Acta Neurochir (Wien) (1994) 129: 100–104

Acta Neurochirurgica

Cavernous Angiomas of the Internal Auditory Canal
A Case Report and Review of Literature

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Summary

Cavernous angiomas of the internal auditory canal (IAC) are rare. They are angiographically occult; and because the clinical symptoms are similar both in intracanalicular cavernous angiomas and acoustic tumors it had been difficult to differentiate pre-operatively both of these pathologies until the advent of magnetic resonance imaging (MRI). Even nowadays the correct diagnosis may be missed if the patient is imaged only with gadolinium enhanced MRI without prior obtaining a non-contrast MRI. These diagnostic difficulties are illustrated by the report of a related case. The importance of thorough neuroradiological investigations stressed and MRI features, surgical management and relevant literature concerning the cavernous angiomas of the internal auditory canal are discussed.

Keywords: Cavernous angioma; internal auditory canal; differential diagnosis; facial nerve; magnetic resonance imaging.

Introduction

Cavernous angiomas are rare vascular malformations of the central nervous system, seldom occurring in the internal auditory canal (IAC); only 15 cases have been previously reported[3, 4, 11, 14, 16, 19, 27]. Indeed, very few cases of cavernous angiomas occurring in the cranial nerves have been described in the literature[7, 15, 32]. Seventy to ninety percent of intracranial cavernous angiomas occur within the cerebral hemispheres[21, 28, 29], and 20–25% of them occur below tentorium[22, 31].

Because the presenting symptoms and audiometric findings are similar in both cavernous angiomas and acoustic tumors, it was difficult to diagnose these angiographically occult vascular malformations, until the advent of magnetic resonance imaging (MRI). Definitive diagnosis was possible only through histopathology. MRI has improved our ability to identify these vascular malformations with greater accuracy preoperatively. We report a case of cavernous angioma within the IAC. Since our patient was imaged preoperatively only with Gadolinium enhanced MRI and no non-contrast MRI was obtained, a diagnosis of acoustic neuroma was made preoperatively.

In this report, we describe the MRI features that differentiate IAC cavernous angiomas from meningiomas, acoustic neurinomas and other lesions. Operative management of IAC cavernous angiomas is discussed and relevant literature is reviewed.

Case Report

A 36 year old man presented with a one year history of progressive right-sided hearing loss and a two month history of vertigo, nausea and unsteady gait. On examination, there was a right-sided sensorineural hearing loss with 20% speech discrimination. The rest of his neurological examination was normal. MRI images with Gadolinium showed a high intensity signal in the right IAC (Fig. 1 a and b). Given the clinical history, physical examination and MRI findings, a preoperative diagnosis of acoustic tumor was made and no further imaging studies were performed.

Operation: The patient underwent a right retromastoid craniectomy in a left lateral position. Once the posterior wall of the internal acoustic meatus was removed, a reddish, soft lesion composed of multiple blood vessels, measuring 4 × 6 × 4 mm, was found posterior to the 7th and 8th cranial nerves and originating in the 7th cranial nerve. Using microsurgical techniques, the lesion was gently separated from the 7th and 8th nerve complex and excised totally.

Pathology: Histopathological examination showed thick, fibrovascular septae delineating spaces of various sizes lined by flattened cells (Fig. 2). These spaces contained erythrocytes, fibrin and neutrophils. Azocarmine stain showed that the walls of the lumina were made up of collagen. No intervening nervous tissue was noted. These findings were consistent with a diagnosis of cavernous angioma.

Postoperative course: In the immediate postoperative period, the patient was noted to have moderately severe mixed hearing loss and normal facial function. At 4 months follow-up, hearing had improved

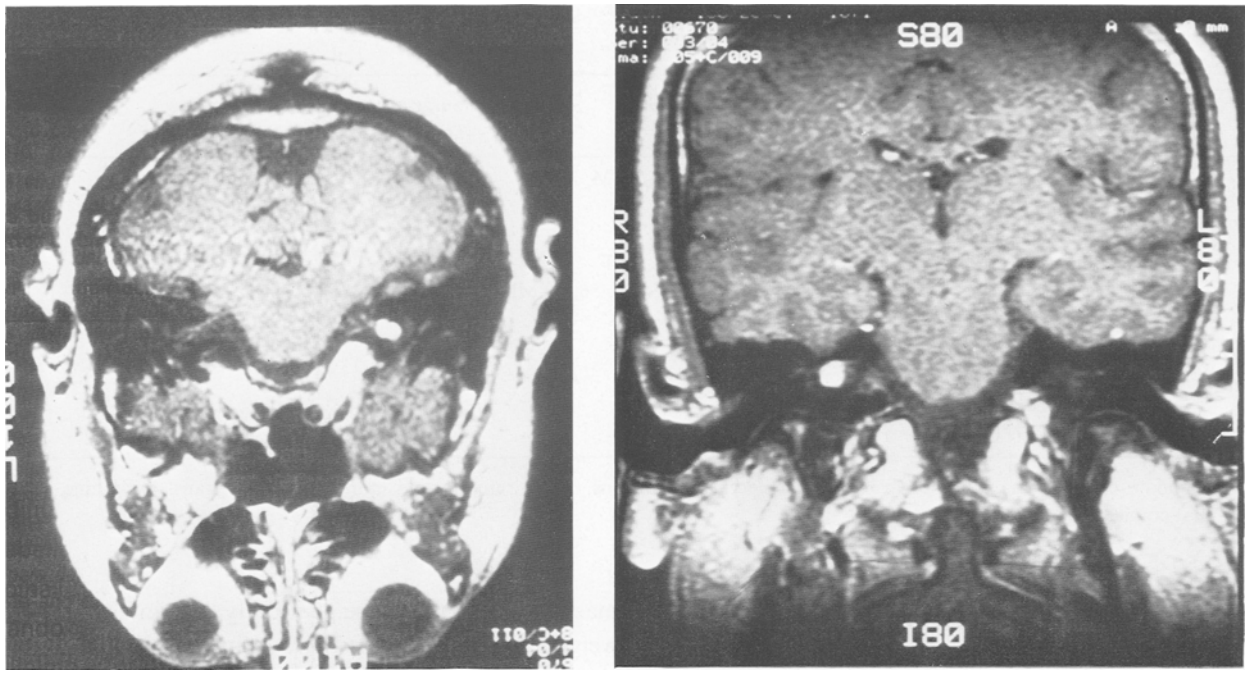

Fig. 1. MRI with Gadolinium showing contrast enhancing lesion in the right internal auditory canal in axial image (a); as well as in coronal image (b)

to mild to moderate sensory neural hearing loss. The remainder of the neurologic examination was normal.

Discussion

Cavernous angiomas (cavernomas) are vascular malformations that may involve the central or peripheral nervous system[15, 22]. They are often sporadic; and familial incidence may be observed[2, 5]. When cavernous angiomas are familial, inheritance is dominant with variable expression[9, 21]. Though solitary lesions are about three times as common as multiple lesions, half of the patients with familial incidence have multiple lesions[21]. Autopsy studies indicated that cavernous angiomas constitute approximately 1% of all intracranial vascular lesions representing 15% of all cerebral vascular anomalies[1–3]. Cranial nerve involvement is rare and only a few cases have been reported[7, 15, 32]. Only 15 cases of cavernous angiomas involving the 7th–8th cranial nerve complex located entirely within the IAC have been reported[3, 4, 11, 14, 16, 19, 27].

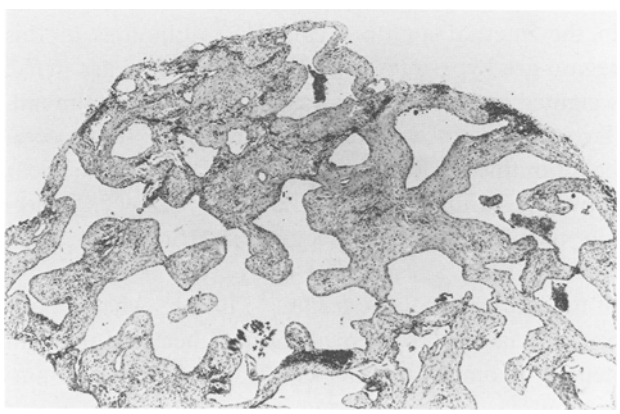

Fig. 2. Cavernous hemangioma showing widely dilated vascular spaces lined by flattened cells (hematoxylin and eosin × 25)

However, there are also occasional reports of cavernous angiomas presenting at the cerebellopontine angle and the middle ear[4, 6, 8, 13, 14, 24]. Iplikcioglu *et al.*[10] also reported cystic cavernous angiomas of the cerebellopontine angle. Penkert *et al.*[20] have reported telangiectasia at the IAC.

Clinical data: When the reported cases were analyzed (including our case), the youngest patient was found to be 24 years[27] and the oldest was 66 years[19]. There was a male predominance (M : F, 10 : 5). Clinically, these patients had small lesions with significant hearing loss. Sundaresan *et al.*[27] noted significant facial weakness in patients harboring small lesions (2/2). They

Table 1. *Cavernous Angiomas Located in Internal Auditory Canal.* Angiomas of other sites including temporal bone, middle ear, cerebellopontine angle; vascular malformations of internal auditory canal are excluded

No.	Author	No. of cases	Age/Sex	Surgical approach	Cranial nerve involved
1	Sundaresan *et al.*, 1976	2	23 M, 50 M	SOC	8th (one each) NI (one case)
2	Brackman *et al.*, 1980	2	NA	NA	NA
3	Mangham *et al.*, 1981	2	29 M, 44 F	TL	7th
4	Pappas *et al.*, 1989	6	36, 39, 44, 56, 66-M; 31/F	TL	7th (5 cases) 8th (one case)
5	Matias-Guiu *et al.*, 1990	1	24 F	NA	8th
6	Bordi *et al.*, 1991	1	29 M	RMC	7th
7	Jacobson *et al.*, 1991	1	41 F	MCF	7th & 8th complex
8	Babu *et al.*, 1993	1	36 M	RMC	7th

MCF middle cranial fossa, *NA* not availabel, *NI* nervus intermedius, *RMC* retromastoid craniectomy, *TL* translabyrinthine, *SOC* suboccipital craniectomy.

stated that patients with marked facial weakness, profound sensorineural hearing loss and the presence of an intracanalicular mass, a diagnosis of cavernous hemangioma (angioma) was likely. After a thorough review of literature, we have found that facial weakness was not a common sign. Pappas *et al.*[19] noted facial weakness in only two of the seven patients, Mangham *et al.*[14] did not report facial weakness in two patients with angioma, and neither did Bordi *et al.*[3], Matias-Guiu *et al.*[16] or our patient. Origin of cavernous angioma was noted to be not constant in relation to the cranial nerve 7th or 8th IAC. At surgery, the lesion was found to be attached to the facial nerve in eight cases, 7th–8th complex in two cases, 8th nerve in three cases, nervus intermedius in one case and information not available in two cases[4] (Table 1).

Radiology and differential diagnosis: Magnetic resonance imaging is the most sensitive and specific diagnostic study for identifying the cavernous angiomas of the IAC. Cavernous angiomas of IAC are hyperdense in non-contrast computerized tomography (CT) and may enhance minimally with contrast. Ocasionally central hypodensity represents old hemorrhage. CT scan with bone algorithms may be used to visualize the enlargement of IAC[17]. However, MRI is more specific and more sensitive than CT scan. Before 1981 only six cases have been reported and after the advent of MRI, 10 more cases have been added to the literature (Table 1) indicating the sensitivity of MRI in identifying these lesions.

In MRI study, both T 1 and T 2 weighted images are hyperintense and the lesion enhances with Gadolinium[19]. In contrast, intracanalicular acoustic neuromas are either isointense or slightly hypointense on T 1 weighted images unless there is intratumoral hemorrhage, which is exceedingly rare. They may be hyperintense on T 2 weighted signal with significant enhancement after intravenous Gadolinium. In the present case, since MRI with Gadolinium was the only study performed and the most common tumor of the IAC is acoustic neuroma, no alternative diagnoses were considered.

In retrospect, had we obtained MRI without Gadolinium in addition to contrast MR images, a diagnosis of cavernous angioma might have been made. Pappas *et al.*[19] have reported cavernous angiomas of the IAC where the preoperative diagnosis was made with the help of MRI.

Differential diagnosis of lesions in the IAC also include facial nerve neuroma, lipoma and meningioma. Facial nerve neuromas often arise from the geniculate, vertical or tympanic segments of the nerve rather than in the internal auditory canal[19, 27]. Lipomas at this region are hyperintense in T 1 and hypointense in T 2 weighted images and don't enhance with Gadolinium. Two percent of the meningiomas occur at the cerebellopontine angle and they are rarely known to occur primarily within the internal auditory canal[26]. Generally, meningiomas at this region arise from the posterior surface of the petrous bone away from or at the edge of the internal acoustic meatus[30]. Rarely, when lesions are associated with elements of both hemangioma and neurilemmoma, the MRI or CT picture may be ambiguous[12].

Histology: Cavernous angiomas are macroscopically reddish, lobulated and well defined. The interior

looks like a honeycomb with variable sizes of spaces filled with blood and separated by fine fibrous strands. Microscopically, there is no capsule seen. The walls of the spaces are lined by a single layer of endothelial cells. There is no intervening nervous tissue among the angiomatous elements and no elastic tissue in the sinusoidal walls[18].

Radiological distinction between capillary telangiectasias and cavernous angiomas is sometimes difficult; sometimes both these can coexist and hence these two often are called telangiectasias[22]. Histopathologically teleangiectasia has intervening nervous tissue. Penkert *et al.*[20] reported IAC angioma as cavernous angioma which was indeed a true telangiectasia.

Surgical approaches: The approaches to IAC are suboccipital[3, 20], middle cranial fossa[11], or translabarinthine[14, 19]. Suboccipital approach has often been advocated by the neurosurgeons to deal with the lesion since they are more familiar with that approach. Though we operated the present case through a suboccipital craniotomy, in patients where there is significant hearing deficit, translabarithine seems to be the best approach. With this approach, cerebellar retraction is avoided and the facial nerve is located easily[19]. Manipulation of the facial nerve is minimized. However, preservation of hearing may be better through suboccipital craniectomy or middle fossa approach[4]. Because these tumors readily peel off from the nerves, rarely the facial nerve may have to be sectioned[14] to achieve a total removal. But even in anatomically preserved facial nerve during surgery, paresis was not an uncommon complication[19].

Total excision is the goal because cavernous angiomas are benign. The preoperative diagnosis of cavernous angioma doesn't change the surgical approach. Though it is tempting to excise these benign lesions, in view of their relatively slow growing nature, a trial of observation is a point of consideration. However, the chances of preserving the facial nerve function are higher in smaller lesions than larger lesions. That may be a point which forces surgeons to intervene as soon as the lesion is identified. Although follow-up studies have not been reported, Matias-Guiu *et al.*[16] have a four year follow-up without recurrence in one case.

Conclusions:

Cavernous angiomas of the IAC are uncommon. They occur most often in men and present with a profound sensorineural hearing loss out of proportion to the size of the tumor. MRI is the best imaging study; hyperintense signal characteristics are seen on T 1 and T 2 weighted images. Cavernous angiomas enhance strongly after intravenous Gadolinium injection. Definitive diagnosis is made by histopathology. Total excision is the goal to prevent recurrence.

Acknowledgement

Thanks to Lynda D'Aquino for typing this manuscript.

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