Arteriovenous malformation of choroid plexus

Case report

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The authors describe a patient with subarachnoid hemorrhage from an arteriovenous malformation of the choroid plexus and present a brief review of related reports.

KEY WORDS • choroid plexus • arteriovenous malformation • subarachnoid hemorrhage

VASCULAR malformations of the choroid plexus are an unusual cause of subarachnoid hemorrhage. This case serves to point out some of the clinical characteristics of arteriovenous malformations of the choroid plexus.

Case Report

A 22-year-old right-handed woman was transferred to State University Hospital following her second subarachnoid hemorrhage. She had been well until 2 years prior to admission when she experienced the first subarachnoid hemorrhage, characterized by headache, stiff neck, and confusion. Bilateral carotid angiography was normal. She was discharged from the local hospital with a diagnosis of schizophrenia. She had remained well until 1 week prior to the present admission when she again experienced sudden severe bifrontal headache and stiff neck, but did not lose consciousness. She was admitted to the local hospital in a confused and agitated state, with hallucinations but no focal neurological findings. Lumbar puncture revealed bloody cerebrospinal fluid (CSF). Her mental status improved and she was referred to the Upstate Medical Center 1 week later.

Examination. On admission, the patient's vital signs were normal. She was confused, disoriented, intermittently agitated, hallucinating, and acutely schizophrenic. She had marked neck stiffness, but no dysphasia; the fundi were normal. There was decreased spontaneous movement and increased tone with hyperreflexia of the right leg. There were no pathological reflexes, and sensory examination revealed no deficit. Lumbar puncture disclosed a pressure of 90 mm H2O; the CSF was cloudy and xanthochromic, with 3700 red blood cells/mm³, and protein of 248 mg%. Transfemoral cerebral angiography the following day revealed an arteriovenous malformation (AVM) near the left trigone, supplied by the anterior and the posterior lateral choroidal vessels on that side (Fig. 1).
There was early shunting to the vein of Galen. The patient was given Thorazine, and her mental status and focal findings gradually improved. Pneumoencephalography 3 weeks after admission (Fig. 1 lower right) demonstrated minimal enlargement of the left lateral ventricle and an irregular floor of the left trigone. Her mental state remained stable.

**Operation.** Four weeks after admission, the patient underwent left parietooccipital craniotomy. A cortical incision near the midline at the parietooccipital junction exposed the left trigone. The ventricle size was normal, while the choroid plexus in the trigone was enlarged and bright red. A large branch of the anterior choroidal artery coursed along the floor of the ventricle at the posterior edge of the choroid plexus; we could readily identify, clip, and divide this artery using the operating microscope at $10\times$ magnification. Multiple small arterioles coursed to the plexus from the medial wall of the ventricle; these were coagulated and cut. We then removed the abnormal plexus from the trigone, leaving normal-appearing plexus in the temporal horn and body of the lateral ventricle in place. There was an area of staining in the ventricular floor due to previous hemorrhage.
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Postoperative Course. The patient had a fluent dysphasia and right homonymous hemianopsia. The speech deficit cleared slowly until 9 days postoperatively, when she rapidly developed a decreased level of consciousness and right hemiparesis. Repeat left carotid and left vertebral angiography revealed no evidence of the previously noted AVM, but there was a large parietooccipital mass which exploration showed to be a dilated, trapped, temporal horn and periencephalic cyst at the site of the previous transcortical incision. After the second craniotomy and decompression were done, she again improved, but a shunting procedure from the temporal horn to the peritoneal cavity was necessary 5 weeks later. She has since shown continued improvement.

Laboratory Studies. The choroid plexus consisted of highly vascular tissue with choroidal epithelium on the surface and numerous blood vessels of various sizes (Fig. 2). The vessels ranged from small, thin-walled channels, 50 to 100 μ in diameter, up to much larger thick-walled vessels with an overall diameter of several hundred μ. The outer portions of these thick-walled vessels showed a non-cellular hyalin type of degenerative change.

Discussion

At least 20 cases of choroid plexus angioma or arteriovenous malformation (AVM) have been reported.\(^1\) Butler, et al.,\(^4\) in their discussion of 17 treated cases of vascular lesions causing mild intraventricular hemorrhage, reviewed eight cases of choroid plexus angioma. Doe, et al.,\(^7\) reviewed the literature describing this lesion and added five postmortem cases. A single case treated with surgery was recently reported by Matsushima, et al.\(^14\) Some of the cases included by other authors\(^2,\)\(^3,\)\(^8,\)\(^10,\)\(^12\)\(^--\)\(^19\) seem to have had primary choroid plexus AVM's have been excluded by us. Several of the patients reported by Liber and Lisa\(^11\) seem to have had primary choroid plexus tumors.\(^9\) Dandy's\(^8\) case, in fact, was an AVM of the lateral periventricular area which apparently drained through an aneurysmally-dilated subependymal vein and shared the deep cerebral venous system with the choroid plexus. Furthermore, it is of some importance to separate choroid plexus vascular lesions from the periventricular malformations although they are thought to have a common embryologic basis, since the surgical treatment of pure choroid plexus malformations should incur less deficit than treatment of a lesion embedded, even superficially, in the basal ganglia or thalamus.\(^4\)

The 20 reported cases show consistent patterns of presentation. Five were infants, four of whom died in the neonatal period from intraventricular hemorrhage associated with multiple congenital anomalies. McGuire, et al.,\(^16\) reported a 3-month-old infant with hydrocephalus, probably secondary to episodic intraventricular hemorrhage; bilateral choroid plexectomies were performed and the vascular malformations identified bilaterally by microscopy. Two other infants had bilateral lesions, and two cases were incidental postmortem findings.\(^7,\)\(^19\) The 13 remaining patients had an average age of 27 years; two patients were over 40 years old and seven were between the ages of 14 and 20 years. Thus, these small angiomas tend to occur in the younger age groups as do the parenchymatous microangiomas.\(^18\) Of the 20 patients, 69% were female. Clinical presentation is usually that of a subarachnoid hemorrhage often without striking focal findings except in patients with associated intracerebral or deep nuclear clots. Although these lesions have been previously described as one of the causes of a "mild" form of intraventricular hemorrhage,\(^4\) 38% of
the adults reported died rapidly from the initial or recurrent intraventricular or intracerebral hemorrhage. If the children are included in this group, 45% died as a direct result of bleeding. Thirty percent of the adults who survived the initial hemorrhage had identifiable recurrent hemorrhages, several of which terminated fatally.\textsuperscript{15,17,18}

Of the 18 patients with symptoms attributable to choroid plexus angiomas, the diagnosis was made antemortem in 10. Two were diagnosed at the time of operative exploration for intracerebral hematoma, seven by angiography, and one after plexectomy for hydrocephalus.

Nine patients, including ours, were operated on for intracerebral hematoma and/or obliteration of the malformation; intraventricular hematomas were encountered in five of these patients and several had clots in the substance of the choroid plexus. The ventricles have often been enlarged as a result of either communicating hydrocephalus or intraventricular hematoma. In four of the patients, the malformation extended into adjacent structures, such as the thalamus, third ventricle, or temporal lobe, making complete removal more hazardous or impossible. In all but one of the cases the malformation was located primarily in the choroid glomus. Patients whose malformations have been removed have generally done well; one patient, whose lesion extended into the third ventricle, died postoperatively. In several patients, including ours, the operative approach caused the major postoperative deficit; in our patient the approach was made more difficult by the small size of the ventricles. No other report mentions the late type of postoperative obstruction of the ventricular system that occurred in our case; perhaps a formal complete choroid plexectomy at the time of the initial procedure would have avoided this problem.

When an AVM or venous angioma fed by the choroidal vessels has been identified in the region of the trigone, pneumoencephalography should be performed as an important preoperative measure to delineate the size and position of the angiomatous mass within the ventricle. In the absence of an intracerebral hematoma (usually temporal lobe) to dictate the operative approach, the medial occipitoparietal approach to the trigone seems most likely to facilitate access to the major feeding vessels along the posterior edge of the lesion. There is clearly no safe way into the trigone area, especially in the dominant hemisphere; however, in view of the incidence of recurrent and fatal hemorrhage from choroid plexus AVM's and venous angiomas, the risk of transcortical ventricular exposure seems warranted.

References
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