Arteriovenous malformation and oligodendroglioma

Case report

ROBERT M. CROWELL, M.D., UMBERTO DEGIROLAMI, M.D.,
AND WILLIAM H. SWEET, M.D., D. Sc.

Neurosurgical Service and Neuropathological Laboratory, Massachusetts General Hospital, Boston, Massachusetts

The coincidence of arteriovenous malformation (AVM) and primary brain neoplasm is rare. We are reporting a case of oligodendroglioma within an arteriovenous malformation.

KEY WORDS • arteriovenous malformation • brain tumor • oligodendroglioma • subarachnoid hemorrhage

REPORTED cases of arteriovenous malformation (AVM) of the central nervous system in association with a primary brain tumor are very rare; only one case of AVM accompanying oligodendroglioma has been previously recorded. We are now reporting a case of oligodendroglioma within an AVM.

Case Report

First Admission. In June, 1971, this 17-year-old right-handed man suddenly felt pain “like an explosion” in the right eye which persisted for approximately 4 hours. Similar headaches recurred almost daily after the first episode. In early July, 1971, nausea and vomiting began to accompany the headaches. On July 23, the patient was admitted to the hospital where physical examination disclosed papilledema as the only abnormal sign. Skull films, sinus films, and electroencephalography were normal. Lumbar puncture yielded xanthochromic cerebrospinal fluid under increased pressure; spinal fluid protein was normal. Technetium brain scan demonstrated abnormal uptake of the isotope in the right temporal region. A ventriculogram demonstrated a lateral temporal lobe cyst which communicated with the ventricular system.

On August 28, the patient was transferred to the Massachusetts General Hospital. The neurological examination showed a left lateral rectus palsy and two diopters of papilledema with flame-shaped hemorrhages and exudates. There was a mild left facial palsy and ataxic gait. A seizure, characterized by unresponsiveness and bilateral tonic elbow flexion, occurred shortly after transfer. The electroencephalogram showed generalized slowing, most marked over the right temporal region. A right carotid arteriogram showed a large, avascular, intratemporal mass.
First Operation. On August 28, 1971, a right temporal craniotomy was performed (W.H.S.). A well circumscribed superficial cyst, 8 × 12 cm in size, was encountered in the temporal lobe. The cyst wall was opened, and along its lining numerous delicate, serpentine, nonpulsatile red vessels were noticed. Xanthochromic discoloration of the interior of the cyst wall testified to prior hemorrhage. Firm, grayish, gritty tissue was encountered along the entire inferolateral margin of the cyst, immediately adjacent to the abnormal vessels. A generous biopsy of the cyst wall and apparent tumor was taken, and communication was established surgically between the cyst and the ventricle that lay just medial to it.

The postoperative course was uneventful except for occasional seizures characterized by a flash of blue light in the right upper quadrant of the visual field.

Histological Examination. The operative specimen consisted of hemorrhagic, soft brain tissue measuring 2.2 × 2.5 × 5.0 cm in aggregate. On microscopic examination (Fig. 1), the specimen contained an AVM composed of vessels of varying caliber and thickness which were separated by gliotic tissue. The vessel walls were frequently grossly distorted either by attenuation and aneurysmal dilatation or by fibrous thickening. The intima was unusually thickened, and the media was either hyalinized or grossly disrupted, showing fragmentation of elastic tissue fibers (Verhoef stain). Pigment-ladened macrophages and hemosiderin pigment gave evidence of prior hemorrhage. Recent hemorrhage was noted in the center of the lesion.

Within portions of the vascular malformation, and to some extent well demarcated from it, were several foci of well-differentiated oligodendroglioma (Fig. 2). The blood vessels within the foci were mostly capillaries showing prominent endothelial cells. There were numerous foci of calcium deposits within the neoplasm. The AVM-oligodendroglioma mass appeared well circumscribed and was bounded by an edge of reactive astrocytes in some areas.

Second Operation. On October 22, a right temporal craniotomy was carried out (W.H.S.), with removal of the anterior 7 cm of the right temporal lobe. Postoperatively the only change in the neurological examina-
Fig. 2. Photomicrograph of oligodendroglioma. Note uniform nuclear size and shape, perinuclear clearing and characteristic vasculature. H & E × 150.

Discussion

The intimate association of AVM and a cerebral glioma has been reported infrequently.\(^1,8,10\) Although both entities are usually sufficiently distinctive histologically seldom to cause serious diagnostic difficulty, their coexistence in the same lesion requires careful scrutiny to avoid over-interpretation. Oligodendrogliomas are well known to be richly vascular neoplasms. The vascularity in such tumors is capillary and not that of tortuous, large vessels with asymmetrically destroyed and thickened walls. Conversely, clusters of atypical glial cells can often be found in cases of AVM and are ordinarily easily dismissed as reactive astrocytes. The presence of swollen oligodendroglial cells in AVM is quite common. Typically, these cells have abundant clear cytoplasm, a distinct plasma membrane, and a centrally placed round nucleus. They are usually scattered through the lesion, admixed with reactive astrocytes, and are interpreted as being part of the reactive process. In the process of reviewing a number of cases of AVM at the Massachusetts General Hospital, we have been impressed with the relative frequency with which these oligodendroglial cells seem to cluster and form aggregates that could easily be interpreted as oligodendroglioma on a given high-power field. Clearly this kind of change never warrants the diagnosis of oligodendroglioma. Kaplan, et al., believe AVM's to be a developmental disorder, and the interpretation of AVM as hamartoma is reviewed by McCORMICK. The unclarified possible significance of the association between AVM's and neoplasms is reviewed by Raynor and Kingman and Russell and Rubenstein.

Subarachnoid hemorrhage together with a parenchymal mass effect seen on contrast studies strongly suggests a vascular lesion associated with hematoma. Occasionally this combination of signs may be seen with solitary brain tumor.\(^2,8,9\) The present case illustrates that subarachnoid hemorrhage with a mass effect may rarely represent AVM with primary brain tumor. This diagnosis can only be made with certainty at surgery or post-mortem examination.

References

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Address reprint requests to: Robert M. Crowell, M.D., Neurosurgical Service, Research Building 505C, Massachusetts General Hospital, Boston, Massachusetts 02114.