Benign astrocytoma associated with arteriovenous malformation

Case report

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An arteriovenous malformation associated with an underlying benign astrocytoma of the right frontal lobe is reported and the radiological and pathological features discussed. Previous reports of multiple primary tumors of the nervous system are reviewed and the few cases of vascular malformations associated with gliomas noted.

KEY WORDS arteriovenous malformation tumor glioma astrocytoma brain multimodal primary tumors of diverse origin in the central nervous system are well known. The subject has been reviewed by Myerson, 8 extensively by Courville, 3 who added 21 cases of his own to 113 cases collected from the literature, and recently by Madonick, et al. 7 The most common combination described are meningiomas and gliomas. Vascular malformations, almost always of the arteriovenous type, are rarely associated with other central nervous system tumors. We are reporting a case in which an arteriovenous malformation and a benign astrocytoma occurred in the same patient.

Case Report

This 17-year-old white boy was admitted to the New York Hospital-Cornell Medical Center because of headache and decreasing vision of 5 weeks’ duration. At age 9 the patient was first seen at the New York Hospital for Jacksonian epilepsy. Neurological examination and skull films then were normal, and an electroencephalogram (EEG) revealed “bilateral high voltage slow wave activity, compatible with a seizure diathesis.” The patient was placed on phenobarbital, 32 mg twice a day, supplemented 1 month later by Dilantin daily, to obtain better seizure control. Thereafter, the patient remained free of seizures for almost 3 years. By age 13, because of refractory seizures, he was placed on Mysoline, 250 mg 5 times a day. The EEG at that time was interpreted as unchanged.

Examination. There was bilateral papilledema. The patient was totally blind in the right eye and had only light perception in the left. Cranial nerves were otherwise intact. His gait was ataxic. There was hyperreflexia on the left side. Plain skull films showed evidence of enlargement of the sella turcica and erosion of the “lamina dura” of the dorsum sellae. The right orbital plate of the frontal bone was depressed (Fig. 1). A brain scan showed an abnormality in the low right frontal region. A bilateral carotid angiogram us-
Fig. 1. Skull films showing sellar changes of raised intracranial pressure and depression of the right orbital root (arrows).

ing a femoral catheter showed that the right external and internal carotid arteries were selectively opacified. A right frontal mass was demonstrated, with large tortuous vessels in the subfrontal region filling from the anterior cerebral artery (Fig. 2). Early venous filling was seen with opacification of the superior sagittal sinus, basal vein, and internal cerebral vein via the anteroseptal vein. The ophthalmic artery was depressed. A nasal branch of the maxillary artery passed into the cranial vault in the region of the cribriform plate to supply a portion of the lesion. Although the very large tortuous vessels were unusual, the lesion was thought to represent a glioma which may have undergone “malignant” degeneration. The small contribution from the nasal branch of the maxillary artery suggested involvement of the dura.

Operation. At craniotomy, an infiltrating tumor occupied the right frontal cortex and involved the underlying white matter and basal ganglia. Along the anterior border of the tumor, a group of vessels in the meninges formed a maze of dilated, twisted vascular channels characteristic of an arteriovenous malformation. Because of the extent of the tumor, only a small portion was obtained for pathological evaluation. The vascular malformation measured $5 \times 2 \times 1$ cm and was removed in toto.

Histological Examination. A small specimen initially received for frozen section was diagnosed as astrocytoma. A small sample of tumor measuring 1 cm in greatest dimension was subsequently submitted in formalin. The tissue was stained with hematoxylin and cosin, trichrome (Masson), phosphotungstic acid hematoxylin (Mallory), elastic (Weigert-van Gieson), and reticulin (Laidlaw) preparations. In the meninges overlying the tumor, a typical arteriovenous malformation was seen (Fig. 3). The majority of the vessels comprising the malformation were abnormal. Several had prominent internal elastic laminae indicating arterial derivation. Many had asymmetrically thickened walls which were focally fibrotic. Reduplication, fragmentation, and absence of the elastic fibers was prominent in many vessels. The caliber of the vessels varied greatly, and aneurysmal dilatation with local attenuation of the wall was observed. Arterial, venous, and capillary structures were identified in the malformation. Numerous smaller vessels and capillary channels showed calcification reminiscent of the mineralization seen near tumors which have undergone hemorrhage. The presence of hemosiderin-laden macrophages indicated previous bleeding.

Adjacent to the arteriovenous malformation, which was confined to the meninges, was a benign astrocytoma (Fig. 4 left). The tumor infiltrated the cortex, but did not in-
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Fig. 3. Low-power photomicrograph of the arteriovenous malformation in the meninges. Vessels are of varying sizes. Some have abnormal configuration of asymmetrically thickened walls. Weigert-van Gieson, × 40.

involve the pia arachnoid. It was composed of astrocyte-like cells, some of which were moderately pleomorphic (Fig. 4 right). Although nuclei varied in size and shape, hyperchromaticity was uncommon and significant atypia was not seen. Necrosis, pseudo-palisading, and mitoses were not features of the tumor. The neoplasm was not vascular microscopically. Proliferative vascular changes often observed in more malignant gliomas were absent. A few small areas of endothelial proliferation were noted.

Postoperative Course. Although the papilledema subsided, vision did not improve. Except for a minor wound infection 4 weeks after surgery, the patient has done relatively well. He has received 6000 R to the brain over a 6-week course.

Discussion

The vascular changes associated with gliomas are characteristic. They usually involve small vessels and are proliferative in nature. The formation of convoluted, glomerular-like channels due to endothelial proliferation is the most common such change. Endothelial proliferation and other vascular abnormalities are more often described in glioblastomas, although they may be found occasionally in astrocytomas, oligodendrogliomas, and metastatic tumors. In an extensive study of the pathology of blood vessels in glioblastomas, Nyström used plastic casts, angiography, light microscopy, and electron microscopy to delineate a multiplicity of vascular abnormalities including lesions that resembled vascular malformations. The vascular lesion in our case does not conform to previously described changes in gliomas and is most appropriately termed an arteriovenous malformation. The lesion was discrete, confined to the meninges, and much larger than any focal vascular changes reported in gliomas. No tumor infiltrated the pia-arachnoid. We have never seen a focus of this size which has been physically separate from tumor cells.

The tumor itself was a benign astrocytoma, or by the classification of Kernohan and Sayre, astrocytoma, Grade II. Microscopically, it was not a vascular neoplasm, and the only vascular change of significance was an occasional small focus of endothelial proliferation.

Arteriovenous malformations are congenital in origin, representing persistent embryonic arteriovenous shunts in which the capillary network has failed to form. These masses of tortuous, ectatic vessels are composed mainly of venous structures lying in the meninges, although they may extend into the cerebral cortex. Most occur in the cerebral hemisphere. They are occasionally associated with other malformations of the nervous system. Mental retardation produced by hydrocephalus or cortical dysgenesis has been described. An association with berry aneurysms is well known. On rare occasion, primary brain tumors have been reported in patients with arteriovenous malformations. Fine, et al., described a 15-year-old boy with headache and signs of subarachnoid bleeding. An arteriovenous malformation was seen in the parietal cortex by angiography. With ventriculography, a mass lesion was demonstrated in the lateral ventricle which by histological analysis proved to be an oligodendroglioma. More recently
Welcker and Seidel\textsuperscript{11} reported a case similar to ours in which an astrocytoma was found in a patient with an arteriovenous malformation. The tumor had not been suspected clinically or radiologically and was discovered only at autopsy.

The radiographic findings in this case were suggestive of a vascular glioma. Early arteriovenous shunting, particularly, is not unusual in a glioblastoma multiforme. Involvement of the dura by superficially situated gliomas can occur and may result in hypertrophy of the meningeal arteries. However, the combination of unusually large, tortuous vessels and an apparent cerebral mass with a peculiar pattern of "tumor vascularity" should suggest the presence of an arteriovenous malformation associated with a glioma. In certain circumstances, it may be difficult to exclude a hematoma which has resulted from bleeding in an arteriovenous malformation.

References

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