Cystic cavernous angiomas of the posterior fossa
Report of three cases

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Three histologically verified cases of cystic cavernous angioma of the posterior fossa are reported. The clinical history of these patients is presented, and the diagnostic and surgical problems arising from the extreme rarity of the combination of cerebellar site and cystic nature of the lesion are discussed.

KEY WORDS • cystic cavernous angioma • angioma • posterior fossa • cerebellum • surgical treatment

CAVERNOUS angiomas are relatively rare vascular malformations; they are generally located supratentorially and confined to one hemisphere. On histological examination they contain closely packed irregular vascular spaces separated by fibrous tissue. The walls of the malformation show hyaline fibrous thickening, old and recent thrombosis in varying stages, and, typically, no elastic or muscle fibers. No cerebral parenchyma lies between the vessels. 1~ Angiography is often normal, 3,7,8,11 and only computerized tomography (CT) scanning has permitted radiological diagnosis and definition of this nosological entity. 3,6,8,11,12 Cavernous angiomas may be cystic 5,13 and have been reported to occur in the posterior fossa. 8,15,16

We describe three such patients who were managed at our institution during the years 1983 and 1984. All three underwent total removal of the cyst, with good results.

Case Reports

Case 1
This 54-year-old man with a history of schizophrenia had for 2 months been suffering from intracranial hypertension of increasing severity. A week before admission the patient, who was totally unable to cooperate, had undergone CT scanning elsewhere, and had had a ventriculoperitoneal shunt placed (his mental status made it impossible to set up an external shunt). This shunt resolved the intracranial hypertension.

On admission, neurological examination showed papilledema and a vague right-sided cerebellar syndrome. Skull x-ray films were negative. A CT scan showed a cystic lesion of the vermis slightly to the right of the midline, with fairly regular enhancement of the cavity wall but no demonstration of an intramural nodule. The tentative diagnosis was a cystic astrocytoma or hemangioblastoma. At surgery, a complete removal of the cystic space-occupying lesion was accomplished. It lay immediately below the cortex and was easily separated from the surrounding parenchyma. It consisted of a cystic component (3 × 2 cm) containing lemon-yellow fluid and a grayish-red intramural nodule (2 × 2 mm). Histological diagnosis was cavernous angioma. A follow-up CT scan 1 month later confirmed total removal of the malformation, definite reduction of ventricular volume, and normal size and position of the fourth ventricle. One year after surgery, there was complete regression of the intracranial hypertension and of the cerebellar symptoms.

Case 2
This 53-year-old woman had had generalized asthenia for some months and increased intracranial pressure with ataxia of increasing severity for 10 days. Neurological examination on admission showed papilledema, with severe right-sided cerebellar signs. Skull x-ray films were normal, and right vertebral and carotid angiography showed marked triventricular hydrocephalus but no abnormal vessels. A CT scan demonstrated a cystic space-occupying lesion of the right cerebellar hemisphere with a small intramural nodule and marked
triventricular hydrocephalus. The initial diagnosis was cystic astrocytoma. At surgery, complete removal of a 4 x 2-cm cyst was achieved, along with a small reddish 2 x 3-mm intramural nodule which lay immediately under the cortex with a clear cleavage plane.

Histological diagnosis was cavernous angioma. The cyst wall was made up of normally structured cerebellar parenchyma, with small patches of recent bleeding. The inner side of the wall was coated with a single layer of flat cells. Histological examination of the lemon-yellow cystic fluid revealed only sporadic basophilic amorphous substance. A follow-up CT scan 7 days postoperatively confirmed total removal of the cyst and a definite reduction of ventricular volume. At follow-up review at 8 months, the patient complained of occasional giddiness. There was complete regression of the intracranial hypertension and cerebellar symptoms.

Case 3

This 44-year-old woman had a progressive 2-month history suggestive of increased intracranial pressure. Neurological examination on admission demonstrated papilledema and vague left-sided cerebellar signs. Skull x-ray films were normal, and left vertebral and carotid angiography showed no pathological circulation. Computerized tomography scans (Fig. 1) showed a cystic space-occupying lesion of the left cerebellar hemisphere, with an intramural nodule and triventricular hydrocephalus. Tentative diagnosis was cystic astrocytoma. An operation resulted in complete removal of the cyst along with the small brownish-red intramural nodule. The cyst contained about 10 ml of lemon-yellow fluid and was easily detached from its subcortical site in the left cerebellar hemisphere. Histological diagnosis (Fig. 2) was cavernous angioma. The cystic fluid contained amorphous material but no cells. A CT scan 8 days later showed that removal of the lesion was complete, and there was a definite reduction in ventricular volume. Follow-up review at 5 months showed persistence of a vague left cerebellar deficit.

Discussion

These three cases of cystic cavernous angioma of the posterior fossa had the same mode of onset: a fairly rapidly developing intracranial hypertension caused by compression of the fourth ventricle. Signs of altered cerebellar function were present. Regular x-ray films and angiography were normal in all three cases. The most useful diagnostic study was CT scanning, which not only defined the exact site of the lesion and its cystic nature but, in two cases, also revealed the presence of an intramural nodule which was slightly hyperdense before and modestly enhanced after injection of contrast medium. In one case the CT scan also showed fairly even enhancement of the cyst wall. Surgical cyst removal was total in each case, helped by the presence of an obvious cleavage plane, and resulted in almost immediate restoration of normal function of the cerebrospinal fluid (CSF) in two cases. In Case 1, ventriculoperitoneal shunting was necessary due to the patient's inability to cooperate. On CT examination 6 to 12 months after their operation, none of the patients had recurrence of the cyst, and the CSF pathways were functioning well.

In our experience and that of others, it is wise to consider the possibility of cavernous angioma in the differential diagnosis of an intracranial supratentorial

![Fig. 1. Case 3. Computerized tomography scans, before (upper) and after (lower) injection of contrast medium, showing a cystic space-occupying lesion with an intramural nodule.](image1)

![Fig. 2. Photomicrograph of the lesion in Case 3. The appearance is of a cavernous angioma. H & E, × 55.](image2)
Cystic cavernous angioma in posterior fossa

Cystic lesion. A pattern suggestive of a cavernous angioma includes a cystic lesion of a cerebellar hemisphere, associated with an intramural nodule that is faintly hyperdense on plain CT scans and enhances slightly on injection of contrast medium, where there are no afferent arteries, drainage veins, or pathological circulation on angiography. This presentation does not, however, rule out other cystic lesions of the posterior fossa, the most likely of which is a cystic astrocytoma. The subtentorial cystic cavernous angiomas that we observed all presented features similar to those of supratentorial lesions, plus a marked mass effect on the fourth ventricle.

The clinical course, in our view, points to a tendency of cystic cavernous angiomas to enlarge, as already documented for the supratentorial variety. None of the patients gave a history of episodes attributable to hemorrhage, notwithstanding histological proof of small bleeding foci, old and recent. We think that this can be explained partly by the extreme unlikelihood of a patient with a massive posterior fossa hemorrhage reaching the operating table and partly, as others have said, by the fact that CT permits earlier diagnosis. In all our patients the skull x-ray films were negative for calcifications, which have been reported in over 30% of cases in some series and were attributed to foci within the malformation or in the surrounding area of gli reaction. The absence of calcifications can probably be explained by the earlier onset of clinical symptoms severe enough to warrant hospitalization, considering that even a modest mass effect in the posterior fossa can cause obstruction of the CSF pathways. Angiography was normal in every case. This, in agreement with other reports, can probably be explained by the extremely fine vessels, the slow circulation, and the frequent presence of thrombi within the vascular spaces of this type of angioma. The results of radical removal of these lesions, helped by the presence of a well defined plane of cleavage and by sparing of the contiguous cerebellar tissue, are excellent.

References

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