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Facial Nerve Tumors

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Objective: To evaluate the surgical results in primary facial nerve (FN) tumors.
Study Design: Retrospective case review.
Setting: Private neuro-otological and skull-base tertiary referral center.
Patients: Twenty eight consecutive patients affected by primary FN tumors that underwent surgery between December 1990 and February 2001.
Interventions: The lesions were removed through a variety of surgical approaches, depending on tumor location and extension, as well as preoperative hearing. In one case, partial removal was performed.
Main Outcome Measures: Preoperative and postoperative FN function; preoperative and postoperative hearing level; and postoperative complications.
Results: Based on histologic examination, tumors were distributed as follows: 18 schwannomas, six hemangiomas, two meningiomas, and two neurofibromas. Tumor location varied, with lesions distributed along the entire length of the nerve. Facial dysfunction was the most frequently recorded symptom, followed by hearing loss. Only five patients presented a preoperative grade 1 facial function. In the remaining patients of the group, the facial deficit lasted from 2 to 120 months, with a mean of 31.2 months. Anatomic integrity of the nerve was preserved in 4 cases; all others required a nerve interruption followed by reconstruction using a sural nerve graft. The complications recorded were: one cerebrospinal fluid leak, one postoperative retraction pocket, and one external auditory canal wall resorption requiring a surgical revision. Preoperative hearing remained unchanged in 8 out of the 15 patients in whom a hearing preservation procedure was attempted. In 25 cases, a follow-up of equal to or longer than 1 year was available, with the FN functions: two grade 1, eight grade 3, nine grade 4, three grade 5, and three grade 6. Patients with a preoperative deficit lasting more than 1 year demonstrated the worst recovery.
Conclusions: Primary FN tumors are rare lesions that include different histologic types. FN deficit represents the most common symptom, but it is not present in all cases. A conservative strategy is often adopted in presence of a normal preoperative facial function. When surgical management is selected, the decision on surgical approach to use depends on tumor size and location, as well as on preoperative hearing. FN integrity may be spared in rare occasions, but more frequently nerve reconstruction is required. Final facial function recovery is mainly dependent on the preoperative presence of FN deficit and its duration. **Key Words:** Facial nerve—Schwannoma—Hemangioma—Meningioma—Neurofibroma.
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Facial nerve (FN) tumors are rare lesions that can involve any nerve segment. Facial paresis represents the most frequent symptom, followed-up by hearing loss. The rare cases that present themselves with normal facial function are much more likely to be misdiagnosed. In any case, once a FN tumor has been correctly diagnosed, deciding what may be the best treatment can sometimes be very demanding. In fact, in most cases requiring surgical removal, the nerve must be sacrificed and, until now, no reconstruction method has been able to guarantee facial function recovery better than a grade 3, according to the House-Brackmann scale (1). The present study reports the experience of the Gruppo Otologico, Piacenza-Roma, Italy, in their surgical treatment of FN tumors.

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MATERIALS AND METHODS

All the charts of the patients affected by FN tumors that were surgically treated at the Gruppo Otologico between December 1990 and February 2001 were retrospectively analyzed. All patients were operated on by the senior author (M.S.). Twenty-eight cases were included in the study. Some of these cases had already been included in other publications (2–4). One additional case, which was diagnosed in the same period, has been conservatively managed through a "wait and see" policy and has been excluded from the current study. Three patients affected by residual tumors had already undergone surgery in other centers; all of them showed a FN deficit. One patient was affected by neurofibromatosis type 2. Preoperative and postoperative FN functions were directly evaluated by one of the physicians of the Gruppo Otologico and, in most cases, documented photographically. No questionnaires or telephone inter-

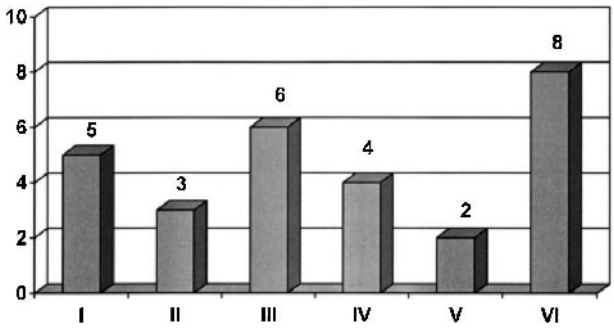

FIG. 1. Preoperative facial nerve function in the 28 cases included in the study.

views were performed. FN function was graded according to the House-Brackmann scale. The repaired facial nerve recovery scale (5) (proposed as reference in FN reconstruction) was not adopted in this study, mainly because most publications still refer to the House-Brackmann scale when reporting results, and so it remains the most suitable for comparing results. In postoperative FN function evaluation, only patients with at least 1 year of follow-up were included. Hearing data were reported as a mean air threshold for the four main frequencies (500–4,000 Hz). Hearing was considered unchanged when postoperative threshold differed between ±10 dB from its preoperative value.

RESULTS

The group included 18 male and 10 female patients, with a mean age of 40.3 years (range: 2–63 yr). Left and right side were involved in 12 and 16 patients, respectively. From a histologic point of view, the lesions were represented by 18 schwannomas, six hemangiomas, two meningiomas, and two neurofibromas. Twenty-three patients presented with FN weakness, 13 with hearing loss, five with tinnitus, and four with hemifacial spasm. On the whole, preoperative FN dysfunction (e.g., weakness and/or hemifacial spasm) was present in 24 out of 28 cases (85.7%). Preoperatively, five cases had normal facial function, 15 presented a partial deficit, whereas eight had complete palsy. The three cases that had already undergone surgery in other centers showed, respectively, a grade 4, 5, and 6. Figure 1 lists the preoperative facial function in all patients. Among the 23 cases affected by a preoperative paresis or palsy, the mean duration of the deficit was 31.2 months, ranging between 2 and 120. FN deficit lasted for more than 1 year in 13 of these cases. Thirteen patients were affected by hearing loss caused by tumor: seven demonstrated a conductive type and three sensorineural, whereas the remaining three cases showed a preoperative dead ear. Two completely asymptomatic cases were discovered accidentally: the first during a canal wall down tympanoplasty for cholesteatoma, the second through a magnetic resonance (MRI) performed for unrelated reasons. Micro-otoscopic evaluation showed a polyp occluding the external auditory canal (EAC) in three cases, whereas a retrotympanic mass was identified in a further six cases. Histologically, these nine cases were represented by eight schwannomas and one meningioma.

Lesions were localized along the entire length of the nerve with a predilection for the geniculate ganglion (GG) and the tympanic portion, each having been invaded in 21 cases. The percentage of involvement of each segment is shown in Figure 2. Most tumors involved multiple nerve segments (23/28); among them, 18 invaded at least three portions. Interestingly, the segments of the nerve running into the cerebellopontine angle or in the parotid gland were never solely affected, but always together with adjacent portions of the nerve. Middle cranial fossa extension was recorded in only one case.

Total removal was performed in all but one case. The partial removal was reserved for an elderly woman affected by a large middle cranial fossa lesion in an attempt to preserve preoperative normal facial function. Many different surgical approaches were chosen according to size and location of the lesion, as well as preoperative hearing level. The middle cranial fossa (MCF) approach was most frequently adopted, alone or combined with the transmastoid route. Table 1 lists the different approaches selected in the series.

The FN was interrupted and reconstructed by means of a cable graft in 24 cases, whereas anatomic preservation was possible in only four cases: two hemangiomas of the internal auditory canal (IAC), a small schwannoma of the second portion accidentally discovered, and the previ-

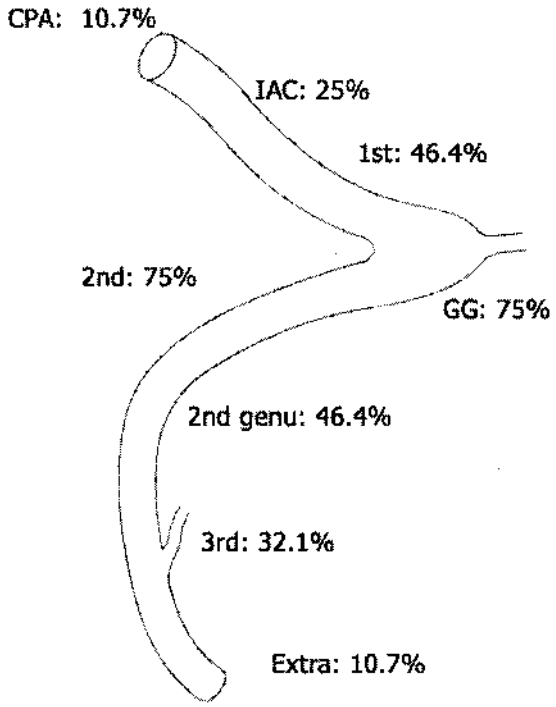

FIG. 2. Rate of involvement of each facial nerve segment. CPA, cerebellopontine angle; IAC, internal auditory canal; 1st, first portion; GG, geniculate ganglion; 2nd, second portion; 3rd, third portion; extra, extracranial portion.

TABLE 1. *Array of adopted surgical approaches*

Subtotal petrosectomy	3
Middle cranial fossa	7
Middle cranial fossa combined with transmastoid	6
Transmastoid	4
Translabyrinthine	5
Modified transchoclear	3

ously mentioned schwannoma that was partially removed. Recorded complications included one rhinoliquorrhea requiring revision surgery, a resorption of the bony posterior canal wall after a combined approach (MCF and transmastoid), and a postoperative epitympanic retraction pocket.

Two patients were lost during follow-up, one follow-up case was incomplete, and in the remaining 25 cases, follow-up was longer than 1 year. Among these, most (16/25, corresponding to 64%) showed a postoperative grade 3 or 4. Figure 3 lists the postoperative FN function of the 25 cases with complete follow-up. Postoperative grade 1 was found in only two cases in which the anatomic integrity of the nerve had been preserved. In the group of patients having undergone nerve reconstruction by means of a cable graft, the best results were witnessed in the cases having no preoperative deficit at all or a deficit that lasted less than 1 year (87.5% of grades 3 and 4 vs. 54.5%) (Fig. 4). Only two patients affected by a preoperative FN deficit for longer than 1 year recovered a grade 3 function.

Hearing was partially (preservation of the inner ear function) or completely sacrificed in 11 cases, either because of tumor location and size or significant hearing loss already present at the time of the preoperative examination. In one case, postoperative hearing was not assessable because of the patient's age (2 yr). In another case, postoperative hearing loss was related to the concomitant ossicular erosion caused by a cholesteatoma. Among the 15 patients in whom a hearing preservation procedure was attempted, one case resulted in postoperative dead ear and six cases showed deterioration in the mean air threshold. In the eight cases remaining, postoperative hearing was unchanged. To date, no recurrence has been identified.

DISCUSSION

Intrinsic FN tumors are rare lesions. Among the different pathologies, schwannomas and hemangiomas are most frequently reported in literature. In our series, schwannomas and hemangiomas accounted for a majority of the group (64.3% and 21.4%, respectively). The remaining lesions included in our series were represented by neurofibromas and meningiomas, both extremely rare at the level of the FN. Histologically, schwannomas and neurofibromas are common neural tumors, whereas it is unclear if hemangiomas should be considered true tumors or vascular malformations (6,7). Meningiomas are common intracranial lesions, but their development at the level of the GG has been reported in only a few cases (4,8).

Tumor location can be distributed along any part of the nerve's course. Our data support findings from other publications showing that multisegment tumors are much more frequent than single-segment tumors. Some lesions show a particular shape with areas of enlargement connected by segments of grossly normal nerve. These segments have been found to be affected by neoplastic cells creating a bridge between apparently independent masses (9). This was evident in some cases in our series, especially in two schwannomas involving the intraparotid nerve segment. A possible explanation for this is that the absence of or the thin consistency of the fallopian canal at the level of some segments of the FN allows the tumor to grow freely without any external resistance. However, the possibility of multicentricity of the lesion has also been proposed (10). In the international literature, the most frequently involved nerve segment is represented by the GG (11). Our data gathered from this study support this finding: both the GG and the tympanic portion appeared affected in 21 out of the 28 cases, corresponding to 75 percent. MCF involvement is particularly rare (11) (one case in our series) and is usually produced by tumors originating from the greater superficial petrosal nerve.

Although preoperative FN weakness is present in the majority of cases (82.1% in the current study), normal FN function does not automatically exclude the possible presence of a FN lesion. In our series, two of the cases with a preoperative grade 1 had some facial symptoms (a previous palsy with complete recovery and a hemifacial spasm); the remaining three cases without preoperative FN deficit were represented by a small schwannoma discovered during a tympanoplasty, a large lesion of the MCF, probably originating from the greater superficial petrosal nerve, and a small hemangioma of the IAC. Among the 23 patients affected by a preoperative FN paresis or palsy, the mean duration of the deficit was 31.2 months, with a range from 2 to 120 months. The deficit lasted less than 1 year in only eight cases. Given that today there is a vast range of diagnostic possibilities, such a long delay between the beginning of

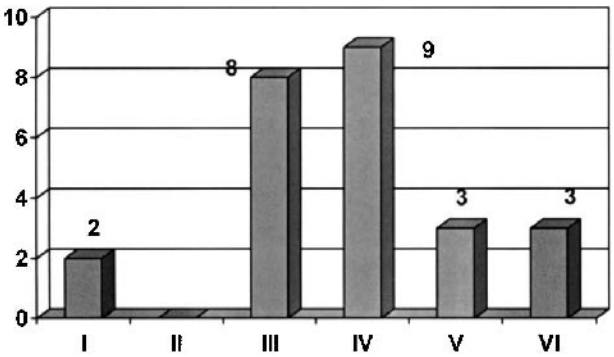

FIG. 3. Postoperative facial nerve function in the 25 cases with a follow-up of ≥1 year.

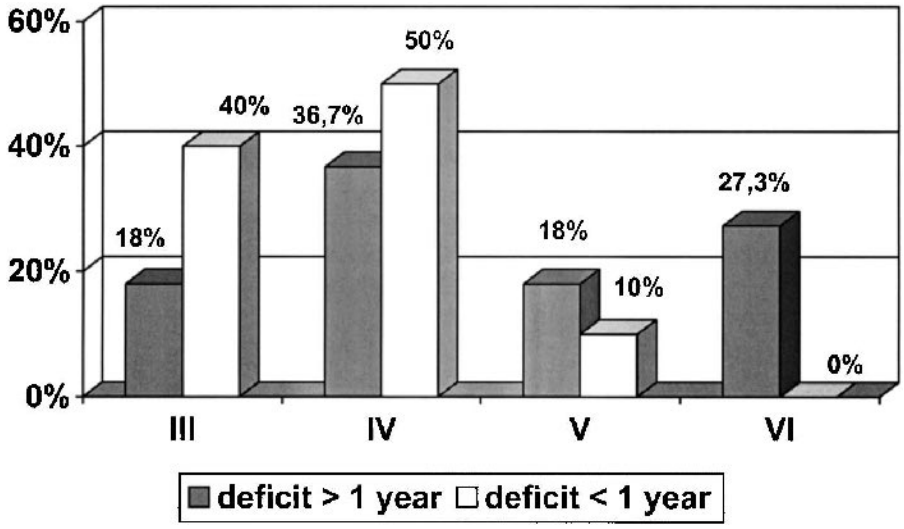

FIG. 4. Facial nerve reconstruction through cable graft interposition. Postoperative facial nerve function versus preoperative deficit duration.

symptoms and proper diagnosis is probably a result of inaccurate evaluation.

Hearing loss is also a common symptom in patients affected by FN tumors. Conductive, sensorineural, or mixed hearing loss may be present, depending on tumor location. Prevalence of conductive hearing loss in our series was correlated to which segments of the FN are most frequently affected (e.g., the GG and tympanic portion).

In cases of FN lesions, the clinical examination mainly focuses on facial function. However, it should be taken into consideration that as the FN runs relatively close to the tympanic membrane and the EAC, some lesions may be detectable by otoscopy. Our series includes six retrotympanic masses and three polyps of the EAC, corresponding to 69.2 percent of the tumors involving the second genu and/or the mastoid portion (Fig. 5). From a histologic point of view, these nine tumors were represented by eight schwannomas and one meningioma. None of the hemangiomas appeared with a mass that was otoscopically visible. This finding is consistent with the fact that these tumors produce neurologic deficits when they still are small (6).

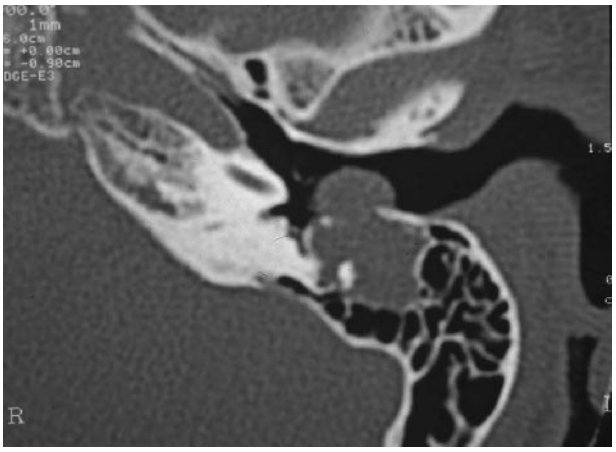

FIG. 5. Schwannoma of the vertical portion of the facial nerve protruding in the external auditory canal as a polyp.

One patient in our series had a FN paresis after an EAC polyp biopsy performed in another center. It should thus be noted that because of the risk of paresis when performing a biopsy, an informed consent mentioning the risk of a FN deficit must be signed by the patient when biopsy is needed.

When a tumor of the FN is suspected, radiologic investigations consist of MRI inclusive of the entire nerve course, from brainstem to the parotid gland. However, in an MRI,a normal nerve can appear as enhanced. A high-resolution CT of the temporal bone is complementary in the case of intratemporal lesions, because the MRI is superior in showing the tumor extension, whereas the CT provides detailed information regarding the relationship with the surrounding bony structures. Enlargement of the geniculate fossa because of anatomic variations should be kept in mind in the presence of normal FN function (12). In these cases, comparison with the contralateral side is often helpful. Differential diagnosis among the tumors on the basis of radiologic investigations can be particularly difficult. The only specific characteristic is the presence of intratumoral bony spiculaes, often visible in the case of hemangiomas (Fig. 6). However, preoperative differentiation among the various tumors that do affect FN is usually of little clinical importance, because it does not have an impact on management. In the presence of a CPA lesion, the possibility of a FN tumor should always be considered. Although in some cases vestibular and FN schwannomas are indistinguishable, sometimes an enhancement of the intralabyrinthine portion of the FN and/or the GG can lead the surgeon to the correct diagnosis.

Once a FN tumor has been diagnosed, its management is particularly demanding, especially in the presence of normal facial function. In fact, in most cases, when attempting a total tumoral resection, the involved nerve segment must be removed. This causes the need for

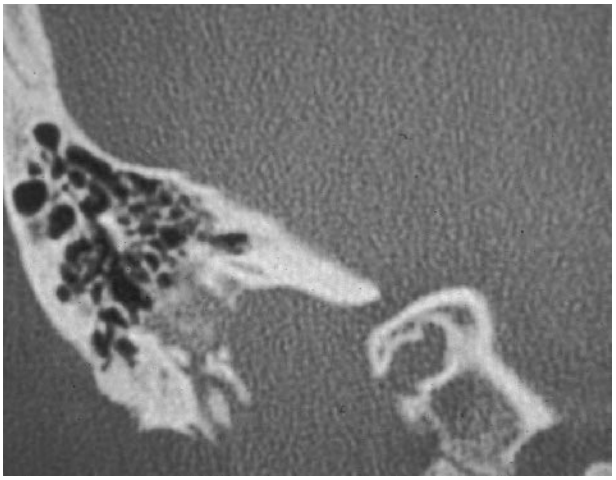

FIG. 6. Hemangioma of the vertical portion of the nerve. The characteristic intratumoral bony spiculaes are easily distinguished.

nerve reconstruction with an immediate FN palsy and leaves no possibility of function recovery better than a grade 3. Hemangiomas, extraneural lesions that originate from the blood supply (6,7), are the least likely to involve nerve sacrifice when removing the lesion. In fact, in some cases, they can be removed and nerve integrity maintained. Because postoperative FN palsy is almost inevitable in tumor removal, most surgeons have adopted a conservative strategy when confronted with a tumor with preoperative normal facial function (13). Periodic radiologic and clinical evaluation seems to be the best option for these patients. They may be able to preserve normal FN function for a long time, because of the slow-growing nature of the tumors. However, depending on tumoral location, the patients should also be informed of the risks of hearing deterioration. It should be also taken into consideration that the appearance of a FN deficit may suggest a surgical treatment, because of the fact that, according to our data, the chance of recovering satisfactory FN function decreases if the surgery is not performed in the first year after the beginning of a preoperative clinical FN deficit.

In our series, five patients with preoperative grade 1 underwent surgery. In two of these, the lesion was preoperatively considered to be a vestibular schwannoma, in one a small prolifecative tumor was discovered during a tympanoplasty whereas in another the lesion was so large that it required surgery. The last patient was a head and neck surgeon who had already experienced temporary FN palsy and decided to undergo tumor removal.

Simple tumor decompression is reported by some authors (14) and may represent another option in presence of a preoperative normal FN function. However, in our center this strategy has never been adopted.

Partial removal, accompanied by intraoperative monitoring, can be justified, theoretically, by the tendency of tumors towards prolifecative growth. The main drawback of partial removal is the difficulty of establishing where the removal should be stopped to preserve the facial function. In addition, FN tumors are usually small lesions, and that makes it difficult to justify a partial removal. In our series, this option was selected only once, because of the patient's normal facial function, advanced age, and tumor size (Fig. 7). This patient showed an immediate postoperative facial palsy with a further recovery to a grade 3, whereas the residual tumor did not show any regrowth on radiologic follow-up.

In most cases with preoperative FN deficit, total removal with FN reconstruction is normally adopted, usually by means of a cable graft. This occurred in 85.7 percent of all of our study population and in 100 percent of the cases with preoperative FN deficit. Unfortunately, FN recovery was not completely satisfactory in most patients with adequate follow-up (15/23). Naturally, these results have been influenced by the prolonged preoperative FN deficit (>1 year) in 9 out of 23 patients, with only two of them reaching a postoperative grade 3.

An array of different surgeries may be selected depending on tumor size and location, as well as preoperative hearing. The presence of cochlear erosion (Fig. 8) should always be evaluated, because in this situation hearing preservation is practically impossible. The MCF approach, alone or with the transmastoid route, was most frequently adopted in our series. Hearing preservation is a feasible target in a large number of FN tumors. In the present series, hearing was preserved in 8 out of the 15 (53.3%) cases in which a hearing preservation procedure was attempted. Sometimes, preoperative good hearing has to be sacrificed because of the particular location and/or size of tumor. In some occasions, the ossicular chain can be dismantled and successively reassembled to completely remove the lesion.

CONCLUSIONS

Even though FN tumors represent only a small percentage of all FN palsies, the possibility of their presence

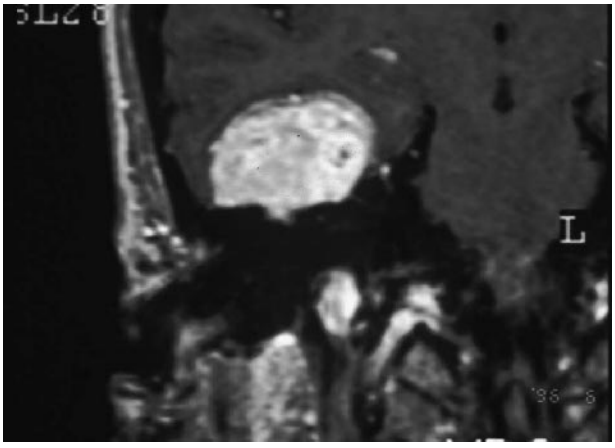

FIG. 7. Large schwannoma that developed at the level of the middle cranial fossa. The tumor was only partially removed because of patient's age.

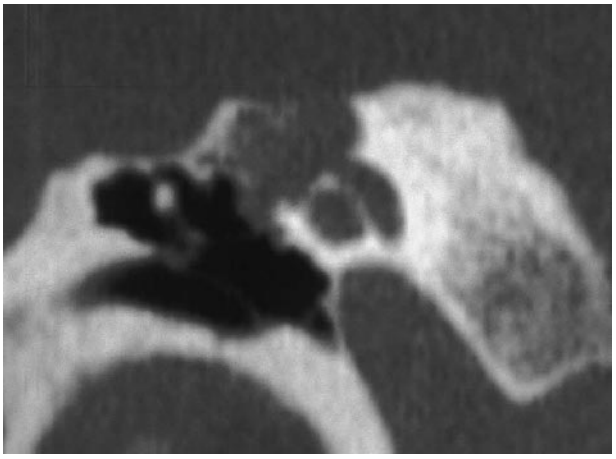

FIG. 8. Hemangioma originating at the level of the geniculate ganglion, with erosion of the cochlear turns.

should always be taken into account to ensure early diagnosis. In rare cases, a FN tumor may not produce any FN deficit. Moreover, differentiation between FN tumors involving the CPA and IAC and vestibular schwannomas is sometimes impossible. Therefore, enhancement of the intralabyrinthine portion and GG must always be carefully evaluated. Once a FN tumor has been diagnosed, different therapeutic options have to be discussed with the patient, taking into consideration preoperative FN function, tumor size and location, hearing level, and patient's age. When surgical removal is selected as treatment, each procedure must be tailored to suit each single case. Anatomical preservation of the FN is extremely rare and in most patients, FN reconstruction is required. According to our data, the chance of recovering satisfactory FN function decreases if the surgery is not performed in the first year after the beginning of a preoperative clinical FN deficit. This again emphasizes how important early diagnosis is.

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