Dural arteriovenous malformation of the skull base with intraosseous vascular nidus

Report of two cases

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Intracranial arteriovenous malformations (AVM’s) have been classified as pure pial, pure dural, and mixed pial and dural. Dural AVM’s are relatively uncommon, with 377 cases documented up to 1990. These lesions were believed to be situated within the walls of the sinuses, but during the last decade researchers discovered a small subgroup of dural AVM’s in extrasinusual locations such as the skull base and tentorium. Two of the 17 patients who were studied between 1976 and 1993 had dural AVM’s that were entirely intraosseous except for their venous drainage, which was via the dural venous sinuses. Although such intraosseous dural AVM’s have not been previously described, the authors elected to group these malformations with dural AVM’s because their venous drainage was intracranial and angiograms revealed identical features.

KEY WORDS: dural arteriovenous malformation • intraosseous nidus • skull-base lesion

Although a few cases of dural arteriovenous malformation (AVM) were reported in the 1930’s, the concept of an arteriovenous communication developing between a dural artery and a venous sinus was first introduced in 1951. During the last 40 years, more patients with this condition were diagnosed because of better understanding of the clinical manifestations and improvements in diagnostic techniques. It was found that, in addition to draining into a venous sinus, these AVM’s drain into dural venous lakes and cortical veins. Such AVM’s were initially described as located solely in the dura mater, which contains dural venous sinuses, but during the last decade researchers have discovered them in other locations such as the tentorium and skull base.

From 1976 to 1993 at our institution, the senior author (G.M.M.) performed surgery for 257 cases of AVM, 17 of which were dural. Two of these cases were very unusual in that angiograms revealed all the features of dural AVM’s except that both the arterial feeders and the actual AVM nidus were situated in the bone around the foramen magnum, although the venous drainage was via intracranial dural venous sinuses. However, after bone drilling was completed, no AVM was found either on the outer or inner surface of the dura.

Case Reports

Case 1

This 48-year-old man had experienced pulsatile tinnitus in his left ear for 6 months and also complained of headaches, concentrated mainly on the left side. His medical history was unremarkable, with no report of any significant head trauma.

Examination. Examination showed the patient to be neurologically intact. On auscultation a loud bruit was audible over his left mastoid area. An angiogram had been performed at another institution; the repeat angiogram obtained at our institution revealed a large dural AVM on the left side of the foramen magnum, craniovertebral junction, and inferior aspect of the clivus. This malformation was fed mainly by meningeal branches of the left occipital, posterior auricular, and both middle meningeal arteries, the meningohipophyseal artery, and the anterior meningeal branches of both vertebral arteries. These arteries drained into numerous veins over the clivus, which drained into the inferior petrosal sinus and subsequently into the cavernous sinus. The major venous drainage was into large venous channels situated in the soft tissues of the neck; these channels then drained into internal and external jugular veins. The patient had also undergone a cervical my-
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**Fig. 1, Case 2.** Selective angiograms showing a dural arteriovenous malformation (AVM). **Left:** Right vertebral angiogram, anteroposterior view, showing the dural AVM. **Right:** Right occipital angiogram, lateral view, showing filling of the AVM.

cologram that was unremarkable except for some prominence of epidural space from the C1–3 level.

**Operation.** A suboccipital craniectomy was performed on the left side, extending to the mastoid region. At the craniocaudal junction, a large arterialized vein was encountered laterally. The dura was vascular, but there was no localized collection of blood vessels. The dura was opened but no malformation was found on the inner surface. The C-1 arch was removed on the left side and the bony covering of the vertebral artery was unroofed. The large arterial feeders arising from the vertebral arteries outside the dura were coagulated. The farther portion of the occipital bone next to the condyle was removed. This bone was very vascular with large vessels; bleeding was controlled with bone wax. After the drilling was completed, the dural opening was extended to C–2, and again no definite intradural abnormality was noted. The large arterialized vein that was encountered soon after exposure had collapsed. Dissection was performed laterally into the muscles; however, no major vessels were encountered.

**Postoperative Course.** The patient experienced some nausea, which subsided, and he recovered without any complications. There was an immediate cessation of bruit and a follow-up angiogram revealed that the AVM was completely gone. Unfortunately, pre- and postoperative films are no longer available for Case 1.

**Case 2**

This 49-year-old right-handed woman developed a syncopal episode at approximately the same time she noticed pulsatile tinnitus behind her right ear. She also complained of pain and stiffness on the right side of her head and neck. Her medical history was unremarkable except for minor surgery.

**Examination.** Examination showed the patient to be neurologically intact. A loud bruit was audible over the right mastoid area. A cerebral angiogram revealed a dural AVM at the level of the foramen magnum and craniovertebral junction on the right side fed by the occipital, posterior auricular, and ascending pharyngeal arteries. The malformation was also supplied by the posterior meningeal branch of the right vertebral artery (Fig. 1), and collateral flow via branches of thyrocervical and costocervical arteries. The AVM drained mainly into the right internal jugular veins, the right paraspinal vein, and right epidural venous plexus within the spinal canal.

The patient underwent embolization and all feeding vessels were successfully occluded except for a small residual supply from the right vertebral artery. After embolization, the patient reported feeling much better. The pulsatile tinnitus had disappeared. After discussion with the patient, it was decided to delay surgery for 6 weeks and to obtain a repeat angiogram at that time. This was planned to give the AVM an adequate chance of undergoing thrombosis. If there were still significant arteriovenous shunting, surgery would be performed. After the 6-week delay, the repeat angiogram was performed, which revealed that the AVM had increased to pre-embolization size and had recruited some new vessels. It was now supplied by cavernous branches of both internal carotid arteries, branches from the right vertebral artery, and the ascending pharyngeal artery, which crossed the midline to supply the AVM. The patient was scheduled to undergo surgery the next day.

**Operation.** A right retrosigmoid craniectomy was performed. Bone was removed from the transverse sinus superiorly to the foramen magnum inferiorly. The C-1 arch was also removed on the right side. The dura was opened but then closed as no AVM was seen intradurally. Because the AVM nidus was medial to the jugular bulb, the sigmoid sinus was gradually skeletonized until the jugular bulb was reached; bone removal was continued to the lateral edge of the occipital con-
Fig. 2. Case 2. Three-dimensional reconstruction of postoperative computerized tomography scans. *Left:* Inferior view showing the extent of craniectomy (arrows). *Right:* Superior view showing bone removal (arrows), which was continued to the lateral edge of the occipital condyle.

dyelo (Fig. 2). Bleeding from the large blood vessels in the bone was controlled with bone wax. No vessels were seen on the outer surface of dura.

Postoperative Course. Postoperatively the pulsatile tinnitus immediately disappeared, and the patient recovered without any problems. A follow-up angiogram did not reveal any residual AVM (Fig. 3).

Discussion

Review of the Literature

Dural AVM's represent 10% to 15% of all intracranial AVM's, and 6% of supratentorial and 35% of all infratentorial AVM's are dural.\(^8\) Since our review of 213 cases in 1984,\(^7\) more reports have appeared in the literature, with the most recent major review in 1990 citing the total number of dural AVM's as 377.\(^2\) Dural AVM's are interesting because of their variable clinical features as well as their pathogenesis. Early reports emphasized that they were congenital,\(^1\) resulting from enlargement of intradural arteriovenous shunts. Other investigators\(^4,6\) proposed that they were acquired lesions arising after sinus thrombosis and trauma. Central to this hypothesis is the concept of physiological arteriovenous fistulas in the dura, which may increase in size secondary to traumatic disruption or because of com-

Fig. 3. Case 2. Postoperative selective angiograms. *Left:* Right vertebral angiogram, anteroposterior view. *Right:* Right common carotid angiogram, lateral view, showing complete excision of the arteriovenous malformation.
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pensatory hypertrophy after sinus thrombosis. Still
other researchers believe that sinus thrombosis arises
secondary to the turbulence of blood resulting from de-
velopment of arteriovenous fistulas and is the effect
rather than the cause of dural AVM. All studies agree
that there are arteriovenous communications in the dura
that for varied reasons may increase in size and lead to
formation of a full-blown dural AVM. Dural AVM's
were initially described only in association with the ve-
nous sinuses but subsequently have been found in other
locations such as the tentorium, skull base, and the in-
traorbital area. It is understandable how they can
develop in these different locations because numerous
arteriovenous shunts normally occur all over the dura.

Other researchers have postulated that dural AVM's can
develop wherever veins follow a transosseous trajec-
tory. They believe such veins are similar to the vessels
in the walls of the dural sinuses; therefore, it is easy
to understand how an AVM can be located entirely
within the bone. Also, the vascular supply of the head
and neck initially appears as one unit and only during
the third stage of development of cerebral circulation
does this supply separate into superficial, dural, and
pial vessels. It is probable that at this time the location
(pial, dural, or superficial to the dura) of a potential
AVM is determined.

Both intracranial AVM's in our patients were situ-
ated around the foramen magnum. Whether this is a
coincidence is difficult to determine. Extranasial dural
AVM's located at the skull base commonly occur
around the edges of the foramen magnum, possibly
due to a large number of transosseous emissary veins
at this location. Extranasial lesions as well as tentorial
AVM's may have a worse prognosis. In the series re-
ported by Pierot, et al., 67% of such lesions were asso-
associated with intracranial hemorrhage and 17% with
paraparesis.

Conclusions

Although little is known about these lesions, our
cases illustrate that some of them may be situated en-
tirely within the bone, which can cause considerable
hemorrhage during bone removal. Preoperative em-
bolization can help to reduce the amount of bleeding;
however, embolization alone is seldom curative as
demonstrated by our Case 2. It is, therefore, important
to recognize these lesions intraoperatively: the com-
plete obliteration of the AVM and relief of symptoms
depends on the extent of bone removal. If such an AVM
is operated on and no distinct malformation is seen out-
side or inside the dura, bone removal should continue
until the entire extent of the malformation (based on
preoperative studies) is exposed and the dura is in-
spected. This can be judged to a certain degree by the
extent of bleeding from the involved bone.

References

   arteriovenous malformations: factors predisposing to an
   aggressive neurological course. J Neurosurg 72:839–850,
   1990
   neous carotid-cavernous fistulas. Surg Neurol 17:
   282–286, 1982
   venous malformation of the major venous sinuses: an ac-
   quired lesion. AJNR 3:13–19, 1982
5. Fincher EF: Arteriovenous fistula between the middle men-
   ingeal artery and the greater petrosal sinus. Case report. Ann
   Surg 133:886–888, 1951
6. House OW, Campbell JK, Campbell RJ, et al: Arteriove-
   nous malformation affecting the transverse dural venous
   1979
   malformations and intracranial hemorrhage. Neurosurgery
   15:332–339, 1984
8. Newton TH, Cronqvist S: Involvement of dural arteries in
   intracranial arteriovenous malformations. Radiology 93:
   1071–1078, 1969
   of dural arteriovenous malformations of the lateral and sig-
   moid sinuses based on histopathological examinations. J
   Neurosurg 76:600–606, 1992
    alas of the posterior fossa draining into subarachnoid veins.
    AJNR 13:315–323, 1992
11. Piske RL, Lasjaunias P: Extranasial dural arteriovenous
    malformations. Report of three cases. Neuroradiology 30:
    426–432, 1988
12. Rowbotham GF, Little E: Circulations of the cerebral hemi-

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