Cerebral Arteriovenous Malformations Causing Cerebrospinal Fluid Fistula

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Cerebral arteriovenous malformations (AVM) are congenital lesions in which arteries are connected to veins through an interposed nidus.[1] This aberrant vascular conglomerate acts as a high-flow and low pressure shunt between the feeding arteries and the draining veins thus creating an overload of the venous outflow from the lesion.[2]

Patients with AVMs become symptomatic through various manifestations, the most frequent being hemorrhage and seizures. Elevated intracranial pressure and papilledema are unusual modes of presentation which may be consequent to the venous outflow overload caused by the AVM.[2] A cerebrospinal fluid (CSF) fistula secondary to this phenomenon, however, has not been previously reported. The case presented herein concerns such an association.

Case Report

A 23-year-old male with a history of cystic fibrosis and hepatic transplantation for which he had received immunosuppressive therapy for the last six years, notably cyclosporine, presented with *de novo* blurred vision and headache. For many years this patient had complained of a left-sided pulsatile tinnitus. The patient's general physical examination was normal and he had a body mass index of 20.7. A thorough neurological examination was normal except for bilateral papilledema without optic nerve dysfunction. Computed axial tomography scanning (CT-scan) and magnetic resonance imaging suggested an AVM (4 cm by 3 cm) without any significant mass effect or hydrocephalus (Figure 1).

Four-vessel angiography confirmed it to be a right posterior temporal AVM supplied by the middle cerebral artery and to a lesser degree by the posterior cerebral artery. The nidus drained into the superficial temporal vein towards the junction between the ipsilateral transverse and sigmoid sinuses (Figure 2). This represents a grade 1 AVM on the Spetzler-Martin grading scale.

Presumptive diagnosis was pseudotumor cerebri (PTC). Two causes were evoked: the AVM with its associated increased intracranial venous pressure, and the cyclosporine, which has been associated with PTC.[3,4] Cystic fibrosis has also been associated with PTC as a result of hypervitaminosis or refeeding syndrome during correction of malnutrition, which was not the case in our patient.[5,6]

Because of the patient's comorbidities and the possibility that cyclosporine therapy may account for the PTC, the AVM was managed conservatively even though it was not excluded as causing the increased intracranial pressure. Owing to a series of lumbar punctures, with the opening pressure varying from 32 cm H_2O to 46 cm H_2O, the patient's condition evolved favourably and shunting was not deemed necessary.

Three years later and four months following the last lumbar puncture, the patient reported intermittent clear rhinorrhea and right auricular fullness. A right myringostomy was performed by an otolaryngologist and yielded a profuse effusion of CSF. Computed axial tomography scanning and MRI were suggestive of 1 mm right tegmen mastoidi erosion adjacent to the AVM. This was not present on the initial CT-scan. A repeated angiography revealed stability of the vascular malformation.

Surgical intervention was indicated to occlude the CSF fistula. Given that the AVM was possibly responsible for the PTC and the secondary CSF fistula, and because it was also adjacent to the bony defect, its removal, during the same surgical procedure, was indicated. After its partial endovascular embolization with Histo-Acryl through the three right sylvian pedicles and Onyx through the pedicle leading to the AVM, it was surgically removed without incident. During the same procedure, the tegmental dehiscence was repaired using a temporal fascia and fat graft.

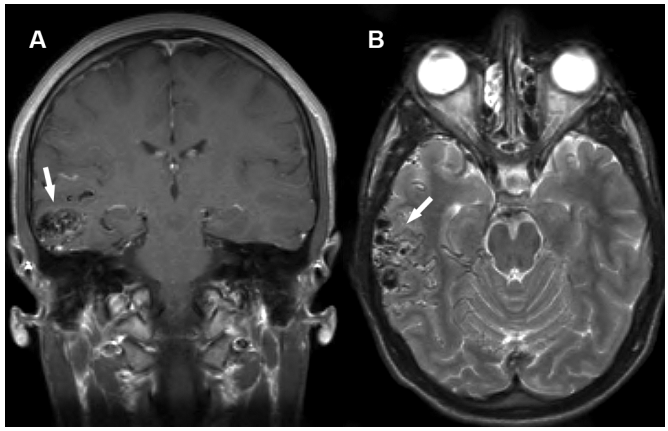

Figure 1: *Magnetic resonance imaging (coronal [left] and axial [right] views) showing the arteriovenous malformation (white arrow).*

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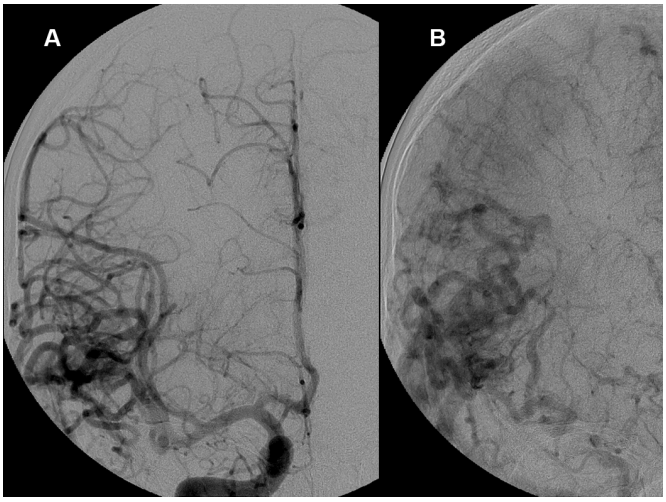

Figure 2: *Angiogram (axial view; arterial phase [left] and venous phase [right]) showing the arteriovenous malformation.*

Following the surgery, the patient was neurologically intact. However, the CSF otorrhea persisted. A mastoidectomy and exploration of the middle ear and the tegmen was undertaken and the fistulous opening found in the supratubal recess was sealed using a fat graft and temporalis fascia. Later review of the preoperative CT-scan demonstrated a dehiscent pneumatised right petrous apex cell and a fistulous tract leading to the middle ear through the supratubal recess just superior to the Eustachian tube meatus. This fistula corresponded to the intraoperative site of CSF leak (Figure 3). Complete excision of the AVM was confirmed by post-operative angiography. Cerebrospinal fluid otorhinorrhea did not recur and the patient remained asymptomatic after a 16 months follow-up, with no evidence of PTC and complete resolution of papilledema.

Discussion

Arteriovenous malformations commonly present with hemorrhage and seizures and more rarely with progressive neurological deficits including dementia.[7,8] To our knowledge, there has not been a report of an AVM causing a CSF fistula. The AVM in our patient produced symptomatic PTC and eventually presented with CSF fistula causing otorhinnorhea.

A review of the English and French literature revealed 23 cases of patients with an AVM symptomatic of raised intracranial pressure without evidence of hydrocephalus or hemorrhage (see Table). These patients had all been diagnosed with PTC. Only two of them showed evidence of venous sinus thrombosis.[19] None of these patients presented with CSF otorhinorrhea. Arteriovenous malformations rarely cause intracranial hypertension without hydrocephalus or hemorrhage, and occurrence of a CSF fistula is even rarer. Conservative treatment without surgery or embollization was noted in only three patients: Chinowitz et al patients 3 and 5[19] and Johnston et al.[20]

Additional review of the literature concerning the association between PTC and CSF otorhinorrhea revealed numerous accounts associating these two conditions, yet there was no instance where an intracranial AVM was also present. However, Willems et al reported a case where a dural arteriovenous fistula presented with a CSF leak.[24]

The physiopathological sequence between our patient's AVM and raised intracranial pressure is proposed to be as follows: Presence of the AVM caused a high volume of blood to be shunted from the arterial feeding branches to the venous draining system. This effectively increased venous blood pressure and cerebral blood volume.[19-21] This caused a decrease in CSF re-absorption due to a diminished pressure gradient between the intracranial pressure and the venous sinuses through the arachnoid granulations.[15,18,20,24,25] In our case, the intravenous increase in pressure was not due to stenosis or occlusion of a major venous sinus, as it is not infrequently seen in association with AVMs. Intracranial pressure increased to match increase in cerebral sinus venous pressure and re-establish the normal gradient.[20,25]

Two theories are most prevalent in explaining the etiology of these spontaneous, non-traumatic, CSF fistulas. The first involves the presence of a congenital defect which can give rise to a CSF leak.[26,27] The other theory contends that arachnoid granulations aberrantly located outside of a dural venous sinus may come into contact with the skull. With the additive effect of the pulsations through these arachnoid granulations over time, erosion of vulnerable bony structures may take place.[28]

In order for CSF leak to occur, dura mater breech is necessary. Increased intracranial pressure and inflammation are both suggested mechanisms.[26,27] Repeated lumbar punctures may have contributed to this inflammatory process. Intracranial hypertension over many years can lead to dural thinning and penetration.[27,29,30]

We hypothesise that, in our case, the combination of the AVM-related raised intracranial pressure and the AVM transmitting pulsations to draining veins of the temporal lobe over the dura and a critical bony structure, are responsible for dural and bone erosion and fistulisation. However, our patient's CT scan revealed a pneumatised right petrous apex and a large cell containing fluid, which in conjunction with the increased intracranial pressure could be sufficient to explain the CSF leak (Figure 3). Aeration of the petrous apex is a normal variant, Yetiser et al having found 12 cases upon review of 430 temporal bone CT-scans taken for diagnostic imaging.[31] From this cell, a fistulous trajectory can be defined due to the presence of liquid along a very narrow tract which abuts into the Eustachian tube and to the middle ear through the supratubal recess (Figure 3).

Intraoperative and radiological observations did not reveal the presence of either a pre-existing congenital defect or of an aberrant arachnoid granulation protruding into the tegmen mastoidi.

We contend that the long standing venous outflow overload, the increased intracranial pressure and the anatomical location of the AVM and its draining veins contributed to the formation of the CSF fistula described above. This is supported by the complete resolution of raised intracranial pressure symptoms and of CSF otorhinorrhea following surgical excision of the AVM and occlusion of the CSF fistula. For this reason, we do not believe that the AVM was an incidental finding. After a follow-up of one year and four months, the patient remained symptom free.

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Table: Review of literature concerning the association between intracranial AVMs and PTC

Series (year)[ref. no]	Age (yr) / Sex	Location of AVM	Arterial Supply Venous Drainage
Paterson and McKissock (1956)[9]	Unknown	Unspecified	Unspecified
Kosary et al. (1973)[10]	25/M	R parietal	-R ACA via the L ACA through the Acom -SSS
Lamas et al. (1977)[11]	42/F	R posterior fossa	-R occipital artery and R posterior branch of middle meningeal artery -Transverse sinus
Weisberg et al. (1977)[12]	40/M	R parieto-occipital with R lateral ventricle compression	-R PCA, R MCA and R ACA - Unspecified
Vassilouthis (1979)[13]	11/M	R posterior frontal	-R pericallosal artery (branch of ACA) -SSS and ISS
D'Avella et al. (1980)[14]	55/M	L occipital	-L occipital artery, L posterior branch of superficial temporal artery, L tentorial vessels from L ICA
Van den Bergh et al. (1980)[15]	65/M	L posterior fossa, along transverse sinus	-Occipital, anterior auricular, posterior auricular, middle meningeal, ascending cervical and vertebral (via PICA and segmental branch C2) arteries -Transverse sinus, sinus rectus and internal jugular vein
Schiffer et al. (1984)[16]	31/F	R mid-rolandic	-R ACA and R MCA -Internal cerebral vein and SSS
Barnett et al. (1987)[17]	21/F	R fronto-parietal	Unspecified
Barrow (1988)[18]	21/F	R frontal parasagittal	-R ACA, R pericallosal and R callosomarginal -Large superficial vein to SSS
Barrow (1988)[18]	33/F	L medial parietal lobe	-L MCA (angular branch), ACA and PCA (parietal, occipital and calcarine branches) -SSS and vein of Galen
Chimowitz et al. (1990)[19]	19/F	R parieto occipital	-R MCA, R ACA and R PCA -SSS
Chimowitz et al. (1990)[19]	21/F	R parietal	-R MCA -SSS
Chimowitz et al. (1990)[19]	32/F	L frontal	-L MCA and L ACA -SSS
Chimowitz et al. (1990)[19]	28/M	R parietal	-R MCA and R ACA -SSS (stenosis of R transverse sinus)
Chimowitz et al. (1990)[19]	44/F	R parieto-occipital, mass effect displacing R lateral ventricle	-R MCA and R PCA -SSS
Chimowitz et al. (1990)[19]	24/M	R occipital-parietal	-R MCA, R PCA and R ACA -SSS (stenosis of R transverse sinus at junction with sigmoid sinus)
Johnston et al. (1991)[20]	32/M	L post frontal	-Unspecified -SSS
Rosenfeld et al. (1991)[21]	32/M	R temporo-parietal	-R MCA (2 distal branches) and R anterior choroidal artery -SSS and R transverse sinus
Cockerell et al. (1993)[22]	35/F	L frontoparietal	-Cortical branches of L ACA and L MCA -SSS
Cockerell et al. (1993)[22]	65/M	R occipital	-R occipital artery -unspecified dural sinus
David et al. (1995)[2]	29/M	L posterior temporoparietal	- L MCA branches -Superficial and deep drainage (unspecified) (venous outflow obstruction in both transverse sinuses and a cortical vein)
Adelman et al. (1998)[23]	21/M	L posterofrontal-anteroparietal region	-Bilateral ECA branches (STA and occipital and middle meningeal Arteries), anterior falcine artery from the ophthalmic, tentorial branches from the ICA, posterior meningeal from the vertebral artery, and posterior falcine branches from the PCA. -SSS

*ACA = anterior cerebral artery; Acom = anterior communicating artery; AVM = arteriovenous malformation; ECA = external carotid artery; F = female; ICA = internal carotid artery; ISS = inferior sagital sinus; M = male; MCA = middle cerebral artery; PICA = posterior inferior cerebellar artery; PCA = posterior cerebral artery; PTC = pseudotumor cerebri; R = right; SSS =superior sagital sinus; STA = superior temporal artery L = left

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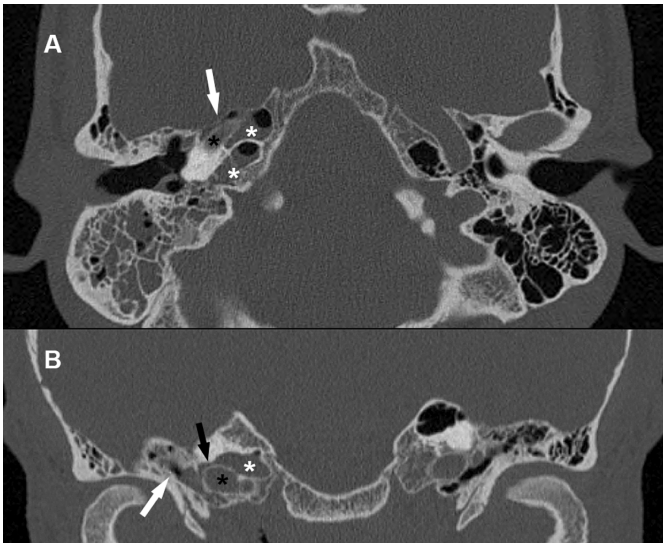

Figure 3: *Computed tomography (axial [upper] and coronal [lower] views) demonstrating the pneumatized petrous apex cells filled with cerebrospinal fluid (white stars) communicating to the Eustachian tube (white arrows) through a small bony dehiscence (black arrow). Black stars in the upper and lower panels show the internal carotid artery.*

We report for the first time an AVM presenting with a CSF fistula related to secondary increased intracranial pressure. The importance of these findings resides in the necessity to surgically excise the AVM in order to address the CSF fistula. Failure to do so may perpetuate pseudotumor cerebri symptoms and prevent effective treatment of the CSF fistula.

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