Orbital Drainage from Cerebral Arteriovenous Malformations

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- OBJECTIVE: To describe the neuro-ophthalmic findings in patients with orbital drainage from cerebral arteriovenous malformations (AVMs).
- METHODS: We reviewed the records of 100 consecutive adult patients with cerebral AVMs who presented to our institution during a 4-year period. All patients with orbital drainage were identified, and their neuro-ophthalmic evaluations were reviewed.
- RESULTS: Three patients (3%) were identified with orbital drainage from a cerebral AVM. The first patient presented with typical chiasmal syndrome (reduced visual acuity, bitemporal hemianopia, and optic atrophy). Magnetic resonance imaging demonstrated a large left temporal and parietal lobe AVM with compression of the chiasm between a large pituitary gland and a markedly enlarged carotid artery. The second patient presented with headaches and postural monocular transient visual obscurations. Examination revealed normal visual function with minimal orbital congestion and asymmetrical disc edema, which was worse in the left eye. Magnetic resonance imaging revealed a large right parietal and occipital lobe AVM without mass effect or hemorrhage and an enlarged left superior ophthalmic vein. The third patient had no visual symptoms and a normal neuro-ophthalmic examination; a right parietal lobe AVM was discovered during an examination for the cause of headaches.
- CONCLUSION: Orbital drainage from cerebral AVMs is rare. Manifestations may include anterior visual pathway compression, dilated conjunctival veins, orbital congestion, and asymmetrical disc swelling. (Neurosurgery 46:820-824, 2000)
- Key words: Asymmetrical optic disc edema, Cerebral pial arteriovenous malformation, Chiasmal syndrome, Elevated intracranial pressure, Orbital drainage

ntracranial arteriovenous malformations (AVMs) are broadly categorized as dural, pial, or mixed dural-pial on the basis of their arterial supply. Dural AVMs receive their blood supply from the meningeal branches, and pial AVMs are supplied by the cerebral or cerebellar arteries (16). Dural AVMs and direct fistulae, particularly those involving the cavernous sinus with drainage into the orbit, have a characteristic neuro-ophthalmic presentation resulting from arteriolization of the orbital venous system. Symptoms include increased intraocular pressure, cranial nerve palsy, conjunctival chemosis, arterialized conjunctival vessels, bruit, and venous stasis retinopathy (14). Patients with pial AVMs commonly present with headaches, seizures, and intracranial hemorrhage (7, 14, 15, 18). Depending on location, size, and pattern of venous drainage, the ophthalmic signs and symptoms of pial AVMs may include visual field defects and positive visual phenomena (4, 5, 14). Venous drainage of pial AVMs usually occurs into the deep venous system associated with the ventricles, or it may follow the pattern of the transcerebral venous system (14). We examined three patients with orbitally draining cerebral AVMs, two of whom exhibited distinct neuro-ophthalmic signs and symptoms.

Orbital Drainage from Cerebral Arteriovenous Malformations

PATIENTS AND METHODS

We reviewed the clinical records of 100 consecutive adult (>18 yr) patients with pial AVMs who presented to the Hospital of the University of Pennsylvania between January 1991 and December 1994 for neurosurgical consultation and who subsequently underwent cerebral angiography. We also analyzed the cerebral angiograms of these 100 patients with AVMs. Three patients with orbital drainage were identified, two of whom had relevant neuro-ophthalmic signs and symptoms. All three patients with orbitally draining AVMs underwent complete neuro-ophthalmic examinations, including assessment of visual acuity, color vision, intraocular pressure, and pupillary reactions, orbital and slit lamp examinations, formal examination of visual fields, and dilated funduscopy. Angiograms of the three patients included in the study displayed features of classic AVM architecture: an arterial feeding vessel or vessels, a nidus of arterial and venous tissue, and an early draining vein. No patients with solely venous angiomas, dural AVMs, or dural fistulae were included. Various angiographic characteristics of these lesions were considered. Venous drainage was characterized as occurring through the deep venous system alone, combined superficial and deep, or superficial alone. The orbitally draining AVMs were detected by abnormal filling of the superior ophthalmic veins.

RESULTS

Orbitally draining cerebral pial AVMs were identified in only 3 of the 100 patients observed during this period. The first patient had a 7.5-cm AVM in the left temporal and parietal lobes. The second had a 3-cm AVM in the right parietal and occipital lobes. The third patient had a 3.5-cm lesion in the posterior aspect of the right side of the splenium of the corpus callosum.

Case reports

Patient 1

A 40-year-old right-handed woman presented with blurred vision in both eyes. Approximately 10 years before, she had developed seizures and been diagnosed with an AVM. Her adherence to her anticonvulsant regimen was poor, and she had experienced approximately one seizure per year for many years. She intermittently used cocaine.

Her visual acuity was 20/20 in the right eye and 20/25 in the left eye. When given the Ishihara test, the patient correctly identified 10 of 15 plates on the right side and 3 of 15 plates on the left side. A left afferent pupillary defect and a mild bilateral proptosis were observed. The globes were nonpulsating. Mild soft tissue swelling of the eyelids along with dilated veins in the conjunctival fornices was observed. Bilateral temporal optic disc pallor was observed (*Fig.* 1), and a bruit was heard over the left orbit. A computerized visual field test revealed complete right homonymous hemianopia. In addition, a superotemporal visual field defect was evident in the left eye. The rest of the neurological examination results were normal. Magnetic resonance imaging revealed a large left hemispheric AVM involving the temporal and parietal lobes as well as the left optic tract. The superior



FIGURE 1. Patient 1. Fundus photograph of the left eye demonstrating temporal pallor of the optic disc and corresponding loss of nerve fiber striations (*arrows*).

ophthalmic vein was dilated on both sides (*Fig. 2*). The intracavernous carotid artery was enlarged, and veins around the chiasm were dilated. The chiasm was observed to be compressed between the carotid artery and an enlarged pituitary gland (*Fig. 2*). A cerebral angiogram confirmed the presence of large veins around the chiasm, which were draining into the orbit. The AVM was noted to be supplied by the middle cerebral and lenticulostriate arteries. We decided to follow the patient unless evidence of progressive visual field loss emerged. Her automated fields have remained stable for 3 years.

Patient 2

A 38-year-old right-handed woman with a long history of seizures developed an intermittent "pulling" pain around her left eye. She also experienced episodic visual graying in the same eye. The graying was diffuse in the left eye, was not hemianopic, and was not present in the right eye. Each episode, which was precipitated by standing, lasted for only a few seconds. Other symptoms included colored spots in her left visual fields, *déjà vu* phenomenon, and unusual smells.

As revealed by examination, visual acuity was 20/15 in each eye and color vision was normal. There was no proptosis or evidence of orbital congestion. Intraocular pressure was 14 mm Hg in each eye, and no afferent pupillary defect was detected. No bruit was detected, and Goldmann visual fields were full. The right optic nerve appeared normal, with possible fullness to the nerve fiber layer at the 6 and 12 o'clock positions. The left optic disc was markedly swollen without hemorrhages or exudates (*Fig. 3*). There were no visible spontaneous



FIGURE 2. Patient 1. *A*, coronal T1-weighted magnetic resonance image demonstrating a large left hemispheric AVM (*short arrow*) and optic chiasm (*long arrow*) compressed between the enlarged pituitary gland (*arrowhead*) and enlarged carotid artery. *B*, axial T1-weighted magnetic resonance image demonstrating the enlarged left superior ophthalmic vein (*arrow*).



FIGURE 3. Patient 2. Color fundus photograph of the left eye demonstrating disc edema at presentation.

venous pulsations in either eye. The rest of the neurological examination results were normal. Magnetic resonance imaging revealed a 3-cm right posterior parietal and occipital lobe AVM without hemorrhage, mass effect, or edema. The left superior ophthalmic vein was dilated. These findings were confirmed by cerebral angiography, and the AVM was noted to have a dual blood supply from both the middle and posterior cerebral arteries. The AVM drained initially into the superior sagittal sinus. As a result of congestion within the sinus, abnormal flow into the left orbit was noted in the venous phase of the angiogram (*Fig. 4*). This patient underwent embolization and complete surgical excision of the AVM. Postoperatively, her disc edema resolved completely in 3 months. She has a residual inferior left-sided homonymous visual field defect, and she occasionally experiences formed visual hallucinations in this area.

Patient 3

A 57-year-old right-handed woman presented with chronic daily headaches. Neuro-ophthalmic examination results were normal, including the absence of a bruit. Magnetic resonance imaging revealed a right parietal AVM. A cerebral angiogram confirmed the presence of the AVM and revealed orbital drainage (*Fig. 5*).

DISCUSSION

We identified three patients with orbital drainage from cerebral pial AVMs. The retrospective nature of this study and the referral patterns of our tertiary care facility have the potential to bias any conclusions regarding the prevalence of this type of drainage pattern. These three patients, however, were part of a series of 100 consecutive patients with pial AVMs. Their presentations were similar to those of other patients with pial AVMs (headaches and seizures), but two of the patients had unique neuro-ophthalmic manifestations of orbital drainage.

The most common neuro-ophthalmic symptoms and signs found in patients with hemispheric pial AVMs are headaches, visual field defects, and visual hallucinations. Involvement of the chiasm and the optic nerve with pial AVMs is rare (11, 14, 21). Two of our three patients with orbitally draining AVMs developed symptoms and signs that were attributable to the abnormal drainage pattern and its effect on the anterior visual pathways.

Patient 1 had dilated (nontortuous) conjunctival vessels and eyelid soft tissue swelling, and she developed a typical chiasmal syndrome with reduced visual acuity, bitemporal hemianopia, and optic atrophy. Visual loss resulted from compression of the chiasm between an enlarged carotid artery, dilated veins, and an enlarged pituitary gland. The venous drainage



FIGURE 4. Patient 2. *A*, lateral view of the arterial phase of an angiogram showing an occipital AVM. The supply is via the middle cerebral artery (*arrow*). *B*, lateral view of the venous phase of the angiogram demonstrating flow to the superior ophthalmic vein (*short arrow*), and missing the superior sagittal sinus anteriorly (*long arrow*).

into the orbit caused the dilation of conjunctival vessels and eyelid soft tissue swelling. Despite the markedly enlarged superior ophthalmic vein, it did not result in an elevation of intraocular pressure, arterialization of conjunctival vessels, conjunctival chemosis, or venous stasis retinopathy. This patient's visual loss was probably the result of optic tract and



FIGURE 5. Patient 3. Lateral view of the venous phase of an angiogram demonstrating flow to the orbital venous system (*black arrows*), and location of the AVM in the posterior portion of the splenium of the corpus callosum (*white arrow*).

chiasmal involvement by the AVM and compression of the chiasm between the vascular elements of the AVM and an enlarged pituitary gland. Sibony et al. (21) reported two patients and identified four others in the literature with chiasmal syndrome secondary to AVMs (3, 5, 6, 22). This series represents a heterogeneous group composed of patients with Wyburn-Mason syndrome, venous angiomas of chiasm, and AVMs intrinsic to the chiasm. Only one of these six patients was similar to ours, with chiasmal compression caused directly by vascular elements of a large pial AVM.

In a series of 202 patients with pial AVMs, Kupersmith (14) found only two with significantly reduced vision. One patient had bilateral optic atrophy and severely reduced vision as a result of venous drainage and damage to the optic chiasm and intracranial optic nerve. Although the degree of visual loss was different, our Patient 1 also presented with bilateral optic atrophy.

Patient 2 had transient visual obscurations and asymmetrical disc edema, which was worse on the side with orbital drainage. Elevated intracranial pressure (ICP) is commonly associated with dural AVMs because they frequently shunt arterial blood into the transverse or other major venous sinuses. The associated venous sinus thrombosis also causes elevated ICP (8, 10, 11, 13, 14, 17). Elevated ICP associated with unruptured pial AVMs is rare (1, 2, 11, 18, 19, 23, 24). One mechanism of raised ICP in this type of AVM is a massive runoff from the large shunting vessels, which increases blood volume and decreases cerebrospinal fluid absorption (1, 2, 23). Large pial AVMs per se, or as a result of an aneurysmal dilation of the venous system draining the AVM, may also exert a mass effect. Although patients with markedly asymmetrical and even unilateral papilledema have been reported to have elevated ICP (9, 12, 20), the unusual venous drainage pattern of this AVM may have contributed to this finding. Angiographically, there was evidence of missing superior sagittal sinus anteriorly (because of maldevelopment or acquired thrombosis), which, by compromising the normal venous outflow of the brain, probably worsened the effect of venous hypertension caused by the high-flow arteriovenous shunting. Postulated mechanisms of the asymmetrical disc swelling include direct compression of the optic nerve by a dilated and enlarged superior ophthalmic vein, or disc edema resulting from increased venous pressure. Finally, reduced pliability of the optic nerve sheath, caused by engorged pial vessels as a result of venous back pressure, may be an additional factor in causing asymmetrical papilledema on the side of orbital drainage.

Two patients with intracranial hypertension secondary to AVMs, the first with a mixed dural-pial AVM and the second with a pure pial AVM, were reported by Kashii et al. (11). The first patient had asymmetrical papilledema with markedly asymmetrical vision loss. Although a steal phenomenon from the ophthalmic artery to the AVM was postulated as a cause for the asymmetrical vision loss, no specific explanation was offered for asymmetrical disc swelling. The AVM drained through the straight sinus into the sigmoid sinus in their first patient, whereas, in the second patient, the AVM drained into the superior sagittal sinus. Orbital drainage of the AVMs was not described in either of these patients.

Patients with pial AVMs that are draining orbitally may present with a more heterogeneous constellation of ophthalmic signs and symptoms compared with the arteriovenous connections that are dura-based in the cavernous sinus. For example, despite flow from the AVM into the superior ophthalmic vein in all three of our patients, none had the classic findings of episcleral arteriolization, glaucoma, retinal venous congestion, and proptosis. However, two of our three patients manifested signs and symptoms related to the orbital drainage.

CONCLUSION

Three percent of patients with cerebral pial AVMs were found to have orbital drainage. Patient 1 exhibited orbital signs and symptoms related to the ophthalmic venous system drainage and congestion. Two of the three patients had visual symptoms caused by involvement of the anterior visual pathway; Patient 1 exhibited direct compression of the chiasm, and Patient 2 had asymmetrical papilledema. Our patients demonstrate that orbital drainage from cerebral pial AVMs may be associated with visual manifestations by heterogeneous mechanisms.

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COMMENTS

Volpe et al. point out that although orbital drainage from a cerebral arteriovenous malformation (AVM) is rare, it can produce neuro-ophthalmic problems. This is useful and interesting. However, the authors fully recognize that this is a

retrospective study of 100 patients with AVMs who presented to their institution. Only three of these patients were identified as having cerebral AVMs that drained the orbit, and only two experienced visual loss.

First, we must deal with the statistics of small numbers. It would be inappropriate to conclude that visual loss is a more common manifestation than ophthalmoplesia for cerebral AVMs that drain into the orbit. Furthermore, no conclusions as to incidence can be established, given the retrospective nature of this study. Issues such as the type of hospital and the types of referring physicians would significantly slant any interpretation. For example, had the authors' hospital been an eye and ear infirmary, most of the remaining patients might have experienced hearing loss or other symptoms associated with involvement of the petrous bone.

In summary, the authors add three more case reports of cerebral AVMs that produced visual symptoms. They also add the information that orbital drainage from cerebral AVMs may produce symptoms via a variety of mechanisms.

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Although dural AVMs and dural arteriovenous fistulae, especially those located anteriorly or in the cavernous sinus region, commonly demonstrate venous drainage into the orbit, pial AVMs rarely demonstrate orbital drainage. Volpe et al. present three patients with moderate to large pial AVMs that demonstrate this unusual pattern of retrograde orbital venous drainage. Two of the patients presented with ophthalmic symptoms and signs. It has been suggested that AVMs develop in the setting of maldeveloped cerebral venous drainage (1), and this might provide one explanation for retrograde orbital venous drainage. A more probable explanation is simply that elevated venous pressure in association with an AVM forces venous blood out through available venous channels, including the orbital veins via the cavernous sinus. Once this occurs, visual symptoms result from elevated intraorbital pressure, pressure on the visual pathway by engorged veins, and papilledema. The second patient presented here did not seem to have a definite venous pathway from the AVM to the orbital veins; elevated cerebral venous pressure or maldeveloped veins (such as absence of the anterosuperior sagittal sinus) might be the explanation for the abnormal drainage in this patient. This study is a reminder that pial AVMs occasionally drain into the orbit and cause symptoms and signs related to anterior visual pathway compression or orbital congestion. This report should prompt careful review of cerebral angiograms and thorough ophthalmological assessment of patients with cerebral AVMs.

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