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ARAŞTIRMA / RESEARCH ARTICLE

Facial nerve schwannomas: management and facial function results

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Fasiyal sinir schwannomları: tedavi ve fasiyal fonksiyon sonuçları

Amaç: Fasiyal sinir schwannomlarının klinik özellikleri, tanısı, tedavi seçenekleri, cerrahi yaklaşımları ve postoperatif fonksiyonel sonuçlarını değerlendirmek.

Yöntem: 1992-2003 yılları arasında, La Paz Hastanesi'nde cerrahi tedavileri gerçekleştirilen fasiyal schwannomlu hastaların dosyaları retrospektif olarak incelendi. Çalışmaya 8 olgu alındı. Hastaların hepsi için şu değişkenler kaydedildi: cerrahi girişimin olduğu tarihteki yaşı, cinsiyet, başlangıç semptomları, fasiyal paralizinin gelişimi, süresi, ameliyat öncesi ve sonrası işitme düzeyleri, tümörün yerleşim düzeyi, cerrahi yaklaşım, fasiyal onarım tekniği, göz yumma yeteneği ve takip süresi. Ameliyat öncesi ve sonrası fasiyal sinir fonksiyonları House-Brackmann skalasına göre derecelendirildi.

Bulgular: Bütün hastalarda cerrahi girişimin olduğu tarihte çeşitli derecelerde fasiyal disfonksiyon vardı ve 4 hastada fasiyal paralizi ilk semptomdu. Yedi hastada, ameliyattan önce iletim tipi ya da sensörinöral işitme kaybı söz konusuydu. Beş hastada tinnitus, 3 hastada instabilite, 3 hastada ağrı, ve bir hastada da otore saptanmıştı. Lezyonlar sinirin bütün uzunluğu boyunca yerleşim göstermekteydi. Dört olguya ameliyat öncesi doğru olarak fasiyal schwannom tanısı konmuştu. Olguların hepsinde fasiyal sinir tümör tarafından tutulmuştu ve eksize edilerek rekonstrükte edilmişti. Hastaların hepsi postoperatif III. ya da IV. fasiyal fonksiyon derecesi göstermekteydi.

Sonuç: Fasiyal sinir schwannomları fasiyal sinir paralizilerinin ayırıcı tanısında dikkate alınmalıdır. Bununla birlikte normal fasiyal motor işlev olasılığı da düşünülebilir. Farklı tedavi seçenekleri benimsenebilirse de son karar, tedavi hastaya göre kişiselleştirilerek önerilmelidir.

Anahtar Sözcükler: Fasiyal sinir schwannomu, fasiyal paralizi, cerrahi tedavi.

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Abstract

Objectives: To evaluate the clinical features, diagnosis, treatment options, surgical approaches, and postoperative functional results of facial nerve schwannomas.

Methods: The charts of patients affected by facial nerve (FN) schwannomas that were surgically treated at La Paz Hospital between 1992 and 2003 were retrospectively analyzed. Eight cases were included in the study. For all of the patients, the following variables were recorded: age at surgery, sex, initial symptoms, evolution of the facial palsy, lenght of facial palsy, preoperative and postoperative hearing levels, extent of the tumor, surgical approach, facial nerve repair technique, eye closure achievement and length of the follow-up. Preoperative and postoperative FN function was graded according to the House-Brackmann scale.

Results: All patients had some degree of facial dysfunction at the time of surgery, facial palsy being the first symptom in 4 patients. Seven patients were affected by conductive or sensorineural hearing loss before surgery. Five patients presented with tinnitus, three with instability, three with pain, and one with othorrea. Lesions were located along the entire length of the nerve. A correct preoperative diagnosis of facial schwannoma was made in 4 cases. The FN was involved by the tumor and had to be interrupted and reconstructed in all the cases. All patients showed a postoperative facial grade III or IV.

Conclusion: FN schwannomas should be considered in the differential diagnosis of facial nerve paralysis. However the possibility of normal facial motor function may be considered. As different therapeutic options may be adopted, the management must be individualized, and implication of the patient in the final decision is recommended.

Key Words: Facial nerve schwannoma, facial paralysis, surgical treatment.

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Introduction

Facial nerve (FN) schwannomas are uncommon benign lesions, and less than 500 cases were reported in the literature. They have been described to originate from all segments of the facial nerve, along the cerebellopontine angle (CPA) to the parotid gland.[1] Depending on tumor location, a variety of symptoms and signs may be produced. Progressive facial palsy represents the most frequent symptom, followed by sensorineural or conductive hearing loss, depending on tumor location.[2] The rare cases that present with normal facial motor function are much more likely to be misdiagnosed. Once a FN schwannoma has been correctly diagnosed, decision making for the best treatment options can sometimes be very demanding. In most cases requiring surgical removal, the nerve must be sacrificed and no reconstruction method has been able to guarantee a postoperative facial function recovery better than a House-Brackmann Grade III.[3] When the patient presents with a moderate or complete facial palsy, complete resection is usually recomended. However, when there is mild or no facial dysfunction, the best choice of treatment is less clear.[4] In cases of facial nerve resection and reconstruction, the duration of facial nerve paralysis and preoperative facial function have been reported to affect the postoperative results.[5]

This study reports our experience in the surgical treatment of FN schwannomas. We discuss the clinical presentation, the diagnostic tools, the surgical management, and postoperative functional results.

Materials and Methods

All the charts of patients affected by FN schwannomas that were surgically treated at La Paz Hospital between 1992 and 2003 were retrospectively analyzed. Eight cases were included in the study. For all patients, the following variables were recorded: age at surgery, sex, initial symptoms, evolution of the facial palsy, lenght of facial palsy, pre and postoperative hearing levels, extent of the tumor, surgical approach, facial repair technique, preoperative FN grade, postoperative FN grade, eye closure achievement and lenght of the follow-up. The age at the time of presentation ranged from 29 to 59 years, with a mean of 41 years. Left and right sides were involved in 3 and 5 patients respectively. Four patients were men, and the other four patients were women. Total removal of the tumor was performed in all cases. Lesions were located along the entire length of the nerve (Table 1). Different surgical approaches were chosen according to the size and location of the lesion, as well as preoperative hearing levels. The middle cranial fosa approach was performed in 2 cases, one of them combined with the transmastoid route. A transmastoid approach alone was performed in 2 cases, and a retrosigmoid approach was performed in 4 cases, one of them combined with a transmastoid route. The FN was interrupted and reconstructed in all cases, by means of a cable graft in four patients or XII-VII anastomosis in other four cases. In the nerve grafting cases, the great auricular nerve was used in three ocasions and the sural nerve was used in

Table 1. Data of 8 cases with facial nerve schwannoma.

Case no.	Age/ Sex	Initial symptoms	Facial palsy (months)	Length of FNP	Extent of tumor	Surgical approach	Facial repair	Initial grade	Final grade	Eye closure	Follow-up
1	57/F	HL	Insidious	13	Intracran	Retrosig.	Sural	II	IV	C	44
2	21/M	FNP	Insidious	13	Lab-Tymp	MCF-transmastoidoid	GAN	VI	III	C	149
3	56/M	FNP	Insidious	6	Mast-EMT	Transmast.	GAN	V	IV	C	12
4	29/F	FNP	Sudden	27	Lab-Tymp	MCF	XII-VII	VI	III	C	96
5	35/F	HL	Insidious	18	Intracran-GG	Retrosig.	XII-VII	VI	IV	C	48
6	57/F	HL	Facial spasm	41	Intracran	Retrosig. - transmastoid	XII-VII	VI	IV	C*	24
7	17/M	HL	Sudden	25	Intracran	Retrosig.	XII-VII	VI	IV	C	91
8	59/M	FNP	Sudden	1	Tymp	Transmastoid	GAN	VI	IV	C	34

no: number; **F:** female; **M:** male; **HL:** hearing loss; **FNP:** facial nerve paralysis; **CPA:** cerebellopontine angle; **Intracran:** intracranial segment; **Lab:** labyrinthine segment; **Tymp:** tympanic segment; **Mast:** mastoid segment; **EMT:** extratemporal main trunk; **GG:** geniculate ganglion; **GAN:** greater auricular nerve; **XII-VII:** hypoglossal-facial nerve anastomosis, **C:** complete. ***A gold weight implant was place in her eyelid.

just one patient. No complications were recorded in any case. The XII-VII anastomosis was performed as previously described.[6]

Preoperative and postoperative FN functions were graded according to the House-Brackmann scale.[7] In the postoperative FN functional evaluation, only patients with at least 1 year of follow-up period were included in the study. Hearing was considered unchanged when the postoperative air conduction threshold difference was in +/- 10 dB level of the preoperative value. Follow up ranged from 12 to 149 months, with a mean of 62 months.

Results

Clinical details of the patients are presented in Table 1. All patients had some degree of facial disfunction at the time of surgery, facial palsy being the initial symptom in only 4 patients. The mean duration of preoperative FN palsy was 18 months, ranging between 1 to 41 months. FN deficit lasted for more than one year in 6 cases, and for more than 2 years in 2 cases. Seven patients were affected by hearing loss before surgery; two demonstrated a conductive type, and two had sensorineural hearing loss, whereas the remaining three cases had a preoperative dead ear. Sensorineural hearing losses and dead ears were seen predominantly in tumors of the intracranial segment of the FN, whereas conductive losses were associated with involvement of the geniculate, horizontal and vertical portions of the FN. Five patients presented with tinnitus, three with instability, three with pain, and one with othorrea. Lesions were located along the entire length of the nerve (Table 1). Most tumors (7/8) involved multiple nerve segments. A correct preoperative diagnosis of facial schwannoma was made in 4 cases (Figure 1). In 4 cases the preoperative diagnosis had been vestibular schwannoma, due to the lack of FN symptoms, and

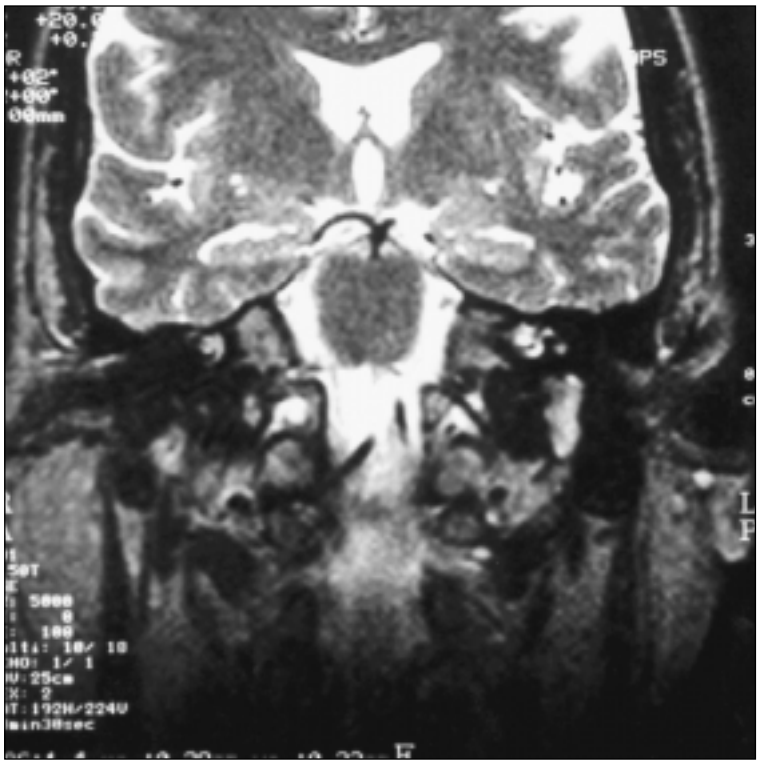

Figure 1. Coronal MRI scan showing an hyperintense and homogeneous mass affecting the mastoid segment of the FN. The diagnosis of FN shwannoma was suggested preoperatively.

therefore the FN involvement by the tumor was discovered during surgery.

All patients had a postoperative facial nerve functional result of Grade III or IV. Complete eye closure was initially achieved in 7 of the 8 cases. A gold weight implant was placed in the eyelid of a patient with an incomplete eye closure. In two patients hearing remained unchanged postoperatively, one had a sensorinerual hearing loss, and the other had a conductive loss (Table 1. Cases 3 and 8, respectively). Three patients already had a preoperative dead ear (cases 5, 6 and 7). The hearing level was improved in one patient, who had a conductive hearing loss preoperatively (case 4). Two patients suffered a dead ear as a consequence of surgery (cases 1 and 2). To date, no recurrence has been identified in any patient.

Discussion

FN schwannomas are rare lesions. Since the first report by Schmidt in 1931, less than 500 cases have been reported in the literature.[8,9] They have been estimated to be the cause of facial palsy in 5% of cases.[10] The rarity of FN tumors and the diversity of their clinical picture together with the fact that their presentation may mimic other more common pathologies, make the diagnosis of these tumors a difficult task.[5] The tumor may be located along any part of the nerve´s course.[1] Our data support findings from other publications showing that multisegment tumors are more frequent than single-segment tumors.[2,11]

There are no specific symptoms for FN schwannomas, and the diagnosis may be difficult. Symptoms may be different depending on the involved segments.[9] Although FN paralysis is the most common presenting symptom,[2] normal FN function does not automatically exclude the possibility of a FN lesion.[5] A gradual onset of facial paralysis is highly suggestive of a tumor, and a complete neurotologic investigation is warranted.[10] Moreover, it has been suggested that patients with facial palsy that does not show recovery in 6 months and patients with recurrent facial paralysis should be investigated for the presence of a tumor.[1,8]

Audiovestibular symptoms (hearing loss, tinnitus, or instability) are also common in patients with FN schwannomas,[12] hearing loss being the second most common complaint. In our study it was the initial symptom in four cases. Conductive, sensorineural, or mixed hearing losses may be present, depending on the tumor location.[7] External auditory canal mass, pain and otorrhea are the other reported symptoms.[2] When a FN tumor is suspected, a brain and temporal bone MRI examination with gadolinium enhacement must be performed depicting the entire course of the nerve, from the brainstem to the parotid gland. Difficulties may arise differentiating FN schwannomas from other tumors of the IAC and CPA, most notably vestibular schwannomas. In many cases, vestibular and FN schwannomas are indistinguishable.[13] In the presence of a CPA lesion, the possibility of a FN tumor should always be considered. There are no specific clinical or radiological features to differentiate a CPA vestibular schwannoma from a CPA facial schwannoma. The presence of a FN dysfunction in a patient suspicious for a vestibular schwannoma, should arise the possibility of a FN tumor, especially if it is a small lesion.[14] Apart from the few cases in which tumor extension to the geniculate ganglion may establish the diagnosis, most patients with CPA FN schwannomas are diagnosed intraoperatively.[15] That was the case in four patients of our study (cases 1, 5, 6 and 7), in which MRI suggested preoperatively that the lesion was a vestibular schwannoma (Figure 2).[16]

Depending on the involved segment; chronic otitis media with cholesteatoma, dermoid cysts, and benign parotid masses must also be taken into consideration for the differential diagnosis of FN schwannomas.[12,17]

Once a FN tumor has been diagnosed, its management is particularly demanding, especially in the presence of a normal or mild facial motor dysfunction. In most cases, when attempting a total tumoral resection, the involved nerve segment must be removed. This causes an immediate FN palsy and the need for nerve reconstruction, leaving no possibility of a functional recovery better than Grade III.[3]

Because postoperative FN palsy is almost inevitable following resection of FN tumors, most surgeons adopt

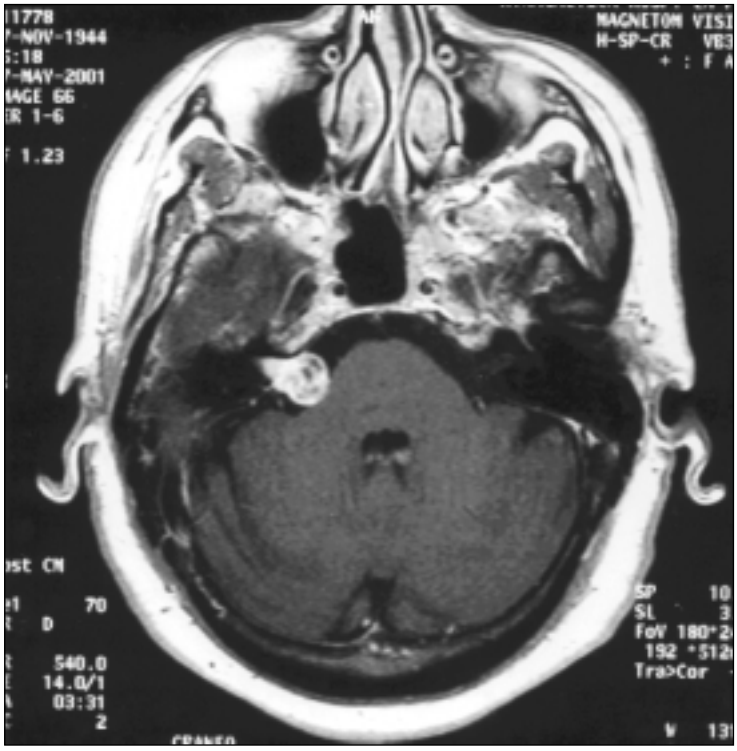

Figure 2. Axial MRI scan showing a 12x20 mm CPA mass with intracanalicular extension. The mass suggested a vestibular schwannoma. FN was identified intraoperatively as the origin of the tumor.

a conservative strategy when confronted with a patient with a preoperative normal function.[4] A follow up strategy with serial MRI and hearing tests may be considered in patients with no, or mild preoperative facial nerve dysfunction.[18] It may be able to preserve a normal FN function for a long time in these patients, because of the slow growing nature of these tumors, and surgery may be delayed until the facial function deteriorates to at least a HB Grade III.[2,4] However, depending on tumor location, the patient should also be informed concerning the risks of progressive hearing loss.[3]

Simple meatal decompression is reported by some authors[19] and may represent another option in the presence of preoperative normal FN function. Angeli and Brackmann[20] reported that wide decompression is an alternative to surgical excision, especially in cases of unsuspected facial schwannoma in patients with normal facial function. They reported 4 patients treated with decompression only, who achieved postoperative HB grade I and II facial nerve functions. Other authors have also reported partial removal of the schwannoma for patients with normal facial function. The main draw–back of partial removal is the difficulty of establishing where the removal should be stopped to preserve the facial function.[3,21]

The age at presentation is another factor to be considered. If the patient is young, early surgical resection is usually recommended, as these tumors tend to grow causing subsequent intracranial or extratemporal extensions, making the future approach more difficult and postoperative complications more likely. Moreover, tumor growth causes progressive degeneration and regeneration of the facial nerve fibers leading to scarring of the distal part of the nerve with consequent poor recovery of facial function following reconstruction.[22] On the other hand, in an elderly patient with no, or mild facial nerve paresis, facial nerve decompression may be sufficient, if surgery is to be performed.[5]

In all the schwannoma cases of our study, total tumor removal with FN reconstruction was performed. Interpositional nerve grafting is the most preferred reconstructive treatment.[23] Hypoglossal-facial nerve anastomosis is used in those cases in which there is an inadequate nerve stump remaining at the brainstem.[24] There is no advantage for any particular type of reconstruction, with the best recovery being HB grade III function.[3] Some authors have argued against the utility of the House-Brackmann scale after a FN reconstruction; because in most cases, the forehead movement which is required for a House-Brackmann Grade III is not recovered. Therefore, other classification scales have been proposed.[25] Unfortunately, the majority of publications still refer to the House-Brackmann scale in reporting results, and therefore it remains the most valid way to compare results.

FN recovery was satisfactory in most of our patients, with facial symmetry at rest in all of them and complete eye closure in 7 out of the 8 patients. A gold weight implant was placed on the affected side's eyelid of the patient with an incomplete eye closure.

As the main factors influencing postoperative recovery are the preoperative facial nerve function and the duration of the preoperative FN deficit, early diagnosis and proper timing of surgery are fundamental to increase the chances of a good recovery.[5,21] In our series the only patient with an incomplete postoperative eye closure had the longest period of preoperative FN deficit (41 months). Recognition of the preoperative deficit duration as the main prognostic factor has important implications in patient counseling. In fact, in particular situations, such as a FN schwannoma with normal facial function, the surgeon and the patient should be aware that, if prolonged for more than one year after beginning of clinical FN disfunction, a conservative policy may have a negative influence on the final surgical result.[26]

In conclusion, FN schwannomas should be considered in the differential diagnosis of facial nerve disfunction. Different therapeutic options may be adopted, taking into consideration preoperative FN function, tumor size and location, hearing level and patient's age. In cases in which surgery is selected as the treatment of choice, anatomical preservation of the FN is very rare. This leads to nerve reconstruction that will give no possibility of function recovery better than a grade III. The chance of recovering satisfactory FN function increases if the surgery is performed within the first year after the beginning of the FN deficit. Therefore, the management of a patient with a facial nerve schwannoma must be individualized and the patient must receive complete information about the nature of his/her pathology and the implication of the different treatment strategies.

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Conflict of interest statement:
No conflicts declared.

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