Mixed Pial-Dural Arteriovenous Malformation in the Anterior Cranial Fossa
—Two Case Reports—

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Abstract
Most arteriovenous malformations (AVMs) associated with the meningeal artery in the anterior cranial fossa are the pure dural type, and mixed pial-dural AVMs are rare. Two types of mixed pial-dural AVM occur in the anterior cranial fossa according to the shunting point: one with the nidus in the brain parenchyma of the frontal lobe, and the other with the shunting point in the dura mater. We describe two patients with AVMs fed by the anterior ethmoidal arteries and the persistent primitive olfactory artery, with the nidus located in the pure brain parenchyma of the inferior aspect of frontal lobe, and drained via an abnormal cortical vein into the cavernous and superior sagittal sinuses. The importance of occluding the venous outflow to obliterate intracranial dural arteriovenous fistula (AVF) is emphasized. However, removal of the nidus in the brain parenchyma is required. The presence of a pial feeder should be considered before diagnosis of dural AVF of the anterior cranial fossa, and preoperative detailed evaluation for the pial supply and shunting point is mandatory.

Key words: mixed pial-dural arteriovenous malformation, anterior cranial fossa, meningeal artery, pial artery, surgical resection

Introduction
Intracranial arteriovenous malformations (AVMs) can be classified on the basis of their arterial supply into pure pial, mixed pial-dural, and pure dural AVMs. Dural arteriovenous fistulas (AVFs) are usually associated with venous hypertension due to sinus thrombosis, and frequently occur in the regions of the transverse and sigmoid sinuses. Dural AVFs are now known to be entities distinct from AVMs. Pial AVMs are generally considered to be a developmental malformation, whereas dural AVFs usually develop secondary to sinus thrombosis, trauma, and surgery. The incidence of pure dural AVFs is lower in the anterior cranial fossa than in other parts of the brain, but the clinical characteristics have been clarified to some extent. Mixed pial-dural AVM in the anterior cranial fossa is rare, with only 11 reported cases. Two types of mixed pial-dural AVM occur in the anterior cranial fossa: one with the nidus in the brain parenchyma of the frontal lobe, and the other with the shunting point in the dura mater.

We report two cases of mixed pial-dural AVM in the anterior cranial fossa with the nidus in the brain parenchyma.

Case Reports

Case 1: A 68-year-old man presented with severe headache and vomiting. He was hospitalized 2 hours after the onset of the symptoms and became comatose at the time of admission. Computed tomography (CT) revealed intracerebral hemorrhage in the left frontal base with ventricular rupture and acute hydrocephalus (Fig. 1). The patient underwent ventricular drainage, and gradually regained consciousness. Subsequently, angiography was performed. Three-dimensional (3D) digital subtraction angiography revealed a mixed pial-dural AVM fed by an anomalous frontoorbital artery and the bilateral anterior ethmoidal arteries (AEAs). The malformation drained into the cavernous sinus via the dilated cortical vein and superior sagittal sinus (Figs. 2 and 3). The patient underwent bifrontal craniotomy 7 days after admission. Opening of the dura mater revealed several draining veins and a dilated venous sac on the surface of the frontal lobe. The patient underwent bifrontal craniotomy 7 days after admission. Opening of the dura mater revealed several draining veins and a dilated venous sac on the surface of the frontal lobe. However, no drainage vein was recognized between the dura mater and the brain surface. Shunts from the anomalous frontoorbital artery on the brain surface, the AEA, and small arteries passing through the dura mater around the olfactory groove were observed and coagulated (Fig. 4). The nidus was located in the pure brain parenchyma of the anterior cranial fossa. Thereafter, the nidus with the venous sac on the surface was removed.
Postoperative angiography showed no evidence of residual vascular malformations (Fig. 5). Histological examination revealed vessels of various sizes in the brain parenchyma and the diagnosis was AVM (Fig. 6). The postoperative course was uneventful. Complete recovery of the impaired higher cognitive functions was noted after the sur-

Fig. 1 Case 1. Preoperative computed tomography scan showing intracerebral hemorrhage in the left frontal base with intraventricular hematoma and acute hydrocephalus.

Fig. 2 Case 1. A: Preoperative left carotid angiogram showing the arteriovenous malformation supplied by the left anterior ethmoidal artery (arrowhead), and the anomalous left frontoorbital artery (arrow). B: Preoperative three-dimensional angiogram showing the arteriovenous malformation supplied by the left anterior ethmoidal artery (arrowhead), and the anomalous left frontoorbital artery (arrow).

Fig. 3 Case 1. A: Preoperative right carotid angiogram showing the arteriovenous malformation supplied by the right anterior ethmoidal artery. B, C: Preoperative left carotid angiogram showing the malformation drained into the cavernous sinus via the dilated cortical vein and superior sagittal sinus.

Fig. 4 Case 1. A: Intraoperative photograph showing the anterior ethmoidal artery (arrow) and the small meningeal artery (arrowhead) through the dura mater around the olfactory groove. The venous drainage between the dura mater and brain surface was not recognized. B: Intraoperative photograph showing the anomalous frontoorbital artery (arrow) as a pial supply.

Fig. 5 Case 1. Postoperative left carotid angiograms, anteroposterior (A) and lateral views (B), showing disappearance of the mixed pial-dural arteriovenous malformation in the anterior cranial fossa and vasospasm of the main arteries.

Fig. 6 Case 1. Photomicrograph showing vessels of various sizes in the brain. Victoria blue hematoxylin and eosin stain, ×20.
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and the brain surface was recognized. Postoperative mag-
frontal lobe. No venous drainage between the dura mater

cated in the brain parenchyma of the inferior aspect of the

the nidus with venous sac was removed. The nidus was lo-

perior sagittal sinus via the dilated cortical vein. The

internal carotid artery as a persistent primitive olfactory

an anomalous left AEA and a pial artery originating from

hemorrhage and subarachnoid hemorrhage (Fig. 7A). 3D

onset and became comatose. CT revealed left frontal base

A 76-year-old man was hospitalized one hour after

Case 2: A 76-year-old man was hospitalized one hour after

images.7,14,15,28) Acquired pial AVMs may develop after si-

vasorum of normal pachymeninges, and may stimulate

arteriovenous shunts, which are found within the vasa

hypertension may promote the growth of microscopic

artery, anterior falx artery, sphenopalatine artery, and su-

anomalous branches of the anterior cerebral artery in 8

patients and arterial supply from the internal carotid ar-

tery in one patient were found to supply blood to the nidus

preoperatively. No pial arterial supply was found in the

preoperative angiograms of 4 of the patients, but was

found during the operation. 3D CT and 3D digital subtrac-

angiography are useful to evaluate the presence of

pial supply as in our cases. The AVMs drained into the su-

uperior sagittal sinus through the pial vein in 10 patients, the
cavernous sinus in 3, the transverse sinus in 1, and the

straight sinus in 1. Venous pouches were present on the
draining cortical vein in 11 of the 13 patients. The nidus

was located in the brain parenchyma in 10 patients, in the
dura mater in 2 patients, and the location was not de-

scribed in 1 patient.

The pathogenesis of mixed pial-dural AVMs in the an-
terior cranial fossa remains unclear. The presence of ab-
normal communications between the pial and meningeal

arteries in the embryonic stage leads to the development of

mixed pial-dural AVMs.27) The anomalous, persistent

primitive olfactory artery that arises from both internal
carotid artery and anterior cerebral artery has a connec-

tion with the ethmoidal artery.21) However, pial arteries

have an embryologic origin different from that of dural ar-
teries, so the angiogenic mechanisms involved in the de-

velopment of these two arterial systems may not necessari-

ly be identical.20)

Venous hypertension occurring secondary to sinus

thrombosis is believed to be the primary mechanism

responsible for the formation of dural AVFs.23) Venous

hypertension may promote the growth of microscopic

arteriovenous shunts, which are found within the vasa

vasorum of normal pachymeninges, and may stimulate

the release of angiogenic factors in experimental

models.7,14,15,28) Acquired pial AVMs may develop after si-

nus or cerebral venous thrombosis.22) The mechanism

causing these malformations originates in blocking of the
communications between the cortical veins and dural

sinuses as a result of retrograde thrombus propagation,
resulting in increased cortical venous pressure, leading to
the development of pial AVMs.22) Thus, the occurrence of

venous hypertension after venous obstruction, the expres-

sion of angiogenic factors, and other angiogenic mechan-
isms may lead to the development of both pial and dural

malformations in the anterior cranial fossa.

Twelve of the 13 patients were managed surgically by

either frontal or bifrontal craniotomy, whereas the other

patient was conservatively observed. Among the feeding

arteries, the dural meningeal arteries were present in the

cribiform plate, and the pial arteries were present either in
the brain parenchyma or on the brain surface. The clas-

sification of AVM into pure dural, pure pial, and mixed

pial-dural types depends on the blood supply and not the

location of the shunting point. Two previous patients with

mixed pial-dural AVM of the anterior cranial fossa had the

shunting point in the dura mater. In this type, occlusion

on the venous side adjacent to the shunt is sufficient.16)

Discussion

Table 1 shows the 13 patients including our two patients,
of whom 11 were men with mean age of 61.8 years at on-

set. A new embryological classification of dural AVFs sug-
gested that fistulas located in the lateral epidural space,

which includes the lamina cribrosa ossis ethmoidalis, are

strongly predominant in men, whereas those located in

the ventral epidural space are more common in women.5)

Dural AVFs in the anterior cranial fossa occur

predominantly in men.24) Further, mixed pial-dural AVMs

in the anterior cranial fossa also show the same level of

male predominance. Most patients with mixed pial-dural

AVMs in the anterior cranial fossa were symptomatic at

the time of diagnosis (11 of 13). However, the lesion was

discovered incidentally in the remaining 2 patients. Eight

patients presented with intracranial hemorrhage, 1 patient

with epilepsy, 1 patient with fourth nerve palsy, and 1

patient with pain in the forehead.

The AVMs were fed by the AEA in all 13 patients: by the

ipsilateral AEA in 10 patients, and by the bilateral AEsAs

in the remaining 3. The AVMs were supplied by the exter-

nal carotid arteries via branches of the middle meningeal

artery, anterior falx artery, sphenopalatine artery, and su-

perficial temporal artery in 3 of the 13 patients. The

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<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Age (yrs)/Sex</th>
<th>Clinical symptoms</th>
<th>Location of nidus</th>
<th>Feeding arteries</th>
<th>Draining vein</th>
<th>Vascular pouch</th>
<th>Operation</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Terada et al. (1984)</td>
<td>64/F</td>
<td>ICH</td>
<td>cortex</td>
<td>ipsi AEA frontal pial artery</td>
<td>SpV → TS</td>
<td>yes</td>
<td>open surgery</td>
<td>MD</td>
</tr>
<tr>
<td>2</td>
<td>Tiyaworabun et al. (1986)</td>
<td>57/M</td>
<td>ICH, SAH</td>
<td>cortex</td>
<td>bil AEAs ACA branches</td>
<td>pial vein → SSS, CS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>3</td>
<td>Martin et al. (1990)</td>
<td>48/M</td>
<td>ICH dura (falx)</td>
<td>parenchyma</td>
<td>ipsi AEA, STA ACA branches</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>4</td>
<td>Tanaka et al. (1991)</td>
<td>60/M</td>
<td>incidental</td>
<td>dura, olfactory bulb</td>
<td>ipsi AEA FOA</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>5</td>
<td>Yamamoto et al. (1993)</td>
<td>62/M</td>
<td>IVth nerve palsy</td>
<td>parenchyma</td>
<td>ipsi AEA, ipsi MMA, bil AFAs ACA branches</td>
<td>pial vein → dural vein</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>6</td>
<td>Yoshida and Yamamoto (1993)</td>
<td>51/M</td>
<td>ICH</td>
<td>parenchyma</td>
<td>ipsi AEA FBA</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>7</td>
<td>Yoshimoto et al. (1993)</td>
<td>66/M</td>
<td>ICH</td>
<td>cortex</td>
<td>ipsi AEA FPA</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>8</td>
<td>Hashimoto et al. (1996)</td>
<td>65/M</td>
<td>epilepsy</td>
<td>cortex</td>
<td>ipsi AEA FPA</td>
<td>pial vein → SSS</td>
<td>no</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>9</td>
<td>Gliemroth et al. (1999)</td>
<td>69/F</td>
<td>ICH, IVH</td>
<td>cortex</td>
<td>ipsi AEA, ipsi STA, ipsi MMA ACA branches</td>
<td>SSS, CS</td>
<td>no conservative</td>
<td>not described</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>Kawaguchi et al. (1999)</td>
<td>60/M</td>
<td>headache</td>
<td>not described</td>
<td>ipsi AEA, FPA, FPA</td>
<td>pial vein → SSS, CS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>11</td>
<td>Kanai et al. (2000)</td>
<td>57/M</td>
<td>incidental</td>
<td>cortex</td>
<td>ipsi AEA FOA</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>12</td>
<td>Present Case 1</td>
<td>68/M</td>
<td>ICH, IVH</td>
<td>cortex</td>
<td>bil AEAs FOA</td>
<td>pial vein → CS, SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>GR</td>
</tr>
<tr>
<td>13</td>
<td>Present Case 2</td>
<td>76/M</td>
<td>ICH, SAH</td>
<td>cortex</td>
<td>ipsi AEA PPOA</td>
<td>pial vein → SSS</td>
<td>yes</td>
<td>open surgery</td>
<td>MD</td>
</tr>
</tbody>
</table>

However, in the type with the nidus in the brain parenchyma, removal of the nidus with/without the venous sac is necessary. The outcome was favorable in 11 of the 13 patients (good recovery in 9 patients, and moderate disability in 2). One patient showed severe disability, and the outcome was not reported for the other patient. Surgical resection is the most reliable procedure for the treatment of mixed pial-dural AVMs in the anterior cranial fossa. However, the limited space around the cribriform plate difficult to identify and ligate. Therefore, a combination of endovascular embolization and open surgery is a possible therapeutic option.

Two types of mixed pial-dural AVM occur in the anterior cranial fossa according to the location of the shunting point. In the type with the shunting point in the dura mater, simple venous out flow disconnection is sufficient. However, in the other type with the nidus in the brain parenchyma, removal of the nidus is required. The presence of a pial feeder should be considered before diagnosis of dural AVF of the anterior cranial fossa, and preoperative detailed evaluation of the pial supply and shunting point is necessary.

References


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