

Cavernous hemangioma of the internal auditory canal encasing the VII and VIII cranial nerve complex: case report and review of the literature

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Abstract Cavernous angiomas originating in the internal auditory canal are very rare. In the available literature, only 65 cases of cavernomas in this location have been previously reported. We describe the case of a 22-year-old woman surgically treated for a cavernous hemangioma in the left internal auditory canal, mimicking on preoperative magnetic resonance imaging MRI an acoustic neuroma. Neurological symptoms were hypoacusia and dizziness. The cavernous angioma encased the seventh and, partially, the eighth cranial nerve complex. A “nearly total” removal was performed, leaving a thin residual of malformation adherent to the facial nerve. Postoperative period was uneventful; hearing was unchanged, but the patient had a moderate inferior left facial palsy (House-Brackmann grade II) slightly improved during the following weeks. On the basis of the observation of this uncommon case, we propose a revision of the literature and discuss clinical features, differential diagnosis, and treatment.

Keywords Cavernous angioma · Internal auditory canal · Acoustic neuroma · Facial nerve · Cochlear nerve

Introduction

Intracranial cavernous angiomas or cavernomas are angiographically occult low-pressure cerebral vascular

malformation. They have an estimated prevalence of 0.1–4 % of the population and account for 8–15 % of all cerebral vascular malformations [1–3]. They occur in a sporadic or familial form; in the last subtype, the presence of multiple lesions is more common [3]. Cavernomas occur usually in the cerebral hemispheres (70–90 %) but may also be found in infratentorial and spinal cord compartments (20–25 %) [1–6]. Cavernous angiomas of the cranial nerves are rare [1], only 65 of which involving the seventh and eighth nerve complex (Table 1). The location inside the internal auditory canal and in the cerebello-pontine angle justifies their inclusion among the unusual lesions to consider in the differential diagnosis of acoustic neuromas.

We describe the case of a young patient with cavernous angioma of the internal auditory canal encasing the VII–VIII cranial nerves, since diagnosis to surgical treatment.

Case report

This 21-year-old Caucasian woman had left-side dizziness and slight hearing loss since 6 months. She was admitted on February 2015, in our Department without any other neurological deficits. Audiometry showed average loss of 20 dB, with 80 % vocal discrimination (AAO-HNS class A) [22]. Magnetic resonance imaging (MRI) showed a medial intrameatal expansive lesion on the left side, slightly debording into the cerebello-pontine cistern, with a maximum diameter of 1.5 cm, isointense to the cerebral parenchyma on T1 and T2 weighted images, with contrast enhancement (Fig. 1). According to Dunn et al. [44], preoperative clinical MRI diagnosis was medial acoustic neuroma.

The patient was operated on in Fukushima’s lateral position, by a key-hole retrosigmoid approach, attempting hearing and facial preservation. After dural opening, lateral basal

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Table 1 Internal auditory canal cavernomas: review of the literature

Author, year	Number	Sex, age	Preoperative symptoms and signs	MRI	Nerve(s) involved	CPA ext	Surgical removal	Results
[7]	1	M, 37	HL	—	7cn–8cn	No	?	ImprFP
[8]	1	M, 23	HL, FP	—	8cn	No	?	ImprFP
	2	M, 50	HL, FP	—	7cn interm	No	?	ImprFP
[9]	1	?	?	—	7cn	7cn	no	STR
	2	?	?	—	7cn	No	STR	?
	3	?	?	—	7cn	No	TR	?
	4	?	?	—	7cn	No	TR	?
[10]	1	M, 29	HL	—	7cn	No	TR	HL + FP
	2	F, 44	HL	—	7cn	No	NTR	Unch
[5]	1	F, 30	HL, FP	—	7cn, 8cn	Yes	TR	?
[11]	1	M, 26	HL	Hyper, Ca	7cn	No	NTR	Transient FP
	2	F, 31	HL, FP	Hyper, Ca	7cn	Yes	NTR	Unch
	3	M, 29	HL	Hyper, Ca	7nc	Yes	TR	HL + FP
	4	M, 39	HL	Hyper, Ca	?	No	?	Unch
	5	M, 56	HL	Hyper, Ca	7cn	No	NTR	HL + FP
	6	M, 44	HL	Hyper, Ca	7cn	No	NTR	HL + FP
	7	F, 66	HL, FP	Hyper, Ca	?	No	?	ImprFP
[12]	1	F, 36	HL, FP, HFS	Iso, CE+	7cn	No	TR	ImprHFS,
[2]	1	F, 24	HL	?	8cn	No	TR	HL
[13]	1	M, 29	HL	Iso, CE+	7cn	No	TR	HL + FP
[14]	1	M, 39	HL, FP	Iso, Ca	?	No	TR	Unch
[15]	1	F, 41	HL, FP	Iso, CE+	?	No	TR	Impr
[16]	1	M, 39	GD	Iso, CE+, Ca	?	No	?	?
[17]	1	M, 58	HL, GD	Hypo, Ca	7cn–8cn	No	TR	HL + HBIIIFP
[18]	1	M, 44	HL, FP	Iso, CE+	7cn	No	TR	Unch
[4]	1	M, 36	HL, GD	Iso, CE+	7cn	No	TR	Impr
[19]	1	M, ?	HL, HFS	?	?	No	?	?
	2	M, ?	HL	?	?	No	?	?
	3	M, ?	HL, HFS	?	?	No	?	?
	4	M, ?	HL	?	?	No	?	?
	5	M, ?	HL, HFS	?	?	No	?	?
[20]	1	M, 34	HL	Iso, CE+	7cn–8cn	No	TR	Unch
[21]	1	M, 51	HL	Iso, CE+	7cn	No	TR	Unch
[22]	1	M, 60	HL	hypo	?	No	STR	Unch
[23]	1	M, 32	HL	Hyper, CE+	7cn–8cn	No	TR	Impr
[24]	1	?	HL, FP	Iso, CE+	7cn–8cn	No	STR	Impr
[25]	1	M, 45	HL, FP	Iso; Ca	7cn	No	TR	Unch
[6]	1	M, 32	HL, FNumb	Iso	7cn	Yes	TR	Unch
[26]	1	F, 34	HL	Iso, CE+	7cn–8cn	No	TR	HL + FP
	2	F, 62	HL	Iso, CE+	7cn–8cn	Yes	TR	HL + FP
[27]	1	F, 24	HL	Iso, CE+	8cn	Yes	TR	Unch
[28]	1	F, 39	HL	Iso, CE+; Ca	7cn	No	PR	?
[29]	1	F, 30	HL, FP	Iso, CE+; Ca	7cn–8cn	No	TR	ImprFP
[30]	1	F, 43	HL	Iso, CE+; Ca	IVN	No	TR	Unch
[31]	1	M, 53	HL, FP	Iso, CE+; Ca	7cn–8cn	No	TR	Unch
[32]	1	M, 61	HL, FP	Iso, CE+	7cn–8cn	No	TR	Unch
[1]	1	M, 76	HL, FP, dysphagia	?	7cn–8cn	Yes	?	ImprFP
	2	M, 53	HL, FP	Hyper	7cn–8cn	Yes	TR	ImprFP

Table 1 (continued)

Author, year	Number	Sex, age	Preoperative symptoms and signs	MRI	Nerve(s) involved	CPA ext	Surgical removal	Results
[33]	1	F, 29	HL, FP	Iso, CE+	7cn–8cn	No	TR	Unch
[34]	1	M, 21	HL	Iso, CE+	SVN	No	TR	Unch
[35]	1	M, 47	HL, FP	Iso, CE+	?VN	No	TR	Unch
[36]	1	M, 21	HL	Iso, CE+	7cn–8cn	No	TR	Unch
[37]	1	F, 23	HL, FP	Iso, CE+	7cn–8cn	Yes	TR	ImprFP
	2	M, 28	HL,	Iso, CE+	7cn–8cn	No	TR	HL + FP
	3	M, 29	HL,	Iso, CE+	7cn–8cn	Yes	TR	HL + FP
	4	M, 40	HL, HFS	—	7cn–8cn	No	TR	No HFS
	5	M, 42	HL,	Iso, CE+	7cn–8cn	No	TR	Unch
	6	M, 53	HL, FP	Iso, CE+	7cn	No	TR	Unch
	7	F, 53	HL, HFS	Iso, CE+	7cn	No	TR	HL + FP
[38]	1	F, 45	FP	Hyper, CE+	?	Yes	?	?
[39]	1	F, 51	HL	Iso, CE+	?	?	TR	?
[40]	1	M, 51	HL, FP	Iso, CE+	7cn–8cn	No	TR	Unch
[41]	1	M, 40	FP	Iso, CE+	?	Yes	?	?
[42]	1	F, 38	HL, FP	Iso, Ca	?VN	No	TR	Unch
[43]	1	M, 47	HL, FP	Iso, Ca	7cn	No	TR	Unch
Present case 2015	1	F, 21	HL	Iso, CE+	7cn–8cn	Yes	NTR	HBIIFP

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? not specified, — not done, 7cn facial nerve, 8cn vestibular-cochlear nerve, Ca calcifications, CE+ contrast enhancement, CPA ext extension in cerebello-pontine angle, Fnumb facial numbness, FP facial palsy (HB: House-Brackmann), GD gait disturbances, HFS hemifacial spasm, HL hearing loss, hyper hyperintense on T1, hypo hypointense on T1, Impr Improved, interm intermediate, IVN inferior vestibular nerve, Iso isointense on T1, NTR near total removal, PR partial removal, STR subtotal removal, SVN superior vestibular nerve, TR total removal, Unch unchanged, VN vestibular nerve

arachnoid cistern was entered and CSF drained for brain de-tention. With a gentle retraction of the cerebellum, an atypical red-bluish mass was observed close to the superior vestibular nerve at the entrance of the internal auditory canal (Fig. 2). During the opening of the acoustic meatus, a highly vascular

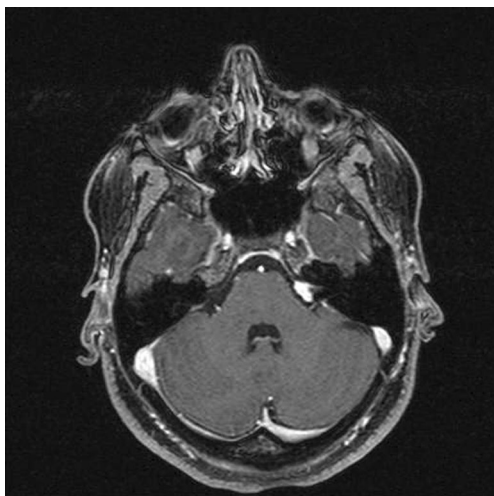


Fig. 1 Preoperative MRI, axial T1 with contrast medium: left medial intrameatal mass, slightly debording into the cerebello-pontine cistern, with a maximum diameter of 1.5 cm, isointense to the cerebral parenchyma in T1, with uptake of contrast medium

soft-tissue mass encasing the seventh and eighth nerve branches was identified, adhering mainly to the facial nerve. The lesion was piecemeal “near-totally” resected using microsurgical technique; a 2-mm residual lesion was left in order to avoid an interruption of the facial nerve.

The postoperative period was uneventful. A CE MRI (Fig. 3) confirmed the nearly total excision of the lesion. The patient was discharged on the fifth postoperative day, showing the same hearing she had in the preoperative examination and a slight left facial paresis (grade II of the House-Brackmann scale), without other neurological deficits.

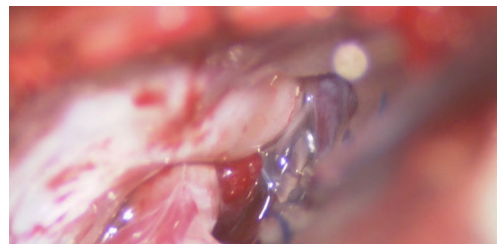


Fig. 2 Intraoperative picture: after gentle retraction of the cerebellum, an atypical red-bluish mass was observed close to the superior vestibular nerve at the entrance of the internal acoustic canal (Fig. 2). During the opening of the acoustic meatus, a highly vascular soft-tissue mass encasing the seventh and eighth nerve branches was identified, adhering mainly to the facial nerve

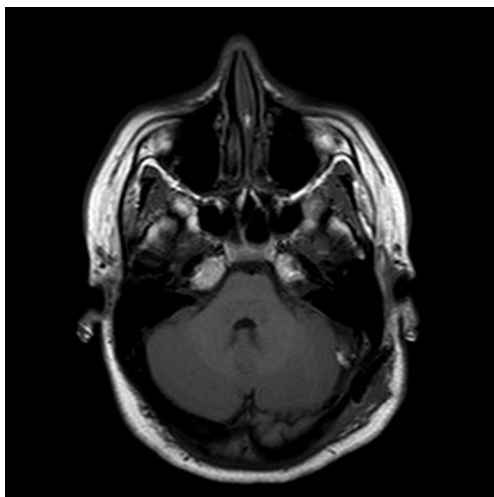


Fig. 3 Postoperative MRI, axial T1 with contrast medium: absence of pathologic contrast enhancement in the internal auditory canal

The histopathological exam revealed benign vascular lesion formed by proliferation of blood vessels and light wall juxtaposed, fibrous, thick, often with hyalinization, lined with simple layer of endothelium, typical characteristics of the cavernous hemangioma, with some remnants of nervous fibers (Fig. 4a and b).

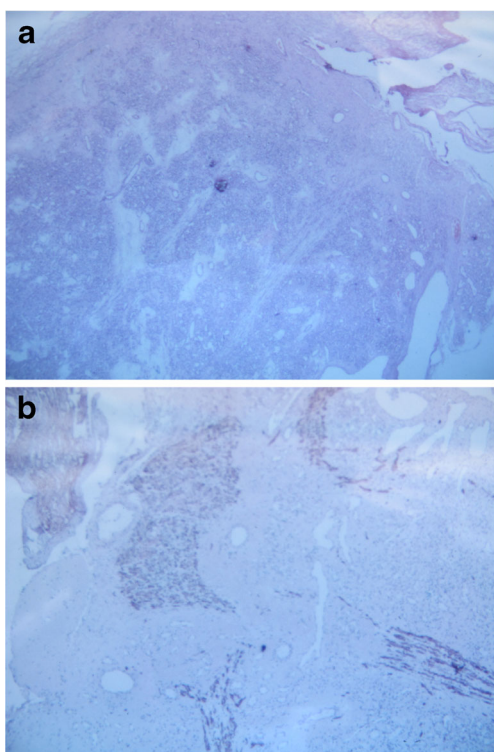


Fig. 4 **a** Histopathological photomicrographs revealed benign vascular lesion formed by proliferation of blood vessels and light wall juxtaposed, fibrous, thick, often with hyalinization, lined with simple layer of endothelium, typical characteristics of the cavernous hemangioma, with **b** some remnants of nervous fibers

At 6 months follow-up, MRI was unchanged; the slight facial paresis completely recovered and hearing was stable.

Discussion

This unusual observation led us to review analytically the international literature to date. Adding our case to the 65 previously described in detail from 1968 to 2014 (Table 1), cavernous angiomas of the internal auditory canal (CAIAC) usually affect young patients (mean age 35.8 years, age range 21–76), with a male prevalence (41 out of 61 cases, 67.2 %). They tend to manifest through decreased hearing acuity (59 out of 62 cases, 95.2 %) and may have insidious, rapidly progressive, or sudden development. Facial paresis may also be present at diagnosis (23 out of 62 cases, 37.1 %). According to some authors [1, 4, 11, 20, 45], the presence of facial paresis and severe hearing deficit in patients with small intracanalicular lesions could be suggestive of a vascular nature, enough to consider it as a possible alternative in the differential diagnosis of acoustic neuromas, although there is no way to differentiate them from clinical data. Hemifacial spasm was present in 6 out of 62 cases (9.7 %). Other symptoms (gait disturbances, facial numbness, dysphagia) were less common.

CAIAC usually are heterogeneous on MRI, appearing mainly isointense in relation to brain parenchyma in T1 and T2 weighted images in 34 out of 48 cases (70.8 %), with marked contrast enhancement in 31 (91.2 %). It appeared hyperintense in 10 cases, with unusual contrast enhancement in 2, and hypointense in only 2 cases [17, 22]. Calcifications may be present in 35.4 % of cases (17 out of 48) and destruction of the internal auditory canal may also be observed, especially on CT with bone window [4, 6, 11, 20]. The differential diagnosis has to be done particularly with acoustic neuroma, which is usually hyperintense on T2 and hypointense on T1 with marked contrast enhancement. In our case, the lesion appeared to be isointense on T1 and T2 with moderate intake of contrast medium (Fig. 1) and determined a slight enlargement of the internal auditory canal, suggesting a preoperative diagnosis of acoustic neuromas. Analogously to other 12 out of 65 patients, in our case too, the cavernous angioma showed a slight extension out of the meatus in the cerebello-pontine cistern, without any difference from vestibular schwannomas.

Inside the canal, except for the four cases in which the CAIAC involved a vestibular nerve [30, 34, 35, 42], the cavernoma encased the acoustic and/or the facial nerve: in 23 out of 52 cases the facial (44.2 %), in 3 the cochlear, and in 22 both (42.3 %). Therefore, CAIAC involved facial nerve in 86.5 % of reported cases.

Microsurgical resection is the treatment of choice, even if the encasement of VII–VIII nerve complex can justify the

possibility of the emergence of important neurological deficits [1–5, 11]. On the other hand, partial resection can lead to recurrence [1]. In the cases described in the literature, a total or nearly total resection was possible in 47 out of 52 (90.4 %) in which this data is reported, whereas in 5, a subtotal or a partial removal was performed.

As regards the postoperative results, the preoperative condition improved in 14 out of 51 cases (23.5 %) and unchanged in 23 (43.1 %). In the remaining 14 patients, including our case, a slight or severe transient or permanent facial palsy was observed.

Even if our case confirm that preoperative differential diagnosis between a CAIAC and a vestibular schwannoma is usually impossible, from the analysis of the literature, it seems to be reasonable to state that, when evaluating a small intra-auricular canal tumor in a young patient, surgeons have to take into account the rare possibility of a cavernous hemangioma. Early diagnosis and surgery may improve the functional outcome, with higher possibilities of preserving neural functions, especially on considering that facial nerve is encased in about 90 % of cases.

Conclusions

CAIAC encasing VII–VIII nerves are rare lesions that should be considered in the differential diagnosis of lesions of the cerebello-pontine angle and internal acoustic meatus. Intrameatal lesion with very high intensity on T2-weighted MR imaging and stippled patterns of calcification on CT is more likely to be cavernous angioma than acoustic neuroma [31]. Total microsurgical resection is the treatment of choice, even if nearly total excision could be sometimes a reasonable option in order to avoid severe and permanent facial nerve palsy.

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