Enterogenous cyst of the fourth ventricle

Case report

FARHAD AFSHAR, M.D., F.R.C.S., AND CARL L. SCHOLTZ, F.R.C.P.A.

Departments of Neurosurgery and Neuropathology, The London Hospital, Whitechapel, London, England

A case of a benign cyst within the fourth ventricle is described. The histology of the cyst wall lining resembled that of an enterogenous cyst of the spinal cord. The relationship between this cyst, colloid cysts of the third ventricle, and enterogenous cysts of the spinal cord is discussed.

KEY WORDS • ventricular cyst • colloid cyst • enterogenous cyst • neuroectodermal cyst

Intraventricular epithelial-lined cysts of the fourth ventricle are extremely rare, and the few reports found in the literature have suggested that they may be related by origin to the colloid or neuroepithelial cysts of the third ventricle. The present report describes a patient with a fourth ventricular cyst in which the histology of the cyst wall resembles that of an embryonic enterogenous cyst of the spinal cord. To our knowledge this characteristic has not been described before.

Case Report

This 48-year-old woman presented in July, 1979, with a 9-month history of progressive unsteadiness after two attacks of vertigo, each lasting 3 days. She was also subject to intermittent bouts of vomiting and had suffered from morning headaches for 3 months.

Examination. She was a healthy Caucasian woman with normal optic discs. She had a coarse nystagmus on right lateral gaze and a finer nystagmus on left lateral gaze. Vertical nystagmus occurred upon upward gaze. The right corneal reflex was depressed. No other cranial nerve deficits were noted. There was no upper limb ataxia, but she had mild ataxia of both legs on heel-to-shin testing, more on the right than the left. The reflexes were all brisk but symmetrical, and the plantar responses were downgoing. Heel-to-toe walking was unsteady with a tendency to fall to either side.

Skull x-ray films were normal. Computerized tomography (Fig. 1) showed a low attenuation area, approximately 3 cm in diameter, in the region of the fourth ventricle, with no contrast enhancement around the lesion. The third and lateral ventricles were of normal size. Vertebral angiogram (Fig. 2) showed an avascular mass situated low in the posterior fossa in the midline, with displacement of the basilar artery.
Enterogenous cyst of the fourth ventricle

Fig. 2. Anteroposterior (left) and lateral (right) vertebral angiograms showing displacement of supra- and retrotonsillar segments arising from the posterior inferior cerebellar artery on the left side (large arrows) and from the anterior inferior cerebellar artery on the right side (small arrows).

forward against the clivus. On the left side the supra- and retrotonsillar segments arose from the posterior inferior cerebellar artery (PICA), but on the right side these segments originated from the anterior inferior cerebellar artery (AICA). These segments were widely displaced from the midline and stretched backward. This finding was consistent with a mass lying in the fourth ventricle and extending down to the foramina of Luschka.

Operation. A midline cerebellar craniectomy was performed. The lower part of the inferior vermis was more prominent than normal and canted to the left. Between the two tonsils, a bluish thin-walled cyst was noted protruding from the foramen of Magendie. Further separation of the tonsils showed the choroidplexus to be stretched over its inferolateral surface. On either side of the midline, the PICA and its branches and the AICA were widely separated but closely related to the cyst wall. The cyst occupied the entire fourth ventricle, reaching as far as the aqueduct. The cyst wall was attached to the roof of the fourth ventricle and slightly to the right of the midline. During the dissection, the cyst ruptured, causing a thin clearish fluid to escape. The cyst wall was readily separated away from the floor of the fourth ventricle, except for an area on the right side laterally where it was lightly attached. The cyst wall extended into the substance of the brain in the roof of the fourth ventricle, but a plane of dissection was present. After removal of the cyst wall in one piece, the floor of the fourth ventricle and the aqueduct could be seen to be normal.

Postoperative Course. Postoperatively, the patient was mildly dysarthric at first. She exhibited spontaneous rotary nystagmus and marked rotary nystagmus on upward and lateral gaze to either side. The right corneal reflex was depressed, and a mild right facial weakness and sixth nerve palsy were noted. Mild dysmetria of the right lower limb was initially present.

At the time of discharge on the 12th postoperative day, the sixth and seventh nerve palsies were resolving, and a month later the patient’s only complaint was minimal unsteadiness of gait. The nystagmus had disappeared, and there were no cranial nerve deficits. No further limb ataxia was elicited.

Pathological Examination. The specimen consisted of an opened cyst, 1.5 cm in diameter, with a wall 0.3 cm thick. Microscopically, the cyst wall was composed of an outer coat of collagen supporting a thin basement membrane and layer of cuboidal epithelium which in places was pseudostratified (Fig. 3). The nuclei were round and basally situated. Many cells contained periodic acid-Schiff-positive mucin, which also covered the epithelial surface. The wall also contained several microcysts, and several argentophil cells were noted with silver staining. Cilia and blepharo-plasts were not identified. No mitoses were seen.

Discussion

Few reports of epithelial-lined cysts of the fourth ventricle can be obtained from the literature. Parkinson and Childe* reported two fourth ventricular cysts in one patient, who presented with formication over the left occiput, unsteadiness of gait, and blurring of vision. The cyst walls in their case showed several epithelial-lined tubules at a distance from the ciliated columnar epithelial lining of the cavity. The two cysts contained a homogenous clear gelatinous fluid. These
FIG. 3. Photomicrograph of the cyst wall showing pseudostratified living cells (arrows) and basement membrane. Bodian, × 115.

authors considered that the cysts were identical with the typical colloid cyst of the third ventricle, but believed this was not the site of origin in their case since passage of a large cyst through the aqueduct would be difficult. Kappers considered that most colloid cysts of the third ventricle were of paraphyseal origin. However, an origin from the choroid plexus, ependyma, or endoderm has also been suggested. Hirano and Ghatak stressed the presence of mucin on the surface and the prominent basement membrane, which to them suggested that these cysts have an origin similar to the enterogenous cysts of the spinal cord.

Shuangshoti and Netsky reported two further cysts in the fourth ventricle at autopsy (Cases 5 and 6), and considered both of these to be neuroepithelial in type, consisting of a thin fibrous wall with inner and outer linings of simple cuboidal epithelium. These cysts were subdivided into irregular spaces by cords of loose connective tissue fibers, and by the presence of lymphocytes and psammoma bodies scattered throughout the cyst wall. These authors considered that these cysts arose from foldings of the neuroepithelium lining in or out of the ventricular system of the brain or central canal of the spinal cord. They postulated that if the neck of these epithelial sacs became occluded ("pinched off") they might become separated and have no communication with the ventricle. If the neuroepithelium were to fold into the neural tissue it would become parenchymatous, whereas folding away from the neural parenchyma would result in a ventricular cyst. It was further considered that the inner lining of these cysts might be derived from ependyma, choroid plexus, or both.

The histological appearance of the present case has many features in common with colloid cysts of the third ventricle. The absence of cilia contrasts with the fourth ventricular cysts reported previously by Parkinson and Childe. These authors did not comment on mucin production, but this is a common feature of colloid cysts.

In our case, the findings of epithelial mucin on the epithelial surface and in the cells, as well as a thin basement membrane, are similar to the observations of Hirano and Ghatak, who interpreted these characteristics as giving colloid cysts an endodermal origin. The absence of cilia and blepharoplasts in our case makes the origin of the cyst from ependyma or choroid plexus unlikely. The presence of scattered argentophil cells in the region of the basement membrane supports an origin from endodermal tissue characteristic of the intraspinal cysts lined with epithelium of intestinal origin. These latter cysts are most commonly seen in the low cervical or upper thoracic region, although histologically similar-appearing cysts have been reported in the pons and upper medulla. The contents of these have been variable, ranging from mucoid to a thin cloudy fluid. In all these cases, the cyst wall has been lined with low columnar or cuboidal cells with evidence of mucous-containing cells.

We, therefore, consider the present case an example of an enterogenous cyst forming a continuum between the spinal enterogenous cysts and those cysts of the third ventricle which have an endodermal origin.

References


F. Afshar and C. L. Scholtz