Surgical treatment of cryptic AVM's and associated hematoma in the brain stem and spinal cord


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Most surgically treated cases of brain-stem hematomas have been attributed to rupture of cryptic arteriovenous malformations (AVM's); however, very few cases have been histologically proven. Similarly, there are very few reports of surgically treated spontaneous hematomyelia, in which the hemorrhage has been histologically confirmed as being due to a purely intramedullary AVM. The authors report three cases with surgically treated, histologically confirmed AVM's, of which two were in the brain stem and the third was in the spinal cord. In all these cases, abnormal vascular tissue in the wall of the hematoma cavity was recognized at operation and excised.

KEY WORDS arteriovenous malformation □9 brain stem □9 spinal cord □9 cryptic lesion surgical treatment

Crawford and Russell 13 in 1956 first gave the designation "cryptic arteriovenous malformation (AVM)" to a cluster of abnormal arteriovenous connections that measure less than 2 to 3 cm in maximum size. Since then, a few major pathological studies have reported such lesions as incidental findings in the brain and spinal cord at autopsy in cases of fatal intracranial hemorrhage, the pons being the commonest site. 13,42,45,46,68,70 Although the term “cryptic” was originally applied to lesions of 2 to 3 cm, AVM's of a much smaller size (that is, a few millimeters in diameter) form the majority of cases reported, particularly in the brain stem and spinal cord.

We present three cases of cryptic AVM, two in the brain stem and one in the spinal cord. All three cases were treated surgically and the AVM’s were confirmed histologically.

Case Reports

Case 1

This 14-year-old girl was well until mid-January, 1982, when she experienced dizziness on moving her head, and intermittent headaches associated with vomiting and photophobia. Her initial symptoms subsided over the following 5 days, but recurred 10 days later. Following this, she noticed progressive drooping of the eyelids and complained of double vision in all directions of gaze, and of difficulty in swallowing.

Examination. The patient was dysarthric. She had bilateral internuclear ophthalmoplegia, rotatory nystagmus, mild bilateral ptosis, mild bilateral facial weakness, and bilateral ataxia, worse on the left side. Her left plantar response was extensor. Electroencephalography showed evidence of a deep midline lesion. A computerized tomography (CT) scan (Fig. 1) revealed a hematoma in the right posterolateral part of the pons displacing the fourth ventricle backward and to the right. There was a thin rim of contrast enhancement in the medial aspect of the lesion. Angiography showed no abnormal vessels but did demonstrate appropriate displacement.

Operation. At operation through a midline posterior fossa exposure, the entire floor of the fourth ventricle had yellowish discoloration and was seen to be bulging posteriorly. Blue hematoma was visible through the upper two-thirds of the ependyma, which was incised. This revealed a well-encapsulated hematoma composed of solid clot, liquid blood, and a margin of fibrin. The hematoma was evacuated together with a small angioma, 2 to 3 mm in diameter, which was encountered.
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Fig. 1. Case 1. Computerized tomography scan showing a posterior lateral pontine hematoma with fourth ventricular displacement. The lateral ventricles were of normal size.

Fig. 2. Case 1. Photomicrograph of the pontine angioma excised at surgery showing ectatic thin-walled vessels, together with areas of blood clot. H & E, × 12.

in the left anterior aspect of the lesion. Histological examination revealed several ectatic thin-walled vessels, together with portions of blood clot (Fig. 2).

Postoperative Course. The patient showed immediate improvement in her eye movements and dysarthria, and she was walking unaided 3 days later. When examined 1 year after the operation, she had no deficit apart from slight diplopia on extreme lateral gaze in either direction.

Case 2

This 28-year-old woman was well until November, 1984, when she developed severe occipital headache over a period of 12 hours, accompanied by vomiting and photophobia. She had no neck stiffness or pyrexia. The following day she awoke with a gross right hemiplegia. Over the next 24 hours, she proceeded to develop dysarthria and limitation of upward gaze.

Examination. She was drowsy and dysarthric, with a left sixth nerve palsy, right seventh nerve palsy, limitation of upward gaze, upward gaze nystagmus, and a dense right hemiplegia involving the face, arm, and leg. Her right plantar response was extensor. Electroencephalography showed a brain-stem disturbance, and a CT scan revealed a hematoma involving the midportion and left half of the body of the pons, displacing the slit-like fourth ventricle backward (Fig. 3). There was no evidence of abnormal contrast enhancement. Her condition deteriorated before cerebral angiography was contemplated, and she became unconscious with irregular breathing. She was referred immediately for surgery.

Operation. At operation through a retromastoid craniectomy, the entire pons appeared swollen, and the intrapontine hematoma was found to be close to the surface below the seventh nerve. The hematoma was reached through a pial incision, and a moderate quantity of liquid and solid clot was evacuated. A nodule of abnormal-appearing vascular tissue, 3 to 4 mm in diameter, was removed from the wall of the cavity. Histopathological examination revealed blood clot and portions of connective tissue containing a number of thin- and thick-walled blood vessels (Fig. 4). These findings were consistent with an AVM.

Postoperative Course. The patient's level of consciousness improved dramatically, but she was left with a dense right hemiplegia, diplopia, and lack of palatal movements. These gradually improved with intensive therapy. Seven months later she was able to speak and swallow, and could live independently at home. However, she is still dysarthric with a modest hemiparesis, but can walk with the help of a cane. Her left upper limb is normal but her right upper limb is functionless.

Case 3

This 38-year-old man presented with paresthesias and
numbness in both lower limbs of 7 months' duration. For 4 months before admission he had experienced progressively increasing weakness in both legs, first noticed in the right foot when he was driving. For 1 week prior to admission he had noted urinary hesitancy and urgency, impotence, and constipation. At no stage had he experienced back pain.

Examination. The patient had no systemic abnormalities, vascular skin lesions, or spinal deformity. He could stand only with support and had a moderately severe spastic paraparesis, worse on the right side. The knee and ankle jerks were exaggerated and both plantar responses were extensor. Sensory examination revealed impairment of light touch and pinprick to the level of the T-10 dermatome. Proprioception in both legs was severely disturbed. Plain x-ray films of the spine were normal. Myelography (Fig. 5 upper) demonstrated a localized swelling of the spinal cord at the T-7 level without complete obstruction to the flow of contrast material. There was no feature to suggest abnormal vascular channels around the spinal cord. This intramedullary mass was also shown on spinal CT scanning (Fig. 5 lower).

Operation. A midthoracic laminectomy was performed. No obvious extradural abnormality was seen. A localized bluish mass presented at the surface of the cord (Fig. 6 left), and a midline posterior myelotomy was made, causing old liquid hematoma to extrude under pressure. The hematoma cavity was lined with areas of solid clot. A nodule of abnormal-appearing vascular tissue was subtotally excised (Fig. 6 right), the residue being obliterated with bipolar diathermy. Histological examination of this vascular tissue confirmed the presence of an AVM (Fig. 7).

Postoperative Course. In the 48 hours immediately following surgery, the patient experienced an increase in weakness in his left leg, but he then rapidly improved. He was discharged on the 10th day, able to walk with the aid of crutches. At 5 months he was walking unaided, but still had some mild residual proprioceptive disturbances confined to his right leg. He also claimed some improvement in sexual function.
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Fig. 6. Case 3. Intraoperative appearance of the spinal cord. Left: A localized bluish mass is visualized at the surface. Right: Appearance following drainage of the intramedullary hematoma and excision of the abnormal vascular tissue.

Fig. 7. Case 3. Photomicrograph of the abnormal vascular tissue excised from within the spinal cord. Thin- and thick-walled vascular channels are noted within the areas of blood clot. H & E, x 13.

Discussion

Cryptic intracranial AVM's have an equal distribution in both the supra- and infratentorial compartment, in contrast to large AVM's which most often occur supratentorially. According to most authors there is a male:female preponderance of 2:1. The incidence of hemorrhage as the first presentation, reported in series that include both large and cryptic AVM's (both supra- and infratentorial) is variable. This ranges from 20% to 75%. The risk of a second hemorrhage is between 23% and 45%, ranging over a few days to many years. Most authors have found a higher risk of hemorrhage in small AVM's.

There are 42 cases in the literature, including our previous series, and numerous isolated case reports where brain-stem hematomas have been surgically treated. In the vast majority of these cases, the cause of hemorrhage was not identified and in some cases it was attributed speculatively to rupture of a cryptic AVM. Histological confirmation was reported in only two cases, in which vessel fragments were seen in aspirated clot. In our present series, the AVM's were found on careful inspection of the wall of the hematoma cavity with the operating microscope. The lesions were firmly attached to the wall and would not have been removed by simple aspiration. In our previously reported series, evacuation of the clot without evidence of removal of an underlying AVM was not followed by recurrent hemorrhage during follow-up periods from 4 to 14 years; however, it is difficult to avoid the conclusion that failure to excise a cryptic AVM that has bled once must carry a risk of further hemorrhage. Furthermore, the presenting clinical picture is commonly that of repeated incidents, suggesting that cryptic AVM's tend to rebleed, like the larger lesions.

The clinical presentations in our Cases 1 and 2, with brain-stem AVM's, showed relapsing deterioration in Case 1 and a rapidly progressive course in Case 2. The variation in course supports the view that there is no typical clinical picture of brain-stem hematomas due to rupture of cryptic AVM's. However, this entity tends to occur in children and young adults.

It is clear that while some patients with brain-stem hematomas may survive when treated conservatively, the incidence of significant disability among such patients is high. While stereotaxic evacuation of such hematomas is now a recognized method of treatment, the possibility remains that a cryptic AVM may be missed and that there may be a risk of further hemorrhage. We have previously demonstrated that surgical evacuation of a brain-stem hematoma is safe and effective, and offers a high incidence of recovery of neural function. We now offer further confirmation of this and suggest that an associated AVM within the brain stem may be safely excised, thus minimizing the risk of further hemorrhage.

Cryptic AVM's in the spinal cord have received less attention than those in the brain stem, although spontaneous hematomyelia is a well-recognized entity. The commonest reported causative lesion is a spinal AVM.
of a size large enough to be demonstrated angiographically.16 Hematomyelia occurring in the absence of a large AVM has commonly been associated with other systemic diseases, such as blood dyscrasias, syphilis, and arteriosclerosis.30 There have been few reports of surgically treated spontaneous intramedullary hemorrhages.13,34,35,50,61 When present, AVM's are usually large obvious lesions,7,23,34,39,50,64 but the possibility of “micro-angiomas” in cases of hematomyelia without a large AVM and in the absence of other recognized causes has been suggested.35 Spontaneous hematomyelia appears to present either acutely with pain and rapid onset of profound neurological deficit, or alternatively in a slower subacute manner, simulating an intrinsic spinal cord tumor, as was the case with our Case 3. Aminoff and Logue2 have analyzed the clinical features in a large series of patients with spinal AVM's, but their analysis was based on lesions that were invariably large enough to be diagnosed neuroradiologically or macroscopically. Therefore, from their report it was not possible to derive a pattern of presentation in cases with purely intramedullary lesions.

The spinal AVM in our Case 3 was identical with the cryptic AVM's described in association with brain-stem hematomas. Evacuation of the hematoma and excision of the AVM were equally effective and safe.

References


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