Nonvascular Lesions of the Jugular Foramen: The Gruppo Otologico Experience

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ABSTRACT

Tumors other than paragangliomas in the jugular foramen are uncommon. Of these, schwannomas and meningiomas predominate. Little clinical data are available in the literature on these tumors at this site. The purpose of this article is to review our experience at the Gruppo Otologico of the management of these tumors. A retrospective series is presented of 32 consecutive patients affected by jugular foramen schwannomas and meningiomas in which their clinical and radiological signs, together with surgical techniques and outcomes, were reviewed. A single-stage resection was possible for the majority of patients when the petro-occipital transsigmoid (POTS) approach was used. This allowed resection of both intra- and extradural components of the tumor with hearing preservation and avoidance of facial nerve transposition. No deaths occurred. Lower cranial nerve palsies constituted the major cause of morbidity, but none of the patients required an adjunctive procedure such as vocal cord medialization, tracheostomy, or percutaneous gastrostomy.

KEYWORDS: Jugular foramen, meningioma, schwannoma, skull base, POTS approach

Tumors arising in the region of the jugular foramen (JF) are rare and pose a challenge for clinicians from both the diagnostic and therapeutic points of view. The most frequent tumors are jugular paragangliomas, schwannomas of the lower cranial nerves (LCNs) (jugular foramen schwannomas [JFSs]), and meningiomas (jugular foramen meningiomas [JFMs]). Rarely, other primary malignant or benign tumors may be encountered in this anatomical region, for example, squamous cell carcinomas, chondrosarcomas, and chordomas.[1]

Schwannomas of LCNs IX, X, and XI represent ~3 to 4% of all intracranial schwannomas, and to date ~200 cases have been reported in literature.[2,3] Similarly, < 100 cases of JFMs have been documented.[4–11] Most of the published articles are of solitary case reports or very small series. It has been hypothesized that JFSs may develop

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from the ganglia of the IX and X cranial nerves (CNs) that are situated close to the JF; the exact nerve of origin remains, for the most part, unknown.[12] Jugular foramen meningiomas are thought to arise from the arachnoid-lining cells of the jugular bulb within the jugular fossa.[13] From the JF, tumors follow the path of least resistance and may invade the posterior fossa superiorly, the parapharyngeal space inferiorly and spread to the skull base around the JF area. Thus, dumbbell-shaped tumors have both intra- and extracranial extensions with a "passing through" component in the JF.

The differential diagnosis of JF neoplasms is made on the basis of the imaging characteristics assessed by high-resolution computed tomography (HRCT), magnetic resonance imaging (MRI), and four-vessel digital subtraction angiography (DSA).[14,15]

Regardless of type, JF tumors present with similar symptoms and signs. Deficits of cranial nerves IX-XII and hearing disturbances are common. By contrast, facial nerve dysfunction is rarely encountered at an early stage.

The ideal treatment of these tumors is total surgical removal. However, the complexity of the tumor location increases the risk of both intra- and postoperative complications, and acquisition of additional neurological deficits. Radical removal is more difficult with meningiomas because of their highly infiltrative nature and intimate relationship to the LCNs.

The infratemporal fossa type A (IFT-A) approach, introduced by Fisch in 1978,[16] allows the surgeon to control the JF area and surrounding structures but necessitates sacrifice of the middle ear structures and anterior transposition of the facial nerve. In our experience, this is the best approach for the resection of more vascular tumors such as JF paragangliomas.[17] When dealing with nonvascular tumors (i.e., JFSs and JFMs) with normal facial nerve function and hearing, a more conservative approach may be used. In such cases, the approach of choice at the Gruppo Otologico is the petro-occipital transsigmoid (POTS) approach. This approach entails a suboccipital craniectomy combined with a retrolabyrinthine petrosectomy and dissection of the retrofacial and infralabyrinthine air cells, partial drilling of the occipital condyle, and transection of the sigmoid sinus. The POTS approach offers the surgeon direct exposure of the JF area while preserving middle and inner ear function without the need for facial nerve transposition[18–20] (Fig. 1).

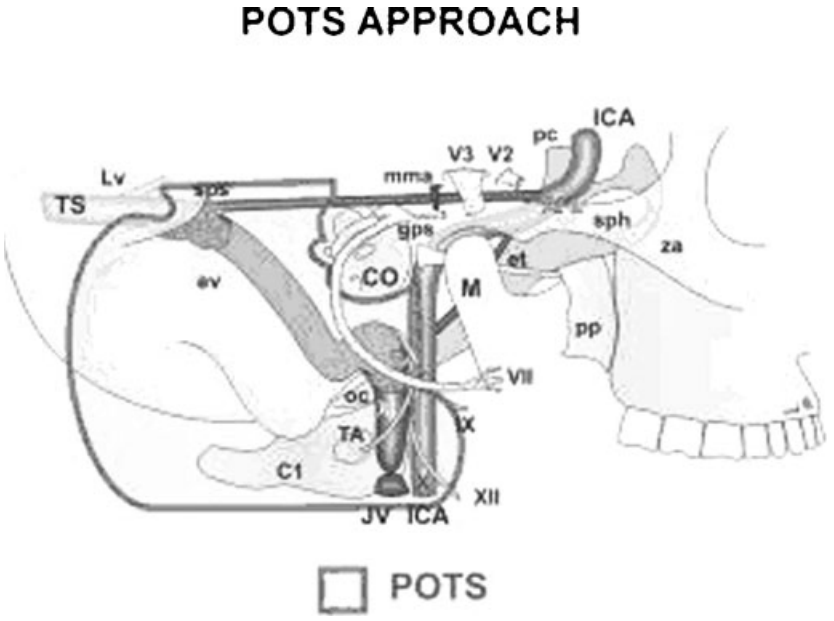

Figure 1 Schematic representation of the petro-occipital trans-sigmoid (POTS) approach.

In this article we present a series of 32 patients managed at our institution for primary nonvascular JF lesions. Clinical manifestations, radiological features, various operative techniques, the pros and cons, and the resultant outcomes of these cases are discussed.

PATIENTS AND METHODS

The charts of all patients with primary tumors arising from the JF that were managed at our institution from January 1988 to January 2005 were reviewed. Patients with jugular paragangliomas were excluded from this series. The data were reviewed specifically for age, sex, presenting symptoms, operative procedures, location and extent of the tumors, postoperative outcomes, and follow-up findings.

Clinical assessment included a complete head and neck examination, neurotological and audiological evaluation and imaging both before and after surgery. Pure tone average was calculated as the mean of 500, 1000, 2000, and 4000 Hz thresholds. Facial nerve function was graded according to the House-Brackmann (HB) scale.[21]

All patients had gadolinium-enhanced MRI of the head and neck, HRCT of the skull base with bone windows was performed in 25 patients, magnetic resonance angiography (MRA) was undertaken in 18 cases, and four-vessel DSA was performed in 10 patients. None of the patients underwent preoperative tumor embolization.

In patients with meningioma, the extent of tumor resection was classified according to Simpson's grading, for which gross total resections correlated to grade I or II and subtotal resections to grade III or IV.[22]

Follow-up consisted of clinical evaluation, hearing tests, flexible fiber optic laryngoscopy, and serial computed tomography (CT) and/or MRI scans. The first postoperative MRI was performed 6 months after surgery and then annually.

RESULTS

Demographic

The study group consisted of 32 patients. Twelve patients had JFM and 20 patients had JFS. All but two patients(one JFM and one JFS) underwent surgical resection of their tumors. These two patients were managed by "watchful waiting" with regular clinical and radiological follow-up because of their advanced age (> 70 years) and absence of symptoms. There were 21 (65.6%) females and 11 (34.4%) males. The mean age was 42.5 years (range, 21 to 77 years). Four patients (all affected by JFS) presented with recurrences following previous tumor removal performed elsewhere by a suboccipital approach. No patient had neurofibromatosis type 2.

Clinical Manifestations

The main presenting features of patients affected by JFS and JFM are detailed in Table 1. The majority of the patients presented with two or more signs and/or symptoms. Dysphonia (in 14 patients [43.7%]), dysphagia (in 12 [37.5%]), and shoulder weakness (i.e., dysfunction of the LCNs) (in 7 [21.8%]) were the presenting symptoms. The first presenting symptoms included hearing loss in 12 patients (37.5%) and tinnitus in 8 patients (25%). Chronic imbalance or true vertigo was the initial symptom in 12 patients (37.5%). Wasting of the tongue was present in six patients (18.7%). Two patients with JFS were found to have a retrotympanic mass, and one patient with JFM had a polypoid lesion in the external auditory canal.

The period of postoperative follow-up ranged from 7 months to16 years (mean, 24.2 months). Every patient underwent the first postoperative MRI 6 months after surgery. The patients who were managed conservatively had interval clinical and audiological evaluation together with MRI on a 6-month basis and showed no evidence of enlargement of their tumors after 2 and 4 years of follow-up.

Table 1 Clinical Features in Patients Affected by Jugular Foramen Meningiomas and Schwannomas

Presenting Signs/Symptoms	Total ($n=32$)	JFM ($n=12$)	JFS ($n=20$)
Hearing loss	12 (37.5%)	6 (50%)	6 (30%)
Tinnitus	8 (25%)	4 (33.3%)	4 (20%)
Dysphonia	13 (43.7%)	5 (41.6%)	8 (40%)
Dysphagia	11 (37.5%)	4 (33.3%)	7 (35%)
Shoulder weakness	7 (21.8%)	2 (16.6%)	5 (25%)
Middle ear mass	3 (9.3%)	1 (8.3%)	2 (10%)
Vertigo/instability	12 (37.5%)	7 (58.3%)	5 (25%)
Glossal atrophy	6 (18.7%)	3 (25%)	3 (15%)

JFM, jugular foramen meningioma; JFS, jugular foramen schwannoma.

Radiology Findings

On MRI, JFMs were iso- or hypointense tumors on T1-weighted images and intermediate signal on T2-weighted sequences with homogeneous enhancement after contrast administration. "Dural tails" were present on nine scans (75%). The six patients who underwent HRCT scan of the skull base and temporal bone had evidence of irregular erosion of the bony margin of the JF with characteristic mixed permeative-sclerotic appearance; signs of hyperostosis and intratumoral calcification were present in one case.

The JFSs appeared as smooth-marginated, round, or lobular masses and were iso- or hypointense on T1-weighted images and iso- or hyperintense on T2-weighted sequences with enhancement after gadolinium. In this series, none showed widening or erosion of the internal auditory canal. On HRCT, JFSs showed regular erosion of the bony margins of the JF without infiltration. Both tumor types appeared dumbbell-shaped when both intra- and extracranial spread occurred.

Tumor Location and Extent

The location and extent of the tumors were determined on the basis of the radiological appearances in all 32 cases and confirmed by intraoperative findings in the 30 cases that underwent surgery. The frequency, location, and extension of the tumors were as follows: JF in 32 patients (100%), posterior fossa/cerebellopontine angle in 21 (65.6%), upper neck in 18 (56.2%), middle ear in 3 (9.3%), and cavernous sinus in 1 (3.1%). Intracranial tumor size varied from 2 to 7 cm (mean, 3.57 cm). Nearly all the tumors had both intra- and extradural extension, with only one case of JFM being completely extradural.

Treatment

Of the 30 patients that underwent surgical removal, four were recurrences of JFS previously operated on elsewhere by the suboccipital approach. All the surgical procedures were performed by the senior author (M.S.). For practical purposes, it is useful to distinguish between the surgical approaches used for schwannomas and meningiomas.

JUGULAR FORAMEN SCHWANNOMAS

Initially, it was our practice for JFSs to use the IFT-A approach, which we did for three patients. Additional resection of the cochlea was performed in two other patients because of tumor invasion. The surgery was staged in these two patients, the second stage being removal of the intradural component by an enlarged translabyrinthine approach (ETLA). A POTS approach was used for one patient. In this latter case, a first-stage combined transcervical-subtotal petrosectomy approach was undertaken because of the patient's large neck (13 cm in diameter); at the second stage of the procedure 3 months later,

the intradural component was removed through a POTS approach.

From 1994 onward, our team adopted the POTS approach, which was used for 14 patients. Single-stage tumor removal was accomplished in all but one patient using the POTS approach. This approach alone provided sufficient access in 12 cases; however, two patients needed to have the POTS approach combined with a labyrinthine destructive procedure (a transotic [TO] approach in one patient and ETLA in the other). Both of these patients had extensive erosion of the carotid canal and no preoperative auditory function. Total tumor removal was achieved in all but one patient, who is currently awaiting a second stage of surgery to remove the intracranial component. No deaths occurred in the JFS patient group. No patient has developed a recurrence during the follow-up period.

JUGULAR FORAMEN MENINGIOMAS

The POTS approach was used in three patients with JFMs, and the IFT-A approach, combined with an ETLA, was used in one patient. POTS combined with labyrinthine destructive procedures was performed in three cases (two with TO and one with ETLA) because of a preoperatively dead ear in one case and to gain control of the carotid canal in the remaining two cases. An enlarged translabyrinthine approach was performed in a case of JFM in which the sigmoid sinus could not be ligated because preoperative MRA showed it to be the dominant sinus; a TO approach was adopted in two cases with preoperative dead ear. A transcochlear (TC) approach was performed in one case that had 270-degree encasement of the vertical and horizontal portions of the intrapetrous carotid artery and involvement of the clivus and cavernous sinus.

Of the 11 patients treated surgically, 9 underwent single-stage removal; in the remaining 2 patients, surgery was staged. The second-stage surgery consisted of a combined IFT-A and ETLA approach in one case and TC approach in the other. Total removal (Simpson's grades I and II) was achieved in six cases. In one patient, residual tumor was left around the intrapetrous portion (genu) of the internal carotid artery (ICA); the patient refused further surgery and was consequently managed with serial MRI scans that showed no further growth after 3 years of follow-up. In one patient, tumor was left surrounding the posterior inferior cerebellar artery (PICA), one patient had residual tumor left in the cavernous sinus, and in another case a small residue was left around the anterior inferior cerebellar artery (AICA). In these three patients with incomplete resection (Simpson's grades III and IV), contrast-enhanced postoperative MRI failed to show the residual tumor and to date there has been no sign of recurrence.

The last of the 11 patients underwent removal of the extradural portion of the tumor as a first stage; the intradural component had intratumoral calcification and has not increased in size over the follow-up period of 2 years. For this reason, it has been decided to manage this patient conservatively for the time being.

In the six patients for whom total removal was accomplished, no signs of recurrence were detected during the follow-up observation period. None of the patients with JFM died.

In both series of patients (JFS and JFM), a staging strategy had been adopted to avoid the risk of postoperative cerebrospinal fluid (CSF) leak associated with possible communication between the subarachnoid space and a wide cervical exposure. This was later found to be largely unnecessary when the POTS procedure was used.

Surgical Technique—the Petro-Occipital Trans-Sigmoid Approach[18–20]

The POTS approach is described below:

- The patient is positioned supine with the head turned to the opposite side.
- A C-shaped incision is made, starting ~3 cm above the auricle, coursing 4 to 5 cm behind the postauricular crease, and extending inferiorly to the level

of the C-1 vertebra. Its anterior end reaches just posterior to the angle of the mandible.

- An inferiorly based U-shaped, myoaponeurotic flap is raised. The base of the flap is at the level of the C-1 vertebra.
- The sternocleidomastoid muscle is retracted posteriorly, and the internal jugular vein is identified just in front of the transverse process of the atlas. After the vein is freed, a suture is passed underneath it for later ligation, taking care not to injure the accessory nerve.
- An extended mastoidectomy is performed with skeletonization of the sigmoid sinus down to the jugular bulb. The digastric ridge is identified, and the mastoid tip is amputated.
- A 4 × 4-cm retrosigmoid craniotomy is performed. A piece of bone overlying the junction of the transverse and sigmoid sinus is left in place to allow extraluminal compression with Surgicel (Johnson & Johnson, New Brunswick, NJ).
- The sinus, the presinus dura, and the jugular bulb are completely uncovered up to the posterior semicircular canal. The retrofacial air cells are exenterated after identification of the mastoid segment of the facial nerve (Fig. 2).
- With gentle pressure on the sigmoid sinus and posterior fossa dura using the suction irrigator, the endolymphatic sac and duct are identified and sectioned to allow further detachment of the dura from the posterior surface of the petrous bone.
- Next, the infralabyrinthine cells are drilled, taking care not to injure the cochlea.
- The proximal part of the sigmoid sinus is compressed extraluminally with a piece of Surgicel placed between the sinus and the overlying bone at its junction with the transverse sinus. The internal jugular vein in the neck is ligated. Next the sigmoid sinus is closed proximally and distally with two tungsten clips and transected.
- The posterior fossa dura is opened with a horizontal incision starting ~3 cm posterior to the sigmoid sinus, coursing anteriorly, traversing the medial wall of the sinus, and ending at the level of the posterior semicircular canal.
- The upper and a lower dural flap are retracted with stay sutures. The jugular bulb is opened. The bulb is usually compressed by the tumor mass. Bleeding from the inferior petrosal sinus is controlled by gentle packing with Surgicel (Fig. 3). Tumor removal then proceeds accordingly.
- To avoid CSF rhinorrhea, all opened perilabyrinthine cells should be exenterated. The apical and retrofacial air cells should be sealed with bone wax.

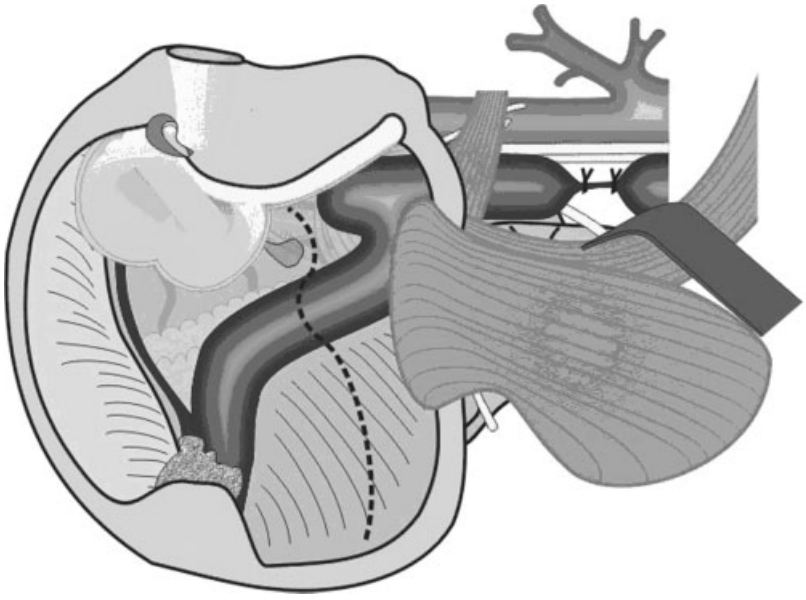

Figure 2 Schematic representation of the petro-occipital trans-sigmoid (POTS) approach before opening of the dura.

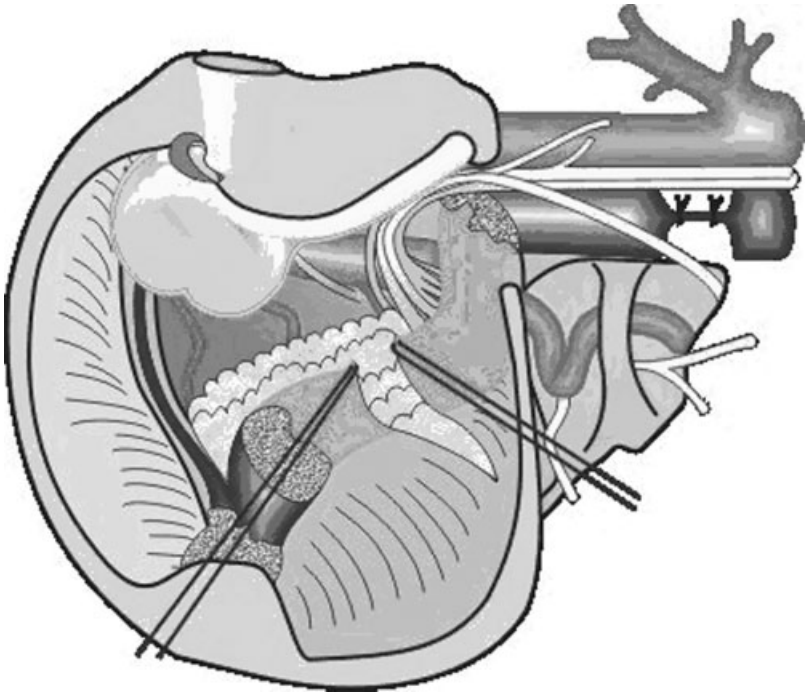

Figure 3 Schematic representation of the petro-occipital trans-sigmoid (POTS) approach after opening the dura.

- At the end of the procedure, the retrosigmoid dura is approximated with sutures, and the rest of the cavity is obliterated with strips of abdominal fat held in place by the repositioned myoaponeurotic flap. No postoperative lumbar drain is used.

Lower Cranial Nerves

JUGULAR FORAMEN MENINGIOMAS

Preoperative examination established that five patients had deficits of at least one CN. All had nerve X deficits, and nerves XI and IX disfunctions were observed in two cases and one case, respectively. Nerve XII was involved preoperatively in three cases, and one patient was affected by VI nerve palsy. One patient had involvement of all four CNs. In all, eight patients acquired additional CN deficits and only two patients maintained their preoperative status (Table 2).

On long-term follow-up, all patients demonstrated good compensation of their LCN dysfunctions; none needed a tracheostomy, vocal cord medialization procedures, or placement of a gastrostomy.

JUGULAR FORAMEN SCHWANNOMAS

Fourteen of 19 patients diagnosed with JFS had at least one LCN deficit preoperatively; IX and X were involved in 12 and 11 cases, respectively, followed by LCNs XI (eight cases) and XII (six cases). Five patients had involvement of all four CNs; five patients had no signs of CN deficits. A new deficit of one or more LCNs was acquired as a result of surgery in 12 patients. In one patient, four LCN palsies were acquired (Table 3).

Facial Nerve

Tables 2 and 3 document the cranial nerve deficits acquired as a result of the management of these tumors. The four patients operated on using the IFT-A approach had permanent anterior transposition of the facial nerve. At the last clinical follow-up, two of them had HB grade I facial nerve function,

Table 2 Pre- and Postoperative Characteristics in Cases with Jugular Foramen Meningiomas

Patient	Surgery (first stage)	Surgery (second stage)	Removal (residue)	Facial Nerve Grade (House-Brackmann)[21]	Cranial Nerve Status (preop/postop)
1	IFT-A + ETLA	IFT-A + ETLA-TA	Subtotal (AICA)	III	Normal/IX,X,XI
2	ETLA + TA	No	Total	II	Normal/Normal
3	POTS + TO	No	Total	I	Normal/IX,X,XI
4	ETLA + TA + POTS	No	Total	III	X/X, XI
5	TO	No	Total	III	X/IX,X,XI
6	POTS	No	Subtotal (intrapetrous ICA [genu])*	I	Normal/IX,X,XI
7	TO + POTS	No	Total	III	X,XII/IX,X,XI,XII
8	POTS	No	Total	I	IX,X,XI,XII/IX,X,XI,XII
9	TC	POTS	Subtotal (cavernous sinus)*	IV	VI, X,XI,XII/VI,IX,X,XI,XII
10	POTS	No	Subtotal (intradural)†	I	Normal/Normal
11	ETLA (dominant sinus)	No	Subtotal (PICA)*	VI	Normal/IX,X,XI

*Residual tumor not visualized on contrast-enhanced magnetic resonance imaging.
†Conservative management because absence of growth after 2 years of follow-up.
IFT-A, infratemporal fossa approach type A; ETLA, enlarged translabyrinthine approach; TA, transapical extension; AICA, anterior inferior cerebellar artery; POTS, petro-occipital trans-sigmoid approach; TO, transotic approach; ICA, internal carotid artery; PICA, posterior-inferior cerebellar artery.

Table 3 Pre- and Postoperative Characteristics in Cases with Jugular Foramen Schwannomas

Patient	Surgery (first stage)	Surgery (second stage)	Removal (residue)	Facial Nerve Grade (House-Brackmann)[21]	Cranial Nerve Status (preop/postop)
1	IFT-A	ETLA	Total	I	IX/IX,X
2	IFT-A (drilling of cochlea)	No	Total	IV	IX,X/IX,X,XI
3	IFT-A (drilling of cochlea)	POTS	Total	I	IX,X,XI,XII/IX,X,XI,XII
4	POTS + ETLA	No	Total	I	Normal/IX
5	POTS	No	Total	I	Normal/IX
6	POTS	No	Total	I	XI/IX,X,XI
7	POTS	No	Total	I	IX,X,XI,XII/IX,X,XI,XII
8	POTS	No	Total	I	IX,X,XI/IX,X,XI
9	POTS	POTS	Total	I	IX,X,XI,XII/IX,X,XI,XII
10	POTS	No	Total	I	IX,X,XI,XII/IX,X,XI,XII
11	POTS	No	Total	I	Normal/IX,X
12	POTS	No	Total	I	Normal/IX,X
13	POTS + TO	No	Total	I	IX,X,XII/IX,X,XI,XII
14	POTS	No	Total	I	Normal/IX,X,XI,XII
15	POTS	No	Total	I	IX,X,XI/IX,X,XI,XII
16	POTS	No	Total	I	X/IX,X,XI
17	POTS	No	Total	I	IX/IX
18	Transcervical + subtotal petrosectomy	POTS	Total	VI	IX,X,XI,XII/IX,X,XI,XII
19	Transcervical	No	Subtotal (intradural residue)	I	IX,X/IX,X,XI

IFT-A, infratemporal fossa approach type A; ETLA, enlarged translabyrinthine approach; POTS, petro-occipital trans-sigmoid approach; TO, transotic approach.

one had grade III, and one grade IV. All the patients who underwent the POTS approach alone ($n = 15$) had an HB grade I facial nerve function at the last clinical follow-up. Among the patients that underwent the POTS approach combined with labyrinthine destructive procedures ($n = 5$), three had grade I and two had grade III at follow-up. Of the remaining patients ($n = 6$), one had HB grade I, one had grade II, two grade III, and two had grade VI. In summary, postoperative facial nerve function HB grades I and II scores were achieved in the long term in 22 patients (73.3%), with 100% of HB grade I in the patients who received only the POTS approach.

Hearing (Tables 4 and 5)

Hearing preservation was attempted in 15 cases using the POTS approach.

JUGULAR FORAMEN SCHWANNOMAS

Of the 12 JFS patients in which hearing preservation was attempted (Table 4), total deafness developed in one case, probably due to intraoperative coagulation of the labyrinthine artery in a tumor with a large intradural component. Hearing was preserved at the preoperative level in seven cases (58.3%), and four patients sustained a sensorineural hearing loss that ranged from 10 to 25 dB.

JUGULAR FORAMEN MENINGIOMAS

Hearing preservation was attempted in three of the JFM cases (Table 5). In one patient, hearing was preserved at the preoperative level. The remaining two cases experienced sensorineural hearing losses; one was 5 dB and the other was 10 dB. The patient who developed a loss of 10 dB sustained a profound sensorineural hearing loss (SNHL) 40 days after surgery that recovered to a moderate SNHL after a course of steroid therapy.

Table 4 Pre- and Postoperative Pure Tone Averages for Air and Bone Conduction for Jugular Foramen Schwannoma Patients

Patient	Approach (first stage)	Approach (second stage)	Preop PTA BC (dB)	Postop PTA BC (dB)	Preop PTA AC (dB)	Postop PTA AC (dB)
1	IFT-A	ETLA	DE	DE	DE	DE
2	IFT-A (drilling of cochlea)		15	DE	30	DE
3	IFTA (drilling of cochlea)	POTS	10	DE	60	DE
4	POTS + ETLA	No	DE	DE	DE	DE
5	POTS	No	20	40	25	45
6	POTS	No	15	15	20	20
7	POTS	No	10	10	10	10
8	POTS	No	10	DE	10	DE
9	POTS	No	10	10	10	10
10	POTS	No	10	10	10	10
11	POTS	No	10	20	10	40
12	POTS	No	15	15	20	20
13	POTS + TO	No	50	DE	70	DE
14	POTS	No	20	35	20	60
15	POTS	No	20	45	30	55
16	POTS	No	10	10	10	10
17	POTS	No	10	10	10	10
18	Transcervical + subtotal petrosectomy	POTS	DE	DE	DE	DE
19	Transcervical	No	25	25*	30	30*

*Hearing level progressively decreased to profound sensorineural hearing loss during follow-up.
PTA, pure tone average; BC, bone conduction; AC, air conduction; IFT-A, infratemporal fossa approach type A; ETLA, enlarged translabyrinthine approach; DE, dead ear; POTS, petro-occipital trans-sigmoid approach; TO, transotic approach.

Table 5 Pre- and Postoperative Pure Tone Averages for Air and Bone Conduction for Jugular Foramen Meningioma Patients

Patient	Approach (first stage)	Approach (second stage)	Preop PTA BC (dB)	Postop PTA BC (dB)	Preop PTA AC (dB)	Postop PTA AC (dB)
1	IFT-A + ETLA	IFT-A + ETLA + TA	20	DE	65	DE
2	ETLA + TA	No	55	DE	55	DE
3	POTS + TO	No	DE	DE	DE	DE
4	ETLA + TA + POTS	No	20	DE	20	DE
5	TO	No	DE	DE	DE	DE
6	POTS	No	20	30	25	35
7	TO + POTS	No	45	DE	45	DE
8*	POTS	No	15	20	20	25
9	TC	No	20	DE	20	DE
10	POTS	No	20	20	25	25
11†	ETLA	No	55	DE	55	DE

*Sudden deafness after 40 days; recovery to moderate SNHL.
†Ipsilateral dominant sinus.
PTA, pure tone average; BC, bone conduction; AC, air conduction; IFT-A, infratemporal fossa approach type A; ETLA, enlarged translabyrinthine approach; TA, transapical extension of ETLA; DE, dead ear; POTS, petro-occipital trans-sigmoid approach; TO, transotic approach.

In summary, in the long term, 8 out of 15 patients (53.3%) who underwent the POTS approach alone maintained their hearing at the preoperative level, and 5 patients (33.3%) had an SNHL of between 5 and 25 dB; one patient had a moderate SNHL and one had a postoperative dead ear.

Complications

There were no peri- or postoperative deaths in this series. Two patients, one operated for JFS and the other for JFM via the POTS approach, experienced postoperative CSF rhinorrhea. One of these patients was managed with the placement of a lumbar drain; the other required surgical revision and obliteration of the middle ear spaces and eustachian tube with abdominal fat and a blind sac closure of the external auditory canal. This patient developed bacterial meningitis and was subsequently managed with high doses of intravenous antibiotics. One JFS patient, who was operated on with the POTS approach, had subcutaneous CSF collection that was managed conservatively with aspiration and tight dressings.

The incidence of CSF rhinorrhea in patients who underwent POTS ($n = 15$) alone was 13.3%, whereas the overall incidence of CSF rhinorrhea in this series was 6.6%. No aspiration pneumonia or problems related to intracranial venous drainage developed in these patients.

ILLUSTRATIVE CASES

Jugular Foramen Schwannomas

CASE 1

Schwannoma of the right LCNs, extending from the cerebellopontine angle (CPA) through the JF into the upper neck. A POTS approach was performed, preserving the preoperative hearing level and facial function (Fig. 4).

CASE 2

Right JFS with neck and posterior fossa extension; the lesion was removed by a combined POTS and ETLA approach because of poor preoperative hearing (Fig. 5).

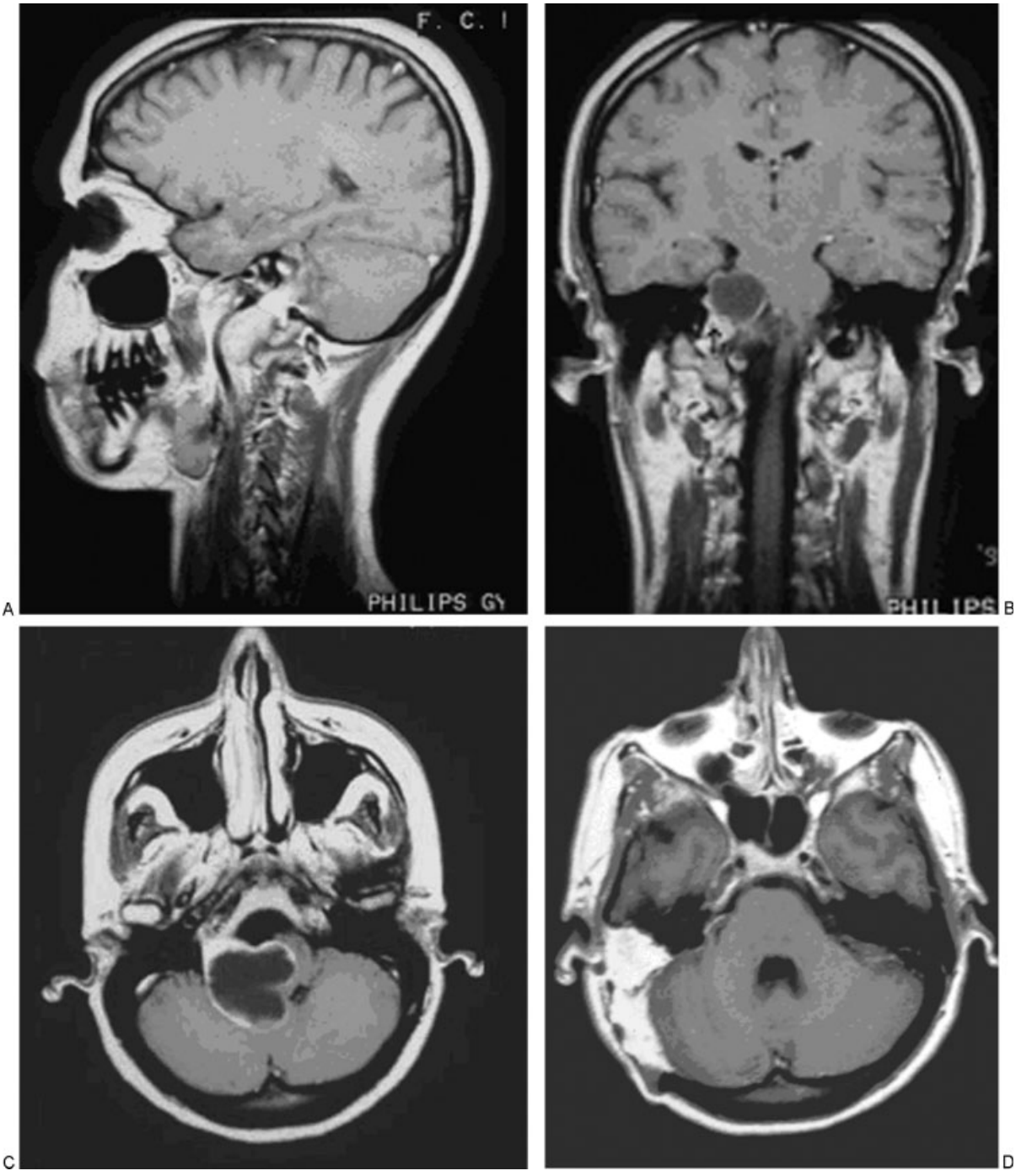

Figure 4 (A) Magnetic resonance imaging (MRI) with gadolinium, sagittal view, showing the tumor extending from the cerebellopontine angle to the neck. The internal carotid artery was in close relation with the tumor. (B) MRI with gadolinium, coronal view, showing the tumor extension into the posterior fossa (arrow: jugular foramen). (C) MRI with gadolinium, axial view, showing the large cystic component at the level of the posterior fossa. (D) Postoperative MRI with gadolinium showing total tumor removal and proper obliteration of the operative defect with abdominal fat.

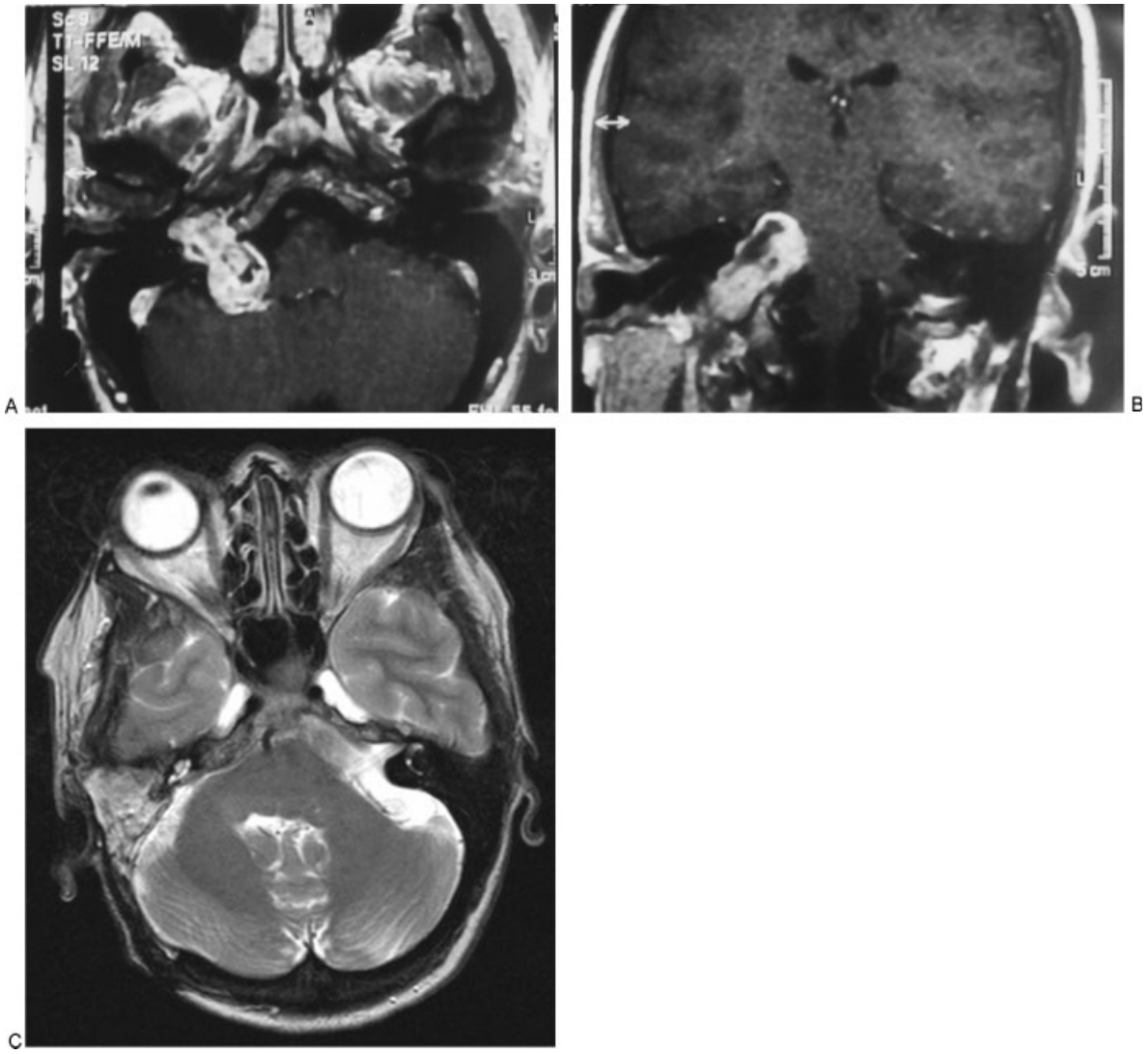

Figure 5 Magnetic resonance imaging (MRI) with gadolinium, axial and coronal views, showing the extension in the (A) posterior fossa and (B) the neck of the jugular foramen schwannoma. (C) Postoperative MRI T2-weighted image, axial view.

CASE 3

Left JFS in a 73-year-old woman, managed conservatively by interval clinical evaluation and MRI scans. The patient showed good compensation—left IX and X CN palsy was nearly asymptomatic (Fig. 6).

CASE 4

Left JFS in a 77-year-old woman. The large cervical component was removed when the patient was 69 years old using a transcervical approach. After 8 years of follow-up, the last MRI scans demonstrated absence of recurrence in her neck or growth of the intracranial component (Fig. 7).

Jugular Foramen Meningiomas

CASE 5

Right JFM with neck extension. Total removal was achieved by means of a single-stage POTS

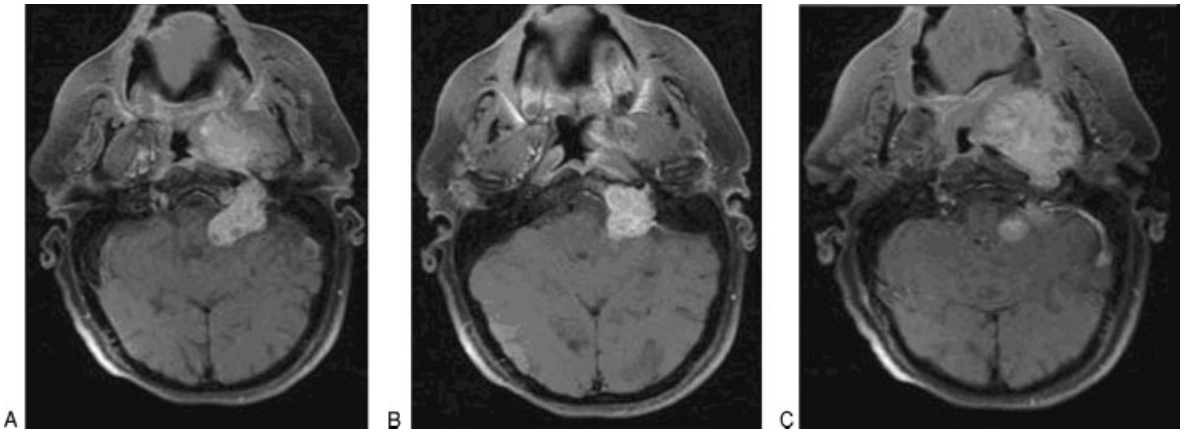

Figure 6 (A–C) Magnetic resonance imaging (MRI) with gadolinium, axial view, showing the tumor extending from the posterior fossa to the neck. The tumor did not grow during 4 years of follow-up.

approach, with preservation of hearing and facial function (Fig. 8).

CASE 6

Right-sided JFM with extension in the posterior fossa and the neck. Surgical excision was performed in two stages using an IFT-A combined with an enlarged translabyrinthine approach (Fig. 9).

DISCUSSION

Nonvascular neoplasms of the JF are rare and are mainly schwannomas and meningiomas. The early diagnosis of these lesions requires a high degree of suspicion that is not often achieved.[1–14] Radiological features, especially those found on MRI, help the clinician to distinguish between the different tumor types.[15,18,21–23] All JF tumors present with similar signs and symptoms, deficits of CNs IX-XII, and hearing loss. Facial nerve dysfunction is rarely encountered in the early stage of the disease process. To the best of our knowledge, only ~200 cases of JFSs have been described in the literature.[2,3,13] Similarly, < 100 cases of JFMs have been reported.[4–11] Only four large series report treatment of JFMs: Ramina et al (10 cases),[10] Molony et al (8 cases),[13] Arnautović and Al-Mefty (8 cases),[8] and Gilbert et al (6 cases).[15]

Jugular foramen tumors usually present with multiple signs and symptoms; in our series the most frequent findings related to LCN deficits and were in the form of dysphonia, dysphagia, or shoulder weakness, followed by hearing loss and vertigo. Hearing loss and balance disturbances were more common in JFM patients, whereas LCN deficits were more frequent in JFSs.

We found that imaging was pivotal in the diagnosis of JF lesions, and this supported the experience reported by others.[23,24] Macdonald et al[23] compared the radiological findings and differentiating features of five cases of primary meningiomas of the JF. Ten cases of JFSs and eight cases of jugular paragangliomas were also reviewed, for comparison. They established that JFMs appeared iso- or hypointense in T1-weighted MRI sequences and had intermediate signal intensity on T2-weighted sequences; all showed intense and uniform signal enhancement after administration of contrast. Centrifugal infiltration of the surrounding skull base in all directions, absence of flow voids, and prominent dural tails were the most characteristic features of these lesions. On HRCT, JF meningiomas produced slight hyperostosis and irregular bony erosion of the JF bony margins. Jugular foramen paragangliomas were associated with typical flow voids ("salt-and-pepper" appearance) on MRI with absence of dural tails and intense enhancement postcontrast. Tumor spread was

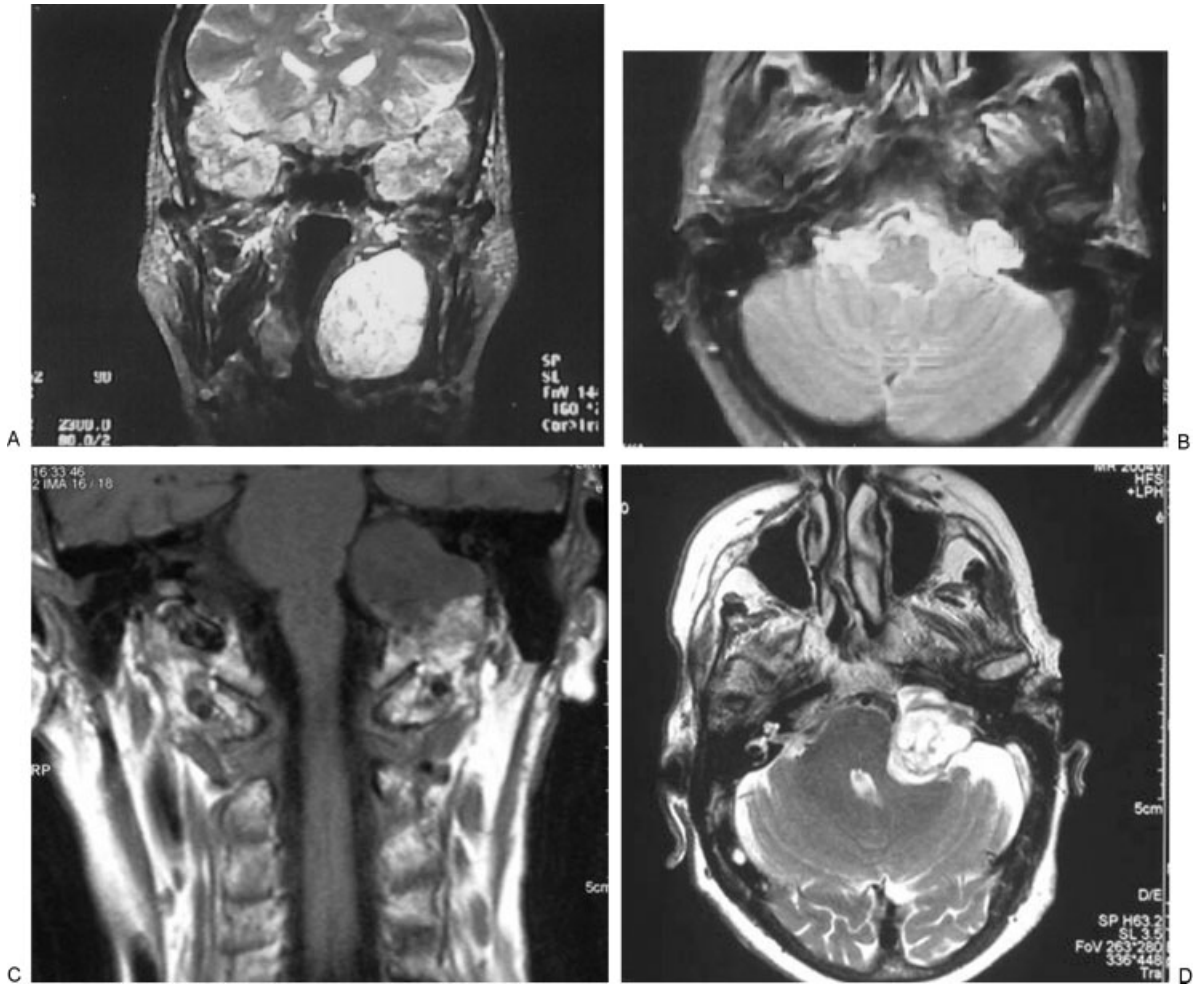

Figure 7 Preoperative T2-weighted magnetic resonance imaging (MRI) scans, coronal (A) and axial (B) views, showing a schwannoma of the left lower cranial nerves with large cervical component and relatively small intracranial extension. T1-weighted coronal (C) and T2-weighted axial (D) MRI images show same patient 8 years after removal of the cervical component. Note the enlargement of the intracranial component.

typically superolateral toward the hypotympanum and the middle ear. On HRCT, paragangliomas caused a distinctive "moth-eaten" erosion of the JF. A JFS on HRCT appeared as a smooth enlargement of the JF without infiltration of the bony margins. On MRI, these tumors were round or lobulated enhancing masses with smooth margins centered on the JF that followed the course of the CN of origin. Meningiomas and schwannomas of the JF assume a dumbbell shape when there is both intra- and extracranial spread.

Complete tumor resection with preservation of adjacent neural structures is recognized to be extremely difficult.[20,25] Various surgical approaches have been described to control the region of the JF and advocated according to the tumor extension, preoperative hearing acuity, facial nerve function, and invasion of the labyrinth as assessed by imaing.[3,9,18,19,25–30] Labyrinthine destructive procedures, such as ETLA with or without transapical extension[31,32] and the modified TC approaches,[27] are indicated when there is a profound hearing loss

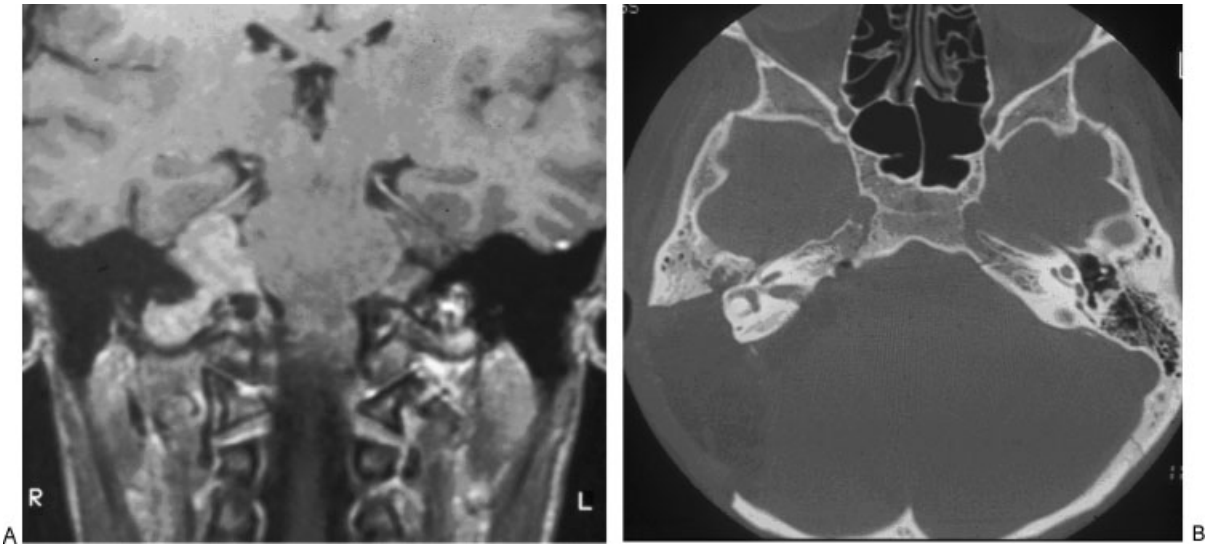

Figure 8 (A) Gadolinium enhanced magnetic resonance imaging (MRI), coronal view, showing a right-sided jugular foramen meningioma. (B) Postoperative high-resolution computed tomography scan with bone window, axial view, showing preservation of otic capsule and middle ear cleft after petro-occipital trans-sigmoid (POTS) approach.

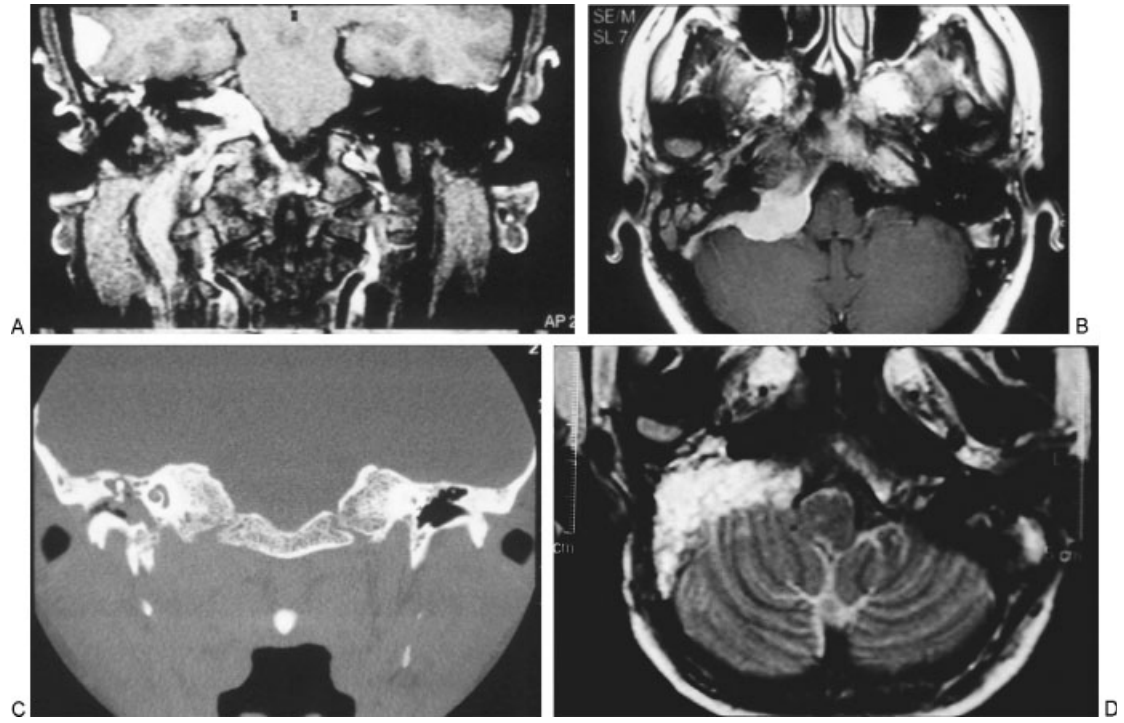

Figure 9 (A) Magnetic resonance imaging (MRI) with gadolinium, coronal view, showing right-sided meningioma of the jugular foramen with a huge cervical component. (B) MRI with gadolinium, axial view, showing posterior fossa extension of the tumor. (C) High-resolution computed tomography scan, coronal view, showing the middle ear completely filled by the tumor. (D) Postoperative MRI, axial view, showing the surgical defect filled by abdominal fat strips.

and image evidence of labyrinthine infiltration or space occupation of the CPA and/or internal auditory canal. The TO approach[33] has the advantage of avoiding rerouting of the facial nerve, but control of the JF with this approach is limited. The IFT-A approach[16] gives excellent control of the JF region; the key to this approach is anterior transposition of the facial nerve. Lack of experience with this technique may result in a 10 to 30% rate of paresis or palsy.[17,34] The patient also acquires a conductive hearing loss as the middle ear is obliterated and a blind sac closure of the external auditory canal is performed.[16,33]

In our experience, the POTS approach is the technique of choice for meningiomas and schwannomas arising primarily in this area. It permits a single-staged, intra- and extradural tumor removal without facial nerve rerouting and also preserves all middle ear structures, otic capsule, and labyrinth. The posterolateral direction of this approach, with sacrifice of the sigmoid sinus and jugular bulb, affords exceptional exposure. This approach was first introduced by Mann et al in 1991[29] as the trans-sigmoid approach and was subsequently modified by Mazzoni and Sanna in 1995.[18]

Functional results presented in this series demonstrate a global postoperative long-term facial nerve function HB grading of I or II in 73% of cases, whereas 100% of HB grade I was obtained in the subgroup of patients who underwent surgical removal using the POTS approach alone. Hearing preservation was possible in 14 of 15 patients who underwent POTS; 8 patients maintained their hearing at the preoperative level (53.3%), and 5 patients (33.3%) had a deterioration of between 5 and 25 dB. One of the remaining two patients sustained a moderate SNHL, and the other had a dead ear.

When feasible, the POTS approach offers the surgeon the opportunity of preserving facial nerve function and hearing level; in our experience, this was possible in the majority of LCN schwannomas. Outcomes are, however, less favorable in JFMs. The higher percentage of hearing and facial nerve preservation obtained in JFS compared with meningiomas can be explained by the higher tendency of meningiomas to infiltrate dura, bone, vessels, and nerves, requiring a more aggressive surgery.

Overall, the major morbidity following surgery was caused by LCN deficits, which seemed to be related more to the pathology rather than to the approach itself. Because of the infiltrative nature of meningiomas, resection of adjacent nerves was often necessary to achieve a complete resection. Preservation of anatomical integrity of the nerves while leaving a portion of the tumor around them is considered acceptable by some authors[30] when dealing with JF tumors, but this obviously carries a higher risk of recurrence and, in any case, does not necessarily guarantee preservation of CN function.[18] When preoperative deficits of LCNs are established, patients in general tolerate the procedure well. In contrast, significant morbidity is experienced by those with normal function before surgery, and they usually require a longer period of recovery. It is interesting to note that none of the patients presented in this series required vocal cord medialization procedures, tracheostomy, or a feeding gastrostomy tube.

Despite the single-stage removal of these intra- and extracranial lesions using the POTS approach, the incidence of CSF leak was quite small, just 13.3%. Several reasons account for this. First, the approach avoids wide contact between the neck and the subarachnoid space; the occipital dura and the medial wall of the sigmoid sinus are sutured, and the remaining small defect is obliterated with strips of abdominal fat. The thick fibrous tissue at the skull base is left undisturbed; this, together with the specially designed myoaponeurotic flap minimizes the resulting surgical cavity and helps to contain the free fat graft. In patients with massive extracranial extension into the neck, staging was necessary to avoid communication between the subarachnoid space and neck dissection because these areas have high risk of CSF leak.[35,36] Therefore, the extracranial part was removed first, followed by second-stage removal of the intracranial component.

Although the POTS approach offers excellent exposure of the jugular fossa, extensive

involvement of the vertical portion of the internal carotid artery is still a relative limitation. This is well illustrated in the case in which a TO approach had to be combined with it to provide the necessary exposure for safe dissection of the tumor from the intrapetrous ICA. Total tumor resection was feasible in all case of JFS, whereas subtotal resection was obtained in four (36.3%) cases of JFM because of infiltration of vital structures such as AICA, PICA, intrapetrous ICA, and cavernous sinus. However, of these only one had an enhancing lesion evident on a postoperative MR scan. One patient affected by JFM was managed conservatively after the first stage of surgery, which involved resection of a huge cervical component that measured 13 cm in its maximum diameter. The small volume of residual intradural disease has failed to grow after 2 years of follow-up. A similar experience was encountered in one patient with a JFS. After a first stage in which a large cervical component was resected, no further growth of the residual tumor was seen on serial MRIs for 8 years; after that it began to grow again. This patient is now 77 years old and is currently awaiting second-stage surgery.

CONCLUSIONS

Primary schwannomas and meningiomas of the JF are rare neoplasms that pose a challenge in their diagnosis and management. Recent advances in neuroradiology have provided clinicians with the ability to differentiate between the tumors preoperatively. The POTS approach has been developed with the aim of removing in one stage most tumors with intracranial (intra- or extradural) and extracranial components while preserving the facial nerve and auditory functions.

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