RESPONSE: I thank Drs. Wilson and Hieshima for their observations on the case report by myself and Dr. Joseph. Although I recognize the senior correspondent’s eminence and experience, I fear he does us an injustice. We could hardly not have considered the diagnosis of dural arteriovenous malformation: a condition well known to all neurosurgeons. We referred to previously published relevant work in our report. We made no claim that our case was unique, and we referred to a previous report of an anterior ethmoidal artery aneurysm.

The radiological findings (absence of tortuous dilated arterial feeders, nidus of a vascular malformation, or draining vein), the operative findings (aneurysm arising from the aforementioned artery, absence of a nidus, vein, or evidence of thrombosis), and the histological confirmation of the nature of the aneurysm were all stressed in our report. We feel that the “label” of giant aneurysm is entirely appropriate in our case.

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Tentorial AVMs

TO THE EDITOR: I read with interest the recent good article by Lewis, et al. (Lewis AI, Tomsick TA, Tew JM Jr: Management of tentorial dural arteriovenous malformations: transarterial embolization combined with stereotactic radiation or surgery. J Neurosurg 81:851–859, December, 1994). I agree with the authors that these lesions are more dangerous than the more common cavernous and transverse–sigmoid dural arteriovenous malformations (AVMs). The reason for their more aggressive natural history is that, as discussed by the authors, they seem to have a peculiar tendency to drain into the pial venous system as opposed to draining into dural sinuses or veins. This pial venous drainage system results in a propensity for hemorrhage because the pial veins are not protected by a dural envelope. In addition, the pial drainage can result, as discussed by the authors, in protean symptomaticity related to venous hypertension, particularly in the posterior fossa and hydrocephalus and/or focal deficits from direct compression of the aqueduct, the brainstem, and cerebellum by venous varices. The authors feel strongly that the treatment of these lesions should be primarily by transarterial embolization, frequently in several stages, followed by either surgical obliteration of the arterial supply or radiosurgery.

The purpose of this letter is to remind the readers of an alternative form of treatment for this particular type of dural AVM. Mullan1 has been discussing for many years the concept that occlusion of the venous side of AVMs can result in complete thrombosis of the lesion; he published some of his reflections on this concept in a recent article in the Journal. The problem with occluding the venous side in parenchymal AVMs is, of course, that arterial hypertension develops acutely in the nidus of the lesion, which can lead to catastrophic swelling and hemorrhage. This is unlikely to occur when the nidus of the AVM (or the fistula in the case of the simpler shunts) is located within the leaves of the dura as it is, by definition, on dural AVMs. Dr. Mullan and others have pointed out that tentorial AVMs offer a uniquely advantageous circumstance for treating the lesion by simple venous occlusion because these AVMs usually drain only through a single vein leading from the fistula; this draining vein is most commonly the petrosal vein. This has been the case in the five patients that I have operated on with tentorial AVMs. In each there was a short, stubby petrosal vein draining the fistula that was located close to the tentorial edge. This vein rapidly communicated with many other veins and the entire posterior fossa venous system was arterialized. After occluding the vein with a clip at the junction of the vein and the tentorium, the previously arterialized pial venous network distal to the clip quickly became blue and soft. In two cases, at the suggestion of Dr. Daniel Rufenacht, I injected glue into the short segment of the vein between the clip and the tentorium to occlude the fistula by retrograde injection of the glue; however, I am not sure that this maneuver is necessary. In all five of my cases, the symptomatology improved markedly and rapidly, and none has bled with a follow-up of between 6 months and 6 years. Each patient had a postoperative angiogram that showed no evidence of residual AVM. This experience has been similar to that at the Mayo Clinic (D. Rufenacht, personal communication, 1994) and at the University of Chicago (S. Mullan, personal communication, 1994). Grisoli, et al.,2 documented four cases treated in an identical manner with good results in 1984.

It is hard to argue with the contention of Lewis and coworkers that in general AVMs should be treated by first reducing the arterial supply to the nidus either surgically or by embolization before obliterating the nidus and the venous drainage to avoid swelling and hemorrhage from the nidus. However, with dural fistulas, as discussed earlier, the nidus is within the leaves of the dura and thus protected from swelling or hemorrhage. It is very difficult, as admitted by the authors, to directly catheterize the fine tentorial feeders to these fistulae. In addition, such embolization carries some danger of embolizing normal capillary beds and frequently requires multiple sessions that add to the morbidity of the procedure. The authors have used radiosurgery to treat seven of the eight lesions. I suspect that the reason for radiosurgery was that transarterial embolization was not successful in eliminating the fistula in these patients. The problem with radiosurgery is that its effect takes time, and these lesions have a relatively morbid natural history. Whether reduction of flow by embolization will reduce the risk of hemorrhage is unknown, as the authors admit. Only three of the seven lesions treated by radiosurgery were eventually demonstrated to be completely obliterated. Direct surgical obliteration of the fistula is difficult because the exact site of the fistula in the tentorium is not always apparent. Surgical obliteration of the arterial feeders in the tentorium can be complicated because these fine vessels usually come from the cavernous portion of the carotid artery (most commonly from the meningohipophyseal trunk), but they can also come from a variety of other dural arteries, as discussed by the authors.

Because of these considerations I currently treat tentorial AVMs by direct surgical occlusion of the venous drainage. In the case of tentorial fistulas draining into the petrosal vein, the relatively simple subtentorial–transientorial approach gives adequate exposure to occlude the vein as it exits the fistula under the tentorium and to coag-
ulate some of the obviously dilated tentorial arterial feeding vessels to the fistula: a maneuver that makes the surgeon feel better but may not be necessary. With this approach, the tentorium is divided behind the fistula, which is usually in relation to the superior petrosal sinus and, incidentally, is almost always thrombosed in these patients. In my opinion, it is not advisable to do the more complicated subtemporal–presigmoid transpetrosal approach to divide the tentorium in front of the fistula; I did this in one of my cases and the approach was very bloody and cumbersome because of the hypervascularized tentorium anterior to the fistula. At surgery it was clear that the same result could have been obtained by the subtemporal–transtentorial approach or even through a simple retrosigmoid craniectomy. I do not agree with the authors that occluding the vein as it exits the tentorium has any likelihood of resulting in venous hypertension or hemorrhagic venous infarction. On the contrary, the venous pressure throughout the posterior fossa is immediately decreased by disconnecting the pial venous system from the fistula.

In brief, I agree with Mullan et al and Grisoli, et al, that most tentorial AVMs can be treated successfully by simple occlusion of the venous drainage of the AVM as it exits the tentorium. In fact, my current opinion is that this is the treatment of choice for this peculiar type of dural AVM.

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References

RESPONSE: Dr. Heros believes that most tentorial dural arteriovenous malformations (AVMs) are best treated with interruption of the venous drainage. His opinion is based on his surgical experience with five of these lesions and the following anatomical considerations. First, unlike parenchymal AVMs that have a nidus, the tentorial dural AVMs have an arterial supply that converges to a simple fistula between an artery and a vein, most commonly the petrosal vein. Second, the shunt is located within the leaves of the tentorium, so it is not subjected to catastrophic swelling and hemorrhage from arterial hypertension caused by ligating the vein. Third, the risk of transarterial embolization of the meningohypophyseal trunk is occlusion of normal capillary beds. Fourth, after radiosurgery, there is a risk of rebleeding while waiting for the dural AVM to thrombose. Fifth, direct surgical obliteration of the fistula is not easy because the arterial supply may originate from several vascular territories, the tentorium is hypervascularized, and the fistula is difficult to locate within the leaves of the tentorium. Dr. Heros cites the corroborating experiences of Drs. Mullan (personal communication), Rufenacht (personal communication), and Grisoli, et al; he advocates ligating the draining vein for most tentorial dural AVMs without prior embolization, surgical obliteration, or stereotactic radiation.

Dr. Heros makes a compelling argument for venous ligation of tentorial fistulas; however, this approach is dangerous for tentorial dural AVMs. Likewise, it is important to distinguish between dural malformations that drain directly into a dural sinus and those with leptomeningeal venous drainage, because the latter are not protected by a dural envelope. In our series, tentorial dural AVMs resembled parenchymal AVMs rather than a simple fistula as Dr. Heros describes. These dural AVMs had a complex arterial supply from several vascular territories with multiple draining veins, and in two cases there were two giant venous aneurysms. To occlude one vein, as Dr. Heros advocates, would have diverted flow into nonarterialized veins and caused a hemorrhage as occurred in Case 1. Moreover, it is difficult to obliterate a tentorial dural AVM by vein occlusion when the malformation is drained by both petrosal veins (Case 2).

Dr. Heros argues that swelling and hemorrhage from the nidus are not encountered after ligating the vein because the nidus is located within the dural leaves. However, there is extensive collateral flow within the dural mater and retrograde flow into cortical veins (Case 9). Dilated leptomeningeal veins and venous aneurysms (Case 5) are frequently the source of hemorrhage as shown in Fig. 4 of our paper. Therefore, if a venous approach is attempted, it is important to ligate the arterialized vein close to the malformation or where the vein enters the subarachnoid space. Dr. Heros correctly states the exact site of the fistula in the tentorium is not always apparent because there is a variety of fine dural arteries and the tentorium is hypervascularized.

Dr. Heros argues that transarterial embolization risks occluding normal capillary beds. Prior to the actual embolization of feeding arteries, amytal or lidocaine was injected to ensure that normal capillary beds were not embolized inadvertently. The feeding arteries were injected with N-butyl cyanoacrylate or polyvinyl alcohol sponge. We preferred the adhesive glue over the polyvinyl alcohol sponge because it was more permanent, more easily controlled, and produced better results. Importantly, no patient suffered a neurological deficit or rebled after embolization.

Dr. Heros believes that radiosurgery is not the optimum treatment because of the risk of rebleeding during the latent interval for dural AVM thrombosis. We believe tentorial dural AVMs are well suited for stereotactic radiation because they are deep seated, usually have tiny feeding vessels, and have a small nidus. Since we submitted the manuscript in September 1993, two more patients (Cases 7 and 9) have had complete obliteration of their tentorial dural AVM. Overall, five (71%) of seven patients have had complete obliteration of the dural AVM and the other two patients have had better than 95% obliteration of the dural AVM. There was one case of transient radiation injury (Case 2), and no AVM rebled after radiosurgery.

We agree with Dr. Heros and others that some tentorial dural fistulas can be treated by a venous approach in carefully selected patients. However, the venous approach should not be used for tentorial dural AVMs. For successful venous occlusion, the dural fistula is usually located at the petrous apex and drains into the petrosal vein. In addi-
tion, the petrosal vein is ligated close to the nidus to avoid diverting flow into collateral veins, which can rupture from high shunt pressure or exacerbate venous hypertension. In our series, there was only one tentorial fistula (Case 5) that fit this criteria, and it was easily obliterated with surgical ligation of the fistula.

Since we submitted our manuscript, we have successfully treated two additional tentorial and one inferior petrosal dural AVMs. The combination of transarterial embolization with either stereotactic radiation or microsurgery has proven safe and effective for the treatment of deep-seated complex dural AVMs.

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References

Gigantism and Somatostatin Neurons

To the Editor: I read with interest the article by Manski and coworkers (Manski TJ, Haworth CS, Duval-Arnould BJ, et al: Optic pathway glioma infiltrating into somatostatinergic pathways in a young boy with gigantism. Case report. J Neurosurg 81:595–600, October, 1994) which posits a causal relationship between gigantism in a 16-month-old boy and a diffuse glioma interspersing with somatostatin (SS) neurons somewhere within the medial temporal lobe (MTL). Although the hypothesis is thought provoking, several questions arise therefrom: 1) Were the MTL SS neurons read as atrophied, hypertrophied, or decreased in number? 2) If diminished, would not the SS neurons in the periventricular anterior hypothalamic with which the corticomedial amygdala communicates be disinhibited and produce a decrease in growth hormone–releasing hormone (GRH) from the arcuate and ventromedial nuclei with which the anterior hypothalamus connects? 3) Would not concomitant infiltration of excitatory amygdaloventromedial pathways produce a decrease in GRH also? 4) Was there evidence of a pituitary adenoma tending to rule out tumor-producing ectopic growth hormone (GH)?

Hypothalamic hamartomas have been reported to cause ectopic overproduction of GRH, and it seems plausible that optic nerve glioma infiltrating the anterior hypothalamus would do likewise; if such effect were secondary to pressure atrophy of local SS neurons, then a glucose challenge would paradoxically increase serum GH (provocative testing was not done in this case), and an SS analog challenge should suppress GH and/or somatomedin C.

Another consideration in this case would seem to be Sotos syndrome, a mostly sporadic congenital syndrome characterized by high birth length and weight, normal GH (as in this case), coarse facial features, hypertelorism, anti-Mongolian epicanthal folds, mild mental retardation, incoordination, ventriculomegaly, increased association with carcinoma (glioma has not been reported), but bone age concomitant with height (not in this case); many of these features still remain in this child despite the endocrine and tumor response to chemotherapy; the specific pathology of this syndrome remains unknown.

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Response: We thank Dr. Dalrymple for her interest in our recent report and hope to clarify her questions. The somatostatinergic neurons from the medial temporal lobe were thought to be slightly atrophied and decreased in number. Stimulation of the corticomedial amygdala has an inhibitory influence on growth hormone (GH) release.5 The corticomedial amygdala sends efferents via the stria terminalis both to somatostatinergic neurons (in the septum and preoptic area) and to GH-releasing hormone (GRH) neurons (in the ventromedial hypothalamic nucleus).8 The corticomedial inhibitory response results from inhibition of GRH from the ventromedial nucleus (VMN);8 the role of the connections to the septum and preoptic area has not yet been determined,8 but somatostatin has been shown to have both excitatory and inhibitory effects in the central nervous system.10 Hence, disruption of this inhibitory pathway by tumor would be expected to result in an increase in GH release. Probably most important in our patient was direct tumor disruption of the periventricular anterior hypothalamic–preoptic somatostatinergic system.7,9

It is the basolateral amygdala that has an excitatory influence on GH release, probably via the ventral amygdalohypothalamic tract that projects to the VMN.8 Tumor disruption of this GH-excitatory pathway would be expected to result in some decrease in GRH and GH release. However, in our patient this was probably more than offset by the expected increases in GH secondary to tumor disruption of the anterior hypothalamic–preoptic somatostatinergic system. Magnetic resonance imaging of the pituitary in our patient showed no evidence of a pituitary adenoma.

Hypothalamic neuronal hamartomas (hypothalamic gangliocytomas) produce GRH and cause overproduction of GH by stimulating GH cells in the anterior pituitary.1,2 The optic pathway glioma in our patient was immunohistochemically negative for GRH and GH. Its infiltration into somatostatinergic pathways in the periventricular anterior hypothalamic–preoptic region and corticomedial amygdalae was thought to disrupt the normal inhibitory influence of somatostatin from these areas, resulting in overproduction of GH.

Although cerebral gigantism (Sotos syndrome) was considered in our patient, he did not fulfill the strict diagnostic criteria to make this diagnosis.3,4,10,12

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