Cerebral Arteriovenous Malformation Causing Benign Intracranial Hypertension

—Case Report—

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Abstract

A 32-year-old male with a large arteriovenous malformation (AVM) in the right temporoparietal area presented with features of benign intracranial hypertension. This association is rare. The pathogenesis is believed to be due to cerebral venous hypertension. The excision of the AVM eliminated the intracranial hypertension.

Key words: arteriovenous malformation, brain swelling, papilledema, intracranial hypertension

Introduction

Cerebral arteriovenous malformations (AVM) present with hemorrhage in 50% of cases, seizures in 30%, headache, hemiparesis, and other focal neurological deficits in 20%.¹⁾ Headache as a sole presenting symptom of AVM is uncommon, comprising 6.36% of Paterson and McKissock's series⁹⁾ and 2.4% of Perrett and Nishioka's series.¹⁰⁾ Papilledema in association with an AVM has been described in a few cases only.^{4,8,9,13)}

However, a cerebral AVM causing benign intracranial hypertension (BIH) and presenting with headache, blurred vision, and papilledema without evidence of intracerebral hemorrhage, subarachnoid hemorrhage, or hydrocephalus is exceedingly rare. Only two cases have been reported in the literature.^{14,15)} We report a patient with a large right temporoparietal AVM who presented with headache and papilledema due to intracranial hypertension.

Case Report

A 32-year-old Sudanese male presented with a 1-week history of severe occipital headache and blur-

red vision but without nausea or vomiting. He had several grand mal seizures 6 weeks previously and had been placed on phenytoin 200 mg per day. On examination, he was alert, orientated and normotensive with gross bilateral papilledema. His visual acuity was 6/60 bilaterally and there was a loud bilateral cranial bruit. The remainder of the neurological and general examination was normal.

A postcontrast computed tomographic (CT) scan showed a right temporoparietal lesion with large vascular channels (Fig. 1). The ventricular size was normal and there was no apparent mass effect. Cerebral angiography demonstrated a large right temporoparietal AVM fed by two dilated branches of the middle cerebral artery and a dilated anterior choroidal artery (Fig. 2 *upper*). The venous drainage was *via* several dilated superficial veins and one deep vein into the sagittal and right transverse sinuses (Fig. 2 *lower*), and was noted to be retarded. Urgent surgery was indicated primarily to save his eyesight, quite apart from preventing hemorrhage in the future.

A right temporoparietal craniotomy was performed. The superficial arterial feeders were coagulated and sectioned. The anterior choroidal branch was

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Fig. 1 Postcontrast CT scan revealing a high-density area in the right temporoparietal region. No ventricular dilatation or apparent mass effect are demonstrated.

Fig. 2 Cerebral angiograms, lateral and anteroposterior views. *upper:* In the arterial phase, a large right temporoparietal AVM supplied by the middle cerebral artery and anterior choroidal artery is shown. *lower:* In the late arterial-venous phase, large venous channels entering the sagittal sinus and an engorged right transverse sinus (arrow) are demonstrated.

occluded when the temporal horn was entered and the most dilated draining vein was removed last. The AVM was completely excised.

Postoperatively he had a left parietal neglect and a left homonymous hemianopia. He was confused for 3 days. The headache and bruit disappeared and the papilledema was subsiding on discharge. When seen 1 year postoperatively his vision was normal and the sole neurological abnormality was a left homonymous hemianopia. He had had a single seizure 9 months postoperatively and this occurred when he forgot his anticonvulsant medication.

Discussion

Headache, papilledema, absence of focal neurological signs and a CT scan showing normal ventricles and no mass, falls into a syndrome known as BIH or pseudotumor cerebri. The etiology of BIH includes endocrine, metabolic, hematological, and connective tissue disorder, increased cerebrospinal fluid (CSF) protein, drugs, toxins, and partial venous sinus occlusion.¹¹ These could all be excluded in our case.

An AVM could cause BIH by several mechanisms. The mass effect of the AVM with its engorged vessels and a focal increase of cerebral blood volume could perhaps increase the intracranial pressure (ICP). Weisberg *et al.*¹⁵ suggested this as a cause in their case, using an isotopic brain scan as supporting evidence. Certainly, mass lesions such as frontal and temporal lobe tumors or chronic subdural hematomas which do not produce focal neurological signs can mimic BIH.¹¹ The chronic hypoxic hypercapnia of chronic pulmonary disease or paralytic hypoventilation, and chronic venous obstruction in the neck or chest have been described as causes of BIH. Vascular engorgement is believed to be the mechanism in these cases.^{5,11} Mathew *et al.*⁷ found a significantly raised cerebral blood volume in two BIH cases using isotopic techniques.

The arterial blood shunting into a major sinus, thus increasing the intraluminal pressure, could significantly impede venous return from the surrounding brain causing cerebral swelling due to venous engorgement and perhaps chronic interstitial cerebral edema. Dural AVMs associated with BIH have been reviewed.^{4,6,13} These lesions may produce raised venous pressure within a large sinus,⁶ resulting in hydrocephalus in some cases and BIH in others.³

Beck *et al.*² described a case of glomus jugulare tumor without intracranial extension associated with BIH, and postulated increased venous pressure in the sinuses and decreased CSF absorption as the cause. We have had one patient with a parasagittal meningioma, who presented with classical features of BIH due to blockage of the superior sagittal sinus

and increased venous pressure. Reduced CSF absorption, due to increased venous sinus pressure or to engorged brain mechanically blocking the arachnoid villi, could cause BIH. Recurrent subarachnoid hemorrhage from an AVM could also impede CSF absorption but in our patient there was no evidence of bleeding preoperatively. Isotopic cisternography demonstrated impaired CSF absorption in the case of Weisberg *et al.*,¹⁵⁾ which also supports this mechanism. Increased CSF production due to venous hypertension affecting the choroid plexus has been postulated as another mechanism of BIH developing in AVM patients.¹³⁾ It is interesting to note the case of Spallone in which a 9-year-old boy with BIH had a CT scan highly suggestive of an AVM but angiography was negative.¹²⁾ The high contrast dose may have produced this appearance.¹²⁾ BIH results from a combination of three abnormalities in varying proportions: increased cerebral blood volume, delayed CSF absorption, and brain edema.⁷⁾

In our case, massive blood shunting into the sagittal and transverse sinuses, causing venous hypertension, increased cerebral blood volume, and reduced CSF absorption, is the postulated pathogenesis of the BIH. The excision of the AVM eliminated the intracranial hypertension.

Finally, this case emphasizes the importance of the venous component of the circulation through AVMs. Too little regard for this has, perhaps, been paid in the literature. Any large fistula is bound to increase intraluminal pressure in the veins and sinuses draining the AVM. Adaptation to this no doubt usually occurs in the majority of patients. In a few, progressive enlargement of the fistula results in an increase in flow which causes an elevation of venous pressure before adequate adaptation of venous return from other areas of the brain can occur, with production of the BIH syndrome: hence its rarity.

Excision of the fistula must equally have profound effects on the venous return postoperatively. No longer is the preoperative adaptation of the venous return from normal areas of the brain necessary. Thus, it is conceivable that stasis and possibly thrombosis may occur in the venous system. This might well be an important factor in producing the postoperative swelling that can complicate surgical excision of large AVMs. We have had one patient in which this swelling occurred in the contralateral hemisphere rather than in the region of the AVM. This postoperative swelling and hemorrhage has been called "normal perfusion pressure breakthrough," and ascribed to impaired vasoreactivity of the adjacent brain. We suggest the venous circu-

lation deserves more attention in this condition.

References

- 1) Adams RD, Victor M: *Principles of Neurology*, ed 3. New York, McGraw-Hill, 1985, pp 622-624
- 2) Beck DW, Kassell NF, Drake CG: Glomus jugulare tumor presenting with increased intracranial pressure. Case report. *J Neurosurg* 50: 823-825, 1979
- 3) Esparza J, Lobato RD, Munoz MJ, Chillon D, Portillo JM, Lamas E: Giant cerebral arteriovenous malformation producing a non-communicating hydrocephalus. *Surg Neurol* 15: 76-80, 1981
- 4) Gelwan MJ, Choi IS, Berenstein A, Pile-Spellman JMD, Kupersmith MJ: Dural arteriovenous malformations and papilledema. *Neurosurgery* 22: 1079-1084, 1988
- 5) Greer M: Ch 101 pseudotumor cerebri, in Youmans JR (ed): *Neurological Surgery*, vol 5, ed 2. Philadelphia, WB Saunders, 1982, pp 3179-3195
- 6) Lamas E, Lobato RD, Esparza J, Escudero L: Dural posterior fossa AVM producing raised sagittal sinus pressure. Case report. *J Neurosurg* 46: 804-810, 1977
- 7) Mathew NT, Meyer JS, Ott EO: Increased cerebral blood volume in benign intracranial hypertension. *Neurology (Minneapolis)* 25: 646-649, 1975
- 8) Mingrino S, Scanarini M, d'Avella D: Treatment of benign intracranial hypertension. *Acta Neurochir (Wien)* 51: 187-193, 1980
- 9) Paterson JH, McKissock W: A clinical survey of intracranial angiomas with special reference to their mode of progression and surgical treatment. *Brain* 79: 233-266, 1956
- 10) Perrett GE, Nishioka H: Arteriovenous malformations. An analysis of 545 cases of cranio-cerebral arteriovenous malformations and fistulae reported to the co-operative study. *J Neurosurg* 25: 467-490, 1966
- 11) Rowland LP: *Merritt's Textbook of Neurology*, ed 7. Philadelphia, Lea & Febiger, 1984, pp 210-215
- 12) Spallone A: Benign intracranial hypertension vs intracranial arteriovenous malformation: A possible CT dilemma. *Acta Neurochir (Wien)* 58: 75-84, 1981
- 13) Van-den-Bergh R, Dralands G, Crolla D, Van-den-Bergh P: Pseudotumor cerebri due to intracranial arteriovenous malformation. *Clin Neurol Neurosurg* 82: 119-125, 1980
- 14) Vassilouthis J: Cerebral arteriovenous malformation with intracranial hypertension. *Surg Neurol* 11: 402-404, 1979
- 15) Weisberg LA, Pierce JF, Jabbari B: Intracranial hypertension resulting from a cerebrovascular malformation. *South Med J* 70: 624-626, 1977

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