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Chondrosarcomas of the Jugular Foramen

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Objectives/Hypothesis: Chondrosarcomas of the jugular foramen are extremely rare tumors. Our review of the literature revealed eleven previously reported cases. The aim of this study is to describe the presenting symptoms, radiographic findings, operative procedures, and postoperative outcome of five histologically confirmed cases of chondrosarcomas arising from the jugular foramen. A review of the literature is also presented.

Study Design: Retrospective study of an quaternary referral otology and skull base private center.

Methods: Five cases of surgically treated and pathologically confirmed jugular foramen chondrosarcomas were identified. The follow-up of the series ranged from 23 to 42 months (mean, 32.8 ± 7.7 months).

Results: A single stage procedure was adopted in all the cases. Two patients underwent type A infratemporal approaches, one patient underwent a transotic approach extending to the neck with ligature of the internal jugular vein, one patient underwent a petro-occipital transigmoid approach, and one patient underwent a combined petro-occipital transigmoid–transotic approach. Gross total tumor removal was achieved in all patients. The most common complications were lower cranial nerve deficits. To date, no recurrence or residual tumors have been observed at radiological controls.

Conclusions: We believe that the primary treatment for chondrosarcomas of the jugular foramen is gross total surgical resection of the tumor. It is our philosophy to reserve postoperative radiotherapy for patients with histologically aggressive tumors, as well as in cases with subtotal resection and recurrent tumors.

Key Words: Chondrosarcoma, jugular foramen, surgical management, outcome.

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INTRODUCTION

Chondrosarcomas (CSAs) of the skull are rare slow growing locally aggressive malignant tumors that constitute 0.15% of all intracranial neoplasms.[1] Approximately three-fourths of all CSAs of the skull occur at the cranial base, accounting for 6% of all neoplasms at this site.[1–3] It has been hypothesized that cranial base CSAs may originate from multipotential mesenchymal cells[4,5] or from embryonal cartilage remnants of skull synchondroses.[2,5–8] The most common tumor sites of origin have been reported to be the petroclival, petro-occipital, spheno-occipital, and the sphenopetrosal synchondroses. Possible locations, in decreasing order of frequency, are as follows: middle fossa (64%), both posterior and anterior fossa (14%), anterior fossa (14%), and posterior fossa (7%).[8] The jugular foramen is an exceptionally rare location for CSAs. In fact, the jugular foramen is located along the posterolateral aspect of the petro-occipital suture. To the best of our knowledge, only 11 well documented cases of CSAs arising primarily in the jugular foramen have been found in the literature worldwide.[2–4,7,9–14] Primary jugular foramen CSAs should be distinguished from those that secondarily spread into the jugular foramen (i.e., CSAs of the petrous apex). Primary jugular foramen CSAs are centered on the jugular foramen (Fig. 1) and may invade the infralabyrinthine temporal bone and the middle ear. They can extend intracranially into the cerebellopontine angle (CPA) and extracranially into the upper neck.

These tumors rarely metastasize, therefore local control represents the goal of therapy. The ideal primary treatment of these tumors is total surgical removal. Radiotherapy may constitute a viable alternative to surgery in selected cases in which there are serious contraindications to surgery as well as in cases with partial excision or with high risk of recurrence. Proton beam radiotherapy, radiosurgery (gamma knife or Cyber knife), or fractioned radiotherapy is often used as an adjuvant treatment.[15,16]

We report a series of five CSAs centered on the jugular foramen, discussing the presenting symptoms, radiographic findings, operative procedures, and postoperative outcome as well as offering a review of the literature. This is so far the largest series of jugular foramen CSAs currently reported in the literature.

PATIENTS AND METHODS

A retrospective analysis of all patients with skull base tumors managed at the Gruppo Otologico Piacenza-Rome between December 1988 and July 2007 was performed. Five patients with CSA centered on the jugular foramen were identified and became the subjects of this study. Chondrosarcomas arising on the petrous apex and spreading into the jugular foramen were excluded. No patients in this series had evidence of Maffucci's syndrome or

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Fig. 1. Axial T1-weighted MRI scan with gadolinium of a chondrosarcoma centered on the right jugular foramen demonstrating heterogeneous enhancement (case 4).

Ollier's disease. The collected data were analyzed for age, sex, presenting signs/symptoms, operative procedures, location and extent of the tumor, histological features, postoperative outcomes, and clinical and radiological follow-up findings. All patients underwent complete otological and neurological examinations, flexible fiber optic laryngoscopy, and hearing assessment by pure-tone audiometry. All patients were evaluated before surgery by high-resolution computed tomography (HRCT) with bone windows, gadolinium-enhanced magnetic resonance imaging (MRI-Gd), and cerebral angiography. The facial nerve function was evaluated before the operation and 1 year after the operation with the House-Brackmann (HB) system.[17] Immunohistochemical analysis was performed on all tumor specimens. All tumors were graded pathologically according to the Rosenberg grading system.[18]

RESULTS

General Characteristics and Clinical Manifestations

The study group was composed of five patients with histologically confirmed jugular foramen CSAs surgically treated by the senior author (M.S.). The relevant patient demographic characteristics, presenting signs/symptoms, location of the tumor, specific operative approaches, histological features, and outcomes for this series of patients are outlined in Table I. Three patients (60%) were female and two (40%) were male. The mean age at the time of surgery was 47.8 ± 12.3 years (range, 35–60 years). All tumors occurred on the right side. The duration of the follow-up period (consisting of clinical evaluation, hearing tests, flexible fiber optic laryngoscopy, and serial CT and/or MRI scans) ranged from 23 to 42 months (mean, 32.8 ± 7.7 months).

The most common symptoms at the time of presentation included: dysphonia (4/5), dysphagia (3/5), pulsatile tinnitus (3/5), HL (2/5), and vertigo (2/5). Facial nerve dysfunction was present in two patients (cases 1 and 2), both of which had HRCT evidence of fallopian canal involvement by the tumor. Other symptoms included unsteadiness (1/5) and weakness of the sternocleidomastoid and trapezius muscles (1/5). All patients had a combination of two or more symp-

TABLE I.
Patient Demographics, Location of Tumor, Type of Operation, and Postoperative Complications in the Five Patients Surgically Treated for JF Chondrosarcoma.

Patient	Sex	Age at Surgery (Yrs)	Presenting Signs/ Symptoms	Preop LCN Deficits	Site of Tumor	Surgical Approach	New LCN Deficits	Gross Total Removal	Preop/Postop	Histology (Grade)	Follow-Up (Mo)
1	F	54	HL, dysphagia, dysphonia, vertigo, FN paresis, EAC mass	IX-X	JF, EAC, ME, IAC, CPA, CC	TO+ Jugular vein ligature	None	Yes	DE/DE	I	38
2	M	60	FN palsy, unsteadiness, pulsatile tinnitus	None	JF, hypotympanum, CC	IFTA-A	IX, X	Yes	NH/DE	I	42
3	M	34	HL, pulsatile tinnitus, dysphonia, vertigo	X	JF, hypotympanum, infralabyrinthine region, N	IFTA-A	IX, XI	Yes	CHL/CHL	I	34
4	F	56	Dysphonia, dysphagia	IX, X	JF, infralabyrinthine region, PCF	POTS	XI	Yes	NH/NH	II	27
5	F	35	Pulsatile tinnitus, dysphonia, dysphagia, shoulder weakness	IX-XII	JF, PA, CC	POTS-TO	None	Yes	NH/DE	I	23

LCN = lower cranial nerve; F = female; M = male; HL = hearing loss; FN = facial nerve; JF = jugular foramen; EAC = external auditory canal; CPA = cerebellopontine angle; IAC = internal auditory canal; PCF = posterior cranial fossa; PA = petrous apex; CC = carotid canal; N = Neck; ME = middle ear; TO = transotic approach; IFTA-A = infratemporal fossa approach-Type A; POTS = petro-occipital-trans-sigmoid approach; NH = normal hearing; DE = dead ear; CHL = conductive hearing loss.

toms at the time of diagnosis. The mean length of symptoms previous to presentation was 17.2 ± 13.6 months (ranging from 3 months to 35 months). Four patients (80%) were found to be suffering from at least one lower cranial nerve (CN) deficit at presentation. Two patients had both preoperative CN IX and CN X deficits. One patient had an isolated CN X deficit. One patient presented with paralysis of CNs IX, X, XI, and XII. One patient had no dysfunction of the lower CNs on initial diagnosis.

On otoscopic examination, one patient (case 1) was found to have a polypoid mass in the external auditory canal. Otoscopic examination of all the other cases was unremarkable.

Location of the Tumor and Neuroimaging Findings

The location and the extent of the lesions were determined preoperatively using the radiology reports (HRCT with bone windows and MRI-Gd), and then subsequently confirmed intraoperatively. In all patients, the tumor was present within the jugular foramen. The frequency of involvement of other structures was as follows: carotid canal, three (60%); CPA, two (40%); infralabyrinthine region, two (40%); middle ear, two (40%); hypotympanum, two (40%); internal auditory canal, one (20%); upper neck, one (20%); external auditory canal, one (20%); petrous apex, one (20%). Three (60%) lesions were exclusively extradural and two (40%) had both intra- and extradural extensions. An extension into the neck was present only in one case (20%). HRCT demonstrated an irregular erosion of the jugular foramen margins in all cases. Intratumoral calcifications or sequestered bony speckles were observed in two cases. An erosion of the vertical segment of the carotid canal was seen in three cases. All lesions had a low signal on T1-weighted images, and a high signal on T2-weighted images. All tumors showed heterogeneous gadolinium enhancement. The preoperative diagnostic suspicion before angiography was CSA (two cases) and paraganglioma (three cases). Angiography demonstrated an avascular lesion in all cases. Preoperative embolization was not attempted in any of the patients. Imaging (HRCT and MRI) and angiography characteristics led to suspect a CSA in all cases. The jugular bulb/vein on the side of the lesion was not patent in all cases, but the collateral venous drainage was well developed.

Surgical Treatment, Tumor Resection, and Pathological Findings

All the patients in our series were informed of the risks and benefits of surgical excision, radiotherapy or simple observation with serial imaging. Four patients underwent a primary operation and one patient (case 3) presented with a recurrent tumor operated on elsewhere by the suboccipital approach. No patients had received preoperative radiotherapy.

A single stage procedure was adopted in all cases. A transotic approach extending to the neck with ligature of the internal jugular vein was performed in one patient (case 1) who presented with a preoperative dead ear and a tumor arising from the jugular foramen with involvement of the CPA, middle ear, and external auditory canal together with erosion of the carotid canal. During surgery, the tumor was found to invade the mastoid portion of the facial nerve and was intentionally transected and reanastomosed with a sural nerve graft. One patient (case 2) with preoperative good hearing and a tumor limited to the jugular foramen spreading into the hypotympanum and the vertical internal carotid artery underwent an infratemporal type A approach (IFTA-A); the facial nerve was sacrificed and a sural nerve graft was performed due to infiltration of the mastoid portion of the facial nerve. One patient (case 3) underwent an IFTA-A with permanent anterior transposition of the facial nerve on account of a tumor extension into the infralabyrinthine region, middle ear, and upper neck. One patient (case 4) with preoperative good hearing and tumor involving the jugular foramen, the infralabyrinthine region and extending into the posterior cranial fossa without middle and inner ear involvement underwent tumor removal through the petro-occipital transigmoid (POTS) approach. One patient (case 5) with a tumor involving the jugular foramen and the petrous apex together with a tumor component located anteromedially to the vertical internal carotid artery, required a combined POTS-transotic approach.

The adventitia of the internal carotid artery wall was not infiltrated in any of the three cases where the carotid canal was involved by the tumor. The lower CNs were infiltrated (four cases) or compressed (one case) by the tumor in all patients. To totally remove the tumor, CN IX was sacrificed in two patients and CNs IX, X, and XI in two other patients. The jugular bulb was already occluded by the tumor in all the cases.

Gross total tumor removal was achieved for all patients. One patient (case 4) underwent postoperative high-dose fractionated proton beam radiation therapy.

The histological examination of the tumors according to the Rosenberg grading system[18] revealed the following subtypes: grade I (four cases), grade II (one case). On immunohistochemical analysis, the tumor cells stained positively for S-100 protein, but negatively for epithelial membrane antigen and cytokeratin.

Facial Nerve Outcomes

Table II summarizes the facial nerve results. Preoperatively, three (60%) patients had grade I facial nerve function, one had grade II, and one had grade VI.

TABLE II.
Preoperative, Immediate Postoperative, and Final Facial Nerve Function According to the House-Brackmann Scale.

Patient	Preoperative	Immediately Postoperative	Follow-Up
1	II	VI*	III
2	VI	VI*	IV
3	I	IV†	I
4	I	I	I
5	I	VI	III

*Sural nerve grafting.
†Anterior rerouting.

In two patients (cases 1 and 2), the mastoid segment of the facial nerve was resected because of tumor infiltration, and the defect was repaired using a sural nerve graft in the same sitting. Both of these patients experienced HB grade VI immediately after surgery; at 1-year follow-up, one patient reached grade III, and the remaining one recovered to grade IV. This patient underwent upper eyelid gold weight implantation. One patient (case 3) underwent a permanent anterior transposition of the facial nerve and had grade VI palsy in the immediate postoperative period but reached grade I after a year.

The patient (case 4) who underwent POTS approach experienced grade I immediately after surgery and at last follow-up still has grade I. The patient (case 5) who underwent a POTS-transotic approach experienced grade VI immediately after surgery and recovered to grade III after a year.

Hearing Outcomes

The preoperative hearing status was as follows: three patients had normal hearing, one patient had conductive HL, and one patient had total deafness. Nonhearing preservation tumor removal was performed in two cases (cases 1 and 5). One of these two patients was already deaf before surgery; in the remaining one the POTS-TO approach was preferred despite the presence of preserved hearing preoperatively. The two patients treated via IFTA-A had preoperative audiometric testing within normal limits. In one of these two patients (case 2) the cochlea was found to be eroded by the tumor and drilled to achieve total tumor removal and avoid recurrence. The remaining one experienced postoperative conductive HL with an average 45 dB air-conduction threshold and 15 dB bone-conduction threshold. Hearing was preserved at the preoperative level (normal hearing) in the patient in whom hearing preservation was attempted via the POTS approach.

Complications and Long-Term Follow-Up

There were no peri- or postoperative deaths, nor cases of cerebrospinal fluid leakage, meningitis or vascular injury in the present series.

After surgery, none of the patients recovered the function of the preoperatively paralyzed lower CNs. Three patients (60%) developed a new deficit of one or more of the lower CNs. One of these three patients experienced a new paralysis of CNs IX and X, and required a prophylactic nasogastric feeding tube in the immediate postoperative period. One patient experienced paralysis of CNs IX and XI. A new isolated deficit of CN XI occurred in one case. On long-term follow-up, all patients were able to compensate well for their lower CN deficits with the help of speech and swallowing rehabilitation. No patient required a vocal fold injection or medialization thyroplasty. Physical therapy was administered in patients with CN XI paralysis to prevent persistent shoulder dysfunction and pain.

Follow-up MRI showed no evidence of tumor recurrence after an average of 32.8 months in all five patients. So far, no patient has developed a distant metastasis.

DISCUSSION

Chondrosarcomas centered on the jugular foramen are extremely rare lesions with only few case reports described in the English literature. Based on compiled literature (Table III) and the present series, jugular foramen CSAs occur more frequently in women. The usual initial presentation ranges between 15 and 77 years of age, with a mean age of 43.1 $\pm$ 19.2.[2–4,7,9–14]

Chondrosarcomas of the jugular foramen may invade surrounding areas (such as the infralabyrinthine region, middle ear, petrous apex, clivus, CPA, and upper neck). Their clinical manifestations, therefore, depend on the size and anatomical location of the tumor. Clinically, jugular foramen CSAs mimic other more common lesions of the jugular foramen such as paragangliomas, schwannomas, and meningiomas.

Including our cases, the most common presenting symptoms include dysphonia (68.7%) followed by HL (62.5%), dysphagia (56.2%), pulsatile tinnitus (31.2%), vertigo (25%), shoulder weakness (25%), and unsteadiness (12.5%). Other symptoms include headache (12.5%) and diplopia (12.5%). Seventy-five percent of the patients initially presented lower CN deficits. Facial nerve dysfunction is usually rare at the early stage of the disease and was present in 31.2% of the cases.[2–4,7,9–14] In our cases affected by preoperative facial nerve dysfunction as well as in those reported in the literature the facial nerve was always infiltrated by the tumor at the level of the mastoid segment.[9,12]

HRCT with bone windows and MRI with gadolinium are essential and complementary investigative techniques used to accurately delineate the extent of the tumor preoperatively and to optimize its management. On HRCT, jugular foramen CSA usually presents as a destructive lesion with enlargement and irregular erosion of the jugular foramen margins. Furthermore, HRCT images may expose varying degrees of intratumoral calcifications. In our series, all patients had an irregular erosion of the margin of the jugular foramen (Fig. 2). Two patients had intratumoral calcifications, and three patients had erosion of the vertical segment of the carotid canal. The profile of these radiological findings is consistent with those in other series.[2,3,12,19]

On MRI, chondrosarcomas are iso- or hypointense on T1-weighted images and hyperintense on T2-weighted images. The tumor enhances either heterogeneously or homogeneously with gadolinium (Fig. 3A, B).[3,5,20] Angiographic studies usually show an avascular mass. Preoperative embolization is not necessary for the majority of these tumors and among the 11 cases reported in the literature only one tumor showed hypervascularity and necessitated preoperative embolization; this tumor was histologically proven to be a mesenchymal CSA.[11] The radiological differential diagnosis of jugular foramen CSAs includes jugular paragangliomas, schwannomas, meningiomas, and chordomas.[21] The other differential diagnoses should include aneurismal bone cyst, cholesteatoma, lymphangioma, inflammatory granuloma, invasive squamous cell carcinoma of the temporal bone, and metastatic disease. Furthermore, CSAs are most difficult to differentiate radiologically from chordomas because they

TABLE III.
Review of the English Literature.

Study	Sex/Age	Presenting Symptoms	Location	Treatment	Postoperative Radiotherapy	Tumor Resection	Histology (Grade)	Comments
Coltrera et al.[4]	F/24	ME mass, deficits of CNs IX, X, XI, XII	JF, ME, anterior foramen lacerum, sphenoid	Temporal craniotomy	No	Partial	NA	Residual tumor at the JF area
								Bronchopneumonia and death 12 days after surgery
Charabi et al.[9]	F/34	HL, vertigo, FN palsy, deficits of CNs X, XII	JF, ME, N	IFTA-type A	No	Total	I	Mastoid segment of FN involved: FN grafting
								Disease free at 1 yr
Reid et al.[10]	F/22	Diplopia, FN palsy, deficit of CNs IX, X, XI, XII	JF, CPA, N	RS (I stage)	No	Total	I	Well compensated IX–XII palsies
				IFTA-type A (II stage)				No F.U.
Reid et al.[10]	F/42	HL, pulsatile tinnitus, ME mass	JF, ME	IFTA-type A	No	Total	I	Lesion of CNs IX-XI
								No F.U.
Harvey et al.[2]	M/38	HL, headache, diplopia, ataxia, deficits of CNs IX, X	JF, PA, PCF, IAC, CC, Lateral clivus	Combined RS and canal wall down mastoidectomy + removal of the vaginal process	Yes	Total	II	Disease free at 4 mo
Watters et al.[7]	M/66	HL, deficits of CNs IX, X, XI, XII	JF, ME	IFTA	Yes	Total	I	Cricopharyngeal myotomy and Teflon injection
								Recurrence 5 yrs after surgery
								Died 5 yrs after surgery (brainstem infarct)
Donaldson et al.[3]	F/77	HL, vertigo, ataxia, deficits of CNs IX, X, XII	JF, PCF	TL+ RS	No	Intracapsular debulking	I	NA
Nozaki et al.[11]	M/15	Pulsatile tinnitus, deficit of CN X	JF	NA	Yes	Subtotal	Mesenchymal	Preoperative embolization
								Residual tumor around the JF
								No regrowth at 35 mo

(*Continues*)

TABLE III.
(*Continued*)

Study	Sex/Age	Presenting Symptoms	Location	Treatment	Postoperative Radiotherapy	Tumor Resection	Histology (Grade)	Comments
Neff et al.[12]	F/25	HL, pulsatile tinnitus, deficits of CNs IX, X, XI, XII	JF, PA, CC, ME, N, PCF, C2	Anterior, middle, posterior fossa craniotomy + IFTA + transcervical	No	Total	II	Sacrifice of ICA and CNs VI-XII
								Mastoid segment of FN involved
								CSF leak treated with ventricular drainage and ventriculoperitoneal shunt
								Medialization thyroplasty
								Gold weight upper eyelid implant
								Disease free at 2 yrs
Raghu et al.[13]	F/45	HL, headache, diplopia, ME mass	JF, ME, PA, Lateral clivus	Transtemporal-IFTA	Yes	Total	I	VI to XII CNs palsies
								Disease free at 6 yrs
Cummings et al.[14]	M/63	HL, gait imbalance	JF, CPA	Far lateral transcondylar	No	Total	I	NA

F = female; M = male; ME = middle ear; HL = hearing loss; CN = cranial nerve; FN = facial nerve; JF = jugular foramen; N = neck; CPA = cerebellopontine angle; IAC = internal auditory canal; PCF = posterior cranial fossa; PA = petrous apex; CC = carotid canal; C2 = second cervical vertebra; IFTA = infratemporal fossa approach; TL = translabyrinthine approach; RS = retrosigmoid approach; POTS = petro-occipital-trans-sigmoid approach; TC = transcochlear; CSF = cerebrospinal fluid; ICA = internal carotid artery; F.U. = follow-up; NA = data not available.

Fig. 2. Axial HRCT scan with bone windows of a right jugular foramen chondrosarcoma illustrating irregular bony erosion of the lateral margin of the jugular foramen and involvement of the mastoid segment of the facial nerve (arrow). Notice the intratumoral calcifications (case 2).

have very similar MRI and HRCT imaging appearances. Chordomas develop primarily in the central clivus area and may extend laterally to the temporal bone. In contrast, CSAs usually arise from structures lateral to the midline.[4] Table IV shows the radiological features of jugular foramen CSAs when compared with other more common neoplastic jugular foramen lesions. Although definitive preoperative diagnosis is difficult to achieve, modern imaging techniques (HRCT and MRI) help to distinguish CSAs from the other more common lesions of the jugular foramen. In uncertain cases, cerebral angiography often guides the surgeon to suspect a CSA. On the basis of HRCT and MRI findings, two of our patients were initially thought to have a CSA and three patients were misdiagnosed as having a paraganglioma. Angiography demonstrated an avascular lesion in all cases and a presumptive diagnosis of a CSA was then made. However, a definitive diagnosis was reached only by histological analysis.

Histopathologically, CSAs are divided into the following subtypes: conventional (myxoid and hyaline types), dedifferentiated, clear cell, and mesenchymal with the conventional form being the most common type within the skull base.[18,20] Dedifferentiated and mesenchymal CSA variants are more aggressive and may metastasize, but account for less than 10% of all skull base CSAs.[16] Rosenberg et al.[18] further subdivided the conventional type into three grades based on cellularity, nuclear size of the cells, chondroid matrix, and nuclear atypia: well differentiated (grade I), moderately differentiated (grade II), and poorly differentiated (grade III). Our cases along with those in the literature report that the majority of jugular foramen CSAs is of the conventional type with 74% of the grade I variety and 20% of the grade II variety. To date, only one case (6.6%) of the mesenchymal variant has been reported.[11] Chondrosarcoma of the skull base is frequently misdiagnosed histologically as chordoma, especially the chondroid variety of chordoma. Immunohistochemical analysis is particularly helpful to differentiate chordomas from CSAs. Chordomas stain positively for epithelial mem-

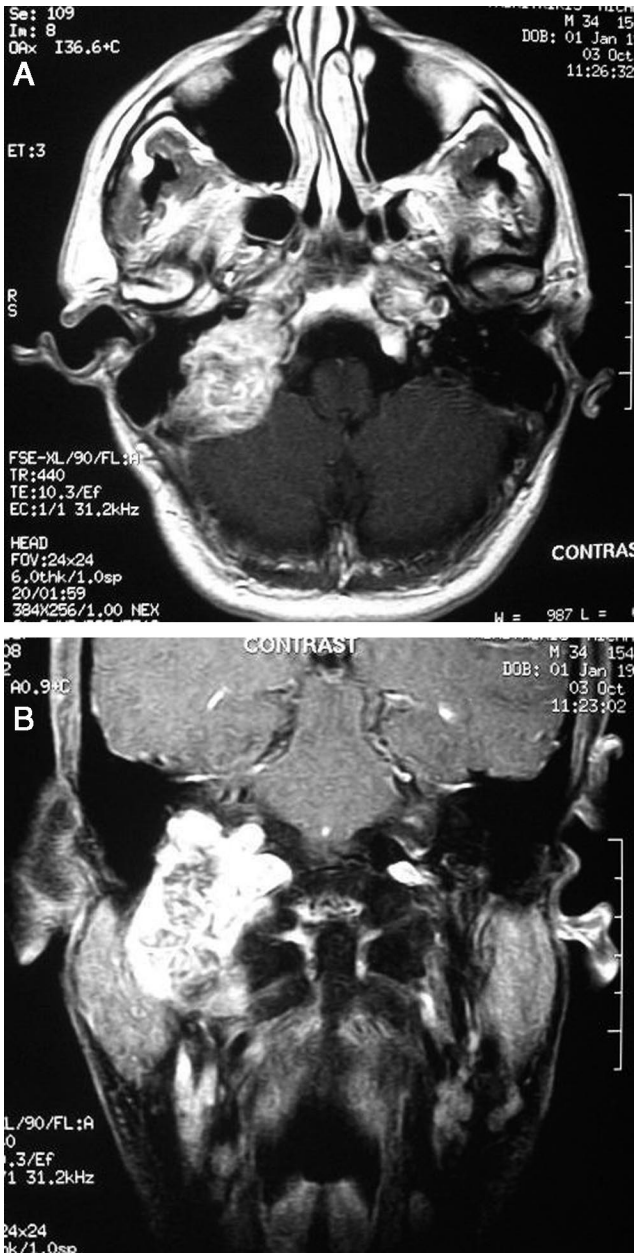

Fig. 3. (A) Axial T1-weighted MRI scan with gadolinium showing a right jugular foramen chondrosarcoma extending to the petrous apex. The tumor shows heterogeneous enhancement and appears confined to the extradural space even if the posterior cranial fossa dura is pushed medially. (B) Coronal T1-weighted MRI scan with gadolinium of the same tumor showing the intra- and extracranial extension of the lesion (case 3).

brane antigen, cytokeratin, vimentin, and S-100 protein. In contrast, chondrosarcomas stain positively only for S-100 protein and vimentin.[12,16]

Because of the rarity of jugular foramen CSAs, there is still no general consensus on the optimal treatment of these tumors. Combinations of surgical debulking, total surgical resection, different radiation techniques or the combination of surgery and postoperative radiation have been proposed for the treatment of chondrosarcomas of the skull base. We believe that surgical gross total resection of the tumor is the mainstay of treatment and management of jugular foramen CSAs. Adjuvant treatments such as proton beam radiotherapy, stereotactic radiotherapy (gamma knife or Cyber knife), or fractioned radiotherapy have also been successfully used.[15,16,22] Postoperative irradiation is usually recommended in patients with high risk of recurrence (tumors more aggressive than grade II) as well as in cases with residual or recurrent tumors. In our series, postoperative radiation therapy was indicated only for one patient with grade II CSA. Some authors propose radiotherapy as a primary treatment for chondrosarcomas reporting results equivalent to surgery and adjuvant radiotherapy[15] but the efficiency of radiotherapy in long-term tumor control is still unknown and further randomized controlled studies with longer follow-up are necessary. Furthermore, valid statistical comparisons between radiotherapy and surgery are difficult to make because of the small number of cases, both in our study and in the literature. Such an analysis in the shape of a prospective, randomized case-control study would necessarily involve multicenter efforts. In our opinion, the risks of radiotherapy should be well balanced against its potential benefits in such slow-growing tumors. We usually do not recommend radiotherapy as the primary treatment in these patients because salvage surgical resection could be extremely difficult in patients who fail primary irradiation. However, radiotherapy may be considered as an alternative primary treatment in selected cases in which there are serious contraindications to surgery (i.e., critical general medical conditions, elderly patients).

The complex anatomical location of these tumors, their frequent encroachment on neurovascular structures, and their potential ability to recur are some of the criteria that make the resection of these lesions particularly challenging. Any surgical approach to the jugular foramen should allow to control the sigmoid sinus and jugular vein. Additional goals of surgery should include preservation of the inner ear, middle ear and facial nerve function, when possible. Single stage total tumor removal should be attempted when possible. Different surgical approaches have been used for single-stage resection of jugular foramen CSAs including the IFTA-A,[7,9–10,23] transtemporal-infratemporal,[13] and far lateral transcondylar approaches.[14] The patient reported by Harvey et al.[2] underwent a combined retrosigmoid-canal-wall-down mastoidectomy, whereas the patient reported by Donaldson et al.[3] underwent a combined translabyrinthine-retrosigmoid approach. In one of the two patients reported by Reid et al.,[10] a planned staged procedure was adopted by using the retrosigmoid approach as the initial surgery and the infratemporal approach for the second stage. In the present series, the IFTA-A approach was used in two patients (cases 2 and 3) with preserved preoperative hearing; in one case the tumor extended into the hypotympanum and into the carotid canal and in the other patient the tumor extended cranially into the hypotympanum and infralabyrinthine region, and caudally into the upper neck. The IFTA-A[23], provides excellent control of the jugular foramen and infralabyrinthine areas, as well as the vertical portion of the internal carotid artery. Despite the excellent exposure of the jugular foramen area, the IFTA-A has the

TABLE IV.
Radiologic Differential Diagnosis in Tumors of the JF.

Lesion	HRCT with bone windows	T1	T2	MRI-Gd	Angiography
Chondrosarcoma	Irregular erosion of JF margins	Iso- or hypointense	Hyperintense	Heterogeneous or homogeneous enhancement	Very low degree of vascularity
	Intratumoral calcifications may be present				
Paraganglioma	Irregular erosion of JF margins	Hypointense	Isointense	Marked dishomogeneous enhancement with multiple flow void areas ("salt and pepper" appearance)	Marked hypervascularity
	Infiltrate surrounding bone				Rapid arterial phase tumor blush and early draining veins
	Caroticojugular spine is characteristically eroded				
Schwannoma	Enlargement of the JF with smooth well defined bony margins	Iso- or hypointense	Iso- or hyperintense	Marked or moderate enhancement	Mild vascularity
	No signs of bony infiltration			Intratumoral cysts may be present	Absence of strong tumor stain
Meningioma	May contain intratumoral calcifications	Iso- or hypointense	Hyperintense	Marked contrast enhancement	Mild/moderate vascularity
	Irregular erosion of JF margins			"Dura tail" sign	Slight arterial tumor blush with prolonged draining veins
	Infiltrate surrounding bone			Broad base	
	Mild hyperostosis may be present				
Chordoma	Irregular erosion of JF margins	Iso- or hypointense	Hyperintense	Moderate to intense enhancement	Low degree of vascularity
	Intratumoral calcifications may be present				
	Midline lesion				

JF = jugular foramen; HRCT = high-resolution computed tomography; MRI-Gd = gadolinium-enhanced magnetic resonance imaging.

disadvantages of postoperative conductive HL due to ear canal closure and facial nerve paresis related to the anterior rerouting of the facial nerve. If the tympanic cavity and facial nerve are not involved we prefer to use the POTS approach instead the IFTA-A. The surgical details of the POTS have already been published.[24,25] The POTS is a posterolateral approach to the skull base and offers a direct, conservative route to the jugular foramen and adjacent areas while keeping the facial nerve in place and preserving the external auditory canal and middle ear (Fig. 4A, B). Areas that can be checked by this approach include the jugular foramen, CPA, occipital condyle, the ipsilateral lower clivus, the vertical portion of the internal carotid artery, and the jugulocarotid space. The approach entails a retrolabyrinthine petrosectomy combined with a retrosigmoid craniotomy and occlusion of the sigmoid sinus and jugular vein. A limited neck approach is required to suture the jugular vein. We used the POTS approach in a normal-hearing patient (case 4) with a tumor involving the infralabyrinthine region extending into the posterior cranial fossa without middle and inner ear involvement. The POTS approach was combined with a transotic approach to resect a tumor (case 5) extending into the petrous apex together with a tumor component extending anteromedially to the vertical internal carotid artery.

Finally, a transotic approach with jugular vein ligation was performed in a patient (case 1) having a preoperative dead ear and a tumor involving the middle ear, external auditory canal, and the carotid canal together with extension into the CPA.

In our experience, the carotid canal was found eroded in three cases but the tumors did not infiltrate the adventitia of the internal carotid artery walls. The jugular bulb was already occluded by tumor in all the five cases.

Postoperative lower CN deficits may represent a serious complication especially in the elderly. Lower CN deficits may be due to intentional sacrifice or to functional loss despite anatomic integrity. On the basis of our data as

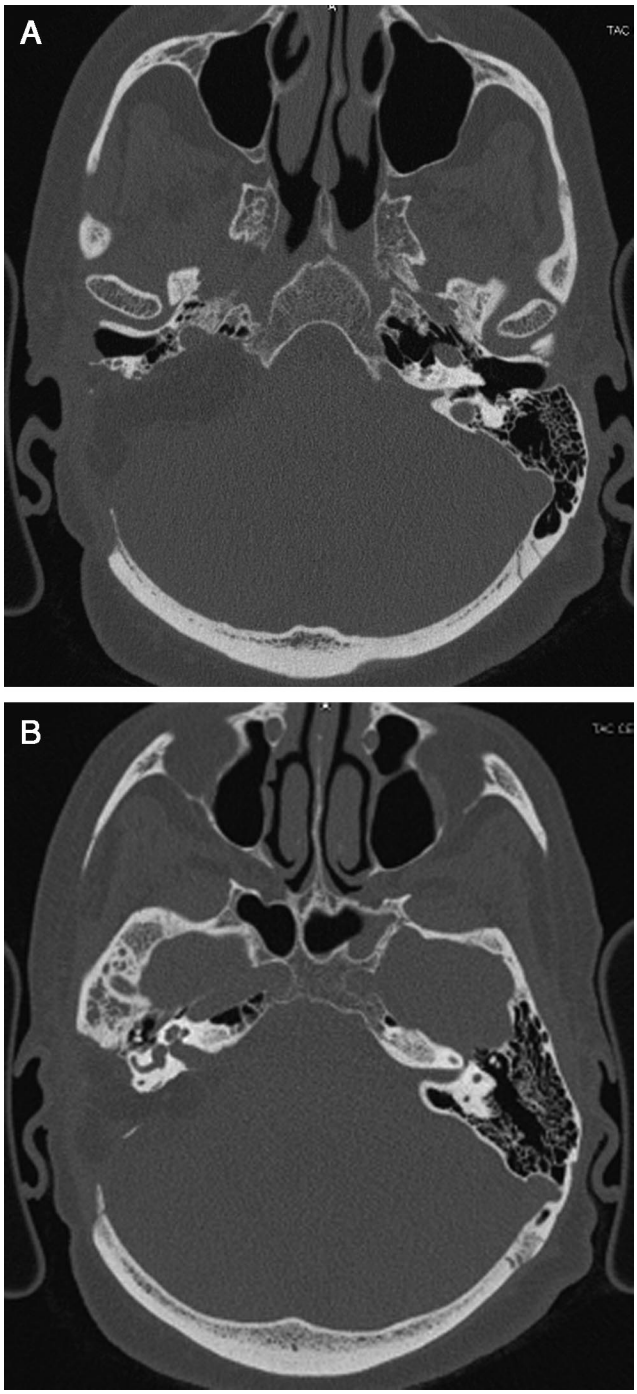

Fig. 4. Postoperative axial HRCT scans with bone window of a tumor (case 4) managed with a petro-occipital transigmoid (POTS) approach. Notice the extent of the bone removal medial to the jugular foramen (A) and the preservation of the hearing system obtained using the POTS approach (B).

well as those reported in the literature a postoperative deficit of CNs IX and X is almost the rule even in previously asymptomatic patients. When lower CN deficits are present preoperatively, the decision to resect the involved nerves is simple. The decision to sacrifice affected lower CN in previously asymptomatic patients is based on several factors. In our opinion, an aggressive removal of the tumor with the risk of lower CN dysfunction may be justified when the patient is young and wishes total tumor removal. Since suddenly palsy of previously functioning lower CNs in elderly patients would be met with serious morbidity, watchful expectancy should be adopted, and surgery is only indicated in the case of CN deficits or impending neurological complications. Though no recovery of lower CN deficits may be expected, on long-term follow-up, patients usually compensate well for their lower CNs loss by the help of speech and swallowing rehabilitation. Physical therapy is recommended in patients with CN XI paralysis to prevent persistent shoulder dysfunction and pain.

In our series, follow-up MRIs showed no evidence of tumor recurrence after an average of 32.8 months. Prolonged follow-up of our patient group is necessary because of the extremely long period it can sometimes take for a recurrence to manifest itself. No distant metastases developed in our and other series.

CONCLUSIONS

Chondrosarcomas of the jugular foramen are very rare tumors and represent an important diagnostic challenge. In these patients, the skull base surgeon must carefully consider the imaging studies. It is the specific combination of HRCT, MRI, and angiography features as well as a high index of suspicion that help to make the final diagnosis.

We believe that the primary treatment for CSAs of the jugular foramen is gross total surgical resection of the tumor. It is our philosophy to reserve postoperative radiotherapy for patients with tumors more aggressive than grade I, as well as in cases with subtotal resection and recurrent tumor.

BIBLIOGRAPHY

1. Cianfriglia F, Pompili A, Occhipinti E. Intracranial malignant cartilaginous tumours. Report of two cases and review of the literature. *Acta Neurochir* (*Wien*) 1978;45: 163–175.
2. Harvey SA, Wiet RJ, Kazan R. Chondrosarcoma of the jugular foramen. *Am J Otol* 1994;15:257–263.
3. Donaldson DR, Myers LL, Diaz-Ordaz E, Grand W, Paterson J, Wax MK. Pathologic quiz case 2. Chondrosarcoma of the jugular foramen. *Arch Otolaryngol Head Neck Surg* 1999; 125:229, 230–231.
4. Coltrera MD, Googe PB, Harrist TJ, Hyams VJ, Schiller AL, Goodman ML. Chondrosarcoma of the temporal bone. Diagnosis and treatment of 13 cases and review of the literature. *Cancer* 1986;58:2689–2696.
5. Lau DP, Wharton SB, Antoun NM, Bottrill ID, Moffat DA. Chondrosarcoma of the petrous apex. Dilemmas in diagnosis and treatment. *J Laryngol Otol* 1997;111:368–371.
6. Williams PL, Warwick R, Dyson M, Bannister LH. *Gray's anatomy*. New York: Churchill Livingstone; 1989.
7. Watters GW, Brookes GB. Chondrosarcoma of the temporal bone. *Clin Otolaryngol* 1995;20:53–58.
8. Brackmann DE, Teufert KB. Chondrosarcoma of the skull base: long-term follow-up. *Otol Neurotol* 2006;27:981–991.
9. Charabi S, Engel P, Bonding P. Myxoid tumours in the temporal bone. *J Laryngol Otol* 1989;103:1206–1209.
10. Reid CB, Fagan PA, Turner J. Low-grade myxoid chondrosarcoma of the temporal bone: differential diagnosis and report of two cases. *Am J Otol* 1994;15:419–422.

11. Nozaki K, Nagata I, Takahashi A, Haschimoto N. Intracranial mesenchymal chondrosarcoma associated with a left transverse sigmoid dural A-V fistole. *Acta Neurochir* (*Wien*) 1999;141:327–328.
12. Neff B, Sataloff RT, Storey L, Hawkshaw M, Spiegel JR. Chondrosarcoma of the skull base. *Laryngoscope* 2002;112: 134–139.
13. Raghu M, Ioannis M, De R, Moffat D. Chondrosarcomas of the temporal bone: presentation and management. *J Laryngol Otol* 2004;118:551–555.
14. Cummings TJ, Bridge JA, Fukushima T. Extraskeletal myxoid chondrosarcoma of the jugular foramen. *Clin Neuropathol* 2004;23:232–237.
15. Hug EB, Loredo LN, Slater JD, et al. Proton radiation therapy for chordomas and chondrosarcomas of the skull base. *J Neurosurg* 1999;91:432–439.
16. Tzortzidis F, Elahi F, Wright DC, Temkin N, Natarajan SK, Sekhar LN. Patient outcome at long-term follow-up after aggressive microsurgical resection of cranial base chondrosarcomas. *Neurosurgery* 2006;58:1090–1098.
17. House JW, Brackmann DE. Facial nerve grading system. *Otolaryngol Head Neck Surg* 1985;93:146–147.
18. Rosenberg AE, Nielsen GP, Keel SB, Renard LG, Fitzek MM, Munzenrider JE, Liebsch NJ. Chondrosarcoma of the base of the skull: a clinicopathologic study of 200 cases with emphasis on its distinction from chordoma. *Am J Surg Pathol* 1999;23:1370–1378.
19. Seidman MD, Nichols RD, Raju UB, Mehta B, Levy HG. Extracranial skull base chondrosarcoma. *Ear Nose Throat J* 1989;68:626–635.
20. Lustig LR, Sciubba J, Holliday MJ. Chondrosarcomas of the skull base and temporal bone. *J Laryngol Otol* 2007;121: 725–735.
21. Löwenheim H, Koerbel A, Ebner FH, Kumagami H, Ernemann U, Tatagiba M. Differentiating imaging findings in primary and secondary tumors of the jugular foramen. *Neurosurg Rev* 2006;29:1–11.
22. Krishnan S, Foote RL, Brown PD, Pollock BE, Link MJ, Garces YI. Radiosurgery for cranial base chordomas and chondrosarcomas. *Neurosurgery* 2005;56:777–784.
23. Fisch U. Infratemporal fossa approach to tumours of the temporal bone and base of the skull. *J Laryngol Otol* 1978; 92:949–967.
24. Mazzoni A, Sanna M. A postero-lateral approach to the skull base: the petro-occipital trans-sigmoid approach. *Skull Base Surg* 1995;5:157–167.
25. Sanna M, Saleh E, Khrais T, Mancini F, Piazza P, Russo A, Taibah A. Atlas of microsurgery of the lateral skull base. Stuttgart/New York: Georg Thieme Verlag; 2008.