



Orbital venous congestion: Rare manifestation of an intracranial arteriovenous malformation

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ABSTRACT

Intracranial arteriovenous malformations (AVM) are uncommon vascular lesions which typically present with hemorrhage or, in the case of unruptured lesions, with seizure, headache, or focal neurological deficit. The importance of the venous drainage pattern in AVM natural history and treatment outcomes is well documented. It is exceptionally rare for an AVM to drain into the orbital venous system. We present a 42-year-old man with a large, complex AVM of the midbrain, basal ganglia, and thalamus which partially drained into the superior ophthalmic vein. The patient was referred for treatment with radiosurgery. The resolution of orbital venous congestion symptoms accompanied AVM obliteration. An attempt to relieve orbital venous congestion by endovascular obliteration or surgical ligation without successful and complete cure of the AVM will most likely destabilize the nidus and predispose it to rupture. Serial ophthalmologic monitoring by an experienced neuro-ophthalmologist is crucial to monitoring the ophthalmologic effects of AVM progression and treatment.

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1. Introduction

Intracranial arteriovenous malformations (AVM) are rare lesions comprised of a pathological nidus typically supplied by intracranial arteries and drained by the superficial or deep intracranial venous systems. Traditionally, AVM most frequently presented with hemorrhage. However, as advanced neuroimaging modalities have become more widely available and commonly utilized, an increasing proportion of AVM are diagnosed prior to rupture. Unruptured AVM most commonly present with seizure, headache, or focal neurological deficit [1]. Visual symptoms are very uncommon, and symptomatic congestion of the orbital venous drainage system from intracranial AVM is exceedingly rare [2,3].

2. Case report

A 42-year-old man presented with a seizure. MRI and MR angiography demonstrated a large, complex AVM with two separate nidi; an anterior nidus centered in the right midbrain, basal ganglia and thalamus, and a posterior nidus located in the posterior right parasagittal frontoparietal cortex (Fig. 1). Digital subtraction angiography further characterized its angio-architecture including arterial supply from the anterior and posterior cerebral arteries, anterior and posterior choroidal arteries, and lenticulostriate and thalamoperforator branch arteries and venous outflow into the internal cerebral vein, vein of Galen, superior sagittal sinus, cavernous sinus, and superior ophthalmic vein (Fig. 2). The anterior nidus, measuring $4 \times 4 \times 3.1$ cm, was primarily supplied by the anterior circulation, and the posterior nidus, measuring $2.2 \times 1.8 \times 1.7$ cm, was primarily supplied by the posterior circulation. Combining the size of the two nidi, the patient's AVM was considered Spetzler–Martin grade V.

On auscultation, a bruit could be heard over the right eye. The patient's ophthalmologic exam was not significantly changed from his previous evaluation by the same neuro-ophthalmologist 3 years previously. The patient had a dense left homonymous hemianopsia and left afferent papillary defect as well as right sided proptosis and chemosis including episcleral venous engorgement.

The intraocular pressure was slightly elevated in the right eye at 23 mmHg and normal in the left eye at 13 mmHg. Visual acuity was normal. After medical management of the patient's presenting seizure, he was referred to our Gamma Knife (Elekta AB, Stockholm, Sweden) Center for treatment with radiosurgery.

3. Discussion

The vast majority of AVM exclusively drain into the superficial or deep intracranial venous systems. It is uncommon for an AVM to drain into an extracranial vein and extremely rare for extracranial venous drainage of an AVM to present with clinical sequelae. We present a patient with a rare, very large, multinodal Spetzler–Martin grade V AVM with partial drainage of the anterior nidus into the superior ophthalmic vein. Engorgement of the superior ophthalmic vein from aberrant arteriovenous shunting resulted in proptosis, chemosis, and slightly elevated intraocular pressure in the ipsilateral eye on ophthalmologic examination.

Volpe et al. presented a series of three patients with orbital drainage from intracranial AVM [2]. Two of the patients had abnormal ophthalmologic findings but only one was due to orbital venous congestion. Mohd-Tahir et al. presented a 31-year-old patient similar to ours with painful exophthalmos of the contralateral eye due to a Spetzler–Martin grade IV right temporal AVM [3]. The patient had complete resolution of his ocular symptoms 1 year after successful endovascular AVM obliteration.

While it is tempting to suppress the orbital venous drainage in order to reduce the symptoms affecting the eye, this can be quite treacherous in the case of an AVM. It is well known that premature AVM venous outflow occlusion, by any means, accelerates the degeneration of the nidus and predisposes it to rupture. Due to the high rates of morbidity and mortality associated with microsurgical excision of grade V AVM, we decided to refer the patient for treatment with radiosurgery. However, it should be noted that the reported rates of radiosurgical obliteration for high grade AVM are discouragingly low, including with both single session and staged approaches [4].

In conclusion, symptomatic orbital venous drainage from intracranial AVM is rare but should be recognized as a potential cause in

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Fig. 1. Magnetic resonance angiography of the brain with three-dimensional reconstruction in (A) sagittal, (B) coronal, and (C) axial views, showing a very large, complex right-sided arteriovenous malformation (AVM) comprised of two distinct nidus: the anterior nidus located in the midbrain, basal ganglia, and thalamus and the posterior nidus located in the parasagittal frontoparietal lobe. (D) Axial T2-weighted brain MRI sequence demonstrating drainage of the AVM into an engorged superior ophthalmic vein (arrow).

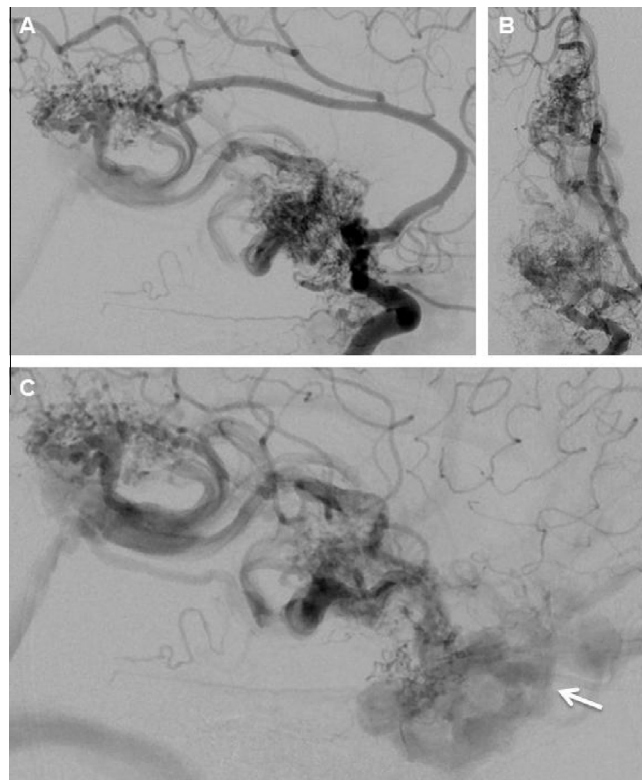


Fig. 2. (A) Lateral and (B) anteroposterior projections of digital subtraction angiography (DSA) with right internal carotid artery (ICA) injection demonstrating a complex arteriovenous malformation with two anatomically separate nidus. Of note, while not shown, the posterior nidus is mainly supplied by the posterior circulation. (C) DSA with right ICA injection, lateral projection of a later arterial phase than seen previously demonstrating early venous drainage of the anterior nidus into a pathologically dilated superior ophthalmic vein (arrow).

patients with proptosis, chemosis, or visual disturbances. The most effective method for relieving symptomatic orbital venous congestion is complete AVM obliteration. For patients with severe ophthalmologic symptoms who have AVM not amenable to standard therapies, palliative occlusion of the draining orbital veins may be considered. However, this must be done with extreme caution as premature AVM venous occlusion elevates the intranidal pressure thereby increasing the risk of AVM hemorrhage.

Conflicts of interest/disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

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Intraoperative aneurysmal subarachnoid hemorrhage after rupture of a previously undiagnosed intracranial aneurysm during Chiari decompression



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ABSTRACT

A type I Chiari malformation occurs when caudal displacement of the cerebellar tonsils below the level of the foramen magnum obstructs the normal flow of cerebrospinal fluid (CSF) between the cranial and spinal spaces, a condition that often needs surgical decompression to restore normal CSF circulation. Abrupt changes in CSF flow dynamics after Chiari decompression can affect the intracranial CSF dynamics to the extent that a previously undiagnosed intracranial aneurysm remote from the site can rupture. The authors describe the development of an intraoperative aneurysmal subarachnoid hemorrhage that occurred as a result of spontaneous rupture of a previously undiagnosed right distal posterior inferior cerebellar artery in a 57-year-old woman with type I Chiari malformation who was undergoing surgical decompression. The mechanism of the aneurysmal rupture appears to be related to the changes of CSF flow dynamics during surgical decompression. Normally, pressure equilibrium between the two sides of the aneurysmal wall prevents its rupture, but factors that significantly affect this equilibrium, such as systemic hypertension, can cause the aneurysm to rupture. To our knowledge, the concept of spontaneous intraoperative rupture of intracranial aneurysm remote from the site of surgery has been described twice previously but under different scenarios. This scenario, to our knowledge, has not been previously described.

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1. Introduction

A type I Chiari malformation occurs when pathological caudal displacement of the cerebellar tonsils below the foramen magnum obstructs cerebrospinal fluid (CSF) flow between the cranial and spinal subarachnoid spaces [1,2]. This acquired anomaly can be associated with spinal abnormalities such as syringohydromyelia, scoliosis, and hydrocephalus [1], and less commonly with cranial abnormalities. Physiologic studies have demonstrated abnormal CSF and blood flow dynamics at the craniovertebral junction in these patients [1,2]. We present a patient with intraoperative subarachnoid hemorrhage (SAH) secondary to rupture of an undiagnosed intracranial aneurysm during suboccipital craniectomy for Chiari decompression. To our knowledge this event has not been reported previously, but intraoperative rupture of an unnoticed

intracerebral aneurysm remote from a surgical site has been reported twice under different scenarios [3,4]. We consider possible reasons for this aneurysmal rupture by discussing CSF and cerebrovascular dynamics of patients with type I Chiari malformation.

2. Case report

A 57-year-old woman presented with a several-year history of positional tinnitus and suboccipital headaches that were precipitated by standing upright and alleviated with supine positioning. MRI of the brain revealed 8 mm of cerebellar tonsillar descent below the foramen magnum, consistent with type I Chiari malformation (Fig. 1).

The patient underwent suboccipital craniectomy, C1 laminectomy, and duraplasty for Chiari decompression. Before incision, she was given 50 g of mannitol for cerebral relaxation. During bone removal at the foramen magnum, her blood pressure elevated acutely coincident with temporary loss of somatosensory evoked

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