Multiloculated Cystic Tumor of the Choroid Plexus of the Fourth Ventricle

Case Report

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Small, benign, asymptomatic cysts occurring within the choroid plexus are well known. Larger cysts, often causing symptoms, also occur but are rare within the fourth ventricle. In this report, we describe an extremely large, cystic mass located within the fourth ventricle which expanded into the adjacent subarachnoid space, producing a variety of symptoms that eventually led to the death of the patient.

Case Report

The patient, a 68-year-old white woman, was admitted to the Montefiore Hospital and Medical Center because of unstable gait and loss of manual dexterity. One year before admission she had had several “syncopal attacks,” and 1 month before admission she first noticed an unsteady gait, a tendency to fall to the right, incoordination of the arms and hands, and vague numbness of the toes and fingers. During the succeeding month she developed slurring of speech, intermittent ringing in the right ear, slight weakness of the left arm, and intermittent opucciinal headaches.

The patient had had arthritis for 30 years, treated with cortisone for the past 7 years, and a supracervical hysterectomy for undetermined reasons 20 years earlier.

Examination. Significant physical findings were confined to the nervous system. Although the patient was well-oriented, she showed mild inability to deal with mathematical calculations or abstract situations. The neck was supple. There was anosmia on the right. The visual acuity and optic fundi were normal. The corneal reflexes were slightly diminished bilaterally. The remainder of the cranial nerve examination was normal.

Muscle strength was good save for a questionable weakness of the left arm. There was a wide-based ataxic gait with a tendency to fall to the right side. Romberg’s sign was present. Performance of heel-to-knee and finger-to-nose tests was impaired. Sensory examination revealed loss of vibratory sensation in the feet and, to a slight extent, in the upper extremities. Position sense in the fingers and toes was also impaired. There was moderate hypalgesia and slight hypesthesia over the right side of the body below the fifth cervical segment. The superficial abdominal reflexes were absent. The remainder of the deep muscle reflexes were brisk but symmetrical throughout, and no abnormal reflexes were present.

A lumbar puncture revealed clear fluid with an opening pressure of 100 mm of water; the spinal fluid contained 120 mg of protein and 56 mg of sugar per 100 ml; no cells were present and the serology was negative. Skull x-rays were normal; spinal column x-rays showed spondylitis deformans affecting the entire column. A cervical myelogram demonstrated a transverse defect at the level of the fifth and sixth cervical vertebrae. Aortic arch and cerebral angiograms showed minimal concavity of the vertebral arteries due to spondylosis with no disturbance of blood flow. Arteriosclerotic plaques were noted in the right internal carotid artery. Tortuosity and segmental dilatation of the basilar artery were consistent with arteriosclerosis. Pneumoencephalograms showed absence of ventricular filling and a dilated callosal sulcus, suggesting a mass lesion in the posterior fossa. An electroencephalogram revealed random delta activity over both occipital lobes.

After admission, the patient rapidly de-
Cystic Tumor of the Choroid Plexus

Cystic Tumor of the Choroid Plexus, becoming increasingly ataxic, dysphagic, and dysarthric. Terminally, she developed signs of bilateral bronchopneumonia and died 2 weeks after admission.

Autopsy Findings. There was bilateral bronchopneumonia, generalized arteriosclerosis, and osteoporosis. The supraccervical portion of the uterus was absent; only a small fibrotic cervical stump remained. No tumor was present in any organ.

The unfixed brain weighed 1370 gm. The cerebral hemispheres were normal on external inspection as well as on serial coronal section. The lateral and third ventricles were of normal size. The choroid plexuses within these ventricles were normal.

External examination of the cerebellum and brain stem revealed multiple, thin-walled, semitranslucent cysts located within the subarachnoid space between the medulla and the left cerebellar hemisphere and obscuring the left foramen of Luschka (Fig. 1).

Fig. 1. Basilar view of the cerebellum and caudal brain stem. Multiple thin-walled semitranslucent cysts are present in the subarachnoid space between the medulla and left cerebellar hemisphere, obscuring the left foramen of Luschka. Note the displacement of the left vertebral artery by a cyst (arrow).
The left vertebral artery and the posterior portion of the basilar artery were displaced laterally by one of the cysts. The ninth through twelfth left cranial nerves were displaced anterolaterally by the cysts to varying degrees. The left seventh and eighth cranial nerves were also displaced, but in an anterosuperior direction and to a lesser degree. The left cerebellar hemisphere and inferior vermis were grossly distorted by the cysts. Cystic masses protruded through the foramen of Magendie, obliterating it. The foramen of Luschka, choroid plexus, and cranial nerves on the right side were normal.

Serial transverse sections through the brain stem and cerebellum revealed multiple cysts filling the caudal half of the fourth ventricle. They severely distorted the medulla and adjacent cerebellum (Fig. 2). The choroid plexus of the left lateral recess of the fourth ventricle could not be identified; the choroid plexus of the right lateral recess appeared normal. The median raphe of the medulla was displaced to the right, and the posterolateral aspect of the left side of the medulla was severely distorted. The ventral portions of the medulla, including the pyramids, appeared unaffected. No evidence of infiltration into the brain substance was present in spite of severe distortion by the cysts. One of the cysts extended across the midline to compress the posteroinferior aspect of the right cerebellar hemisphere.

The cysts ranged from a few millimeters to 1.5 cm in diameter and were distended with clear mucoid material. The inner walls of the cysts were, in general, smooth. The outer walls were apposed to one another, giving the appearance of a multilocular mass.

Microscopically, all cysts were lined by cells ranging from low cuboidal to tall columnar types usually arranged in a single layer (Figs. 3 and 4). In some areas, however, the cells were heaped up into small fronds projecting into the cystic spaces. Absence of parenchymal invasion was confirmed microscopically.

The cytoplasm of the flattened and low
Cystic Tumor of the Choroid Plexus

cuboidal cells was granular and eosinophilic with an occasional vacuole, while the plump columnar cells, when stained with hematoxylin and eosin, contained copious amounts of faintly bluish material in the cytoplasm. This material was intensely mucicarmine and periodic acid Schiff (PAS) positive. Generally, uniform nuclei were oriented basally, and no mitoses were present.

The cystic spaces also contained strongly positive PAS and mucicarmine reactive material with clumps of granular debris. The interstitium between the cysts was composed of reticulated fibrocollagenous tissue of varying thickness.

Fig. 3. Microscopic section of part of one of the smaller cysts. The lining consists of columnar epithelium with vacuolated cytoplasm. Clumps of granular debris are seen in the lumen. Loose connective tissue surrounds the epithelial lining. The space between the basement membrane of the lining cells and the connective tissue wall is an artifact produced during histologic preparation. (H. & E., ×10).

Fig. 4. Microscopic section of two adjacent cysts separated by a layer of connective tissue. Tall columnar cells with vacuolated cytoplasm line one cyst (upper left) while cuboidal epithelium lines the second (lower right). The nuclei are uniform and basally situated. The lumina of the cysts are indicated by C. (H. & E., ×100).

After careful examination of serial sections of the medulla and the caudal part of the fourth ventricle, small remnants of choroid plexus were found buried within the twisted masses of cysts and interstitial connective tissue. Often, the apparently normal epithelium of the choroid plexus fragment was continuous with tall columnar cells which were identical to those lining the cysts (Fig. 5).

The stroma of the choroid plexus, containing specks of calcification, persisted below the cuboidal cells as they became progressively more columnar. The columnar cells were also found to line deep clefts within the stroma. Presumably, the cysts themselves, often visible near the depths of the clefts, were derived by pinching off of the deeper portions of the clefts. The columnar lining of the cysts thus are surrounded by a fibrous stroma identical to that of the choroid plexus.
This cystic mass cannot be properly classified as a papilloma of the choroid plexus because of the absence of significant solid papillary components. Cysts of various sizes, with or without epithelial linings, have been reported in the choroid plexus papillomata. In some instances, the cells composing the papilloma have been found to be columnar in type with intracytoplasmic mucin. Even more rarely, cysts lined by columnar cells with mucin production have been described in association with plexus papillomata. In all such cases, the solid papillary element formed the significant portion of the lesion. If the underlying lesion in our case is indeed a neoplasm of the choroid plexus, then we have to assume that the entire papilloma has undergone these cystic changes.

On the other hand, epithelial-lined cysts, presumably of congenital or developmental origin, are frequently encountered in choroid plexuses undoubtedly unaffected by neoplastic processes. Mucin production by the lining cells is a frequent concomitant finding. Large cysts of this variety, the so-called "colloid" cysts, are, in general, confined to the third ventricle, especially when symptom-producing. One case of symptom-producing epithelial-lined cysts of the fourth ventricle has been reported. Two cysts lined by a layer of ciliated columnar epithelium and containing gelatinous fluid were successfully removed surgically from a 28-year-old man, but the attachment of the cysts was not determined.

The lesion we are reporting is unusual because of its large size, complexity, and the rapid onset of symptoms suggesting a fast-growing cerebellar tumor. If this cystic mass was indeed of congenital origin, one must explain the late and rapid development of symptoms.

It seems likely that the rapid expansion of the cysts by the accumulation of mucin was due to the sudden increase in secretory activity of the abnormal cells within the choroid plexus or to a blockage of drainage of continuously secreted material after many years quiescence. A concomitant slow proliferative process of these abnormal cells perhaps enhanced the enlargement of the mass by forming clefts and cysts repeatedly (Fig. 6). The large multicystic mass, so

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**Discussion**

A number of diagnostic possibilities are suggested by the gross and microscopic appearance of the lesion. Although the cystic mass superficially resembles a so-called "arachnoid cyst," microscopic examination excludes this diagnosis because the cysts are lined by columnar cells, distinctly epithelial in nature, instead of the spindle-shaped cells of the pia-arachnoid.

The possibility that this lesion represents a well-differentiated mucin-producing adenocarcinoma must be considered but the absence of any primary lesion excludes it. The continuity of the cysts and cleft linings with apparently normal choroid plexus epithelium clearly points to the choroid plexus as the site of origin of the lesion. Therefore, the lesion must be either a neoplasm of the choroid plexus or a result of changes within a pre-existing congenital malformation within a portion of the myelencephalic choroid plexus.

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**Fig. 5.** Microscopic section through the choroid plexus of the left lateral recess of the fourth ventricle. Transition between the cuboidal epithelium of the choroid plexus and tall columnar cells, identical to those lining the cysts, is shown (arrows). (H. & E., X20.)
formed, produced severe distortion of the brain stem, ultimately leading to the death of the patient.

Although the precise origin remains obscure, we felt it worthwhile to report the occurrence of so large a multiloculated cystic mass associated with the choroid plexus of the fourth ventricle in an elderly person. So far, we have not been able to find a similar case reported. In addition, it should be noted that such a lesion may be accessible to surgical removal. It may therefore be desirable to include the possibility of such a lesion in the differential diagnosis of expanding masses within the posterior fossa.

Summary

We have described the occurrence of a multicystic benign mass in the fourth ventricle and adjacent subarachnoid space in a 68-year-old woman who showed clinical signs and symptoms of a posterior fossa lesion. We have demonstrated the origin of this lesion from the choroid plexus of the fourth ventricle.

References


