Third ventricular choroid plexus arteriovenous malformation simulating a colloid cyst

Case report

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A case of a cystic encapsulated arteriovenous malformation arising from the tela choroidea of the third ventricle is reported. Although the appearance of this lesion on computerized tomography scan suggested a colloid cyst by its location, it was atypical because of the non-homogeneity both before and after contrast infusion. The increased density on the unenhanced scan was due to hemosiderin pigment in the capsule wall caused by old hemorrhage. The diagnosis in this case was made by histological examination.

Key Words • arteriovenous malformation • choroid plexus • colloid cyst • computerized tomography

Cystic tumors presenting in the anterior third ventricle are usually colloid cysts,3,5,7,33,40 or, less commonly, craniopharyngiomas.9,12,21,20 A case of an encapsulated cystic arteriovenous malformation (AVM) arising from the choroid plexus of the third ventricle is presented. It is believed to be the first such case of this rare lesion presenting in this manner in the third ventricle.

Case Report

This 11-year-old Ceylonese girl presented with a long history of bifrontotemporal headaches. The headaches, throbbing in nature, began to increase in frequency over a year to once a week and, finally, were present constantly for the 3-month period prior to diagnosis. They were often associated with nausea and vomiting. The patient had also had several brief syncopal episodes over the three years before admission, that occurred while she was walking or standing. She denied any visual complaints and had no symptoms of focal weakness or sensory disturbance.

Examination. General neurological examination was completely normal. Funduscopic examination revealed early papilledema. The patient was started on Decadron, which resulted in prompt relief of her headache.

Skull films were normal. Computerized tomography (CT) brain scans were performed before and after the infusion of intravenous contrast medium in the axial and coronal planes (Fig. 1). The axial views revealed a round, well circumscribed mass at the level of the foramen of Monro with symmetrical ventricular enlargement. A small area of increased density was present at the periphery of this lesion on its left anterior aspect. This same region showed minimal contrast enhancement. The center of the lesion was less dense than its periphery, suggesting a possible cystic component. The third ventricle was identifiable below this mass, with its walls displaced laterally and superiorly. The non-homogeneous appearance of the lesion on the CT scan suggested that the lesion was not a colloid cyst.

Operation. The patient was operated on through a right frontal transventricular approach to the third ventricle. Through the operating microscope, an encapsulated lesion was seen protruding through the right foramen of Monro. This was aspirated. It contained several cc’s of dark greenish fluid in which floated shimmering flakes of cholesterol crystals.
AVM of third ventricle choroid plexus

FIG. 1. Axial (A, B) and coronal (C, D) computerized tomography scans localized a well circumscribed round mass to the region of the foramen of Monro. Precontrast (A, C) scans revealed minimal increased density at the periphery of this lesion (white arrow, A). Postcontrast scans (B, D) revealed minimal contrast enhancement at the periphery of this lesion (white arrow, B). The obstructive hydrocephalus was moderate. Coronal views (C, D) showed this lesion to be separate from the infundibulum and within the superior and anterior aspect of the third ventricle.

After the cyst was collapsed, it was possible to dissect it free from the surrounding walls and floor of the third ventricle. The base of the cyst was traced to the roof of the third ventricle. The mass had separated the two columns of the fornix, and was arising from the tela choroidea of the roof of the third ventricle. Here it was coagulated with the bipolar forceps and cut off.

The patient's postoperative course was uneventful except for one focal seizure. She remains free of neurological deficit.

Histological Examination. Microscopic examination of the excised lesion revealed clusters of closely packed, mostly thin-walled blood vessels of various sizes. Most of these vessels had the features of either capillaries or venules, but a number showed a distinct, usually irregular, internal elastic lamina, indicating their arterial nature (Fig. 2 upper left). Some of them also displayed fairly normal muscular media. Their walls often showed irregular thickening due to hyalinization. The abnormal vessels were surrounded by loose fibrous tissue, often accompanied by a moderate cellular inflammatory reaction. A poorly preserved epithelial layer in a papillary configuration suggestive of choroid plexus was present (Fig. 2 upper right and lower left). The vascular malformation extended into neural tissue where it showed significant gliosis, presumably representing the amputated base of the lesion in the roof of the third ventricle (Fig. 2 lower right). Throughout the malformation and the dense cellular fibrosis, there were masses of cholesterol crystals associated with iron pigment, lymphocytes, and foreign body giant cells characteristic of old hemorrhage. Other areas showed foci of intense chronic inflammatory changes consisting of lymphocytes, plasma cells, and macrophages.

These histological features were interpreted as those of an AVM arising in the tela choroidea of the choroid plexus of the third ventricle. This lesion showed evidence of both recent and old hemorrhage.

Discussion

Vascular malformations arising primarily in the choroid plexus are extremely rare. The first documented angioma of the choroid plexus was probably that reported by Guerard in 1833. Liber and Lisa reviewed six cases reported in the literature up to 1940, although at least one case is probably a tumor and not an angioma. Twenty additional cases have been reported since the review of Liber and Lisa. Only two of these cases have involved the third ventricle. In 1859, Förster reported a vascular malformation the size of a pigeon's egg in the roof of the third ventricle, associated with the choroid plexus. The angioma reported by Bonnal, et al., was located primarily in the right lateral ventricle, but did extend through the foramen of Monro into the third ventricle. The majority of these lesions involve the lateral ventricles.

The age of patients with symptomatic lesions ranges from the newborn period to 60 years. The most common age of clinical presentation is in the second decade of life. Females (64%) are more often affected than males (36%).

Clinically, the lesion in our patient produced symptoms similar to those of a colloid cyst, with intermittent headaches, probably produced by transient obstruction to the outflow of cerebrospinal fluid through the foramen of Monro. The patient also had several syncopal episodes lasting seconds, which occurred while she was either walking or standing. Similar episodes have been reported in patients with colloid cysts.

A review of the 26 previous case reports of choroid plexus vascular malformations indicates that these lesions usually presented with nonlocalized complaints, such as headache, nausea and vomiting, decreasing level of consciousness or coma, and dizziness. In general, the symptoms were associated with abrupt onset of subarachnoid hemorrhage. Fourteen
of these patients were operated on, with an operative mortality of 28%. Of the 10 patients who survived after surgery, three had moderate to severe neurological deficits. Seven patients reported in the literature died without surgery, and the majority of these were infants. In two cases, a choroid plexus angioma was an incidental finding at autopsy.

Radiographically, this lesion most resembled a colloid cyst because of its characteristic location. However, its appearance on CT scan was unusual for a colloid cyst because of the presence of peripheral density and non-homogeneous contrast enhancement. Naidich, et al., noted that colloid cysts show homogeneity when absorption coefficients are examined throughout the lesion, whereas craniopharyngiomas do not, even though both appear to be uniformly dense on visual examination. A craniopharyngioma was unlikely in this case because the lesion lacked continuity with the region of the infundibulum. Som, et al., reported a large chronic cystic hematoma in the frontal region which, on CT scan, appeared to have a radiolucent center with a dense rim resulting from iron deposition as a result of hemosiderin breakdown products. Histologically, the wall of this cystic hematoma contained a venous angioma. In the case reported here there was a peripherally increased density on the CT scan, and it was believed that this was due to hemosiderin pigment present in the capsule wall.

The majority of the cases reported of vascular

FIG. 2. Photomicrographs of the excised lesion. *Upper Left:* Three small arteries are revealed within the arteriovenous malformation with a characteristic elastic lamina. Elastic van Gieson, × 480. *Upper Right* and *Lower Left:* Representative areas of epithelial cells in a papillary configuration are suggestive of choroid plexus. H & E, × 480. *Lower Right:* The arteriovenous malformation is seen extending into the neural tissue of the roof of the third ventricle. H & E, × 480.
malformations involving primarily the choroid plexus are true AVM's, as defined by Russell and Rubinstein.\textsuperscript{11} The cases of McConnell and Leonard\textsuperscript{24} and Dandy\textsuperscript{11} were called a cavernous hemangioma and a venous aneurysm, respectively, but, in fact, both were true AVM's because of the presence of elastica within the vessel walls in scattered places. In at least five cases, there was significant extension of the AVM to adjacent brain: to the thalamus,\textsuperscript{6,10,11} temporal lobe;\textsuperscript{6} and posterior parietal lobe.\textsuperscript{4} Two cases of cavernous angioma\textsuperscript{23} and one case of a venous angioma\textsuperscript{8} of the choroid plexus are represented in the vascular pathology of these lesions. In one patient, both an angioma and a papillary adenoma were diagnosed histologically.\textsuperscript{8} In several cases, the pathology is not entirely clear.\textsuperscript{2,4,8,15,18,30,\textsuperscript{30}}

Butler, et al.\textsuperscript{8} postulated that angiomas of the choroid plexus most likely develop at approximately the 30-mm embryonic stage, where large crossing arterial and venous endothelial tubes are most evident near the relatively enormous choroid plexus.\textsuperscript{28} Padget\textsuperscript{28} postulated that fistulas develop in areas where the primitive vessels cross when they are separated by only a double layer of endothelial cells. Choroid plexus angiomas predominate in the glomus, although malformed vessels may extend into the villi. Shuangshoti and Netsky\textsuperscript{35} showed that the glomus is the most heavily vascularized portion of the choroid plexus, both in fetal life and in later years, and hence is the most likely to contain abnormal collections of vessels from developmental errors.

The interesting encapsulation of the lesion presented here likely resulted from hemorrhage into the stroma of the choroid plexus without escape of significant blood in the cerebrospinal fluid. This lesion definitely showed evidence of both old and recent hemorrhages.

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