Excision of a cirsoid arteriovenous malformation of the corpus callosum in a 16-year-old boy

Case report

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CIRSOID arteriovenous malformations, although rare, seem to occur most commonly in the region of the corpus callosum. Nine cases have been reported and of these six have been operated on. A review of the literature suggests a number of common features: 1) in contrast to most arteriovenous malformations, cirsoid lesions of the corpus callosum are well-circumscribed and consist of a tight coil of anomalous vessels that form a localized midline mass; 2) the malformation is usually nourished by the pericallosal arteries from both sides and is drained by the inferior sagittal sinus or great vein of Galen; and 3) the clinical picture is generally that of recurrent subarachnoid hemorrhage without localizing signs.

The current case is of interest because the patient is the youngest to be reported to date and because the case is only the second in which a complete excision of the lesion has been demonstrated.

Case Report

A 16-year-old Negro boy was apparently well until he was 14 years old, when he experienced the first of seven attacks of headache, fever, and stiff neck. The attacks were characterized by an abrupt onset of severe throbbing bitemporal headache unrelated to activity or other precipitating factors. Within 3 to 6 hours, fever and stiff neck were usually prominent, and these complaints would persist for 3 to 4 days. With each attack the patient was forced to bed for several days, but he complained of no other symptoms and was otherwise well. Following each attack he recovered fully and returned to school. During the interval between attacks (the shortest being 3 weeks, the longest 5 months), he maintained his average scholastic record and participated actively in sports (the patient stands 6 ft 5 in., weighs 230 lbs, and has hopes of playing professional basketball). Two days following the patient's seventh and most recent attack of headache and stiff neck, he was admitted to the Children's Hospital of Washington D.C.

Examination. The general physical examination on admission was unremarkable. The neurological examination was also normal except for moderate neck rigidity and bilateral retinal hemorrhages. No bruits were heard. A lumbar puncture showed grossly bloody cerebrospinal fluid which was found...
to be faintly xanthochromic. A bilateral carotid arteriogram demonstrated a midline cri-roid arteriovenous malformation that was richly supplied from both sides (Figs. 1 and 2). A fractional pneumoencephalogram revealed a 3 x 3 x 2 cm mass in the area of the corpus callosum, projecting into the frontal horn of the right lateral ventricle (Fig. 3).

Operation. Under routine general endotracheal anesthesia, a free bone plate was removed from the right parasagittal area, and the corpus callosum was approached by retracting the right frontal lobe away from the falx (Fig. 4). At the base of the longitudinal fissure a racemose arteriovenous malformation was identified overlying the corpus callosum and embedded within it. The lesion was compact and circumscribed and consisted of a tight coil of vessels fed by the pericallosal arteries and drained by a number of large and small veins to the inferior sagittal sinus. The vascular mass was surgically isolated by systematically obliterating the arterial supply with multiple silver clips. This resulted in a modest collapse of the malformation which was then further isolated by clipping the larger venous channels. By this means it was possible to excise the lesion uneventfully and to deliver it in one piece from its bed in the corpus callosum. The callosal defect measured 1 x 3 cm at the conclusion of the procedure and permitted direct examination of the frontal horn of the right lateral ventricle. No vascular communications to the thalamostriate vein, choroidal vessels, or deep cerebral veins were seen.

Postoperative Course. Recovery was uneventful. The patient was up and walking on the 7th postoperative day and was found to have no neurological deficits. On the 21st postoperative day a brachial arteriogram was performed which indicated that the vascular malformation had been completely excised (Fig. 5). The patient has since returned to school where his record remains average. Preliminary psychological and intelligence tests reveal no significant deficiencies.

Discussion

Although the 1966 Cooperative Study on 545 cases on craniocerebral arteriovenous malformations reports a grouping of 15
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"ventricular or paraventricular" lesions occurring in the midline, it is not clear whether some or any of these cases are similar to the case reported here. The literature on cirsoid arteriovenous malformations has recently been reviewed by Dany, et al., who accumulated a total of nine cases including three of their own. Table 1 summarizes the published data. The following points seem evident:

1. The lesions present recurrent subarachnoid hemorrhages which can be mild (current case) or severe.

2. In contrast to the frequent association of epileptic seizures and focal neurological deficits with arteriovenous malformations of the cerebral hemispheres, signs of localizing value have not been reported with cirsoid lesions of the corpus callosum.

Fig. 3. Pneumoencephalograms demonstrating extension of the lesion into the body of the right lateral ventricle. Left: Anteroposterior view. Right: Lateral view.
3. Angiography is diagnostic and in most cases the lesions appear as a walnut-sized or plum-sized midline mass. In general the arterial supply is from the pericallosal arteries of both sides although a lesser supply from the callosomarginal, Sylvian, and anterior choroidal vessels has been also reported.\(^2,3\)

Venous drainage of these lesions is into the inferior sagittal sinus directly or into the vein of Galen via the posterior callosal veins.\(^2,3\)

4. The gross appearance of these lesions is characteristic. The abnormal vessels are arranged in a compact, racemose coil, and the lesion is usually circumscribed and discrete. (The lesion has been referred to as a “cirroid aneurysm” by some authors.\(^2,3\) Although it may occasionally extend from the substance of the corpus callosum to involve the cingulate gyrus or septum pellucidum, the lesion differs from primary malformations in these other areas with respect to gross appearance, blood supply, and operability.\(^2\)

5. Finally, there is nothing about the size, location, or vascular supply of these lesions to suggest that they are inoperable. Of the seven cases operated
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![Postoperative right brachial arteriograms showing surgical clips in place and indicating that the lesion has been completely excised. There is partial filling of the anterior cerebral artery. Left: Anteroposterior view. Right: Lateral view.](image)

**TABLE 1**

*Summary of 10 cases of cirsoid arteriovenous malformation*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author, Year</th>
<th>Age (yrs), Sex</th>
<th>Presenting Signs†</th>
<th>Localizing Signs</th>
<th>Epilepsy</th>
<th>Surgery</th>
<th>Postop Course‡</th>
<th>Repeat Angiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Dany, et al. (1968)</td>
<td>32 M</td>
<td>subarachnoid hemorrhage (1)</td>
<td>none</td>
<td>0</td>
<td>excision</td>
<td>stupor and right hemiparesis necessitating reoperation; eventual resolution of deficits</td>
<td>incomplete excision</td>
</tr>
<tr>
<td>2</td>
<td>Dany, et al. (1968)</td>
<td>45 F</td>
<td>subarachnoid hemorrhages (2)</td>
<td>none</td>
<td>0</td>
<td>excision</td>
<td>uneventful</td>
<td>incomplete excision</td>
</tr>
<tr>
<td>3</td>
<td>Dany, et al. (1968)</td>
<td>42 M</td>
<td>subarachnoid hemorrhages (2)</td>
<td>none</td>
<td>0</td>
<td>excision</td>
<td>deterioration in affect and motivation</td>
<td>complete excision</td>
</tr>
<tr>
<td>4</td>
<td>Leppo, et al. (1956)</td>
<td>37 M</td>
<td>subarachnoid hemorrhages (4)</td>
<td>none</td>
<td>0</td>
<td>0</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>5</td>
<td>Basset, et al. (1951)</td>
<td>25 F</td>
<td>subarachnoid hemorrhages (4)</td>
<td>episodic right hemianesthesia</td>
<td>0</td>
<td>coagulation and clipping</td>
<td>uneventful except for one generalized seizure</td>
<td>none</td>
</tr>
<tr>
<td>6</td>
<td>Basset, et al. (1951)</td>
<td>40 F</td>
<td>subarachnoid hemorrhages (3)</td>
<td>none</td>
<td>0</td>
<td>coagulation and clipping</td>
<td>uneventful</td>
<td>none</td>
</tr>
<tr>
<td>7</td>
<td>Houdart, et al.* (1963)</td>
<td>not given</td>
<td>subarachnoid hemorrhages (5 or 6)</td>
<td>none</td>
<td>0</td>
<td>0</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>Houdart, et al.* (1963)</td>
<td>not given</td>
<td>subarachnoid hemorrhages (5 or 6)</td>
<td>none</td>
<td>0</td>
<td>0</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>9</td>
<td>Houdart, et al.* (1963)</td>
<td>not given</td>
<td>subarachnoid hemorrhages (5 or 6)</td>
<td>none</td>
<td>0</td>
<td>clipping of feeding arteries</td>
<td>*</td>
<td>incomplete excision presumed, data not given</td>
</tr>
<tr>
<td>10</td>
<td>Milhorat (1970)</td>
<td>16 M</td>
<td>subarachnoid hemorrhages (7)</td>
<td>none</td>
<td>0</td>
<td>excision</td>
<td>uneventful</td>
<td>complete excision</td>
</tr>
</tbody>
</table>

* Some data on Houdart's patients can be found in the paper of Dany, et al., 1968 (see ref. 2).
† Subarachnoid hemorrhage documented; six others suggested by the history.
‡ Long-term follow-ups are not available on any of the reported cases.

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on (including our current case), there were no deaths, one complication (cerebral edema mimicking intracerebral hematoma and requiring reoperation), and only one patient with residual neurological deficits (altered affect).

Although the physiological functions of the corpus callosum have not been fully defined, it seems clear that localized lesions involving this structure can be resected uneventfully with few or no neurological deficits. In three of the operated cases, the lesions were incompletely obliterated, and in two others postoperative angiograms were not obtained. Although there are insufficient data to judge the clinical value of these procedures, there is reason to believe that ligation of the feeding arteries alone is inadequate treatment. In view of the tendency for arteriovenous malformations elsewhere to recur after partial ablation, it is likely that total excision, when and where appropriate, is the only way to assure a full and lasting cure.

Summary

This report describes the diagnosis and successful excision of a cirsoid arteriovenous malformation of the corpus callosum in a 16-year-old boy. Because of their size, blood supply, and location, malformations in this region are surgically accessible and may be excised with few or no neurological deficits.

References

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