) they arise from a congenital defect, or b) as a result of tumor degeneration.

### Table 1

<table>
<thead>
<tr>
<th>Author, Year</th>
<th>Age, Sex</th>
<th>History and Findings</th>
<th>Pathology</th>
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<tr>
<td>Antoni (1926)</td>
<td>45 F</td>
<td>5 mos of headache, ataxia, dizziness and vomiting; nystagmus, intention tremor, and dysdiadochokinesis on the right; staggered and fell to right</td>
<td>hydrocephalus secondary to a 4x5 cm cyst of the right cerebellar hemisphere, no gross tumor</td>
</tr>
<tr>
<td>Lindau (1926)</td>
<td>52 M</td>
<td>9 mos of progressive headache, dizziness, and vomiting; severe ataxia, bedridden for 3 mos before admission; nystagmus, stiff neck, right-sided dysdiadochokinesis, left extensor plantar, fell to the right; died prior to surgery</td>
<td>hydrocephalus secondary to a hen's egg cyst of the vermis filled with clear yellow fluid, rich in protein</td>
</tr>
<tr>
<td>Szigethy (1928)</td>
<td>47 F</td>
<td>museum specimen; no history available</td>
<td>cyst wall composed of cell-poor neuroglial fibers; no tumor seen and no lining</td>
</tr>
<tr>
<td>Cushing (1931)</td>
<td>3 M</td>
<td>no history except that the patient suffered from acromegaly and mild cerebellar symptoms</td>
<td>thin-walled cyst filled with xanthochromic fluid</td>
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Congenital Origin. The suggestion that these cysts are congenital in origin was initially championed by Antoni and Szigethy. Antoni drew an analogy between simple cysts of the cerebellum and syringomyelia. Szigethy postulated that the simple cyst arose from a congenital fourth ventricle diverticulum that had lost its connection with the ventricle and subsequently its ependymal lining, presumably from the effect of pressure atrophy. Favoring this theory is the fact that cases of intraparenchymatous cerebellar ependymal cysts clearly exist. The origin of these ependymal cysts is undoubtedly developmental.

As Schenk pointed out, they probably arise from a defect in the normal development of the primitive roof plate of the foetal hindbrain. It is possible that with the passage of time and the development of increasing pressure within the cyst, the ependymal lining might disappear. An analogy appears to exist in the formation of the pineal gland. In 1932, Cooper investigated the development of the pineal in relationship to the formation of symptomatic pineal cysts. She noted that with the growth of the two anlage of the pineal, a diverticulum became pinched off from the third ventricle within the gland which was gradually obliterated by a proliferation of pineal glia. There was a tendency noted for some of these congenital cavities (cavum pineale) to persist. Many retained a lining of ependymal cells, but cysts could be found devoid of a cell lining, displaying only a thick glial wall. However, in the four cases reported, where the cyst had enlarged the pineal enough to cause symptoms, a definite ependymal lining was seen. Cysts of the pineal without an ependymal lining did not appear to enlarge.

Alvord and Marcuse recently drew an analogy between simple cysts of the cerebellum and several cases reported of congenital cysts of the posterior fossa which they termed internal meningoencephaloceles. They speculated that these "meningoencephaloceles" arose from heterotopic glial nests and suggested that simple cysts of the cerebellum were an intraparenchymatous variety of this entity.

There are several objections to this thesis. First, the cases they reported and ones they retrieved from the literature were in infants or children rather than middle-aged adults. Second, one of their cysts clearly was lined by an ependyma-like cell. Third, the multiplicity of Rosenthal fibers so distinctive in our cases was not noted to be present in the walls of their meningoencephaloceles. Fourth, the glial constituent of their cases appeared to be islands of astrocyte nuclei, more cellular and much less fibrous than the glial cysts we are reporting. This glial tissue was closely approximated to a stroma of vascular connective tissue capped by arachnoid cells. This glia-arachnoid and cellular lining organization more suggests a congenital origin than do the simple cysts. Last, it would appear that congenital cysts seem to be mainly midline lesions, although presumed congenital cysts of the lateral recess occur. Simple cysts, often occurring laterally in the cerebellar hemisphere not related to the fourth ventricle or the lateral recess, suggest a different etiology.

Origin from Tumor

Several lines of evidence support the tumor theory of Lindau and Cushing. Foremost is the remarkable similarity, as noted by both authors, of these simple cysts to the non-neoplastic portion of cysts associated with cerebellar astrocytomas. The striking number of Rosenthal fibers, and the cell-poor, dense glial fiber network appear identical to many tumor cysts (Fig. 6). Similar also are the cyst contents. As opposed to the relatively clear fluid, most resembling CSF, found in the majority of congenital cysts, glial cysts much like tumor cysts, contain yellow fluid with a high protein content.

The benign course and occasional unexplained remissions seen in cerebellar astrocytomas suggest the possibility that these tumors may sometimes undergo spontaneous degeneration. Although most cerebellar astrocytomas will recur following incomplete removal, usually within 3 years, Bucy and Thieman have recently reported five patients with survivals up to 36 years following incomplete resection of their tumors. In several of these cases operation was limited to just biopsy and suboccipital decompression. Ringertz and Nordenstam also noted a surprising survival following incomplete resection of cerebellar astrocytomas. In their series, 18 cases had partial resection of the tumor. Ten patients were free of signs or symptoms of progression from 3 to 16 years.
Fig. 6. Case 4. A segment of the non-neoplastic cyst wall from a case of cerebellar astrocytoma presenting with a mural nodule and a large cyst at surgery. Note the similarity to Fig. 4. H & E, ×256.

after the incomplete removal. In one case where only biopsy was performed, the patient had survived eight years free of progression at the time of publication.

It is evident histologically that these tumors are prone to degenerative changes. The tumor astrocytes frequently show evidence of clasmatodendrosis, and their protoplasmic processes often become swollen and tortuous, breaking off to form the hyaline Rosenthal fibers. Indeed, in areas of degeneration it has been noted that the tumor cells may disappear entirely, leaving only a fiber framework. Gross and microscopic cystic degeneration is often seen, and degenerative changes in blood vessels are not uncommon. The areas of cystic glial stroma containing large stellate astrocytes seen in two of our cases are very similar to the degenerative areas seen in some cerebellar astrocytomas.

Against the tumor theory of Lindau and Cushing is the basic fact that no tumor is seen. However, when one considers the large number of degenerating fibers seen in the walls of these cysts it is reasonable to suggest that at one time a large number of cells may have been present. That these are the remnants of a microscopic astrocytoma is a not unreasonable assumption. The usual advanced age of the patient at the onset of symptoms argues against this hypothesis. Cerebellar astrocytomas tend to occur more commonly in children, with a mean of about 13 to 15 years in most series. The range, however, does extend up to about 60 years. Several authors have noted that the midline, more frequently solid tumors, tend to occur at an earlier age than the lateral, usually cystic tumors. In the series from Montreal, the average age of patients harboring a lateral cystic lesion was 34 years. Although cystic tumors are not uncommon in the very young, it is possible that cystic degeneration may be a function of the age of the patient as well as the age of the tumor.

It is tempting to consider a range in cases of cerebellar astrocytoma, from predominantly solid tumors to degenerated tumors with only a simple cyst remaining. Between these extremes would fall cases of cystic tumors with a mural nodule and cystic tumors with only microscopic tumor present, not necessarily as an evolutionary sequence, though this has been suggested. That these morphological variations in cerebellar astrocytomas occur is well accepted. That simple cysts are yet another variant of these tumors seems possible.

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