Turgut Özal Tıp Merkezi Dergisi **6(1):1999**

CONSERVATIVE FACIAL NERVE MANAGEMENT IN JUGULAR FORAMEN SCHWANNOMAS

Yasar COKKESER, M.D.*
Derald E. BRACKMANN, M.D.**

Objective: *Although transposition of the facial nerve is crucial in infiltrative, vascular lesions involving the jugular foramen, it was our objective to show that a conservative approach to management of the facial nerve is sufficient in jugular foramen neuromas because of their non-infiltrative, less vascular nature and medial location in the jugular foramen.*
Patients: *Sixteen patients with jugular foramen schwannoma were treated (18 procedures) between January, 1975 and October, 1995. The eight males and eight females ranged in age from 13 to 66 years, with a mean age of 47.7 years.*
Intervention: *One-stage, total jugular foramen neuroma removal without transposition of the facial nerve, using a variety of surgical approaches.*
Main Outcome Measures: *Facial nerve transposition (yes or no), House-Brackmann facial nerve grade, lower cranial nerve status, complications.*
Results: *One stage total tumor removal was accomplished in all of the cases. In 13 (72%) of the neuromas, removal was accomplished without facial nerve transposition. Transposition was performed in two revision cases to control the carotid artery completely, two cases with large tumor extension anteriorly to the petrous apex and one case with extensive involvement of the middle ear. A House-Brackmann facial nerve grade I or II was obtained in 16 of the 18 procedures, with one grade III and one case which remained grade V, as it was preoperatively.*
Conclusions: *One stage, total tumor removal can be achieved with excellent control of the important vascular structures and without transposition of the facial nerve in the majority of jugular foramen schwannomas.*
Key words*: Facial nerve, schwannoma, jugular foramen*

* Inonu University, ENT Dept. MALATYA, TURKEY.
** House Ear Clinic and House Ear Institute, LOS ANGELES, CA

Juguler foramen nörinomlarının tedavisinde konservatif fasyal sinir yaklaşımı

Amaç: *Juguler forameni tutan, infiltrative, vasküler lezyonların tedavisinde yeterli expojur için fasyal sinir transpozisyonu önemlidir. Bu çalışmadaki amacımız juguler foramen nörinomlarının vaskülaritesi az, non infiltratif ve juguler foramende medialde yerleşmesi özellikleri nedeniyle fasyal siniri transpoze etmeden tedavi etmenin yeterli olacağını göstermektir.*
Hastalar: *Ocak 1975 ile ekim 1995 arasında juguler foramen nörinomu nedeniyle tedavi edilen 16 hasta (18 operasyon). Hastalar 8 erkek ve 8 kadın, 13 ile 66 yaşları arasında idi, ortalama 47.7 yıl.*
Müdahele: *Tek aşamalı fasyal siniri transpoze etmeden, çeşitli operasyonlarla total juguler foramen nörinomu eksizyonu.*
Ana değerlendirme kriterleri: *Fasyal sinir traspozisyonu (evet veya hayır), House-Brackmann fasyal sinir greydi, alt kranyal sinir durumu, komplikasyonlar.*
Bulgular: *Tek aşamalı, tüm hastalarda total tümor eksizyonu gerçekleştirildi. On üç hastada (%72) fasyal siniri transpoze etmeden, juguler foramen nörinomu çıkartıldı. İki revizyon vakada karotid arteri tam kontrol için, diğer olguda petroz apekse ulaşan çok büyük tümörlü iki hastada ve yaygın olarak orta kulağı tutan bir olguda transpozisyon uygulandı. On altı hastada House-Brackmann greyd 1 veya 2 fasyal sinir sonucu elde edildi, bir hasta da greyd 3, bir hastada preoperatif seviyesi olan greyd 5 olarak kaldı.*
Sonuç*: Juguler foramen nörinomlarının tedavisinde, tek aşamalı olarak total tümor eksizyonu, fasyal siniri transpoze etmeden, vasküler yapıların mükemmel kontrolüyle yapılabilir.*
Anahtar kelimeler*: Fasyal sinir, şvannom, juguler foramen*

Correspondance Address:
Yasar Cokkeser, M.D.
Necip Fazıl Kısakürek Cad.
Sancak 1 apt. No 10
MALATYA / TURKEY
Phone: 90 422 341 0660
Fax: 90 422 341 0728
ycokkeser@hotmail.com

Jugular foramen neuroma, is an uncommon pathology, and approximately 100 cases are reported in the literature[1-13], and only two series included more than 10 cases[4,13].

Surgical treatment of lesions involving the region usually results in loss of cranial nerve functions due to either involvement by extensive lesions in the jugular fossa or results from surgical approach used in order to gain access to the region. Introduction and perfection of the infratemporal fossa (ITF) approach into the otoneurosurgical armamentarium for the treatment of extensive glomus jugulare tumors, is a turning point in the treatment of the pathologies involving the jugular foramen area[14]. Classically ITF approach designed to treat glomus tumor includes anterior transposition of the facial nerve in order to obtain good control of the anterior extent of pathology as well as the carotid artery. Routine transposition of a normal facial nerve causes permanent conductive hearing loss and may lead to at least temporary deficit[5,15]. Several features of the jugular foramen schwannomas, unlike glomus jugulare tumors, allow complete tumor removal without transposition of the facial nerve and avoidance of these complications. Cranial nerves occupy the anteromedial portion of the jugular foramen so neuromas originating from these nerves grow medial to the jugular fossa, and they are less vascular and less infiltrative in nature than glomus jugulare tumors.

It is the purpose of this paper to review authors' experience of the surgical management of the jugular foramen schwannomas with special emphasize on conservative facial nerve management.

SURGICAL TECHNIQUE

Patient is placed supine and the head is turned to the contralateral side. The operation begins with a C shaped postauricular incision 3-4 cm posterior to the postauricular sulcus, extending into the upper neck. Inferiorly based periosteal flap is elevated as a second layer of closure. Identification of the extratemporal facial nerve is accomplished after its exit from stylomastoid foramen and control of the neurovascular structures in the neck is obtained. A complete mastoidectomy is accomplished and the bony labyrinth, descending facial nerve, sigmoid sinus (SS) are skeletonized. Posterior fossa dura is exposed anteriorly and 2-3 cm posteriorly to the SS (Figure 1).

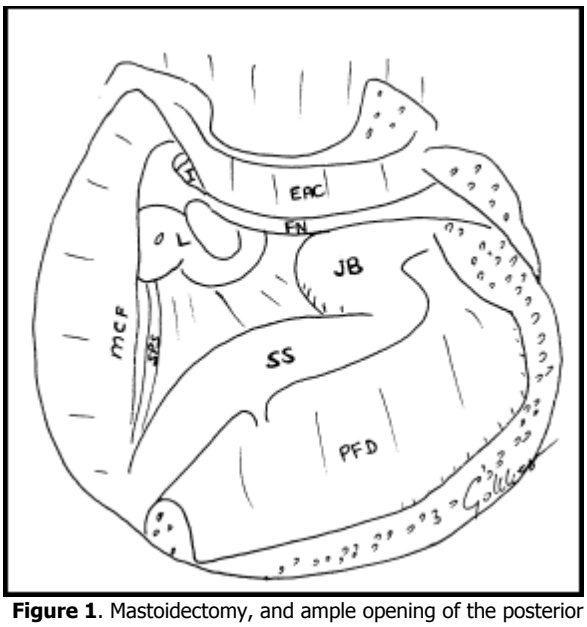

Figure 1. Mastoidectomy, and ample opening of the posterior fossa dura, including sigmoid sinus, upto the occipital condyle and foramen magnum.
SS: Sigmoid sinus, L: Labyrinth, EAC:External auditory canal, PFD: posterior fossa dura, MCFD: Middle cranial fossa dura, I: incus, JB:juguler bulbus, SPS:Superior petrosal sinus, OC: occipital condyle, FN: Facial nerve.

The sternocleidomastoid muscle is detached from the mastoid tip and the mastoid tip is removed. Bone removal over the posterior fossa is extended inferiorly up to the soft tissue around the foramen magnum with care of the vertebral artery in close proximity. Posterior belly of the digastric muscle is detached with care being taken to the facial nerve. The bone around the jugular bulbus is removed. External auditory canal and the facial nerve is left intact. Upper border of the exposure is labyrinthine block and internal auditory canal and the lower border is the soft tissue around the foramen magnum and occipital condyle. Anterior border of the exposure is foramen lacerum and the carotid artery and anterolaterally distal part of the third portion of the facial nerve. The jugular vein is double ligated in the neck and SS is packed extraluminally. Sigmoid sinus and the jugular vein are opened. The inferior petrosal sinus is packed carefully without damaging the ninth nerve which has close relationship to this nerve. The condylar emissery vein is also packed

if present The posterior fossa dura is opened and exposure of the pathology is obtained (Figure 2).

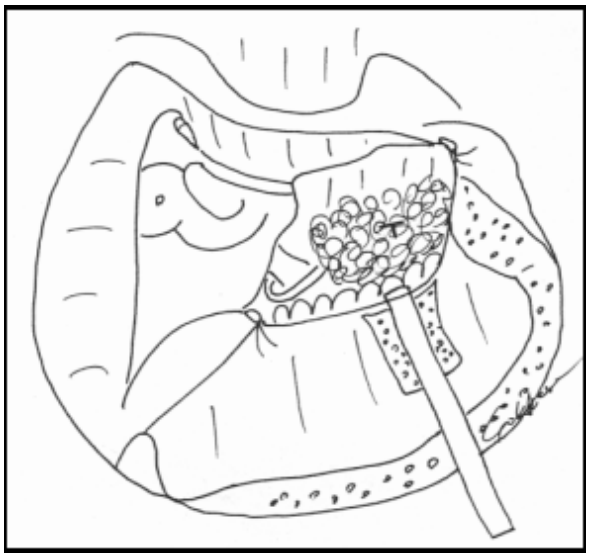

Figure 2. Exposure of the tumor after opening of the posterior fossa dura and occluding the sigmoid sinus. AICA: Anterior inferior cerebellar artery, T: tumor, C: Cerebellum, 8: Intradural part of 8th nerve. Other legends as in figure 1.

Tumor removal is carried out by gutting inside the tumor by taking care of anteriorly located carotid artery. Total tumor removal is accomplished in one stage (Figure 3).

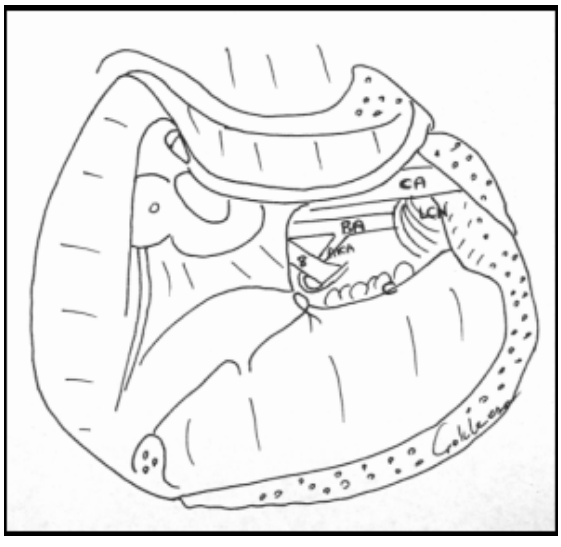

Figure 3. Exposure of the cerebello-pontine angle anatomy after tumor removal. The carotid, basilar artery, AICA, and the 8th nerve are seen. CA: Carotid artery, BA: Basilar artery, LCN: Lower cranial nerves exiting jugular foramen. Other legends as in figure 1 and 2.

Dural closure is carried out and cavity is obliterated with abdominal fat. Two layer closure of the mastoid is carried out. Mastoid dressing is applied. A lumbar subarachnoid drain is established.

Partial facial nerve transposition, distal to the second genu can permit removal of the pathologies near and anterior to the facial nerve at the stylomastoid foramen (Figure 4).

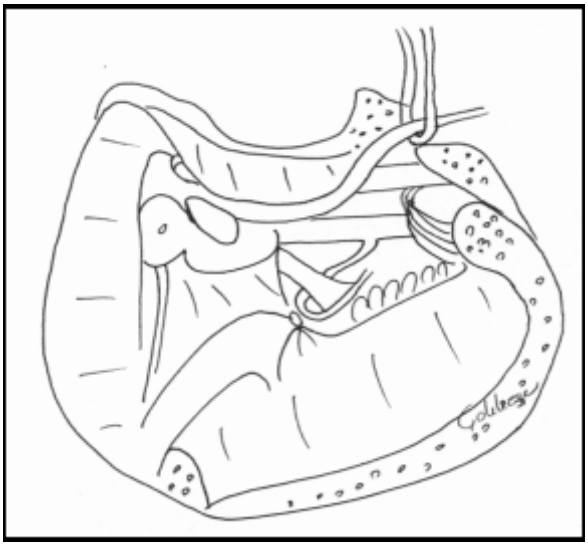

Figure 4. Partial mobilisation of the facial nerve distal to the second genu.

MATERIAL AND METHODS

Subjects: Sixteen patients with jugular foramen schwannoma were surgically treated at the House Ear Clinic from January 1975 to October 1995. Eight males, and eight females ranged in age13 to 66 years with a mean age of 47.7 years. Tumors were on the left in 13 and the right in 3 cases.

Medical records were retrospectively reviewed for patient presenting symptom characteristics and management, treatment outcome, and cranial nerve management with a special emphasize on facial nerve. Because two cases had later recurrences, 18 surgical procedures were performed. Each procedure was evaluated as a separate case and outcomes are presented for the total 18 procedures.

RESULTS

Preoperative symptoms included mainly, audiovestibular and lower cranial nerve (LCN)

findings (table 1).

Table 1. Presenting Symptoms

A- Audiovestibular symptoms	
Vestibular symptoms	10
Imbalance	8
Ataxia	1
Vertigo	1
Hearing loss	9
SNHL	7
Conductive	2
Tinnitus	8
Fullness	2
Middle ear mass	2
B- Lower Cranial nerve dysfunctions	
Hoarseness	10
Shoulder atrophy, pain	6
Tongue atrophy, Hipomotility	6
Headache	3

The most frequent symptoms were vestibular symptoms, hoarseness, tinnitus and hearing loss. Tumor sizes ranged 3 to 8 cm with a mean of 4.5 cm. Tumor location included the jugular foramen, intracranial space and extension to the neck or frequently a combination of these (table 2).

Table 2. Tumor location.

Tumor location	**number of cases**
Jugular foramen/Intracranial space /Neck extension	7
Jugular foramen/Intracranial space	6
Jugular foramen	3
Intracranial space	1
Neck extension	1

A variety of approaches were used in the surgical treatment of these cases (table 3).

Table 3. Surgical approaches used in the management

Surgical approaches	**number of cases**
Infratemporal fossa approach /Facial nerve transposition	5
Infralabyrinthine	5
Translabyrinthine/suboccipital	3
Retrolabyrinthine/suboccipital	3
Retrolabyrinthine/extended facial recess	2

The facial nerve was left in place in 13 cases and transposed anteriorly in 5 cases. In two of the five cases, tranposition of the facial nerve was required due to scar tissue from previous operation which impeded safe removal of tumor from the carotid artery. Two other cases had a large extension to the anterior petrous apex and in one case extensive middle ear involvement necessitated the tranposition of the facial nerve. Postoperative facial nerve results were normal (Grade 1) or the same as preoperatively in 15 cases, and transient paresis (Grade II or III) developed in 3 cases (table 4).

Table 4. Facial nerve grades (House- Brackman)

Facial nerve Grade	**Preoperative #of cases**	**Postoperative #of cases**
I	17	14
II	-	2
III	-	1
IV	-	-
V	1	1
VI	-	-

Two of these three cases underwent an infratemporal fossa approach with anterior transposition of the facial nerve. The other case was operated by an extended facial recess approach. Two of these three cases resulted in grade II and one grade III facial nerve result. Lower cranial nerve management is shown in Table 5.

Table 5. Postoperative lower cranial nerve status compared to preoperative function

Postoperative LCN status	**number of cases***
No LCN function difference	8
All preoperative LCN functions preserved	3
All LCN functions missing preoperatively	5
1 or 2 LCN functions lost	5
1 LCN function loss	3
2 LCN function loss	2
3 or 4 LCN functions lost	3

*Lower cranial nerve results are presented in only the 16 primary procedures.

No changes in LCN function difference occured in 8 cases (50%). In three of these cases succesful preservation of all LCN functions obtained and in 5 cases LCN functions were missing preoperatively. In other patients, varying deficits of single to all LCNs occured. The eight cases with no difference in LCN function compared to preoperative level had an uneventful recovery (table 6), while other patients had postoperative swallowing and speech diffculty. The most prolonged

Table 6. Postoperative lower cranial nerve function compensation.

Postoperative course	
* Normal or slight swallowing or speech difficulty	8
* Swallowing or speech difficulty took mean 6 months to compensate	7
* Swallowing and speech difficulty took more than a year to compensate	3

Compensation occured in two of the three cases with postoperative function loss in which all lower cranial nerves were intact preoperatively. Rehabilitation included swallowing training, vocal cord injection, laryngoplasty in majority and cricopharyngeal myotomy and gastrostomy in one case each (table 7).

Table 7. Additional procedures to rehabilitate swallowing and speech

Procedure	**number of cases**
Vocal cord injection	4
Laryngoplasty	3
Myotomy	1
Gastrostomy	1

Other than swallowing and speech difficulty, complications included three CSF leak requiring surgical closure and in one case wound infection followed CSF leak. Five cases were secondary procedures. One stage, total tumor removal were accomplished in all cases. Two recurrences appeared 7 and 8 years after primary operations. One of these cases died 36 days after revision as a result of complicated CSF leak followed by wound breakdown, meningitis and coma.

DISCUSSION

Succesful treatment of lesions involving the jugular foramen area includes complete tumor removal without additional neurological deficit.The jugular foramen is a key structure in the lower skull base with a complex and variable anatomy[16]. It is formed by anteromedial neural compartment and posterolateral venous compartment into which several venous sinuses enter. The jugular foramen is actually a canal rather than a simple foramen with a changing relationship to each other. It serves as a major port of exit for the cranial nerves 9 through 11 and as the major venous drainage of the brain. The vertical portion of the facial nerve stradles over the jugular bulb. Lesions arising from jugular foramen intimately involves this neural structures and thus surgical treatment of these lesions results in cranial nerve loss. Frequently lower cranial nerves are inevitably lost due to direct involvement by pathology. But the morbidity of facial nerve frequently results from surgical procedures used to reach the pathology. Anterior transposition of the facial nerve is crucial in controlling extensive, infiltrative, vascular lesions involving the hypotympanum and the lateral face of the jugular fossa as well as safe tumor removal from the carotid artery.

Unlike an infiltrative, vascular glomus jugulare tumor or meningioma, neuromas originating from the jugular foramen with their several properties, lend themselves to total tumor removal without transposition of the facial nerve. Jugular foramen neuromas originate from medially located nerves in the jugular bulb, and show expansive growth patterns without infiltration of the carotid artery or jugular vein. The medial location of these tumors provides enough space for tumor exposure and removal while preserving facial nerve in its location especially in cases with well-developed infralabyrinthine cells. In addition, the friable, easy to remove, less vascular nature of the neuromas are helpful factors.

Despite the variety of approaches which were used for different location and sizes of pathology, % 72 of the cases in this series had one stage. Total tumor removal is achieved with excellent control of the important vascular structures without transposition of the facial nerve. However, in five cases, (28 %), transposition of the facial nerve was required either because scar tissue prevented safe removal of the tumor from the carotid artery or tumor was very large, reaching the petrous apex or there was extensive middle ear involvement. In these cases, transposition of the facial nerve was accomplished with preservation of surrounding fibrous tissue at the stylomastoid foramen and continuous intraoperative facial nerve monitoring[15,17].

In our experience, most jugular foramen

schwannomas can be managed without facial nerve transposition. When necessary, a partial mobilization of the inferior half of the vertical facial nerve could be accomplished in order to control anterior extension of the pathology, or the approach be modified into an ITF approach at any time[18]. Following this philosophy, we obtained House-Brackmann grade I and II facial nerve results in 16 procedures. One case was preserved at the preoperative grade and one case had a grade III result.

Postoperative swallowing and speech difficulty was the most significant morbidity resulting from surgical treatment of jugular foramen lesions. Coordinated function of the 9th, 10th and 12th nerve regulates the swallowing and speech. Postoperative swallowing and speech difficulty found to be directly correlated with the acute difference in cranial nerve function. Eight cases 50% had no postoperative difference in lower cranial nerve function from pre- to postoperative periods. Eight cases with no postoperative in lower cranial function had a smooth postoperative course with a quick recovery. The other cases had postoperative speech and swallowing difficulty. In the five cases in which one or two of these nerves were sacrificed, an average of 6 months was required to compensate the deficit. The most prolonged postoperative compensation period was observed in two of the three cases in whom all of the lower cranial nerves were intact preoperatively, but was lost postoperatively. Compensation of these three patients took more than a year with a mean of 15 months. Rehabilitation of these cases was assisted by surgical measures as well as the physical swallowing and speech therapy. Speech and swallowing difficulty benefited from vocal cord injection, laryngoplasty and swallowing training.

Three cases with CSF leak required surgical repair. CSF leak is the most dreaded complication in these cases[5]. Intraoperative establishment of the lumbar drain helped in obtaining total tumor removal in one stage without increased CSF leak. Two cases of recurrences appeared 7 and 8 years after the original operation. One of recurrent cases developed CSF leak and infection, and died 36 days after the operation. Scar tissue resulting from previous surgical operation also made difficult to preserve important neurovascular structures.

A quite variable involvement of locations with variable extension is observed causing varied symptomatology[4,10]. A cranial nerve symptom in a patient should alert the physician for the possibility of cerebellopontine angle pathology, and an adequate evaluation should be performed[19].

One stage, total jugular foramen neuroma can be achieved without transposition of the facial nerve in 72% of the cases in our series. Total tumor removal can be achieved with minimum complications.

REFERENCES

1. Horn KL, House W, Hitselberger WE. Schwannomas of the jugular foramen. Laryngoscope 1985 :761-5.
2. Graham MD, LaRouere MJ, Kartush JM. Jugular foramen schwannomas: Diagnosis and suggestions for surgical management Skull Base Surgery 1991, 1:34-8.
3. Crumley RL, Wilson C. Schwannomas of the jugular foramen. Laryngoscope 1984;772-8.
4. Kaye AH, Hahn JF, Kinney SE, Hardy RW, Bay JW. Jugular foramen schwannomas. J Neurosurg 1984, 60:1045-53.
5. Franklin DJ, Moore GF, Fisch U. Jugular foramen peripheral nerve sheath tumors. Laryngoscope 1989, 99 :1081-7.
6. Arenberg IK,McCreary HS. Neurilemmomas of the jugular foramen. Laryngoscope, 1971, 81:544-57
7. Gacek RR. Schwannomas of the jugular foramen. Ann Otol Rhinol Laryngol 1976;89:215-24.
8. Maniglia AJ, Chandler JR, Goodwin WJ et al . Schwannomas of the parapharyngeal space and the jugular foramen. Laryngoscope1979; 89: 1405-15.
9. Neely JG Reversible compression neuropathy of the eight cranial nerve from a large jugular schwannoma. Arch Otolaryngol 1979:105:555-60.
10. Kinney SE, Dohn DF, Hahn JFWood BG. Neuromas of the jugular foramen. In Neurologic surgery of the Skull Base. Brackmann DE. (Ed.) Raven Press New York, NY 1979;pp.361-7.
11. Raquet F, Mann W, Amedee R Maruer J, Gilsbach J. Functional deficits of cranial nerves in patients with jugular foramen lesions. Skull Base Surgery 1991; 1: 117-9.
12. Mann W, Amedee R, Gilsbach J, Perneczky A, Wolfensberger M. Transsigmoid approach for tumors of the jugular foramen. Skull Base Surgery,1991; 3:137-41.
13. Pluchino F, Crivelli G, Vaghi MA. Intracranial neurinomas of the nerves of the jugular foramen. Acta Neurochirurgica 1975; 31: 201-21.
14. Fisch U, Pilsbury HC. Infratemporal fossa approaches to lesions in the temporal bone and base of the skull Arch Otolaryngol 1979;105:99-107.
15. Brackmann DE. The facial nerve in the infratemporal approach Otolaryngol Head Neck Surg 1987;97:15-7.
16. Saleh E, Naguib M, Aristegui M, Cokkeser Y, Sanna M, Lower skull base: Anatomic study with surgical implications. Ann Otol Rhinol Laryngol 1995;104: 57-61.
17. Leonetti JP, Brackmann DE, Prass RL. Improved preservation of facial nerve function in the infratemporal approach to the skull base. Otolaryngol Head Neck Surg 1989; 101:74-8.
18. Jackson CG, Cueva RA, Thedinger BA, GlasscockME. Conservation surgery for glomus jugulare tumors:The value of early diagnosis. Laryngoscope 1990:1031-6.
19. Leonetti JP, Brackmann DE. Glomus vagale tumor: The significance of early vocal cord paralysis. Otolaryngol Head Neck Surg 1989;100 :533-7.