The Laryngoscope

Meningiomas of the Internal Auditory Canal

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Objective: Identify and define specific preoperative and postoperative characteristics of intracanalicular meningiomas (ICMs) in order to improve their diagnosis and management, and to differentiate them from intrameatal vestibular schwannomas (IMVSs).

Methods: Preoperative symptomatology, magnetic resonance imaging (MRI), and postoperative outcomes of 28 ICMs were analyzed. The results were compared to the literature and IMVSs treated by our group.

Results: Anacusis and progressive hearing loss were more frequent in the present population than the cases reviewed (P = .0064 and P = .0001, respectively). Hearing loss affected more than 90% of the patients, with anacusis in 32.1% of the cases. Facial palsy affected 17.9% of the patients. In comparison to IMVSs, preoperative anacusis was more associated to meningiomas (P = .0037), and the facial nerve was more compromised in ICMs than IMVSs, both preoperatively (P = .0011) and at follow-up (P < .0001).

According to a re-evaluation of preoperative MRIs and comparison with IMVSs, linear tumor borders, and linear morphology along the internal auditory canal wall, but not the presence of a dural tail, were significantly more present in ICMs (P = .0035, P = .0004, P = .1963, respectively). These characteristics could have led to a correct preoperative diagnosis in 61% of our cases.

Conclusion: Contrariwise to IMVSs, the frequent preoperative anacusis and facial palsy demonstrate the more aggressive nature of ICMs, which also carry a higher risk of postoperative facial palsy and difficulty to preserve hearing. An attentive evaluation of imaging should ease diagnosis, and asymptomatic or stable ICMs should be enrolled in a wait-and-scan protocol.

Key Words: Intracanalicular meningioma, internal auditory canal, facial palsy, anacusis.
Level of Evidence: 4

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INTRODUCTION

Meningiomas are the most common intracranial neoplasms, accounting for approximately 18% of intracranial tumors, whereas in the cerebellopontine angle (CPA) and internal auditory canal (IAC) they represent only a minimal part, comprising from 3% to 12% of the cases.[1–3] Most CPA meningiomas derive from the epithelial lining of the arachnoid villi, along dural sinuses of the petrous bone, and may extend into the IAC.[4] However, 1.7% of CPA meningiomas arise directly from the IAC due to the presence of arachnoid cells and granulations at cranial nerve exit foramina.[5]

Although they tend to invade dura and bone, they are benign lesions whose symptoms are mainly caused by compression of adjacent structures, which can lead to nerve atrophy and interfere with the blood supply.[5,6] Because of the similarities in symptomatology and imaging with vestibular schwannomas (VSs) of the CPA and IAC, and due to the higher frequency of VSs in the area,[7] approximately 75% to 90% of the reported cases of intracanalicular meningiomas (ICMs) were preoperatively suspected to be VSs.[8,9] This fact, in association with the rarity of this entity, makes it extremely difficult to conduct prospective studies on CPA and ICMs, and most information derives from retrospective case studies and small case series. Moreover, there has been an increased use of primary stereotactic radiosurgery for the treatment of CPA and intracanalicular tumors,[10] which prevents reliable conclusions due to the lack of a histopathologic analysis.

There have been less than 50 cases of pure ICMs described in literature and nearly 100 IAC meningiomas extending partly in the CPA (largest series are collected in Table I).[6–8,11–36]

We present 28 cases of ICM, operated at our center in the past 30 years, and analyse their symptomatology, preoperative imaging, surgical management, intraoperative findings, histopathologic diagnosis, and postoperative outcomes.

The objective of this study was to identify and define specific preoperative and postoperative characteristics of ICMs in order to improve their future diagnosis and management and differentiate them from intrameatal vestibular schwannomas (IMVSs).

MATERIALS AND METHODS

The study was approved by the institutional review board, and all subjects gave informed consent on the use of their data in accordance with the Declaration of Helsinki.

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TABLE I.
Largest Case Series of Intracanalicular Meningiomas.

Study	N (EM ext.)	PO FN	PO Hearing	FN Outcome	Hearing Outcome
Arriaga et al. 1992	9 (ns)	—	85% HL	1 grade III/IV, 1 grade V/VI	—
Breuer et al. 2000	9 (8)	—	3 AN, 3 HL	3 grade II, 6 grade I	1 PTA worsening, 1 WRS drop
Nakamura et al. 2004	8 (7)	—	1 AN, 6 HL	Normal	2 AN, 6 hearing-preserved
Peyre et al. 2012†	10 (8)	1 grade III	9 HL	1 transient facial palsy, 1 III, 1 IV, 1 VI	4/6 postoperative serviceable hearing
Calzada et al. 2015	15 (ns)	1 grade V	ns	only 1 case described: grade III	—
Present study	28	4 grade III, 1 grade II	26 PHL/ SSNHL	4 II, 11 III, 1 IV, 1 V	2/5 hearing-preserving surgeries

† = 12 cases described in article, but with 2 not involving the IAC. AN = anacusis; EM ext = extrameatal extension; FN = facial nerve; HL = hearing loss; IAC = internal auditory canal; N = number; ns = not specified; PHL = progressive HL; PO = preoperative; PTA = pure-tone average; SSNHL = sudden sensorineural HL; WRS = word recognition score.

A retrospective analysis of all patients with meningioma, managed at our institution between December 1988 and July 2019, was performed. Of the 263 meningiomas identified, petroclival and jugular foramen meningiomas with CPA extension were excluded. One hundred and sixty posterior petrous face meningiomas were analyzed. Of these latter cases, 29 were ICMs with or without minimal extrameatal extension (up to 5 mm). One patient was affected by neurofibromatosis type 2 and was therefore excluded. Twenty-eight patients were finally included in this study.

All patients underwent preoperative audiologic examination by pure tone audiometry and speech audiometry. Pure-tone average (PTA) was calculated as the mean of 500 Hz, 1 kHz, 2 kHz, and 4 kHz thresholds. Hearing results were evaluated according to the American Academy of Otolaryngology–Head and Neck Surgery (AAO-HNS).[37] The preoperative and postoperative facial nerve functions were graded according to the House-Brackmann scale (HB).[38] All patients underwent gadolinium-enhanced magnetic resonance imaging (MRI). In a minority of patients, a high-resolution computed tomography (CT) without contrast administration had also been requested. Particular attention was given to MRI on mass location and morphology of the lesions, contrast enhancement, and the presence of dural changes. CT scans were available in 11 patients and were analyzed for bony alterations and hyperostosis.

Surgery was indicated in patients affected by disabling vertigo or a compromised facial nerve, according to our protocol.[39] Patients affected by anacusis (unmeasurable thresholds) were also given a surgical indication. The rest of the patients presented were given the option to undergo a hearing preserving technique or a wait-and-scan follow-up. In some cases, however, patients preferred surgery even when not strongly indicated. Only two of the patients presented were failures of the wait-and-scan program because of tumor growth.

The surgical approach was selected according to the preoperative audiometry, extent of IAC filling, and extrameatal involvement. Intraoperative facial nerve monitoring was conducted in all patients. The extent of tumor resection was classified according to the Simpson scale, where grade I consists in total resection of the tumor with excision of its dural and bony attachment; grade II consists in a total tumor resection and coagulation of its dural attachment; grade III refers to a total tumor resection without resection or coagulation of its dural attachment and its extradural extension; and grade IV is a subtotal tumor resection.[40] All tumors were histopathologically confirmed meningiomas.

Follow-up consisted of clinical and audiologic evaluations and yearly MRI scans.

Charts of 151 cases of histopathologically confirmed IMVSs were additionally evaluated; and preoperative symptoms, preoperative and postoperative facial nerve function, as well as preoperative imaging, were compared to ICMs.

Finally, the collected data were compared to the data that resulted from a review of the literature.[6–8,11–36]

Statistical analysis was performed using Graphpad Prism 8 software (GraphPad Software, Inc, San Diego, CA). The data collected were analyzed using descriptive statistics; continuous variables were expressed as mean ± standard deviation; and categorical variables were expressed as percentage (frequency). When comparing populations, Fisher exact test and odds ratio (OR) with a 95% confidence interval (CI) (OR, 95% CI) were used. Values were considered significant at $P < .05$.

RESULTS

Among the 28 patients affected by ICM, 57.1% were women. Forty-six percent of the patients were operated on the left side. The mean age at surgery was 48.7 ± 14.04 years old. None of the patients had been previously irradiated.

Twenty-two of the tumors were limited to the IAC, whereas five had an average of 3.7 mm extension in the CPA (minimum extension was 2 mm, maximum 5 mm), and one had a millimetric “tail” extending outside the IAC, in the posterior CPA.

All patients were symptomatic at onset. Hearing loss (HL) was the predominant first symptom, with 16 patients (57.1%) presenting progressive HL and two (6.9%) sudden sensorineural HL (SSNHL). Tinnitus appeared from the beginning in eight cases (28.6%), and in one case it was associated to hyperacusis. Two patients (7%) suffered from vertigo from the beginning, and one (3.6%) complained instability but not rotatory vertigo spells. The facial nerve was interested at onset in four patients (2 with facial weakness, 7.1%; and 2 with hemifacial spasm, 7.1%).

At the time of surgery (Table II), HL affected more than 90% of our series, with 32.1% of the patients being anacusic. According to the AAO-HNS, 15 patients (53.6%) had a class D (dead ear), and six patients (21.4%) had a class C. Four (14.3%) and three (10.7%) patients presented a class A or B hearing, respectively. Mean PTA

TABLE II.
Symptoms at Diagnosis and Surgery of Intrameatal Vestibular Schwannomas and Intracanalicular Meningiomas.

Symptom	IMVS (%)	ICM (%)
SSNHL	32 (21.2)	2 (7.1)
Fluctuating HL	4 (2.6)	0 (0)
Profressive HL	97 (64.2)	25 (89.3)
Total pts with HL	133 (88.1)	27 (96.4)
Anacusis**	12 (7.9)	9 (32.1)
Tinnitus	84 (55.6)	12 (42.9)
Vertigo	40 (26.5)	5 (17.9)
Recurrent vertigo	2 (1.3)	0 (0)
Instability	47 (31.1)	11 (39.3)
Total pts with vestibular impairment	79 (52.3)	14 (50.0)
Facial weakness	2 (1.3)	3 (10.7)
Facial spasm	0 (0.0)	4 (14.3)
Total pts with facial palsy**	2 (1.3)	5 (17.9)
Fullness	4 (2.6)	1 (3.6)

Each patient presents more symptoms. Data presented express the number of patients (percentage in each subpopulation). Symptoms incidence at the time of surgery.

HL = hearing loss; ICM = intracanalicular meningioma; IMVS = intrameatal vestibular schwannoma; pts = patients; SSNHL = sudden sensorineural hearing loss.

**$P < .005$.

(excluding the anacusic patients) of the affected ear was 50.1 ± 21 dB, and word recognition score (WRS) was 43.6% ± 46.4%. Contralateral hearing had a mean PTA of 21.1 ± 8.2 dB. One patient suffered from ear fullness. The majority of patients complaining tinnitus had hearing residues, whereas anacusic patients were rarely affected by tinnitus ($P = .039$, Fisher exact test). The five patients with preoperative facial palsy were classified as grade III and II according to the HB scale, in four and one cases, respectively. None of the patients presented other cranial nerves' impairments.

Preoperative MRI was uncharacteristic in the majority of the cases, and lesions were preoperatively suspected to be VSs in 85.7% of the cases. Only in two cases was the presence of a dural tail inclined toward a meningioma (Fig. 1). In one case, the tumor extended toward the facial nerve, with strong enhancement of the labyrinthine segment of the facial nerve up to the geniculate ganglion, which was suggestive of facial nerve schwannoma. Similarly, another patient suffered from hemifacial spasm and presented an anterior IAC gadolinium enhancement that also alluded to a facial nerve schwannoma. Both lesions were only eventually excluded by histopathology. Only one case, among those with available CT scan, presented intralesional calcifications, whereas none of the cases included evidenced hyperostosis or spurs.

A retrospective analysis of the preoperative MRI evidenced the presence of linear tumor borders (perpendicular to the IAC wall) in 26.3% of the patients, whereas in 36.9% of the cases the borders appeared convex (Fig. 2). Radiologic characteristics were compared in the two populations (Table III): linear tumor borders ($P = .0035$, Fisher exact test) and a linear morphology

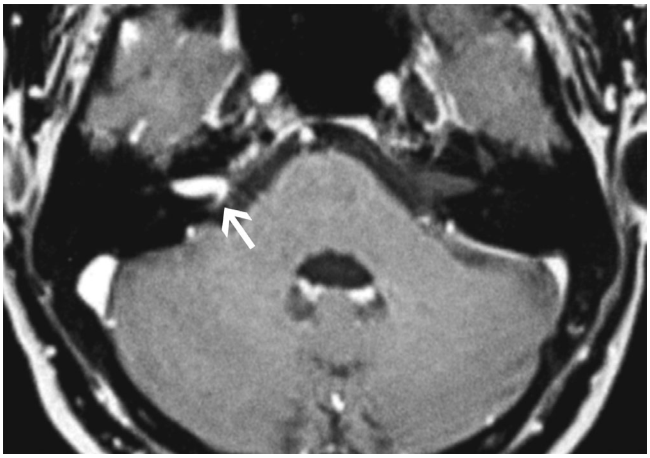

Fig. 1. Preoperative MRI of an intracanalicular meningioma with a posterior dural tail [arrow] (axial T_1-weighted gadolinium-enhanced image). MRI = magnetic resonance imaging; T_1= longitudinal relaxation time.

along the IAC wall ($P = .0004$, Fisher exact test) were significantly more present in ICMs than IMVSs. The presence of a dural tail was not statistically significant ($P = .1963$, Fisher exact test).

Five patients were operated through a hearing preserving approach. Four patients were operated with an enlarged middle fossa approach (EMFA), and one patient, with a 5 mm CPA extension, was operated through a retrosigmoid approach. Two (40%) of these patients maintained their preoperative hearing postoperatively.

All patients with a deaf ear or unserviceable hearing were operated through a translabyrinthine approach (TLA).[41] There were no revision surgeries.

In 10 (36%) cases, a meningioma was suspected intraoperatively because of dural adherence and origin as well as hypervascular consistency. In three (11%) cases, the tumor infiltrated the bone, which was also suggestive of meningioma. In one case, hyperostosis was present intraoperatively. Two patients with preoperative facial weakness and one patient with preoperative hemifacial spasm had intraoperative evidence of facial nerve tumor infiltration. The nerve was therefore sacrificed and simultaneously repaired by a sural nerve cable graft in two cases. In the third case, the patient underwent subsequent hypoglossal-facial anastomosis. The facial nerve was anatomically preserved in the rest of the cases and responded to minimal intraoperative stimulation. Two of these cases had the facial nerve engulfed by the tumor and compressed. Two cases presented a cochlear nerve tumor infiltration.

Total tumor removal was achieved in all cases (classified as I and II according to the Simpson scale).

Histologic evaluation of the specimens revealed five subtypes of benign meningioma, grade I, according to the World Health Organization Classification of Central Nervous System Tumors. The most common subtype was meningothelial in 13 cases (46.4%). Other types, in order of frequency, were: four fibrous (14.3%), four transitional (14.3%), four angiomatous (14.3%), and three

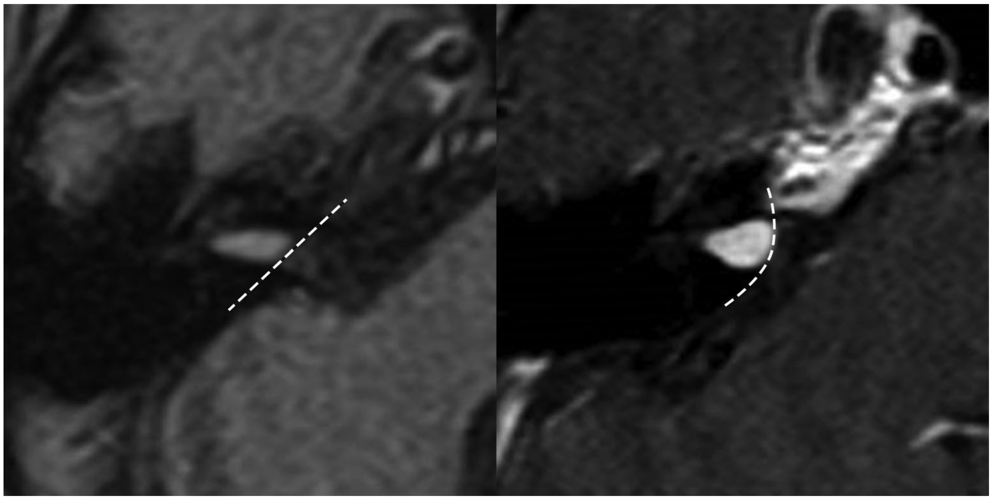

Fig. 2. Preoperative MRI of intracanalicular meningiomas (axial T_1-weighted gadolinium-enhanced image). The dashed line outlines the medial border of the tumor. Linear medial border (left) and convex medial border (right). MRI = magnetic resonance imaging; T_1= longitudinal relaxation time.

TABLE III.
MRI Characteristics of Intracanalicular Meningiomas and Intrameatal Vestibular Schwannomas.

	Convex Border	IAC Wall	Dural Tail	Linear Border
Meningioma	36.8	26.3	10.4	26.3
Vestibular schwannoma	87.2	2.0	3.4	7.4

Data presented are percentages in each subpopulation. IAC = internal auditory canal; MRI = magnetic resonance imaging.

psammomatous (10.7%). Although some symptoms were more frequent in some histopathologies, the limited number of cases of each subtype does not allow valid statistical comparisons. Anacusis was present in most histopathologies without any apparent predilection. Interestingly, all patients with preoperative facial palsy were meningothelial or angiomatous meningiomas, as were all cases with infiltrated facial and cochlear nerves. The prevalence of the different histopathologies in literature (when described) resulted in meningothelial meningioma in 76% of the cases and psammomatous in 19%.[6,8,12,16,21,24,27,30,32,35] Rare cases of transitional meningioma have also been published.[16,24]

Follow-up had an average duration of 42 months (range 1–209 months).

Facial nerve function was repeatedly evaluated after surgery (Table IV). In the immediate postoperative days, nearly 40% of the patients presented a grade VI facial nerve, whereas at the last follow-up the majority of the operated patients presented a functional facial nerve (grades I–III). Only one patient had a grade IV facial nerve palsy and one a grade V facial palsy. This last patient presented facial palsy before surgery and intraoperative evidence of infiltration of the facial nerve by the tumor, which was sacrificed and repaired by sural nerve graft. The other two patients who underwent facial nerve reconstruction resulted in grade III. One patient resulted in a grade IV facial nerve function, with no intraoperative evidence of facial infiltration.

Ten of the 12 patients who suffered preoperatively from tinnitus ceased to refer tinnitus after surgery.

The prevalence of the symptoms of this series of ICMs was compared to information of histopathologically confirmed IMVSs treated at our center (Table II). IMVSs presented in general better hearing, although not statistically significant (PTA 50.1 ± 21 vs. 42.7 ± 20 in ICMs). WRS were significantly worse in ICMs than IMVSs (43.6% ± 46.4% vs. 80.5% ± 33.8%, respectively, $P = .0075$, unpaired t test). Preoperative anacusis was present in less than 8% of IMVS. Moreover, it was significantly more associated with meningiomas ($P = .0037$), with a higher risk of presenting anacusis, because meningioma presented 4 times the risk of anacusis in case of an IMVS (OR 4.137, 95% CI: 1.550–10.30). On the other hand, SSNHL, tinnitus, or vestibular impairment were not significantly more present in any of the two populations. Postoperative hearing outcomes were not compared because they were largely affected by the surgical approach and due to the limited number of hearing preserving techniques employed in ICMs (5 ICM cases, 18%, vs. 45 IMVSs, 30%). The preoperative and postoperative facial nerve functions of ICMs and IMVSs are summarized in Table IV. The facial nerve was significantly more affected in meningiomas (both preoperatively: $P = .001$, as well as in the immediate postoperative evaluation: $P = .0008$, and the final follow-up: $P < .0001$). The risk of a meningioma of presenting preoperative facial palsy was about 16 times the risk of an IMVS (OR 16.1, 95% CI: 3.080–82.90), whereas the risk of having a facial palsy postoperatively (last follow-up) was more than four times the risk of it happening in case of an IMVS (OR 4.241, 95% CI: 1.701–9.825).

TABLE IV.
Facial Nerve According to House-Brackmann Grading.

	I		II		III		IV		V		VI	
	VS	M	VS	M	VS	M	VS	M	VS	M	VS	M
Preoperative	149 (98.7)	23 (82.1)	0 (0)	1 (3.6)	1 (0.7)	4 (14.3)	1 (0.7)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Immediate postoperative	95 (62.9)	8 (28.6)	15 (9.9)	3 (10.7)	8 (5.3)	3 (10.7)	4 (2.6)	2 (7.1)	4 (2.6)	1 (3.6)	25 (16.6)	11 (39.3)
Last FU	116 (76.8)	10 (35.7)	12 (7.9)	4 (14.3)	17 (11.3)	12 (42.9)	2 (1.3)	1 (3.6)	1 (0.7)	1 (3.6)	3 (2.0)	0 (0)

Data presented are the number of patients (percentage in each subpopulation) with intrameatal vestibular schwannomas and intracanalicular meningiomas in three different evaluation times: preoperative, in the immediate postoperative period, and at the last follow-up. FU = follow-up; M = meningioma; VS = vestibular schwannoma.

DISCUSSION

To the best of our knowledge, this is the largest series of ICMs, an entity reaching approximately 100 described cases (including the cases of this article). As previously mentioned, the diagnosis is often completed only after surgery because of the clinical and radiological similarities to VSs. Meningiomas tend to be isointense on T_1-Weighted MRI with strong gadolinium-enhancement, just as VSs, and they tend to be isointense on T_2-Weighted MRI.[42] A recent study suggests that the perilymphatic signal can improve diagnosis,[43] whereas the typical sign of increased thickness of the dura, which appears as a dural tail, was not statistically more present in meningiomas when compared to IMVSs. According to other authors, the borders of meningiomas tend to be more ragged,[8] whereas in this series a linear border was statistically more associated to ICMs as opposed to IMVS. After a re-evaluation of preoperative MRIs, up to 61% of our cases could have been suspected to be meningiomas due to a dural tail, a linear morphology along the bony wall or linear borders of the tumor.

The increased trend to wait and scan or treat with radiosurgery has certainly increased missed diagnoses in the last decade. Intrameatal tumors notably constitute the majority of the patients enrolled in a watchful waiting follow-up because they have the least tendency to grow.[40] We suspect that there are more patients affected by meningioma among the 1,245 patients, with suspected VS enrolled in our wait-and-scan protocol. Moreover, many characteristics typical of meningiomas can be evidenced only intraoperatively, and diagnosis is only possible with histopathological evaluation. In a study on radiosurgery of suspected IAC meningiomas, 63% decreased in size, whereas the rest remained unchanged.[44] On the other hand, in spite of its aggressiveness, surgery confers an 100% tumor removal. In this series, all of the patients had a Simpson I or II tumor exeresis, and none of the patients developed tumor recurrence.

Hearing Function

Preoperative anacusis was by far more associated to ICMs than IMVSs; the latter also presented in general better hearing.

In comparison to the literature, this study includes a significantly elevated number of patients with compromised hearing (86% vs. 52% patients, $P = .0001$, Fisher exact test) and anacusis (32.1% vs. 10%, $P = .0064$, Fisher exact test).[6–8,11–36] The other symptoms had a similar prevalence to the literature. Hearing loss is probably secondary to compression because nerve infiltration is rarely encountered. In our series, the cochlear nerve was infiltrated in two cases, whereas it was engulfed by the tumor in another two cases.

In this series, TLA was the predominant approach, which is concordant to the substantial frequency of anacusis and unserviceable HL. In literature, hearing preserving techniques were used in more than 50% of the patients, despite many patients having suffered from preoperative anacusis or unserviceable HL. Among the remaining patients with preoperative class A to C hearing, fewer than 63% maintained hearing.[8,16,18,20,22–24,27,28,30,33,34] It is thus imperative to decide the appropriate approach, considering the importance of an adequate tumor removal over the confined possibilities of hearing preservation with surgery. Moreover, most of the patients (10 of 12) ceased to complain tinnitus after surgery. Although we are not able to explain the underlying mechanism, given the preoperative negative association between anacusis and tinnitus, we may suppose that the postoperative sensory loss affected also tinnitus.

Radiosurgery can be considered a hearing preserving technique, although hearing continues to be affected by radiosurgery after years of its use. According to Pollock et al., hearing preservation in IAC meningiomas treated with radiosurgery decreased from 93% in the first year to 42% after 5 years.[44] According to another larger series, however, with a review of the literature on gamma knife radiosurgery for CPA meningiomas with IAC invasion, hearing deteriorates in only 11% of the patients, although at the expense of tumor control because tumor shrinking is occurs in less than 40% of the reported cases.[45]

Facial Nerve Function

Facial nerve symptomatology occurred in four patients (14.3%) from the beginning and eventually affected five patients (17.9%) by the time of surgery. Intraoperatively, however, only three cases revealed facial nerve infiltration. The facial nerve was affected in ICMs to a further extent if compared to IMVSs. Preoperative facial palsy occurred more frequently in ICMs.

Moreover, the majority of IMVS presented a grade I facial nerve at the last follow-up, whereas the majority of ICMs presented a grade III. An important fact that emerged is the frequent immediate postoperative grade VI facial palsy in case of ICMs, which could imply a more adherent tumor or the need for more aggressive intraoperative maneuvers in meningiomas versus VS.[46]

In the cases reviewed, where facial postoperative function was described, more than 50% presented a "normal" facial nerve.[6–8,11–12,15–27,31,33–34,36] Some authors opted for a subtotal tumor removal in order to preserve the facial nerve.[14,15,22,27] Among the cases reported in literature, 12.2% presented a transient facial palsy[18,21,23,27,33,36] and 26.5% a permanent facial palsy.[7,11,13,20,22,24,25,31,33] However, an actual comparison between our results and the cases reviewed would be very difficult because most articles either present a limited number of patients or do not use a facial palsy classification. Breuer et al. describe nine ICMs operated by an EMFA resulting in 100% of the cases with a grade I to II facial nerve function; however, two of these tumors were partially resected, and none of their cases presented preoperative facial palsy.[24] Peyre et al. describe 12 meatal and ICMs (one was affected by preoperative grade III facial palsy) who underwent both hearing preserving techniques and TLA. 16.7% of the cases reported in the article resulted in a grade IV or VI facial nerve.[33]

CONCLUSION

Preoperative assessment should include a more attentive evaluation of imaging, especially concerning tumor morphology and position, which should ease preoperative diagnosis and allow a more adapt patient counseling. The particularity of meningiomas, although benign tumors, to invade bone and dura necessitates a wide excision and adequate exposure in order to ensure a total removal, which can only be achieved by surgical intervention. TLA allows an excellent exposure and control of the IAC, and therefore limits residues or recurrences.[47]

Meningiomas differ from other IAC tumors, because of their increased aggressiveness (anacusis and facial palsy), in addition to the frequent postoperative facial palsy and difficulty to preserve hearing. A wait-and-scan protocol should be favored in asymptomatic or stable meningiomas.

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