

# Sudden deafness as a manifestation of the rupture of a cerebral arteriovenous malformation

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**S**udden deafness is defined as sudden or rapidly progressing neurosensory hypoacusis of at least 30 dB in 3 contiguous audiometric frequencies. In most cases the underlying cause is not known; hearing may or may not recover, and recurrence is possible.<sup>1,2</sup>

A number of etiologies have been proposed, including viral infection, particularly following pseudo-influenza processes of the upper airways. The most frequently implicated viruses are adenovirus, coxsackievirus, influenza, rubella, measles, herpes, and parotiditis virus.<sup>3</sup> Vascular pathology is the most frequent cause in elderly patients. In this context, 3 vascular mechanisms have been defined<sup>4</sup>: hemorrhage, thrombosis, and vascular spasm. Other less commonly cited causes include autoimmune diseases and allergies. A characteristic common to all of these mechanisms is cochlear ischemia, which produces hypoxia-anoxia of the organ of Corti.

Occasionally, sudden deafness may be attributable to life-threatening central nervous disorders.

The present study describes the case of a patient with sudden deafness due to an exceptional and serious cause: the rupture of a cerebral arteriovenous malformation (AVM).

## CASE REPORT

A 53-year-old man presented to the Emergency Service with right ear tinnitus and hypoacusis of sudden onset, without other symptoms. Audiometry identified right neurosensory hypoacusis with an auditory threshold of 70 dB. The patient was admitted to the Ear, Nose and Throat Service for

intravenous vasodilatory treatment. In the hours following admission, the patient developed an important headache with vomiting. A brain computed tomography (CT) scan was therefore requested, which showed the presence of a right subarachnoid hemorrhage (Fig 1A). The patient was moved to the Neurology Service for continued vasodilatory treatment and evaluation. Magnetic resonance imaging (MRI) and magnetic resonance angiography showed the presence of an AVM in the right temporo-parieto-occipital region (Fig 1B and C). Nimodipine was administered intravenously, followed by improvement in hearing and headache. The patient is presently stable, with occasional headaches, and is pending surgery or selective embolization.

## DISCUSSION

Sudden-onset deafness is not an exceptional ear, nose, and throat emergency. The symptoms are sudden hypoacusis accompanied by tinnitus. Approximately 30% of patients also present dizziness with a gyrating sensation and instability, associated with nausea and vomiting—generally related to severe loss of hearing.<sup>5</sup> A correct anamnesis is important in such cases, placing special emphasis on the possible antecedents of cardiopulmonary disease, endocrine and metabolic disorders (diabetes, hypercholesterolemia), and neurologic pathology (cerebrovascular stroke, etc).

Audiometry can define the degree of hearing loss. A basic neurologic examination is required to exclude possible neurologic deficits capable of inducing central pathology.

The appearance of intense headache and vomiting in such situations is suggestive of a central neurologic origin, such as acute hemorrhage. The most common presenting symptom related to subarachnoid hemorrhage is described as “the worst headache of a person’s life” in 85% of cases. The onset of the headache may or not be associated with a brief period of consciousness, seizure, vomiting, or meningismus. Hearing loss is related in 5% of patients.<sup>6</sup>

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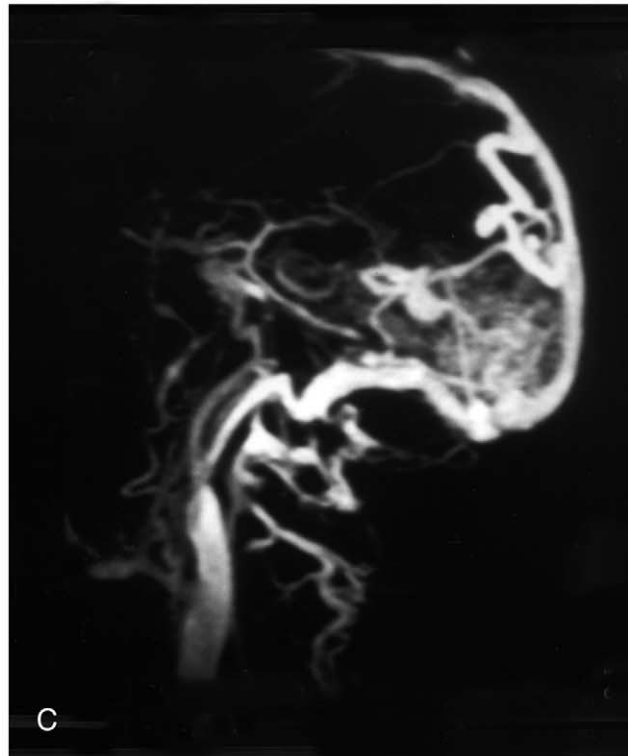
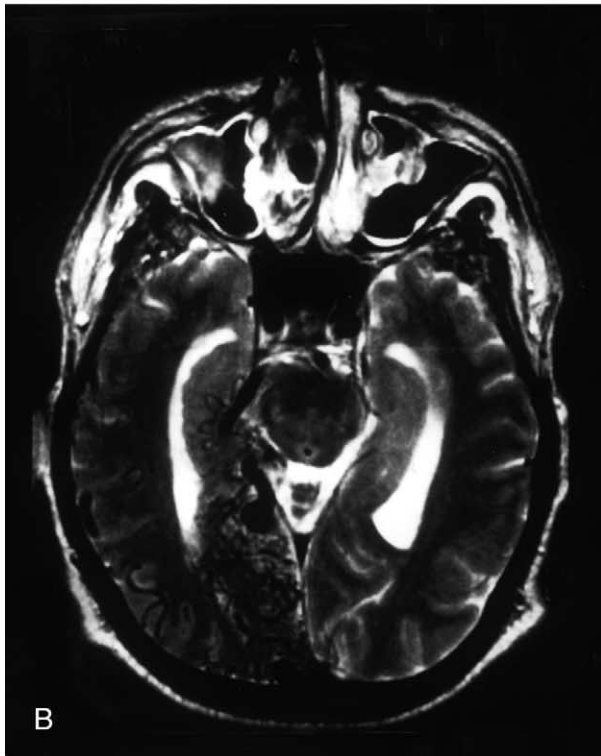
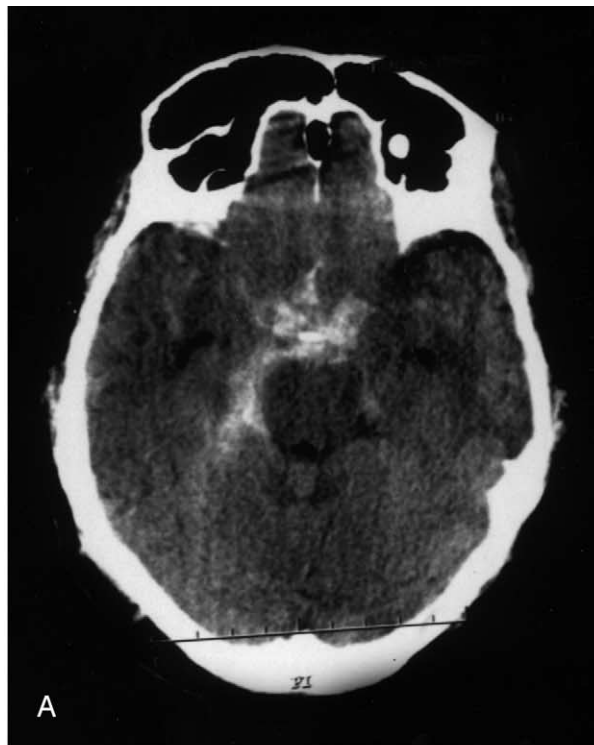
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**Fig 1. A,** Axial CT view showing a hyperdense image corresponding to subarachnoid hemorrhage at the perimesencephalic level. **B,** T2-weighted axial MRI acquisition showing a wavy circumscribed image with signal vacuancy and no mass effect, in the right hemisphere at temporo-parieto-occipital level, corresponding to a very large AVM. **C,** Magnetic resonance angiography (second acquisition, axial plane). Note drainage of the malformation through thick veins that in turn empty into the great cerebral vein, straight sinus, and superior longitudinal sinus.

In our patient, hypoacusis was the manifestation of subarachnoid hemorrhage secondary to rupture of a cerebral AVM. The inner ear may have been damaged in 2 ways: (1) by the presence of erythrocytes surrounding the nerves within the inner auditory canal and following the nerve fibers on entering the Rosenthal canal and in the fluids of the inner ear<sup>7</sup> and/or (2) thorough vasospasm originating at brain level in situations of hemorrhage.<sup>8</sup> The fact that hearing recovered after the provision of vasodilatory medication points to the latter cause in our patient.

A CT study should be made in patients with these symptoms to exclude subarachnoid bleeding. If the presence of hemorrhage is confirmed, angiography or magnetic resonance angiography is indicated after stabilization of the patient, to define the underlying cause.

Most nontraumatic subarachnoid hemorrhages (85%) are due to the rupture of intracranial sacular aneurysms, whereas 10% are attributable to cerebral AVMs and 5% to other causes.<sup>9</sup>

AVMs are the most dangerous congenital vascular malformation. They consist of anomalous vessels that join the arterial and venous systems. Most constitute arteriovenous fistulas in which the participating vessels increase in size and grow over time. Intracranial AVMs occur in about 0.1% of the population, which is one tenth the incidence of intracranial aneurysms.<sup>10</sup> The main clinical signs and symptoms are headache and seizures. In a large percentage of cases, AVMs initially manifest as intracerebral hemorrhage.<sup>11</sup> Such bleeding is generally intraparenchymal and is less commonly found at the subarachnoid level.

The treatment of AVMs is indicated whenever associated bleeding has occurred or the patient has epileptic seizures. The therapeutic options range from surgery<sup>12</sup> to selective embolization and ste-

reotoxic irradiation.<sup>13</sup> Patient monitoring usually suffices in asymptomatic cases.

## CONCLUSIONS

Sudden deafness can be the first manifestation of severe central pathology such as acute subarachnoid hemorrhage. In cases of sudden deafness accompanied by some other atypical sign or symptom (especially neurologic), a central cause should be evaluated as a first etiologic possibility.<sup>1</sup>

## REFERENCES

1. Kanzaki J. Diagnosis criteria or acute profound deafness. *Acta Otolaryngol (Stockh)* 1994;514(suppl):7-8.
2. Lacosta JL, Sánchez-Galán L, Infante JC. Sordera brusca. Experiencia en nuestro hospital. *Acta Otorrinolaringol Esp* 1998;49:189-93.
3. Guiral H, Risco J, Figuerola E. *Anales ORL Iber-Am* 1997; 6: 565-79.
4. Lienhart H, Gouteyron JF, Faugere JM. Sorderas bruscas y fluctuantes. *Encyclopedie Medico-Chirurgicale* E20.183:A10.
5. Nakashima T, Yanagita N. Outcome of sudden deafness with and without vertigo. *Laryngoscope* 1993;103: 1145-9.
6. Adams HP, Jergenson DD, Kassell NF, et al. Pitfalls in the recognition of subarachnoid hemorrhage. *JAMA* 1980;244:794-6.
7. Arnold W, Vosteen KH. Sudden deafness as a consequence of rupture of a basilar artery aneurysm. *HNO* 1977;25:127-30.
8. Qureshi AI, Suarez JJ, Bhardwaj A, et al. Early predictors of outcome in patients receiving hypervolemic and hypertensive therapy for symptomatic vasospasm after subarachnoid hemorrhage. *Crit Care Med* 2000;28:824-9.
9. Van Gijn JJ, Rinkel GJ. Subarachnoid haemorrhage: diagnosis, causes and management. *Brain* 2001;124:249-78.
10. Singer RS, Ogilvy CS, Rordorf G. Congenital vascular malformations of the neuroaxis. *UpToDate* 2001;9:3.
11. Ondra SL, Tropp H, George ED, et al. The natural history of symptomatic arteriovenous malformation of the brain: a 24 year follow-up assessment. *J Neurosurg* 1990;73: 387-91.
12. Heros RC, Korosue K, Diebold PM. Surgical excision of cerebral arteriovenous malformation: late results. *Neurosurgery* 1990;26:A570-8.
13. Ogilvy CS. Radiation therapy for arteriovenous malformations: a review. *Neurosurgery* 1990;26:725-35.