

GLOMUS RADIOCIRUGÍA

Dra. Loreto Yáñez S.
Radio-oncóloga Neurocirujana

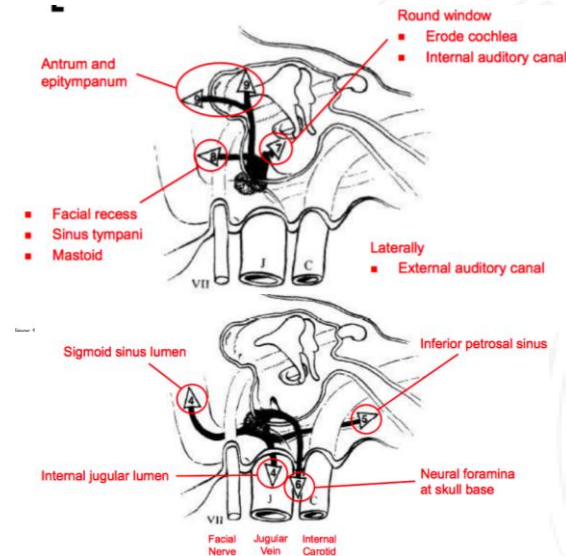
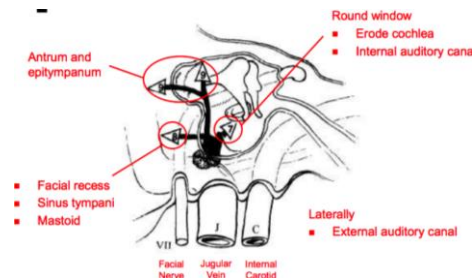
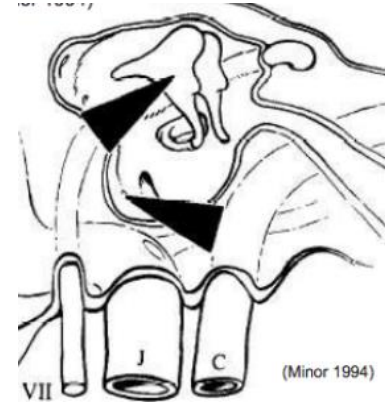


Introducción

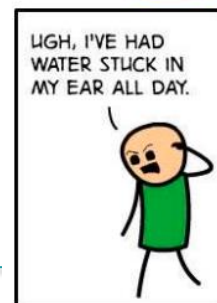
- 1941
- Tumor infrecuente en gral, pero es el mas frecuente en oído medio y f. Yugular
- Lento crecimiento
- Hipervascularizado
- Aparece en f. Yugular del hueso temporal
- Suele afectar PC bajos
- Se le incluye dentro de los paragangliomas:
 - de cuerpo carotídeo, glomus vagal, glomus timpánico.
- 5-10% tu múltiples
- <10% Desorden familiar A. dominante

Patrón de diseminación de los Glomus:

- Región del bulbo yugular (seno petroso inf, vena yugular interna, seno sigmoide)
- Protímpano, hipotímpano, mesotímpano
- Intracraneal



- 6:1 mujeres/hombres en 5 y 6 décadas
- Por el cuadro insidioso: retraso en diagnóstico.
- 4% pueden metastizar a



(<http://www.explosm.net>)



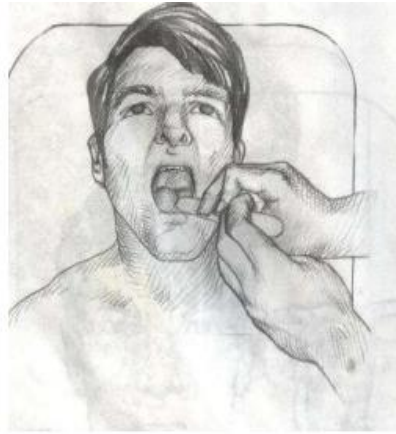
Cyanide and Happiness © Explosm.net



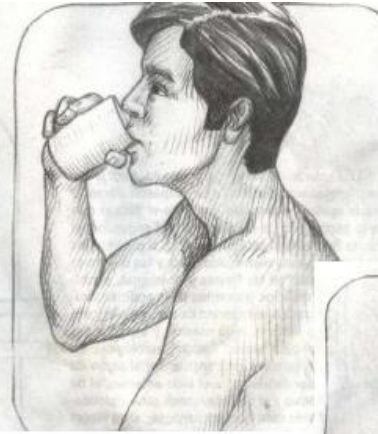
Ser hipocondriaca
me pone enferma.

Viet Pham
MD
Dayton Young
MD

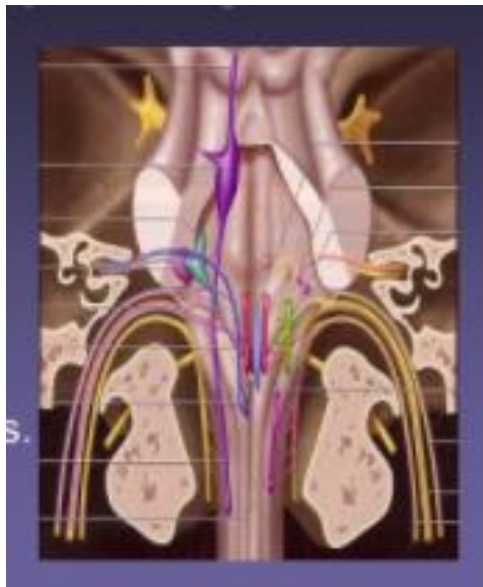
- Síndrome de Foramen Yugular (S. Vernet) es patognomónico:
 - paresia de PC IX a XI [3]



IX par



IX y X par



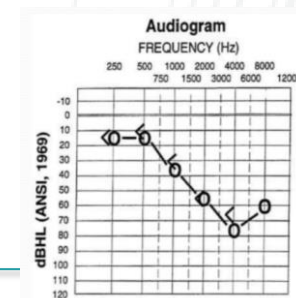
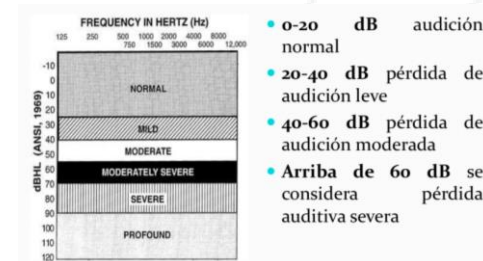
- Náuseas, deglución, elevación de paladar (9, glosofaríngeo)
- Aus. Náuseas, paresia de cuerdas vocales, reflujo nasal (10, vago)
- Elevación de hombros y rotar cuello (11, espinal)

- Hipoacusia conductiva 80%
- Tinnitus pulsátil 60%
- Otros: s. oído tapado, otorrea, hemorragia, soplo, masa de oído medio. Dolor de oreja (raro) .
- Si afecta oído interno: vértigo, hipoacusia sensitivo-neural

Estudio

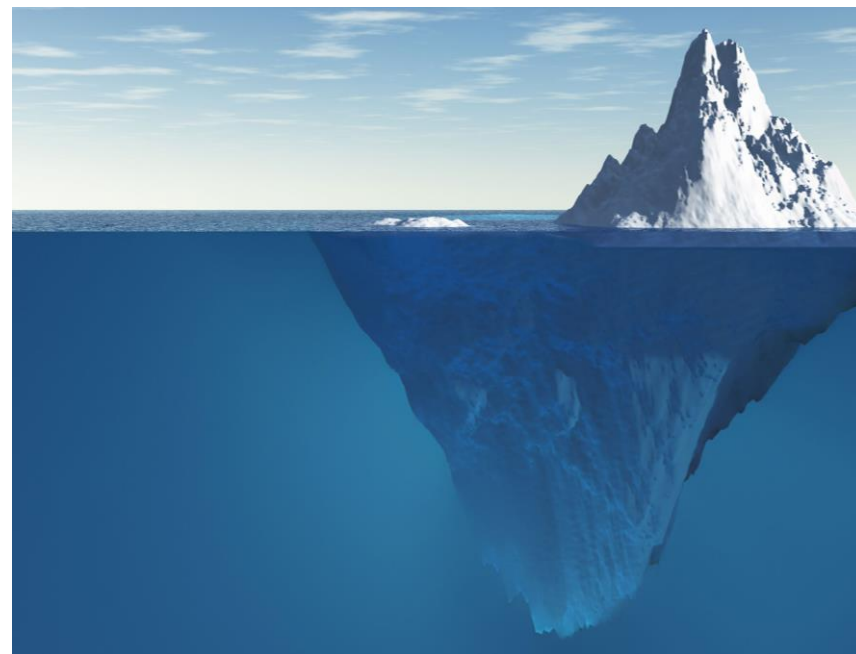
- Otoscopía : tumor rojo-azulado, pulsátil, tras la membrana timpánica (iceberg).
- Audiometría:
 - Hipoacusia conductiva y sensorineural*

Audiometría:
rojo O: oído derecho, azul y X: oído izq.
<> con o sin enmascarar

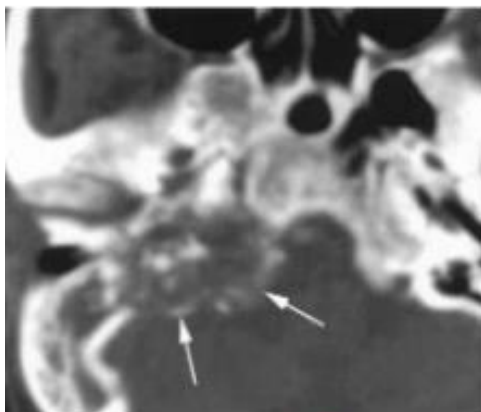




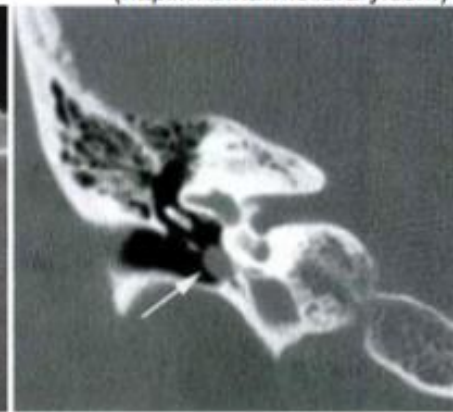
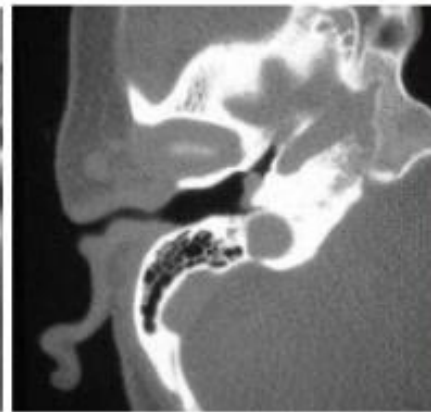
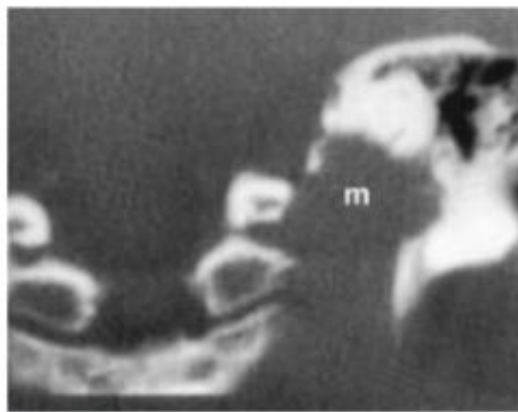
(N Engl J Med 2010; 362:e66.)



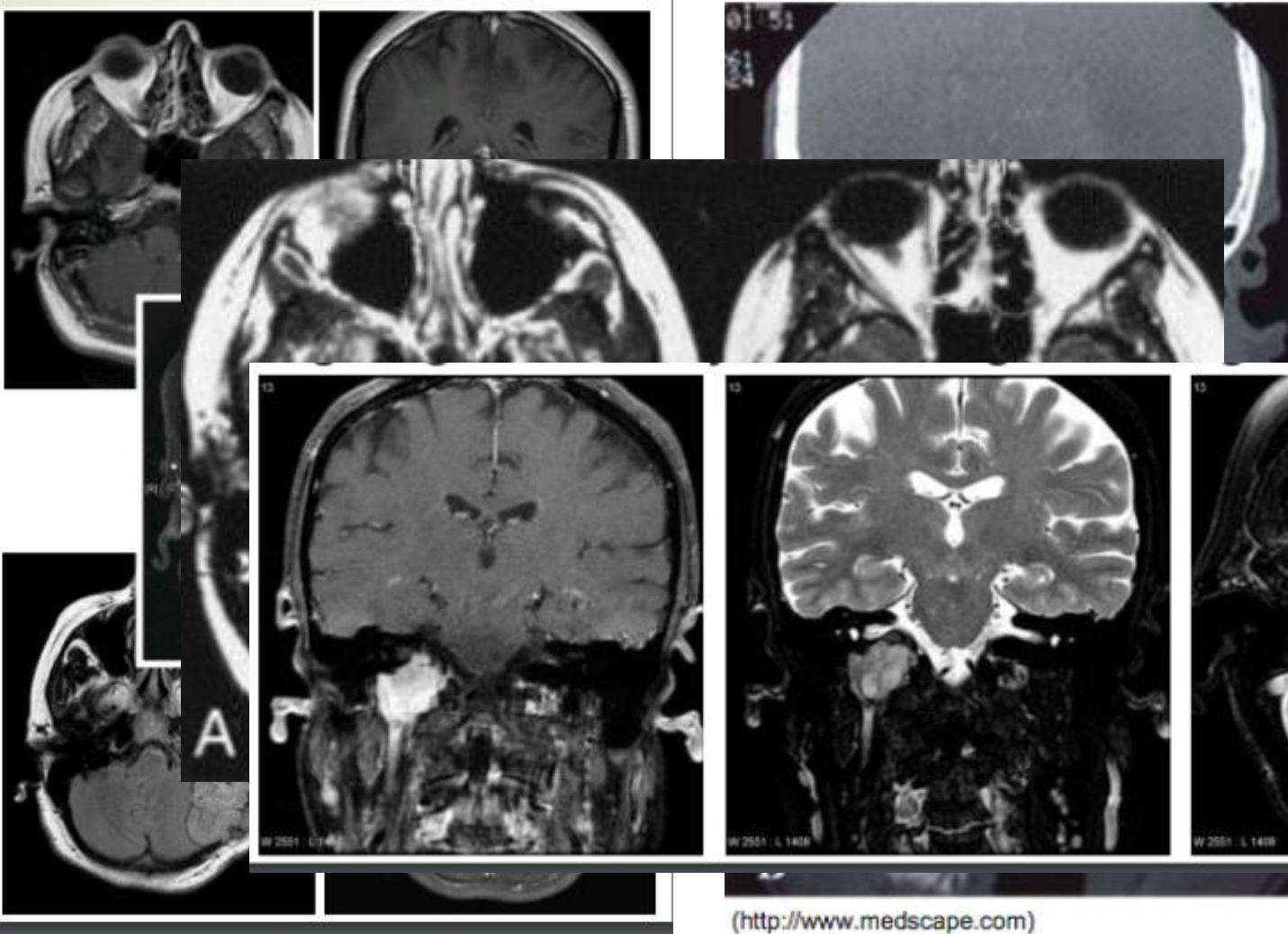
- Rx cráneo:
 - Crecimiento del foramen yugular y fosa.
- TAC Axial, coronal
 - Destrucción ósea.
- RNM con gadolínico (DTPA) , T2 exagera dimensiones.



Glomus Jugulare

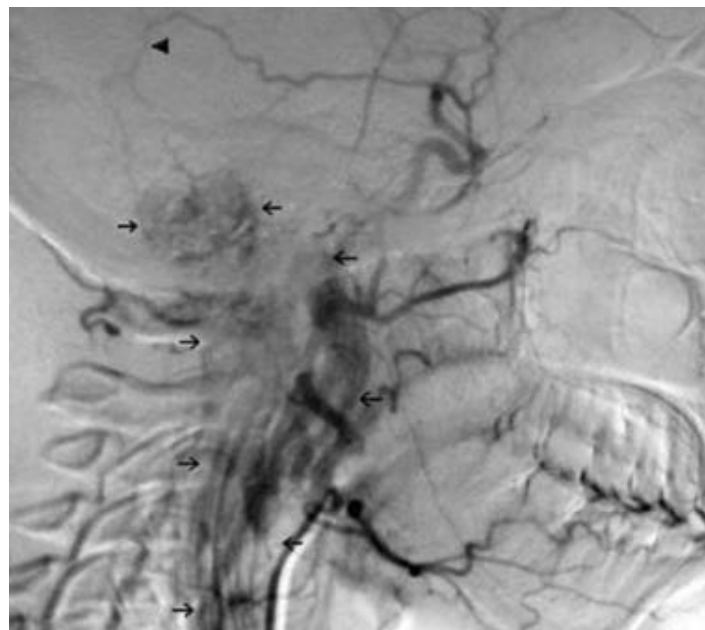


Glomus Tympanicum



(<http://www.medscape.com>)





Clasificaciones:

- Glasscock-Jackson
- Fisch

Clasificación de FISCH

- Se basa en la extensión del tumor a estructuras anatómicas vecinas
- Se relaciona con morbilidad y mortalidad.

GLOMUS TUMORS

TYPE	CHARACTERISTICS
A	Limited to middle ear cleft
B	Limited to tympanomastoid complex No infralabyrinthine involvement
C	Involves labyrinthine compartment Extends to petrous apex
D	Intracranial involvement

Tratamientos



- 1.- Observación, Terapia medicamentosa
- 2.- Neurocirugía
- 3.- Radiocirugía o RDT fraccionada
- 4.-Embolización (no curativa, preop)
- 5.- Mixta

Observación

- U. Vanderbilt en añosos, si no hay compr de tronco, sospecha de malignidad, observar.
- 15 pacs. (80% mujeres; mediana de edad 69.6 años), 5 crecen rx. 0.8 mm/año (0.6-1.6 mm/año), al crecer no aparecen inmediatamente síntomas —————> RNM gd

Tratamiento médico:

- bloqueo adrenérgico si secretan catecolaminas.

Adrenérgicos

Cansancio
Sudación
Taquicardia
Palpitaciones
Temblor
Ansiedad o nerviosismo
Irritabilidad
Sensación de hambre
Náuseas
Vómito
Palidez
Parestesias

Cirugía

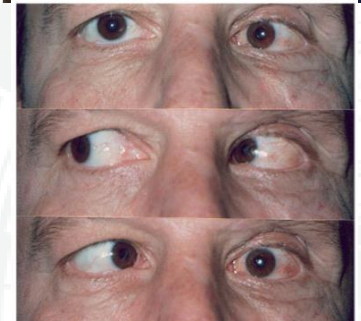


- Terapia de elección, aunque Radioterapia (SRS 1 o hipoFx) permite un control local y manejo de síntomas de paresia de PC.



Cirugía

- Complicaciones quirúrgicas:
- Sangrado
- Nuevas paresias de P.Craneales
- Fístula LCR (riesgo de meningitis)
- Perforación de la Mb. timpánica
- Colesteatoma





RADIOCIRUGÍA VERSUS CIRUGÍA:

- RT MEJOR CONTROL TUMORAL
- RT MENOS COMPLICACIONES
- RT MENOS PROBABILIDAD DE DETERIORO DE PARES CRANEALES.

TABLE 2. Comparison of the treatment outcomes of jugular paragangliomas treated with surgery or radiotherapy.

Parameter	Surgical series, mean rate (95% CI)	Radiotherapy series, mean rate (95% CI)	<i>p</i> value
Tumor control	78.2% (72.2–84.2)	91.5% (85.3–93.3)	.002
Deaths by the tumor	0.8% (-0.2–1.8)	1.4% (-0.6–2.8)	.54
Deaths by treatment	1.6% (0.6–2.6)	1.2% (0.2–2.2)	.56
Major complications	28.2% (19.5–36.9)	11.4% (4.4–18.4)	.003
Cranial nerve palsies after treatment (per patient)	0.9 (0.7–1.1)	0.08 (-0.05–0.2)	< .001

Abbreviations: CI, confidence interval.

Complex tumors of the glomus jugulare: criteria, treatment, and outcome.

Al-Mefty O¹, Teixeira A.

Ⓜ Author information

Abstract

OBJECT: Tumors of the glomus jugulare are benign, slow-growing paragangliomas. Their natural history, surgical treatment, and outcome have been well addressed in the recent literature; however, there remains a subgroup of complex tumors—multiple, giant, malignant, neuropeptide-secreting lesions, and those treated previously by an intervention with an adverse outcome—that is high risk, presents surgical challenges, and is associated with treatment controversy. In this article the authors report on a series of patients with complex glomus jugulare tumors and focus on treatment decisions, avoidance of complications, surgical refinements, and patient outcomes.

METHODS: In this retrospective study, the patient population was composed of 11 male and 32 female patients (mean age 47 years) with complex tumors of the glomus jugulare who were treated by the senior author within the past 20 years. These include 38 patients with giant tumors, 11 with multiple paragangliomas (seven bilateral and four ipsilateral), two with tumors that hypersecrete catecholamine, and one with a malignant tumor. Six patients had associated lesions: one dural arteriovenous malformation, one carotid artery (CA) aneurysm, two adrenal tumors, and two other cranial tumors. All but one patient presented with neurological deficits. Cranial nerve deficits, particularly those associated with the lower cranial nerves, were the prominent feature. Twenty-eight patients underwent resection in an attempt at total removal, and gross-total resection was achieved in 24 patients. Particularly challenging were cases in which the patient had undergone prior embolization or CA occlusion, after which new feeding vessels from the internal CA and vertebrobasilar artery circulation developed. The surgical technique was tailored to each patient and each tumor. It was modified to preserve facial nerve function, particularly in patients with bilateral tumors. Intrabulbar dissection was performed to increase the likelihood that the lower cranial nerves would be preserved. Each tumor was isolated to improve its resectability and prevent blood loss. No operative mortality occurred. In one patient hemiplegia developed postoperatively due to CA thrombosis, but the patient recovered after an endovascular injection of urokinase. In four patients a cerebrospinal fluid leak was treated through spinal drainage, and in five patients infection developed in the external ear canal. Two of these infections progressed to osteomyelitis of the temporal bone. There were two recurrences, one in a patient with a malignant tumor who eventually died of the disease.

CONCLUSIONS: Despite the challenges encountered in treating complex glomus jugulare tumors, resection is indicated and successful. Multiple tumors mandate a treatment plan that addresses the risk of bilateral cranial nerve deficits. The intrabulbar dissection technique can be used with any tumor, as long as the tumor itself has not penetrated the wall of the jugular bulb or infiltrated the cranial nerves. Tumors that hypersecrete catecholamine require perioperative management and malignant tumors carry a poor prognosis.

PMID: 12507134 DOI: 10.3171/jns.2002.97.6.1356

- Retrospectivo de 43 pacientes.
- 24 ptes resección completa
- al embolizar previo a la cirugía o al ocluir A carótida dificulta la cirugía
- Una **trombosis carotídea** (urokinasa)
- 4 fístulas de LCR
- 5 infecciones canal auditivo externo (**2 osteomelitis**)

Jugular and vagal paragangliomas: Systematic study of management with surgery and radiotherapy

HEAD & NECK—DOI 10.1002/HED AUGUST 2013

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- Rev sistemática => 69 artículos:
 - 41 de cirugía = 1310 ptes
 - 20 de RT = 461 ptes
 - 14 de SRS = 261 ptes
- Ningún RCT ni prospectivo

CIRUGÍA:

- n: 1084 ptes
- Seguimiento: **5.5 años**

- **85 % control tumoral**
- **1 % mortalidad por
progresión de
enfermedad**



- Complicaciones:

- **2 x parálisis** de P.Craneales
- **2 % de pacientes falleció** por complicaciones del post-op.

TABLE 3. Preoperative and postoperative cranial nerve impairment.

Cranial nerve	JPG preoperative, no. of patients	JPG postoperative, no. of patients	VPG preoperative, no. of patients	VPG postoperative, no. of patients
VII	189	386	10	31
IX	258	533	19	65
X	296	499	73	215
XI	167	328	19	47
XII	210	328	26	87
Others	21	30	—	—

Abbreviations: JPG, jugular paraganglioma; VPG, vagal paraganglioma.

TABLE 4. Postoperative complications.

	No. of patients	CSF fistula	★ Aspiration	★ Infection	★ Meningitis	Stroke	Other	Dead	Total complications
JPG	937	91	63	36	23	15	20	17	263
VPG	226	6	23	5	1	5	10	3	53

Abbreviations: CSF, cerebrospinal fluid; JPG, jugular paraganglioma; VPG, vagal paraganglioma.

RT fraccionada:

- N: 461
- Seguimiento: **9 años**
- Dosis: ppal% 45 – 50 Gy
(rango de 40 – **65 Gy**)
- **Control Tu: 89 %**
- **3 % mortalidad por
progresión de
enfermedad**

- **Complicaciones:**
 - No aumenta
incidencia de
parálisis de PC
(incluso **la mejoró en
el 0.5 % de los casos**)



- 2% de pacientes
falleció por
complicaciones
tardías de la RT.

TABLE 6. Radiation therapy complications.

	No. of patients	Osteonecrosis	Brain necrosis	Deafness	Other	Dead	Total complications
EBRT	461	12	4	17	15	9	57
SRS	261	—	—	13	3	—	16

Abbreviations: EBRT, external beam radiation therapy; SRS, stereotactic radiosurgery.

Dosis:

- Más complicaciones con dosis > 55Gy
- No hay evidencia que >50Gy sea más efectiva que 45-50 Gy

Radiocirugía:

- n:261 ptes (11 GK y 3 SFRT)
- Seguimiento: **3.5 años** (9 años en RTfx y 5,5 años en cirugía)
- Dosis: no describe rango
- **Control Tu: 94 % (En ~ 1/3 disminuyó el tamaño) => 89% rt y 85% cirugía**
- **0 % mortalidad por progresión de enfermedad (OJO: FU corto)**



- Complicaciones:
 - **No aumenta incidencia** de parálisis de PC (incluso la mejoró en el 9 % de los casos)
 - **No hubo complicaciones** severas tardías de la SRS.
 - Sólo el **7.4 %** de los ptes empeoró síntomas o signos neurológicos

Comparaciones

Radiación (externa o SRS) versus Cirugía

- Mejor **control** tumoral (91 /78%)
- Menos **complicaciones** (11/28%)
- Menor probabilidad de **parálisis** de PC (0,08/0,9%)

TABLE 2. Comparison of the treatment outcomes of jugular paragangliomas treated with surgery or radiotherapy.

Parameter	Surgical series, mean rate (95% CI)	Radiotherapy series, mean rate (95% CI)	p value
Tumor control	78.2% (72.2–84.2)	91.5% (85.3–93.3)	.002
Deaths by the tumor	0.8% (-0.2–1.8)	1.4% (-0.6–2.8)	.54
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Major complications	28.2% (19.5–36.9)	11.4% (4.4–18.4)	.003
Cranial nerve palsies after treatment (per patient)	0.9 (0.7–1.1)	0.08 (-0.05–0.2)	< .001

Abbreviations: CI, confidence interval.

POR QUÉ SEGUIMOS OPTANDO



RT externa v/s Radiocirugía:

- Sin diferencias en control tumoral
- Sin diferencias en complicaciones mayores
- SRS menos mortalidad por tratamiento y mejoría de compromiso de PC , pero en este art.el seguimiento es corto con SRS



TABLE 5. Comparison of the treatment outcomes of jugular paragangliomas treated with conventional radiotherapy or radiosurgery.

Parameter	EBRT series mean rate (95% CI)	SRS series mean rate (95% CI)	p value
Tumor control	89.1% (83.5–91.8)	93.7% (83.4–100)	.32
Deaths by the tumor	3.2% (–0.4 to 5.2)	0% (0–0)	.03
Deaths by treatment	2% (0.4–3.7)	0% (0–0)	.04
Major complications	10.4% (4.6–21.9)	6.5% (–4.3 to 21.9)	.53
Increment in cranial nerve palsies after treatment (per patient)	0.15 (–0.11 to 0.4)	0.002 (–0.002 to 0.053)	.25

Abbreviations: JPG, jugular paraganglioma; EBRT, external beam radiation therapy; CI, confidence interval; SRS, stereotactic radiosurgery.



CLINICAL INVESTIGATION

Brain

RADIOSURGERY OF GLOMUS JUGULARE TUMORS: A META-ANALYSIS

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Purpose: During the past two decades, radiosurgery has arisen as a promising approach to the management of glomus jugulare. In the present study, we report on a systematic review and meta-analysis of the available published data on the radiosurgical management of glomus jugulare tumors.

Methods and Materials: To identify eligible studies, systematic searches of all glomus jugulare tumors treated with radiosurgery were conducted in major scientific publication databases. The data search yielded 19 studies, which were included in the meta-analysis. The data from 335 glomus jugulare patients were extracted. The fixed effects pooled proportions were calculated from the data when Cochrane's statistic was statistically insignificant and the inconsistency among studies was <25%. Bias was assessed using the Egger funnel plot test.

Results: Across all studies, 97% of patients achieved tumor control, and 95% of patients achieved clinical control. Eight studies reported a mean or median follow-up time of >36 months. In these studies, 95% of patients achieved clinical control and 96% achieved tumor control. The gamma knife, linear accelerator, and CyberKnife technologies all exhibited high rates of tumor and clinical control.

Conclusions: The present study reports the results of a meta-analysis for the radiosurgical management of glomus jugulare. Because of its high effectiveness, we suggest considering radiosurgery for the primary management of glomus jugulare tumors. © 2011 Elsevier Inc.

Glomus jugulare, Radiosurgery, Gamma knife, Paraganglioma, Meta-analysis.

Table 1. Data from 19 included studies

Study	Year	Average marginal dose (Gy)	Modality	Patients (n)	Follow-up (mo)	Tumor control (%)	Symptom control (%)
Navarro Martin <i>et al.</i> (9)	2010	14	GK	10	10	100	100
Genc <i>et al.</i> (29)	2010	15.6	GK	18	53	94	94
Miller <i>et al.</i> (12)	2009	15	GK*	5	34	100	100
Ganz <i>et al.</i> (30)	2009	13.6	GK	14	28	100	100
Sharma <i>et al.</i> (19)	2008	16.4	GK	24	26	100	100
Lim <i>et al.</i> (34)	2007	20.4	LINAC [†]	18	60	100	100
Henzel <i>et al.</i> (8)	2007	Fractionated	LINAC	17	40	100	100
Gerosa <i>et al.</i> (17)	2006		GK	20	50	100	90
Varma <i>et al.</i> (13)	2006	15	GK	17	48	76	88
Poznanovic <i>et al.</i> (39)	2006	15.1	LINAC	8	16	100	100
Bitaraf <i>et al.</i> (36)	2006	18	GK	16	19	100	100
Feigl <i>et al.</i> (37)	2006	17	GK	12	33	100	92
Sheehan <i>et al.</i> (40)	2005	15	GK	8	28	100	100
Pollock (43)	2004	14.9	GK	42	44	97	NA
Maarouf <i>et al.</i> (18)	2003	15	LINAC	14	48	100	92
Feigenberg <i>et al.</i> (28)	2002	15	GK	5	27	80	80
Saringer <i>et al.</i> (25)	2001	12	GK	13	50	100	100
Jordan <i>et al.</i> (4)	2000	16.3	GK	8	27	100	100
Liscak <i>et al.</i> (14)	1999	16.5	GK	66	24	100	96

Abbreviations: GK = gamma knife; LINAC = linear accelerator; NA = not available.

* Study used combined microsurgical and radiosurgical approach.

† Conventional LINAC and CyberKnife systems included in study.

J Neurosurg 114:1299–1305, 2011

A meta-analysis of tumor control rates and treatment-related morbidity for patients with glomus jugulare tumors

Clinical article

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AND ANDREW T. PARSA, M.D., PH.D.**

Department of Neurological Surgery, University of California, San Francisco, California

Object. Because of the rarity of glomus jugulare tumors, a variety of treatment paradigms are currently used. There is no consensus regarding the optimal management to control tumor burden while minimizing treatment-related morbidity. In this study, the authors assessed data collected from 869 patients with glomus jugulare tumors from the published literature to identify treatment variables that impacted clinical outcomes and tumor control rates.

Methods. A comprehensive search of the English-language literature identified 109 studies that collectively described outcomes for patients with glomus jugulare tumors. Univariate comparisons of demographic information between treatment cohorts were performed to detect differences in the sex distribution, age, and Fisch class of tumors among various treatment modalities. Meta-analyses were performed on calculated rates of recurrence and cranial neuropathy after subtotal resection (STR), gross-total resection (GTR), STR with adjuvant postoperative radiosurgery (STR+SRS), and stereotactic radiosurgery alone (SRS).

Results. The authors identified 869 patients who met their inclusion criteria. In these studies, the length of follow-up ranged from 6 to 256 months. Patients treated with STR were observed for 72 ± 7.9 months and had a tumor control rate of 69% (95% CI 57%–82%). Those who underwent GTR had a follow-up of 88 ± 5.0 months and a tumor control rate of 86% (95% CI 81%–91%). Those treated with STR+SRS were observed for 96 ± 4.4 months and had a tumor control rate of 71% (95% CI 53%–83%). Patients undergoing SRS alone had a follow-up of 71 ± 4.9 months and a tumor control rate of 95% (95% CI 92%–99%). The authors' analysis found that patients undergoing SRS had the lowest rates of recurrence of these 4 cohorts, and therefore, these patients experienced the most favorable rates of tumor control ($p < 0.01$). Patients who underwent GTR sustained worse rates of cranial nerve (CN) deficits with regard to CNs IX–XI than those who underwent SRS alone; however, the rates of CN XII deficits were comparable.

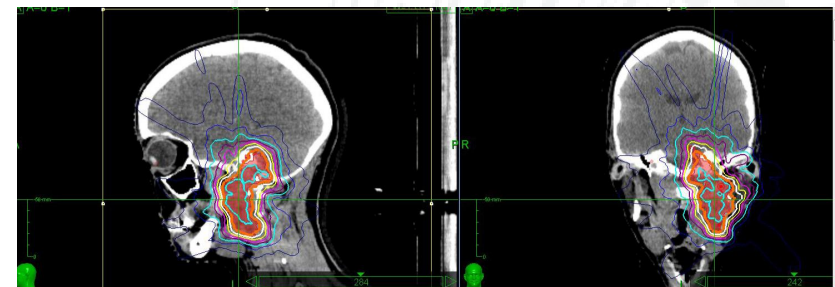
Conclusions. The authors' analysis is limited by the quality and accuracy of these studies and may reflect source study biases, as it is impossible to control for the quality of the data reported in the literature. Finally, due to the diverse range of data presentation, the authors found that they were limited in their ability to study and control for certain variables. Some of these limitations should be minimized with their use of meta-analysis methods, which statistically evaluate and adjust for between-study heterogeneity. These results provide the impetus to initiate a prospective study, appropriately controlling for variables that can confound the retrospective analyses that largely comprise the existing literature. (DOI: 10.3171/2010.9.JNS10699)

Caso Clínico FALP:

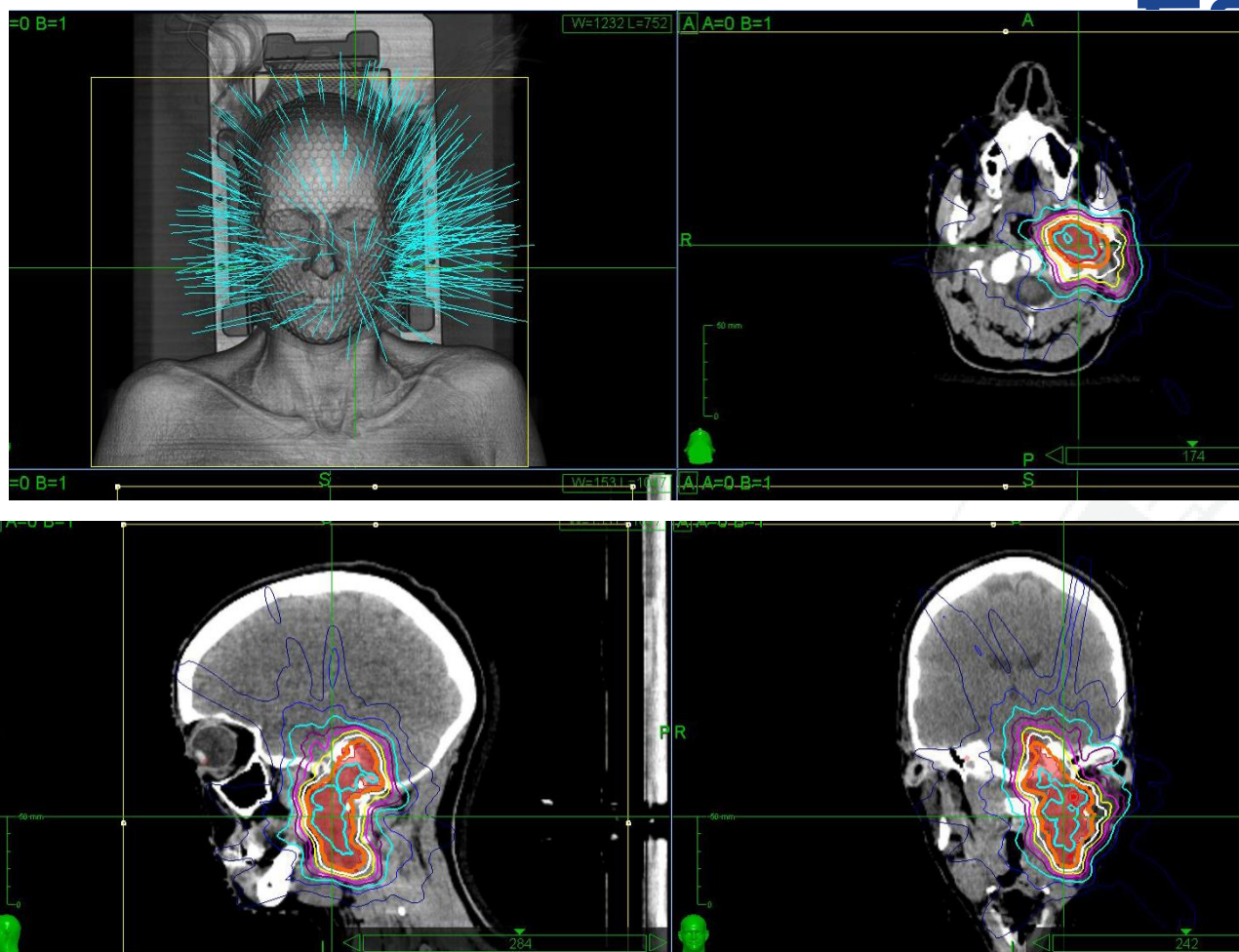
Paraganglioma yugular izquierdo



- Mujer de 45 años, del extremo sur de Chile (difícil acceso a tratamientos)
- Comenzó con vértigo (2 meses), estrabismo por pérdida de la abducción del ojo izquierdo (6º par izq), tinnitus severo a izq. intermitente, hipoacusia izq, atrofia de hemilengua izquierda, cambios en su voz. Se dg inicialmente **Sd Depresivo**.
- Síntomas progresivos, sin taquicardia, sin sudoración ni otros.
- RNM imagen sospechosa de paraganglioma extenso yugular izq **FISCH D** que llega hasta altura C1-C2, por lateral ingresa en el CAI y al canal de Dorello izq.



Es derivada para consideración de Cyberknife 24Gy en 3fx de 8Gy.
Fusión de TAC, RNM, AngioRNM.
Tracking 6Dskull.



- Paciente tratada ambulatoriamente y sin complicaciones en Noviembre 2016, al control a los 6 meses se aprecia mejoría en la atrofia lingual y estrabismo, esta mejoría (parcial) se mantiene a la fecha.

ALGORITMO PROPUESTO

