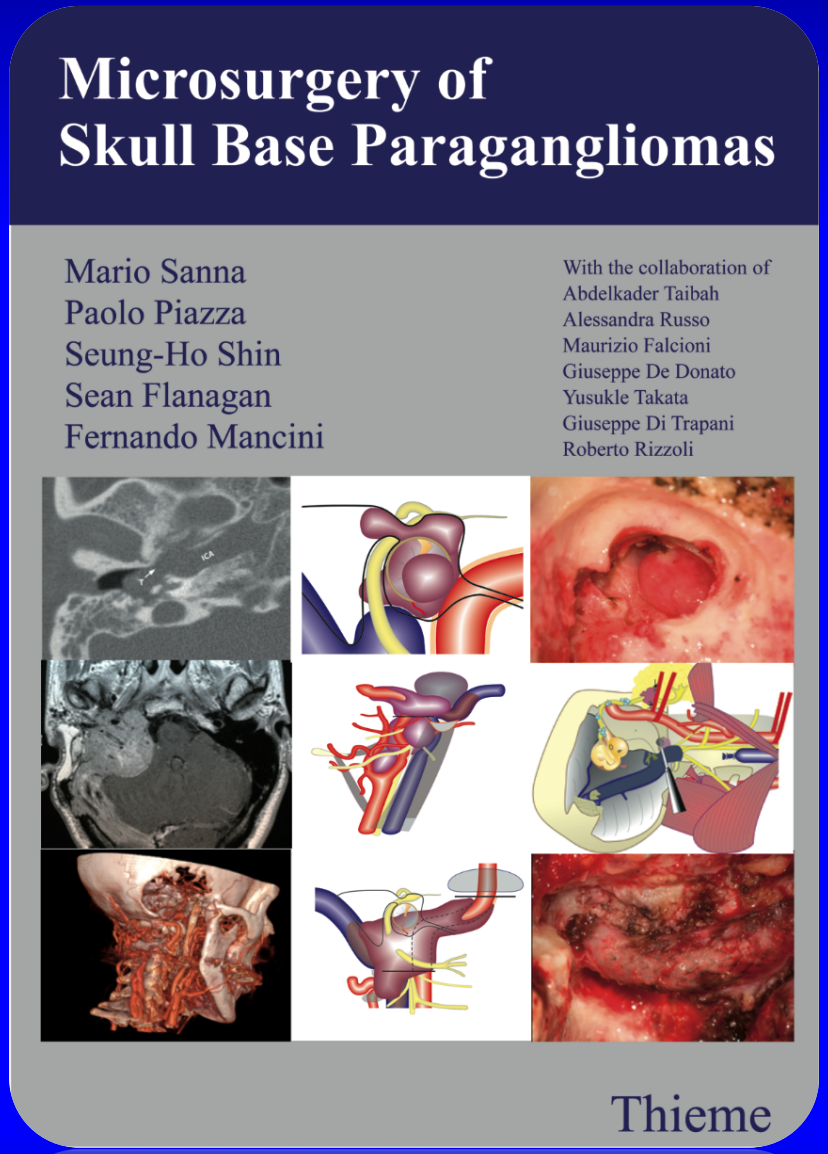


SURGICAL MANAGEMENT OF SKULL BASE PARAGANGLIOMAS

Mario Sanna and Gianluca Piras

GRUPPO OTOLOGICO 1983

Piacenza-Rome
Italy

MARIO SANNA FOUNDATION ONLUS

SKULL BASE PARAGANGIOMAS: A PARADIGM SHIFT

- *Skull Base PGs are a challenge : vascular, locally aggressive, and involve important neurovascular structures: jugular bulb, ICA, FN & LCNs.*

- *Considered inoperable in the past*

SKULL BASE PARAGANGIOMAS: A PARADIGM SHIFT

- *PARADIGM SHIFT: Surgery has emerged as the mainstay of this subset of tumors due to:*

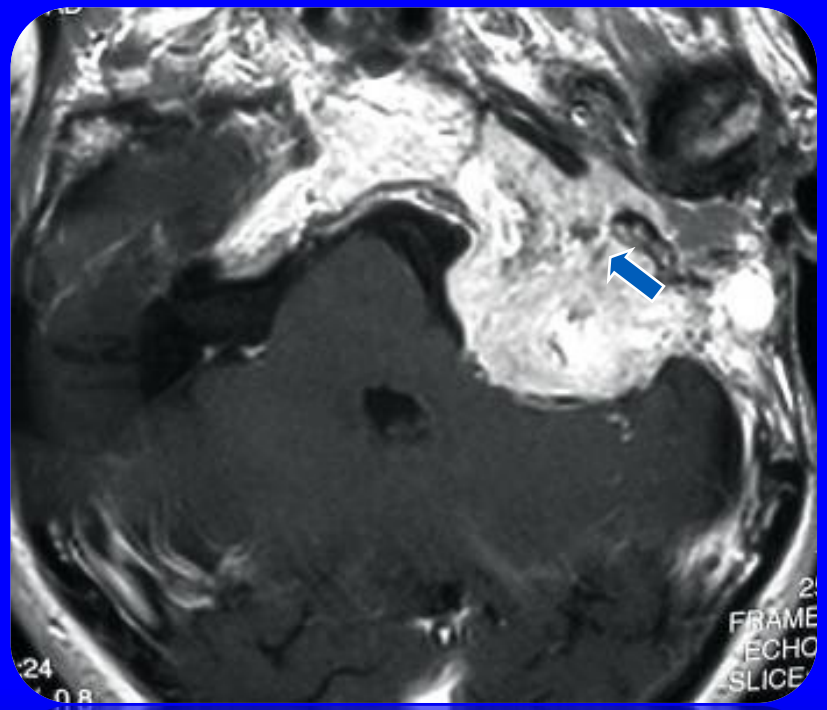
- *A thorough anatomic & surgical mapping of the skull base & description of various rational approaches*

- *Technological improvements in microsurgery, neuromonitoring, neuroanesthesia, & neuroradiology*

INTRODUCTION

- Paraganglioma – Benign neuroendocrine neoplasms along sympathetic NS

- Sites: Head & Neck, thorax and abdomen

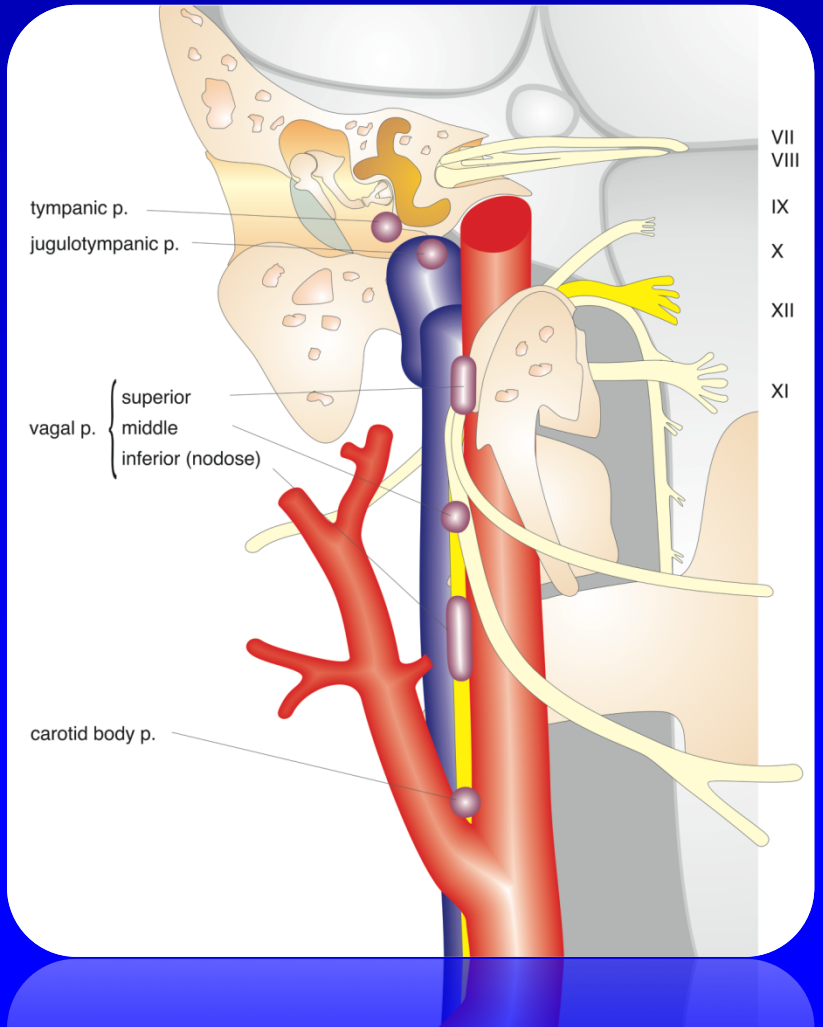


HEAD AND NECK PARAGANGLIOMAS

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

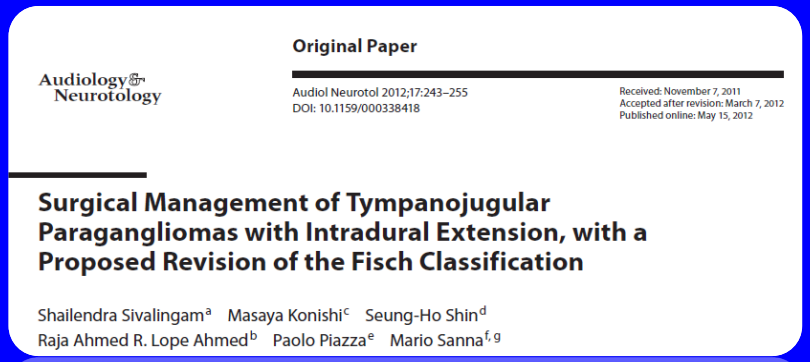
- Middle ear (Tympanic and Tympanomastoid Class A-B)
- Dome of the jugular bulb (Tympanojugular - Class C)
- Along the vagus nerve (Vagal Paraganglioma – VP)
- In the neck in the bifurcation of the carotid artery (Carotid Body Paraganglioma – CBP)



MODIFIED FISCH'S CLASSIFICATION FOR TYMPANOJUGULAR PARAGANGLIOMAS

Modifications classification (Gruppo Otologico)

- Tympanojugular paraganglioma
  - Class C1
  - Class C2
  - Class C3
  - Class C4
  - Di
  - Ve/i



Original Paper

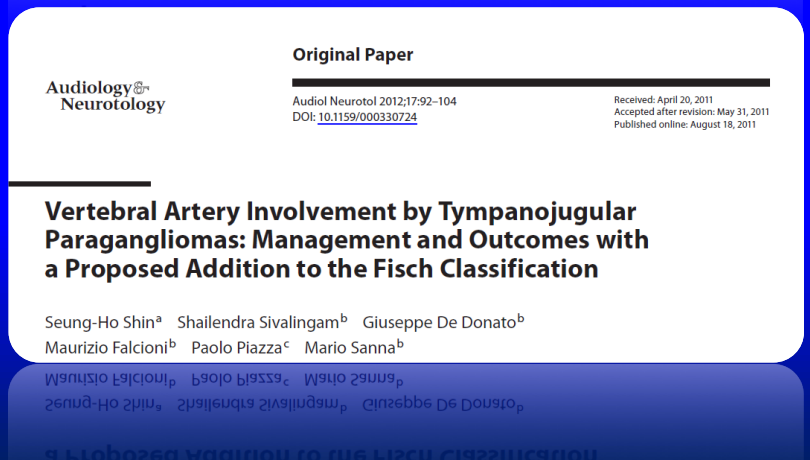
Audiology & Neurotology

Audiol Neurotol 2012;17:243–255
DOI: 10.1159/000338418

Received: November 7, 2011
Accepted after revision: March 7, 2012
Published online: May 15, 2012

Surgical Management of Tympanojugular Paragangliomas with Intradural Extension, with a Proposed Revision of the Fisch Classification

Shailendra Sivalingam[a] Masaya Konishi[c] Seung-Ho Shin[d]
Raja Ahmed R. Lope Ahmed[b] Paolo Piazza[e] Mario Sanna[f, g]



Original Paper

Audiology & Neurotology

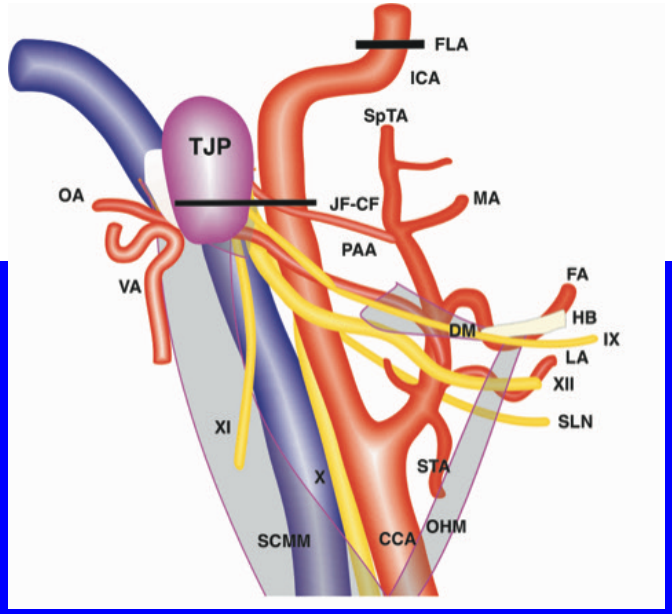
Audiol Neurotol 2012;17:92–104
DOI: 10.1159/000330724

Received: April 20, 2011
Accepted after revision: May 31, 2011
Published online: August 18, 2011

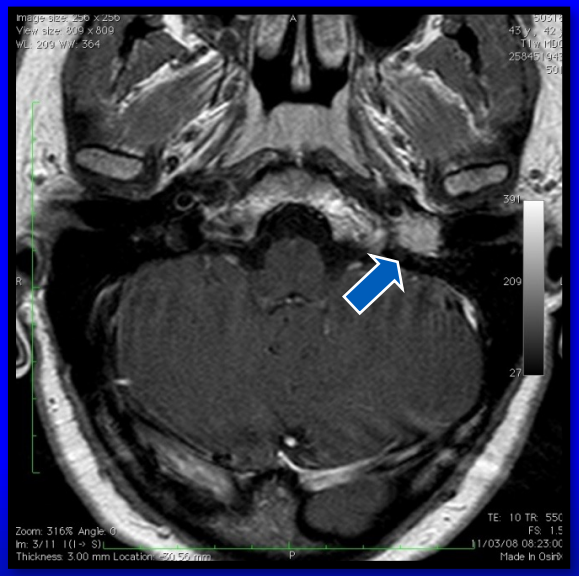
Vertebral Artery Involvement by Tympanojugular Paragangliomas: Management and Outcomes with a Proposed Addition to the Fisch Classification

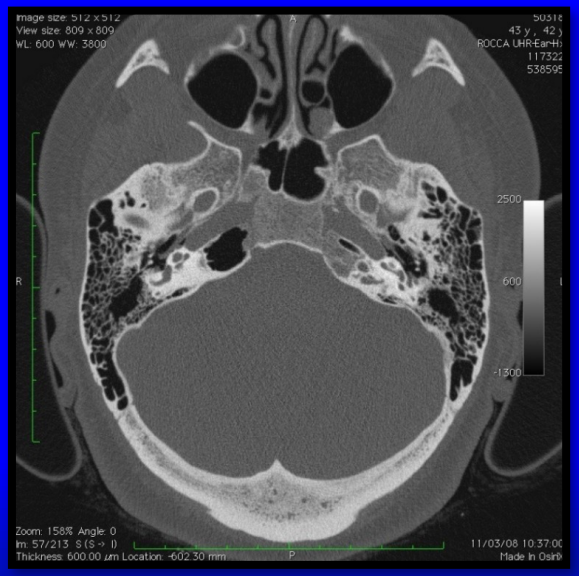
Seung-Ho Shin[a] Shailendra Sivalingam[b] Giuseppe De Donato[b]
Maurizio Falcioni[b] Paolo Piazza[c] Mario Sanna[b]

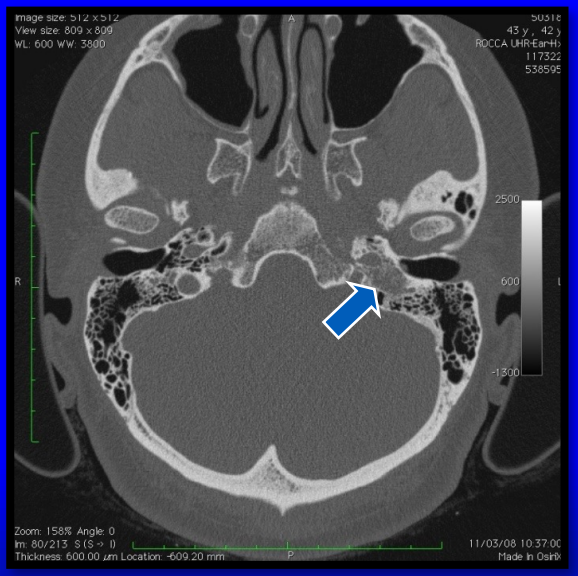
MODIFIED FISCH'S CLASSIFICATION



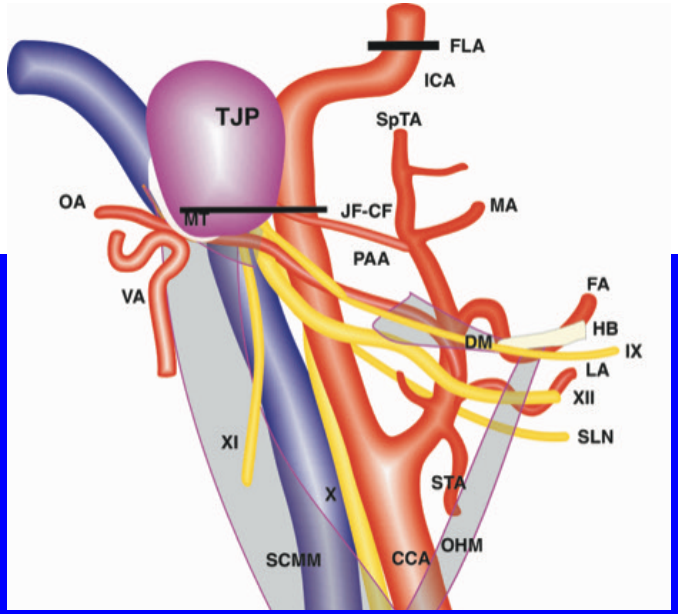
- CLASS C1
  - Tumor limited to the carotid foramen of the petrous ICA



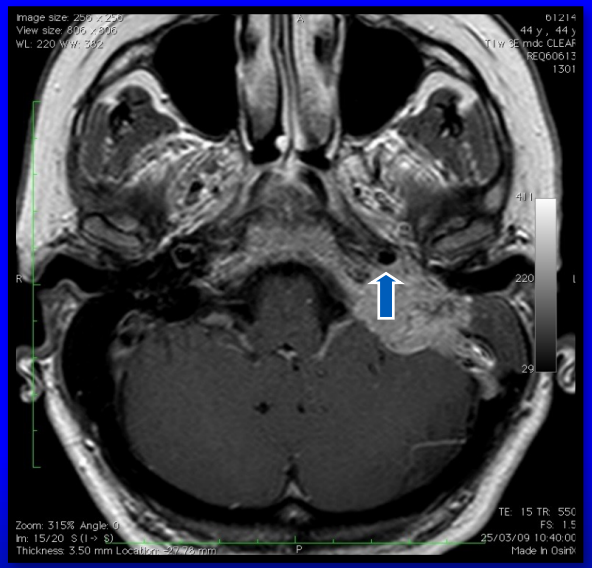


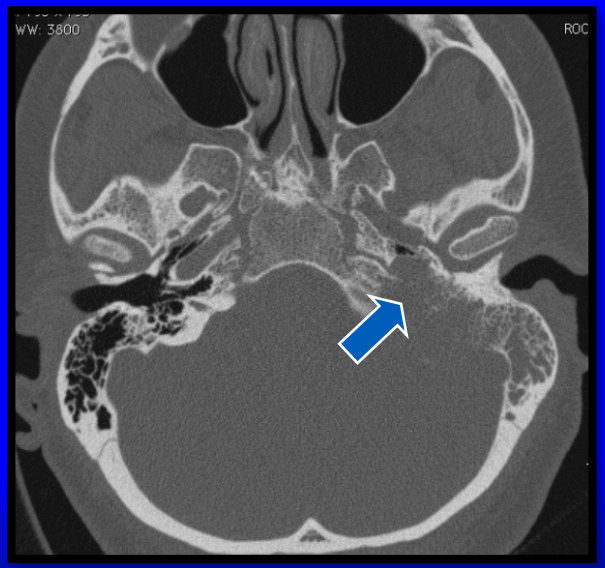


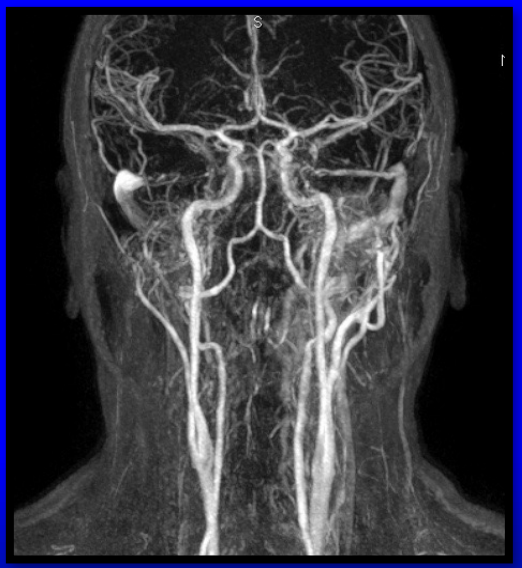
MODIFIED FISCH'S CLASSIFICATION



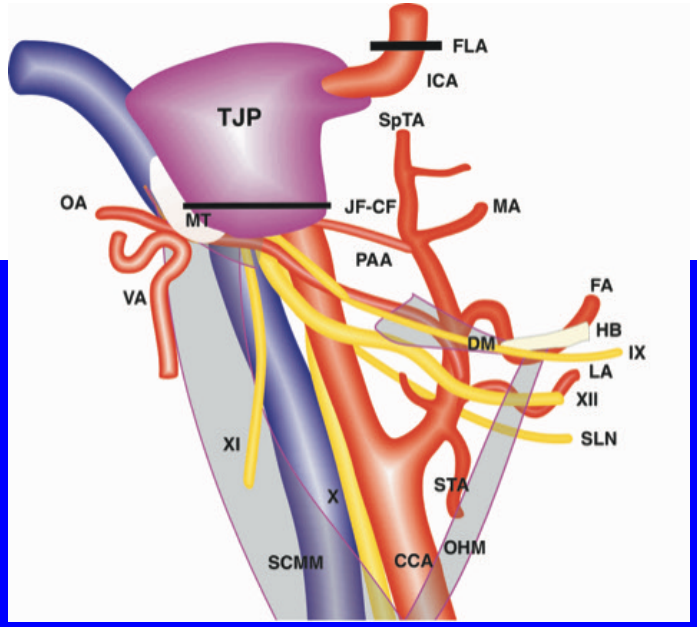
- CLASS C2
  - Tumor extends to the vertical portion of the petrous ICA



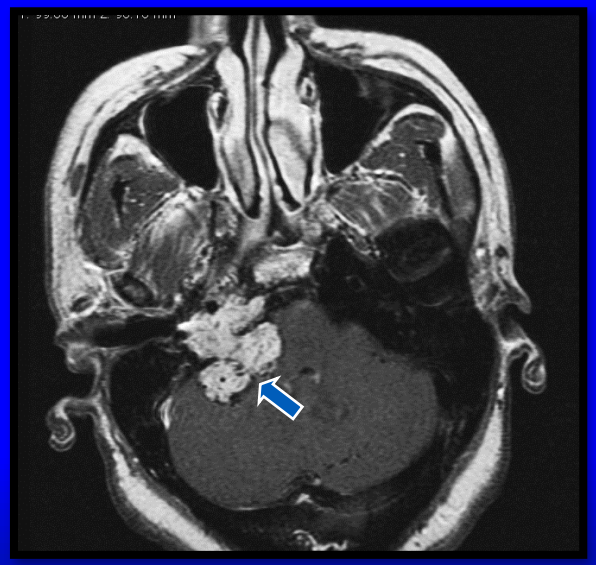


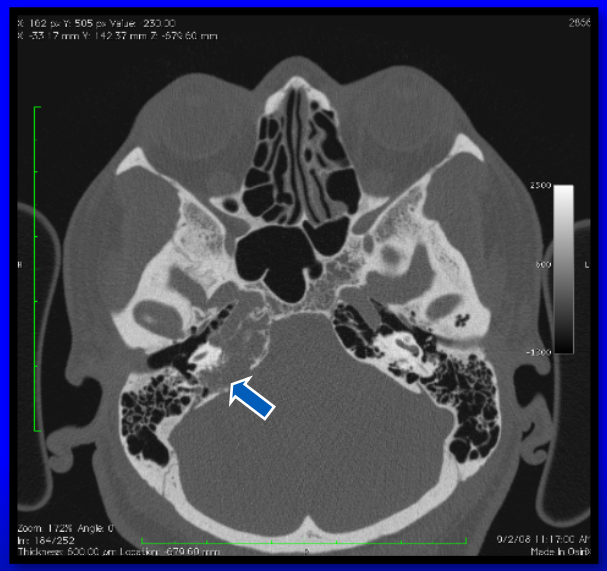


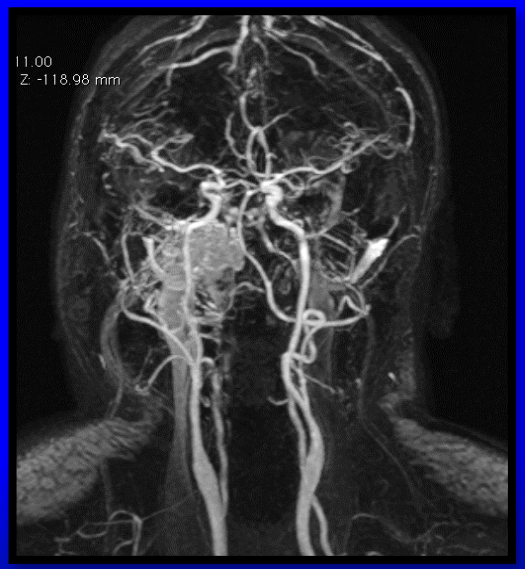
MODIFIED FISCH'S CLASSIFICATION



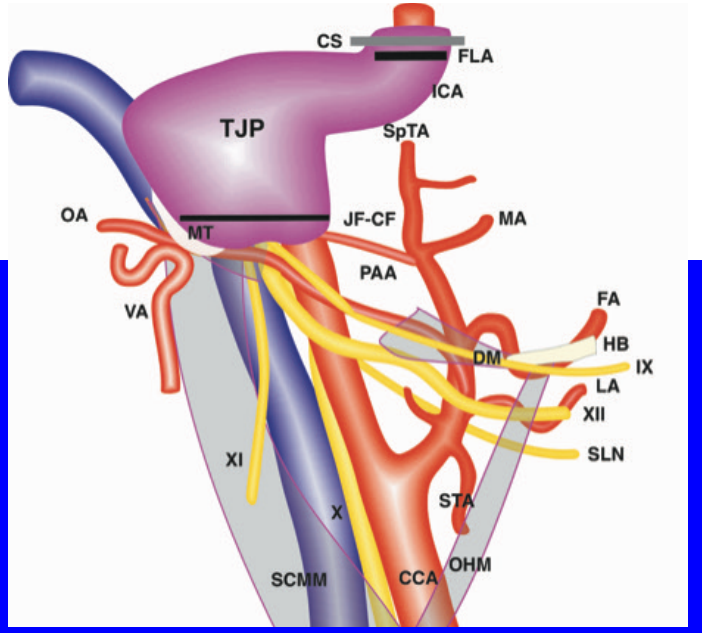
- CLASS C3
  - Tumor extends to the horizontal portion of the petrous ICA



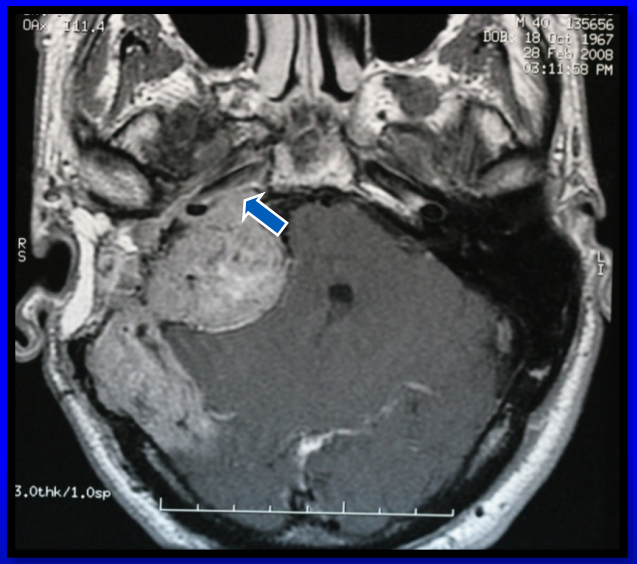


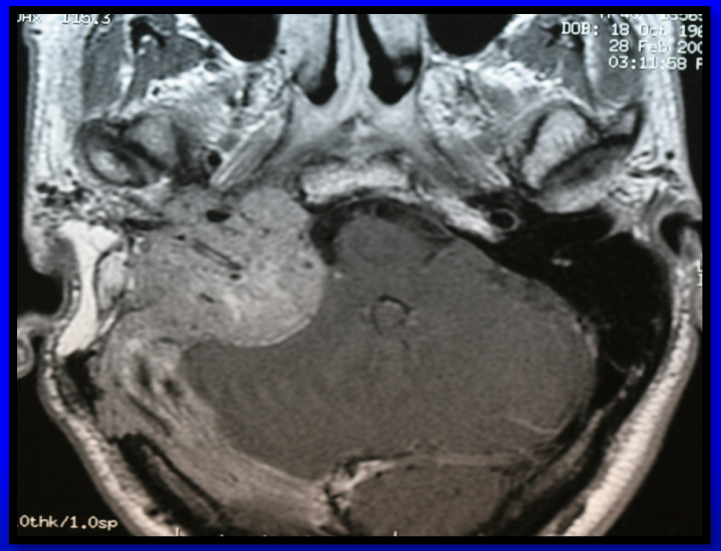


MODIFIED FISCH'S CLASSIFICATION

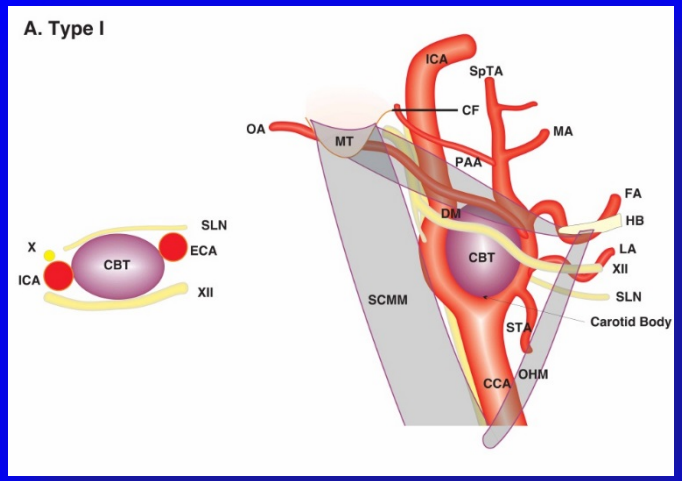


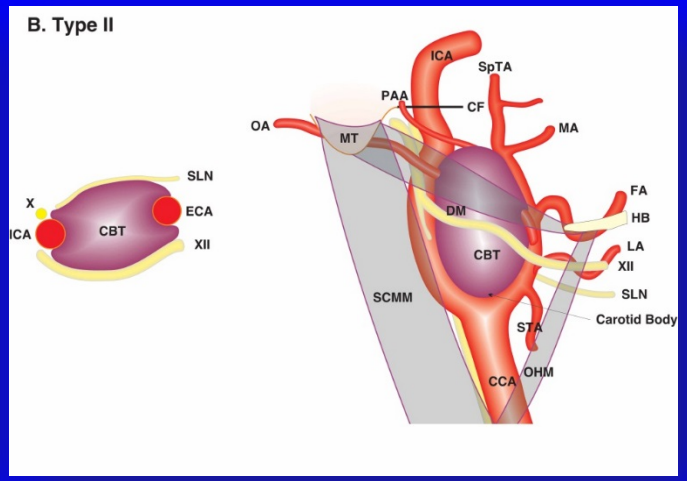
- CLASS C4
  - Tumor extends to the anterior foramen lacerum of the petrous ICA

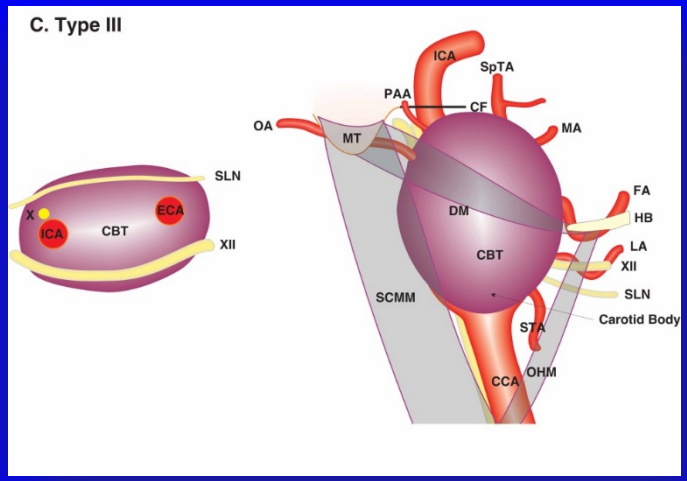




CAROTID BODY PARAGANGLIOMA



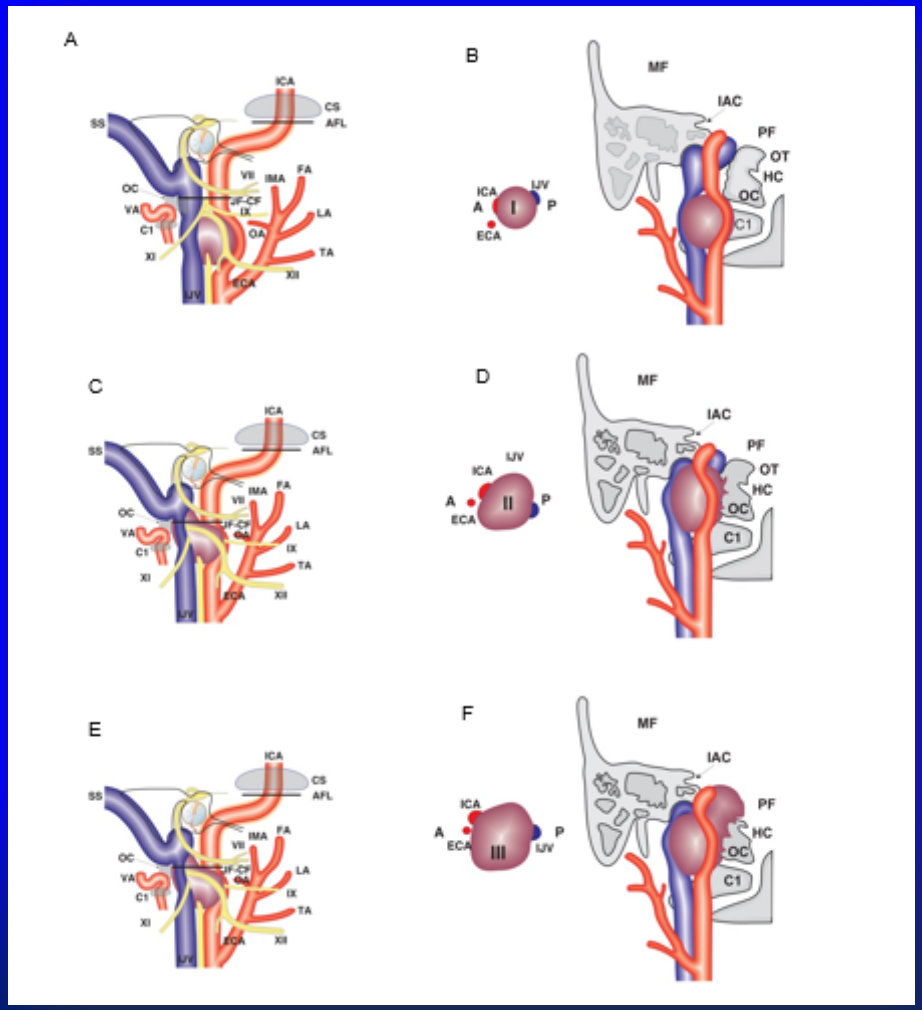




Shamblin Classification:

Type I: Localized and not encasing or infiltrating the arteries.
Type II: Partially adherent to surrounding vessels.
Type III: Intimately surrounding or encasing the vessels.

VAGAL PARAGANGLIOMA



- **Type I:** Do not invade the jugular foramen.
- **Type II:** Invade the jugular foramen but no extensive bone destruction.
- **Type III:** Deeply invade the jugular foramen & middle ear with extensive bone destruction.

- Varying degrees of carotid invasion
- Varying degrees of intracranial extension

MULTIPLE TUMORS

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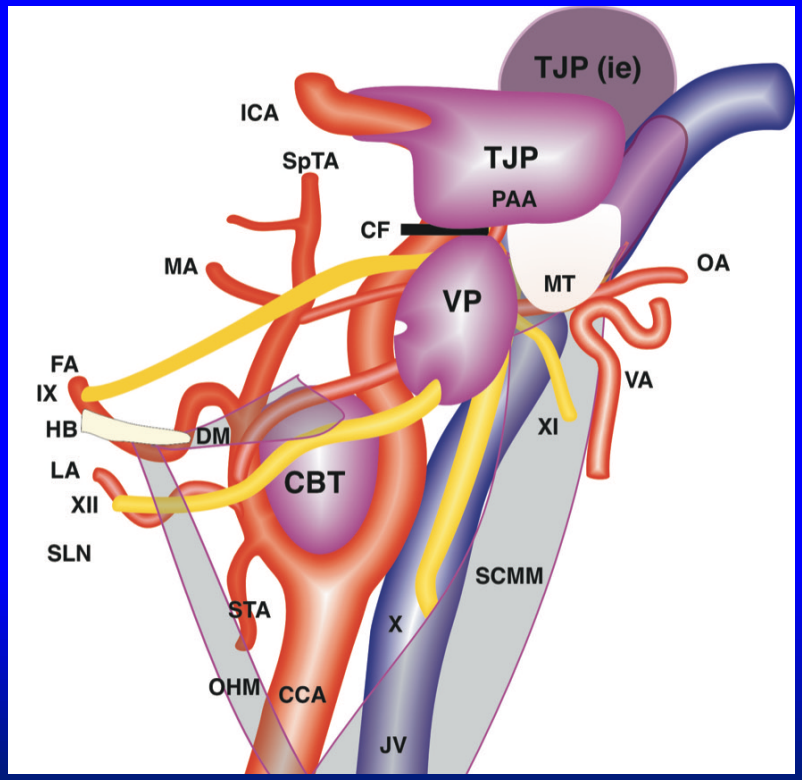
GRUPPO OTOLOGICO 1983

- Considered as COMPLEX cases because: Decision making is extremely difficult



PRE-OP MANAGEMENT

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CORNERSTONES OF PRE-OPERATIVE RADIOLOGICAL INVESTIGATIONS

- Determine type and extent of disease
- Evaluate for associated lesions
- Assess involvement of major vessels
- Evaluate collateral circulation
- Assess intracranial extension

→

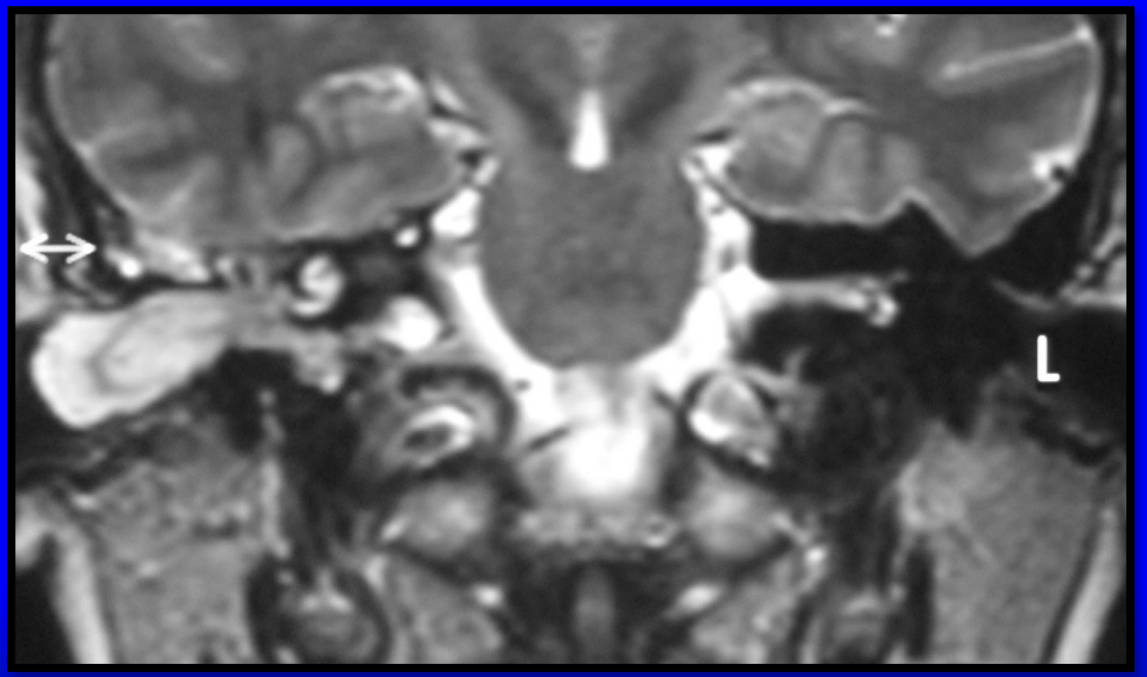
- Skull Base CT scan
- Brain/Neck MRI ± angio
- Arteriography (ICA)

OTOSCOPY

- Red pulsatile retrotympanic mass

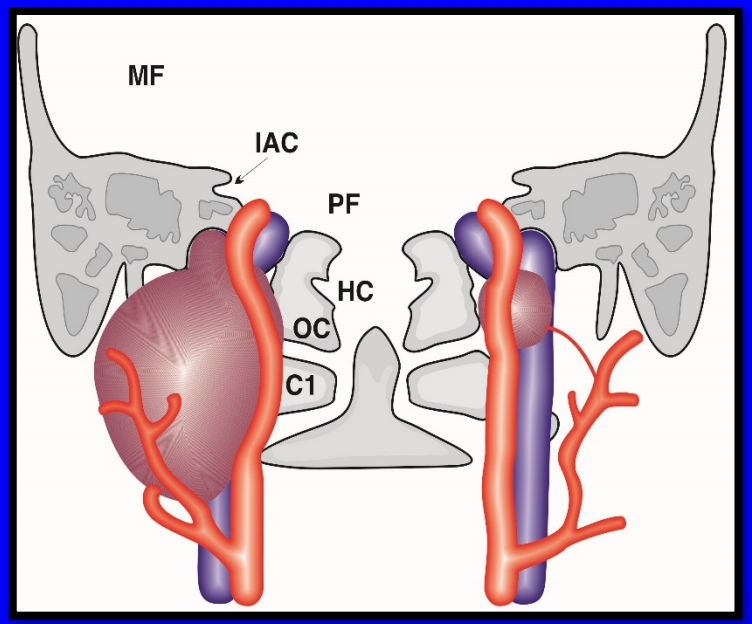
OTOSCOPY

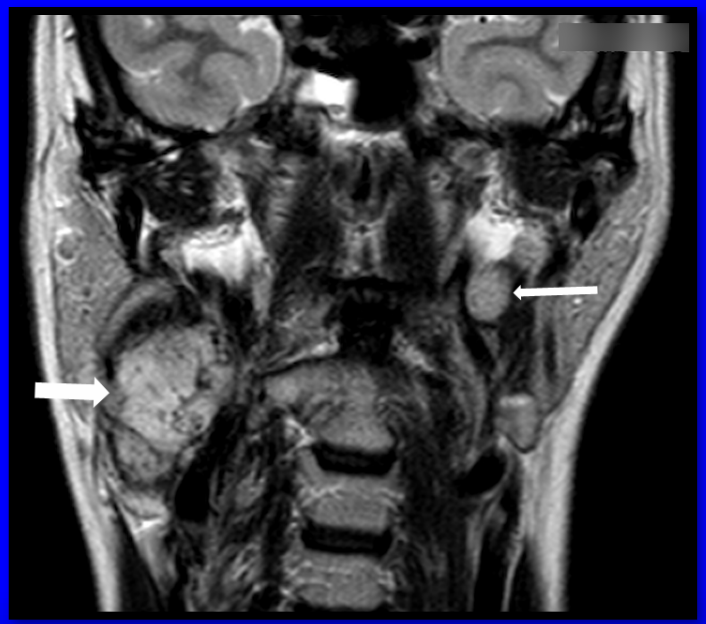
- Sometimes presents as an EAC polyp



CLINICAL EVALUATION

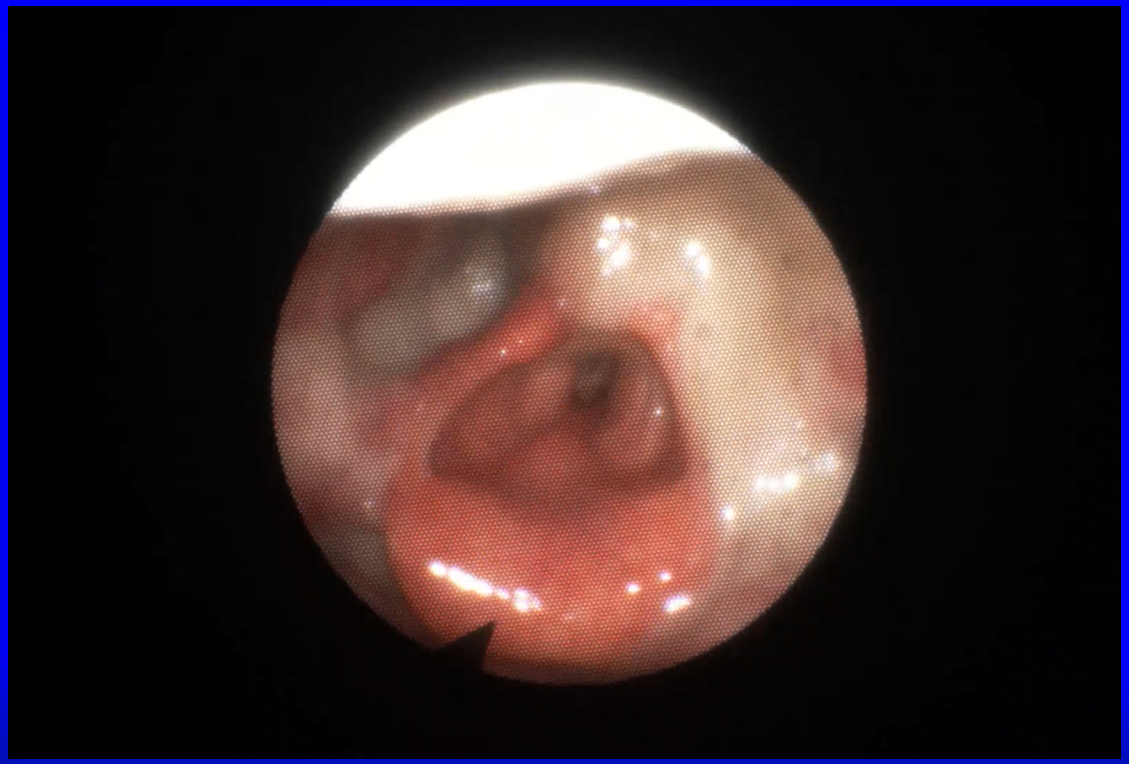
- Rarely a CBP or a VP can present with a neck mass





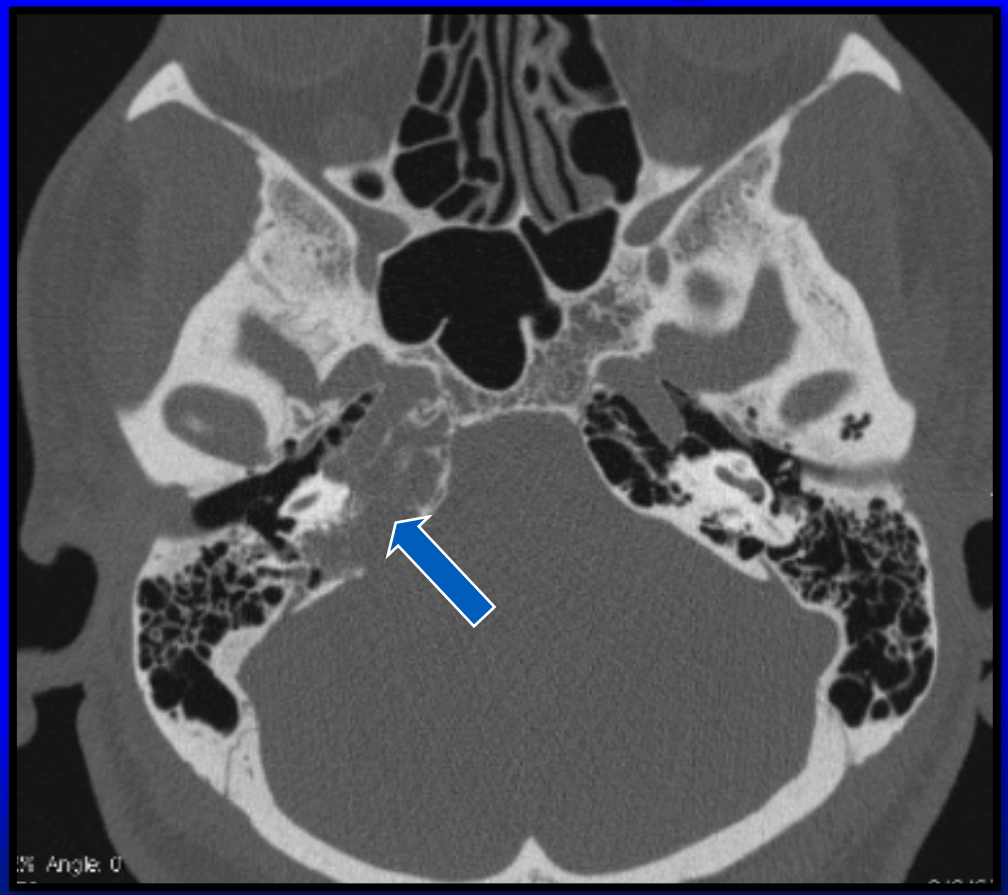
CLINICAL EVALUATION

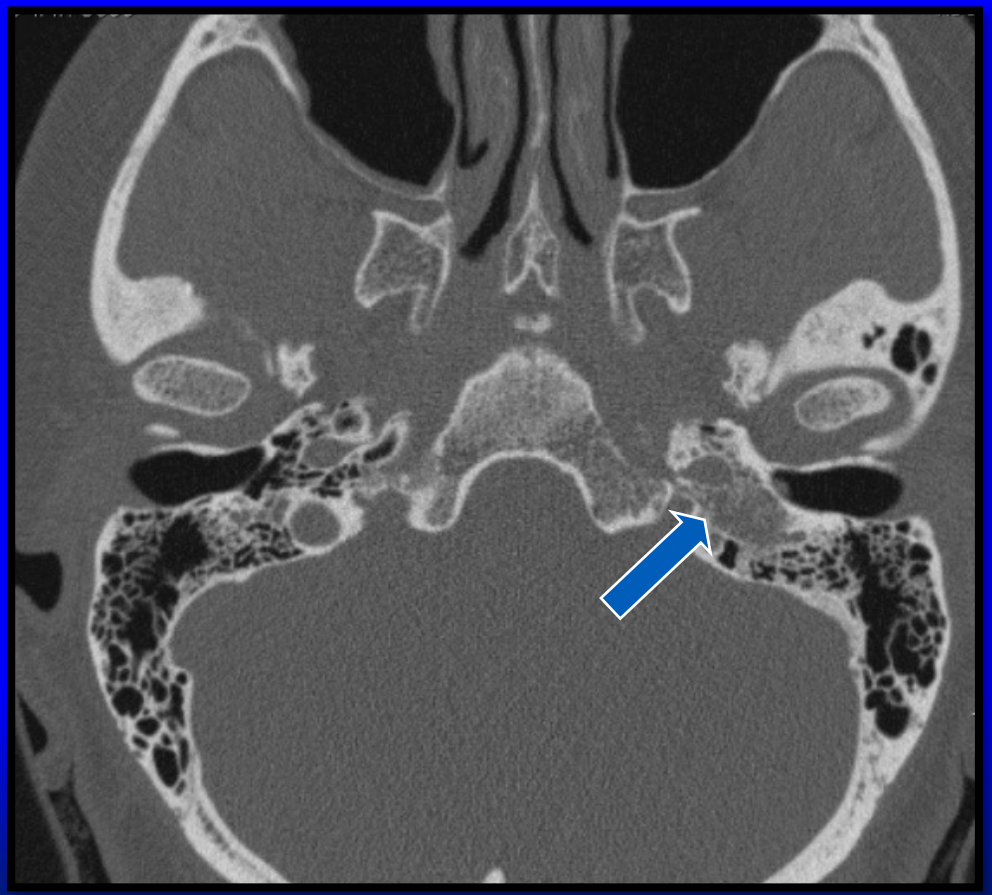
- In more extended cases lower cranial nerves palsies can occur



RADIOLOGY – CT SCAN

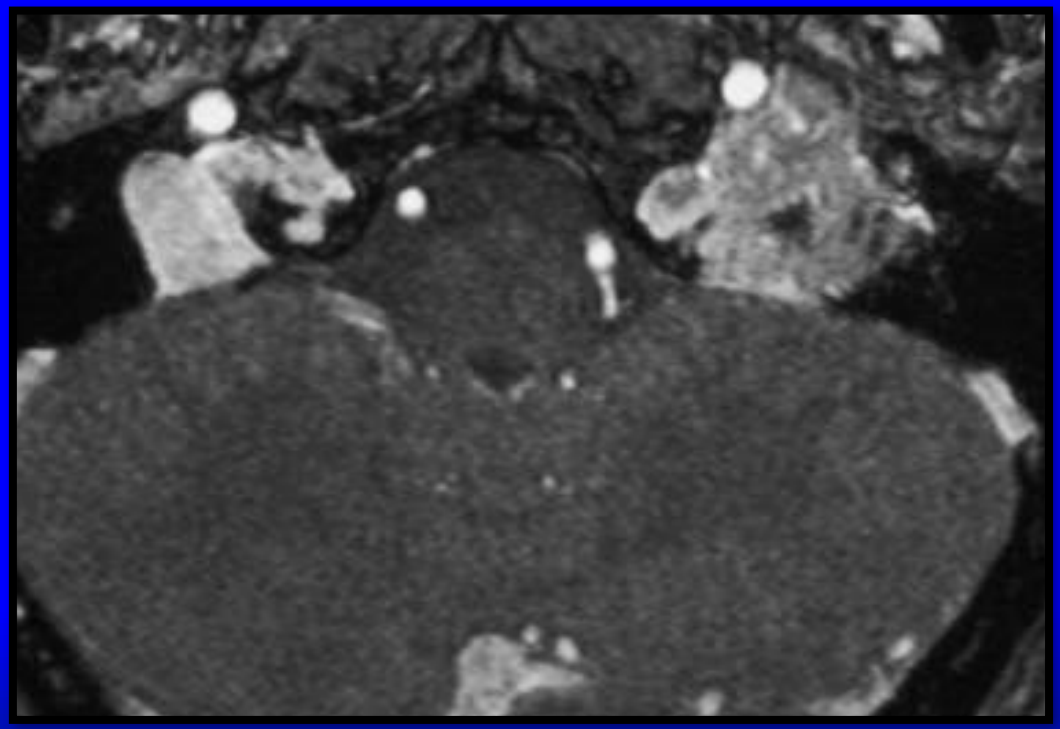
- Bone window: Irregular bone erosion

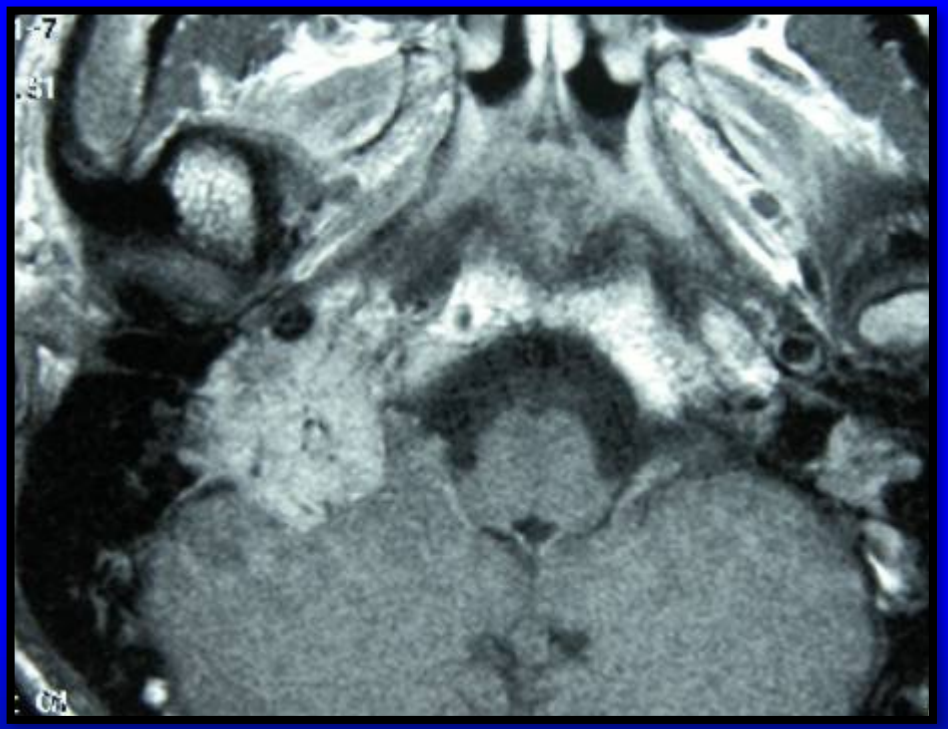




RADIOLOGY - MRI

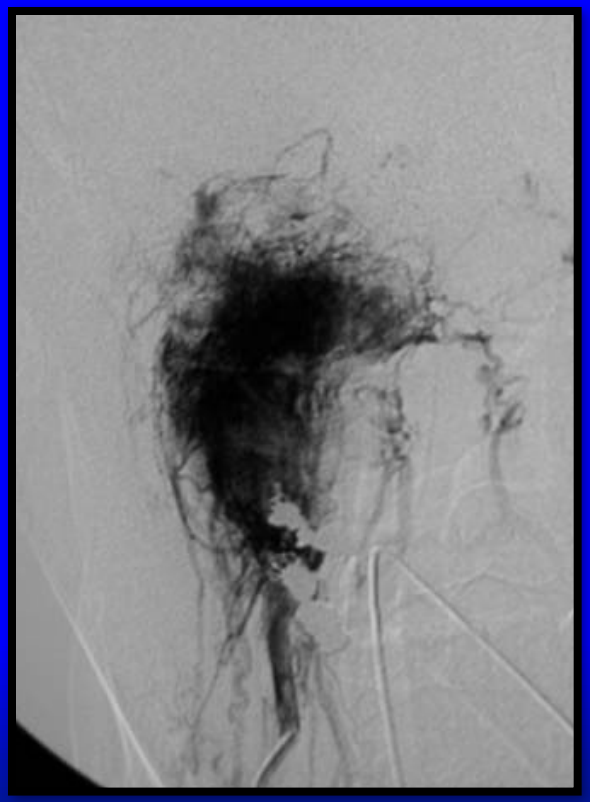
- Salt and pepper appearance with INTERNAL VOIDS

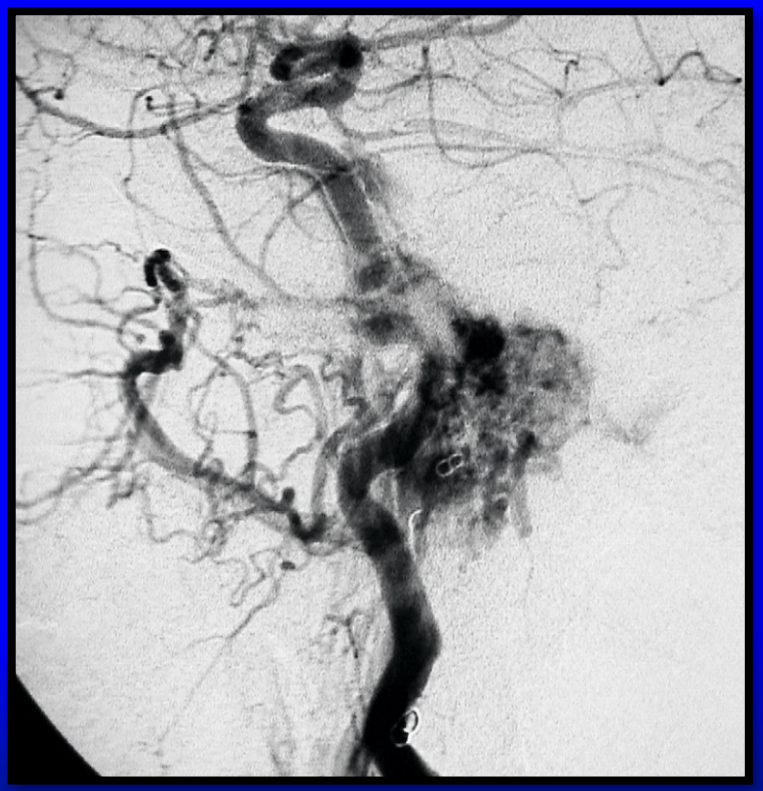




ANGIOGRAPHY

- Tumor blush due to increased vasculature

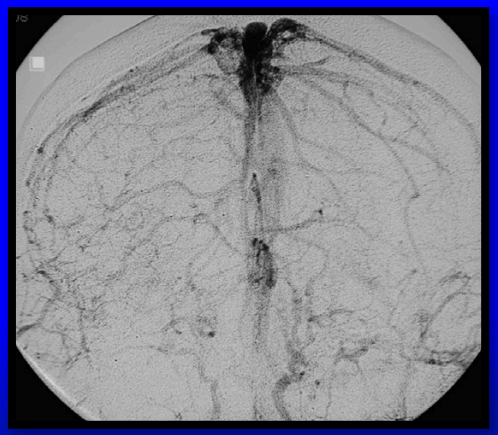


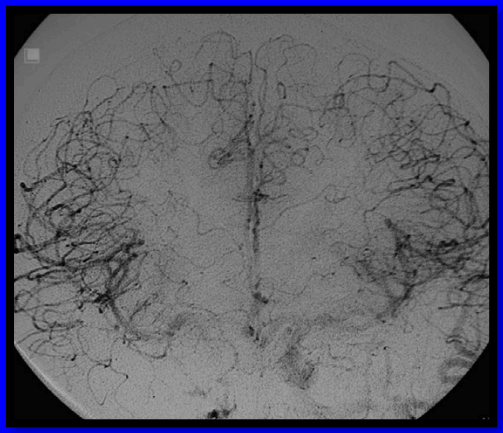


ANGIOGRAPHY

- To evaluate the symmetry of circulation of the two cerebral hemispheres

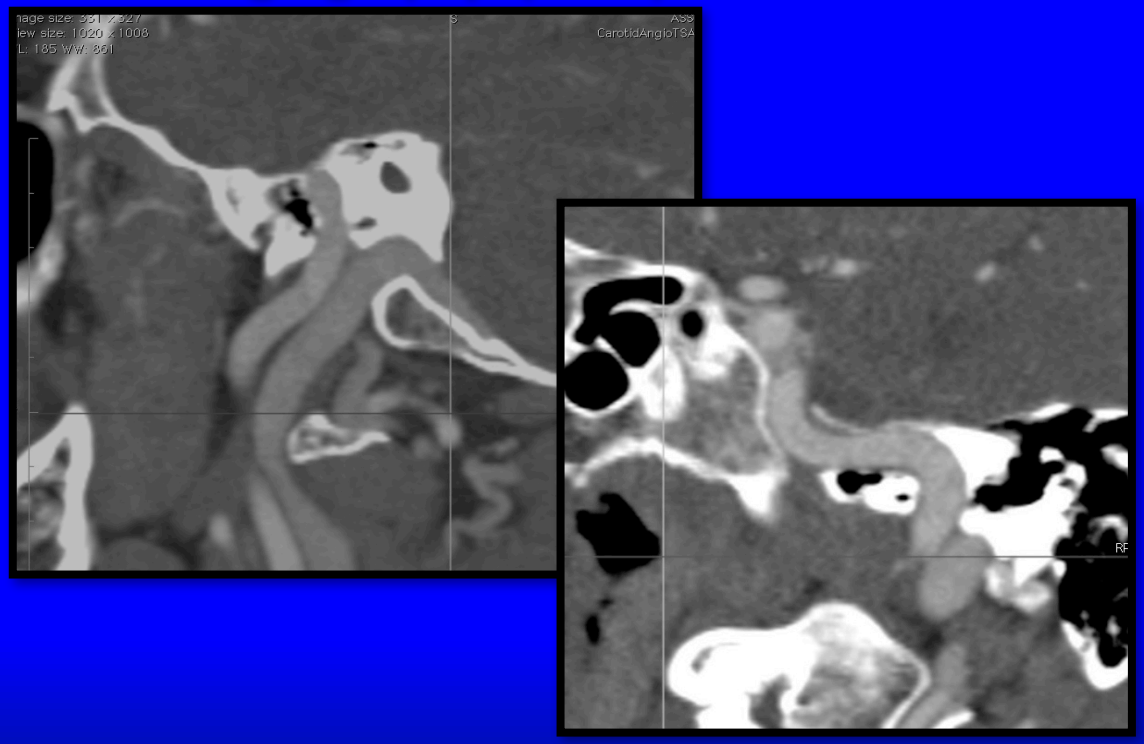
- ARTERIAL PHASE
- CAPILLARY PHASE
- VENOUS PHASE



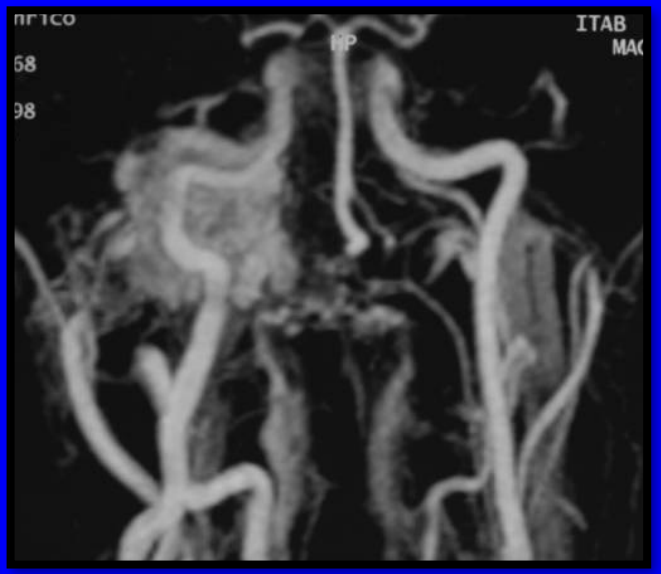


ANGIOCT-ANGIO MRI

- CT angiography (CTA)

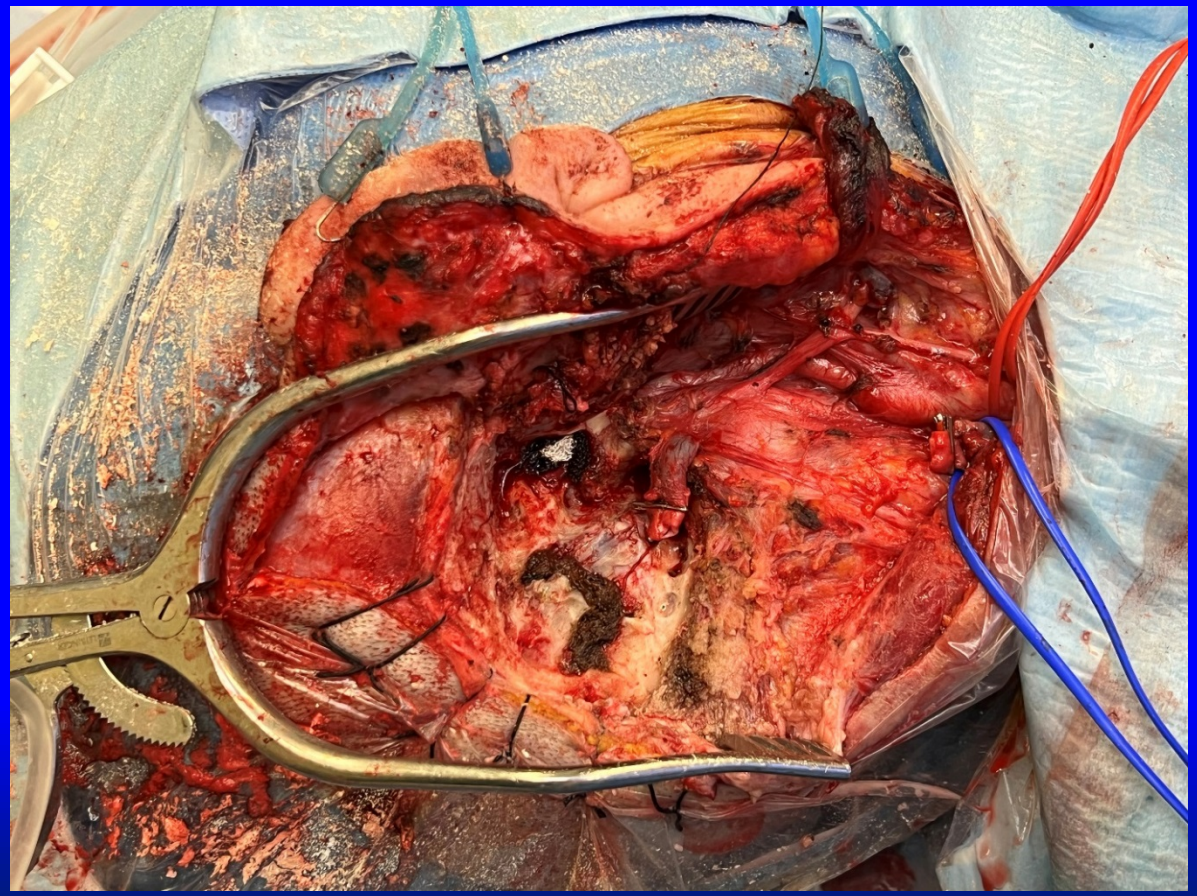


- MR angiography (MRA)



MANAGEMENT

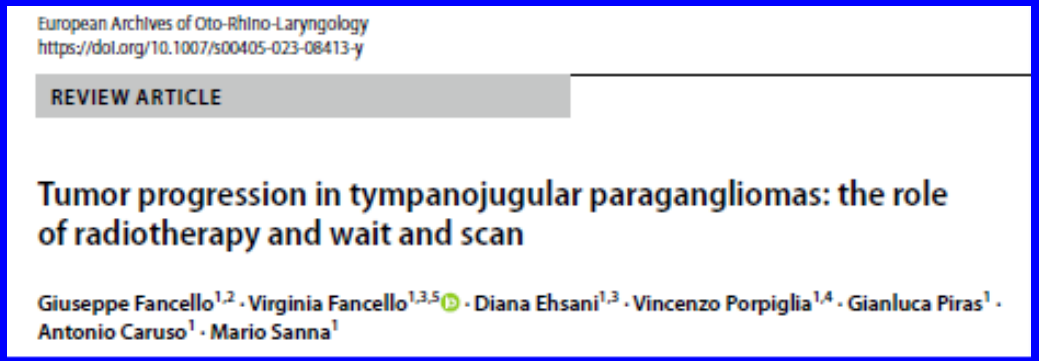
- Three primary options



1. Surgery
2. Wait & scan
3. Radiotherapy

INDICATIONS FOR WAIT & SCAN

- Age >65 yrs
- Normal LCNs function in elderly



European Archives of Oto-Rhino-Laryngology
https://doi.org/10.1007/s00405-023-08413-y

REVIEW ARTICLE

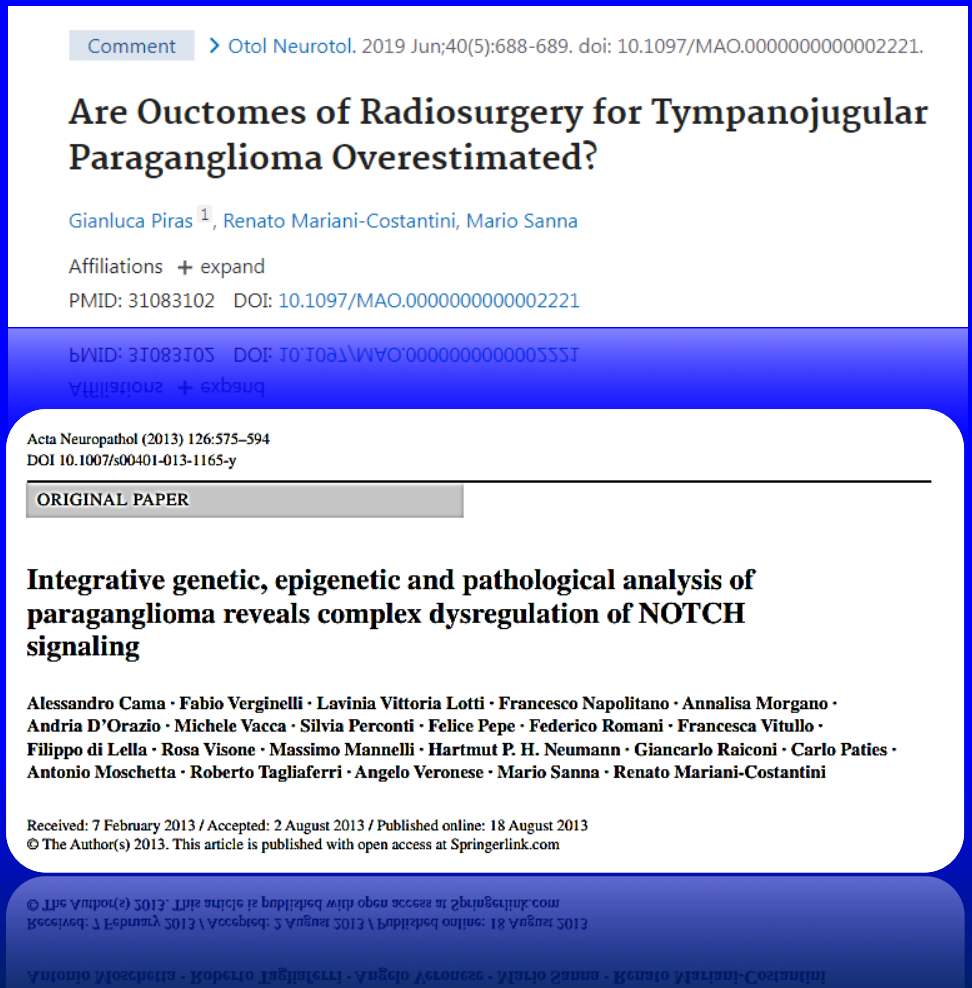
Tumor progression in tympanojugular paragangliomas: the role of radiotherapy and wait and scan

Giuseppe Fancello[1,2] · Virginia Fancello[1,3,5] · Diana Ehsani[1,3] · Vincenzo Porpiglia[1,4] · Gianluca Piras[1] · Antonio Caruso[1] · Mario Sanna[1]

1983-2023 (62 cases)
87.1% showed tumor stability after W&S
(mean FU 75.9 months)

RADIOTHERAPY

- Radiotherapy has a peripheral role in the management of TJPs
- *In our center RT is not indicated as a starting treatment*



Comment › Otol Neurotol. 2019 Jun;40(5):688-689. doi: 10.1097/MAO.0000000000002221.

Are Ouctomes of Radiosurgery for Tympanojugular Paraganglioma Overestimated?

Gianluca Piras [1], Renato Mariani-Costantini, Mario Sanna

Affiliations + expand

PMID: 31083102 DOI: 10.1097/MAO.0000000000002221

Acta Neuropathol (2013) 126:575–594
DOI 10.1007/s00401-013-1165-y

ORIGINAL PAPER

Integrative genetic, epigenetic and pathological analysis of paraganglioma reveals complex dysregulation of NOTCH signaling

Alessandro Cama · Fabio Verginelli · Lavinia Vittoria Lotti · Francesco Napolitano · Annalisa Morgano · Andria D'Orazio · Michele Vacca · Silvia Perconti · Felice Pepe · Federico Romani · Francesca Vitullo · Filippo di Lella · Rosa Visone · Massimo Mannelli · Hartmut P. H. Neumann · Giancarlo Raiconi · Carlo Paties · Antonio Moschetta · Roberto Tagliaferri · Angelo Veronese · Mario Sanna · Renato Mariani-Costantini

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PREOPERATIVE TUMOR EMBOLIZATION

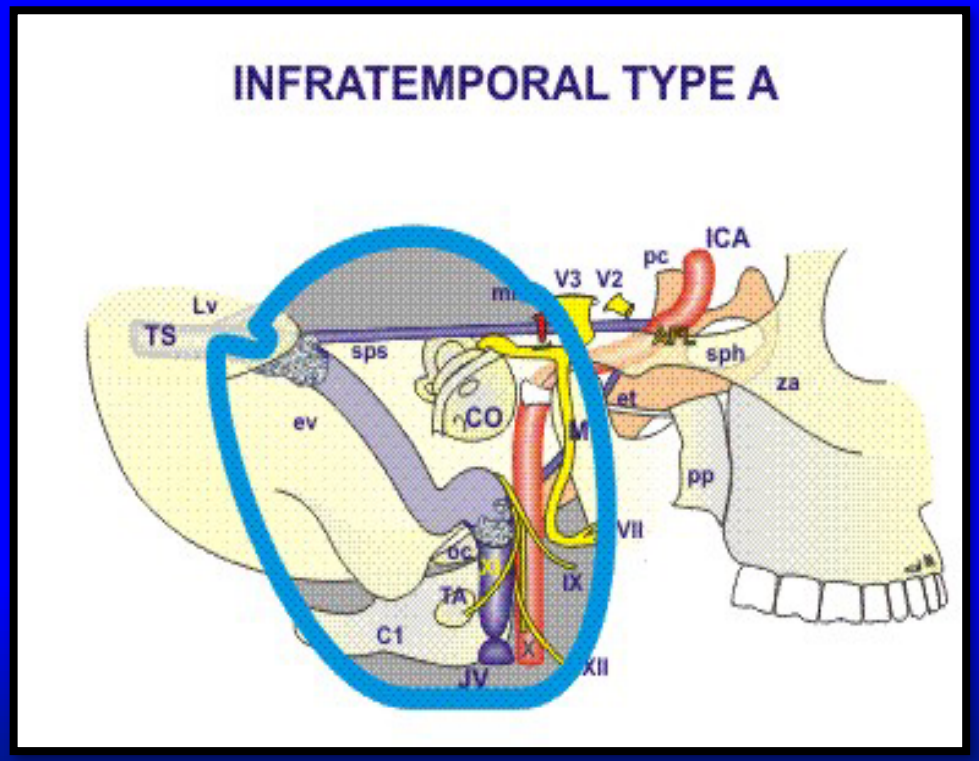
- Embolization: We embolize all the cases 24-48 hrs before surgery

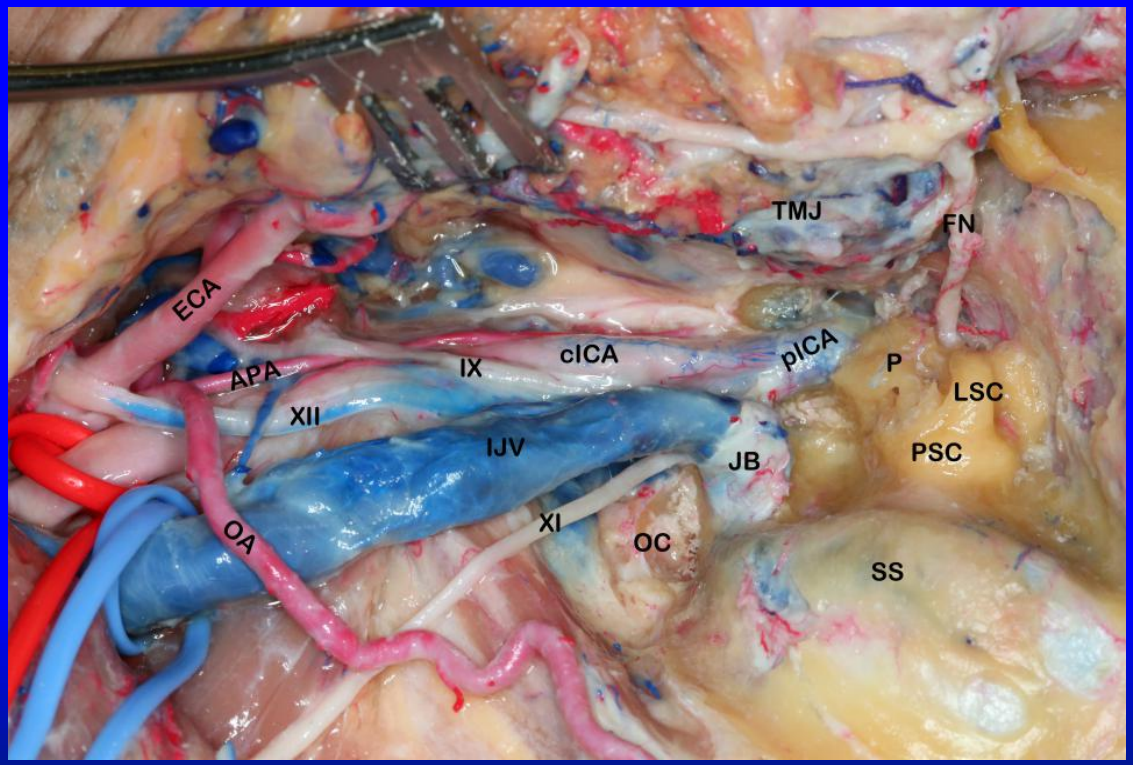
SURGICAL STRATEGIES FOR CLASS C & D TJP

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

- Infratemporal Fossa Approach Type A (WITH FACIAL NERVE RE-ROUTING) is the WORKHORSE OF Class C & D TJPs





CLASS C1

Elderly patients (≥ 65 years) with normal LCNs

Wait and Scan

Elderly patients with paralysis of LCNs

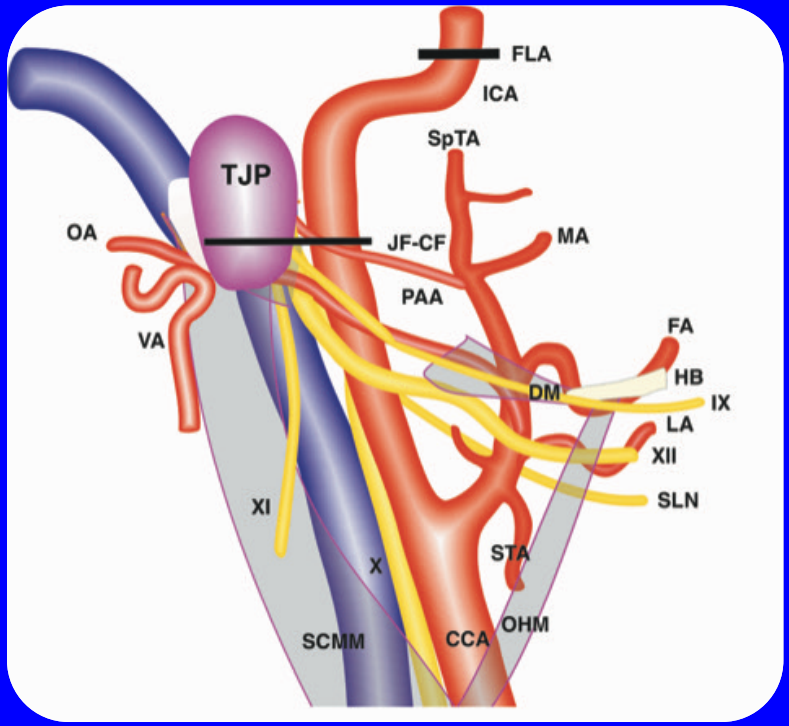
Wait and scan

Subtotal removal + Radiotherapy ?

Radiotherapy ?

Young patients (< 65 years) with normal LCN function

ITFA with preservation of the medial wall of JB if not infiltrated



CLASS C2

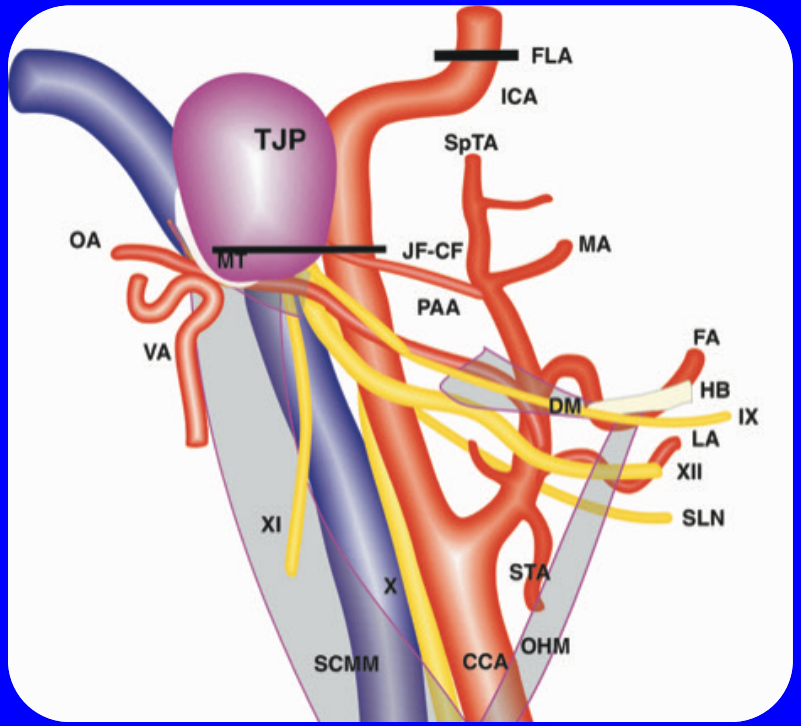
Elderly patients

Wait and scan

If growth: Subtotal removal ± Radiotherapy **?**

Young patients

ITFA with or without removal of the medial wall of JB



CLASS C3

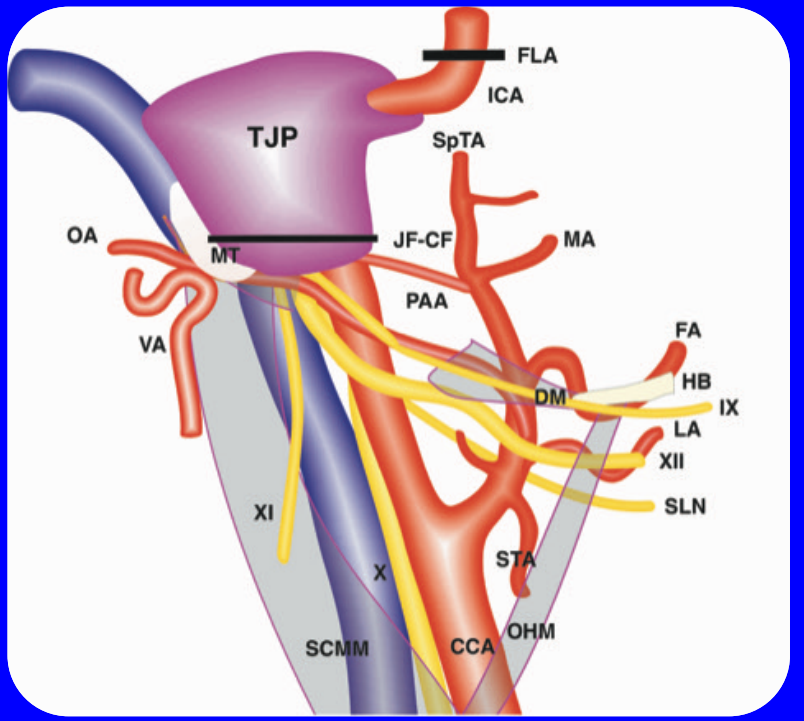
Young patients

Total tumor removal (IFTA)
after Balloon occlusion/Stenting of the ICA

If intradural extension Staged Removal
(Translabyrinthine approach)

Elderly patients

Wait and scan. If growth: Subtotal removal ± radiotherapy ?



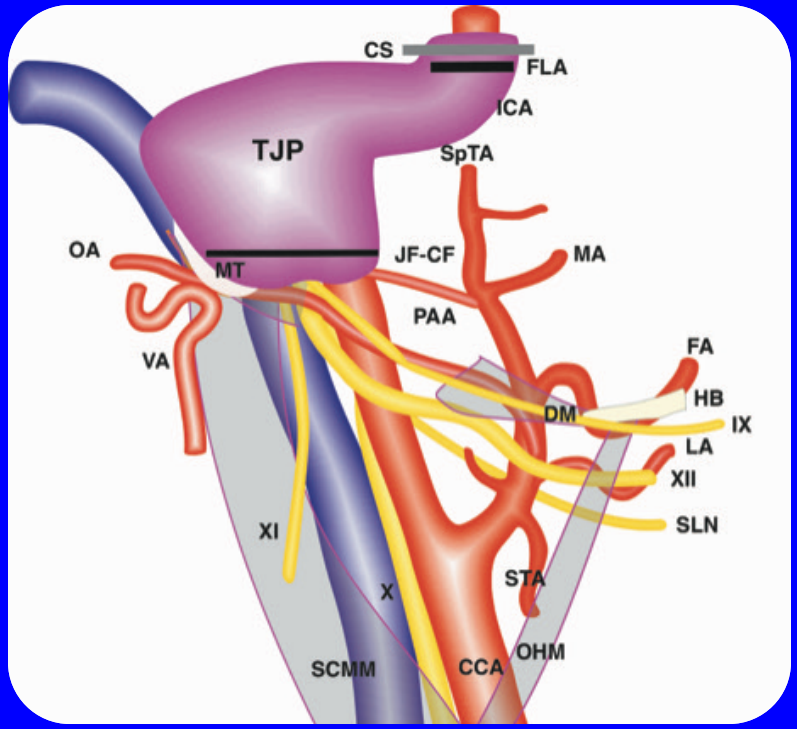
CLASS C4

Staged removal

First stage: subtotal extradural tumor removal (IFTA/ IFTB) after Balloon occlusion/Stenting of the ICA

Second stage: intradural tumor removal (Translabyrinthine approach)

Radiotherapy for the cavernous sinus



CBT - VP

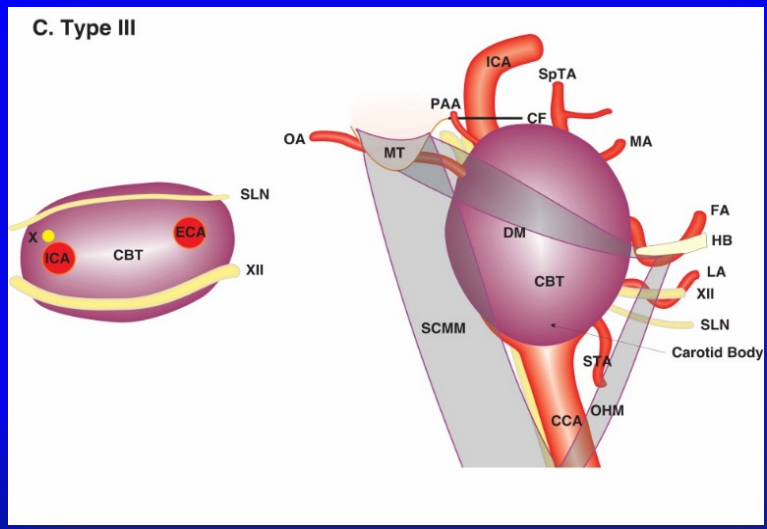
CBT

Almost always surgically operable with preservation of the LCNs with a transcervical approach, ICA stenting if Shamblin 3

VP

Wait and scan if normal X;

If X cn palsy or tumor extended into the skull base removal through transcervical approach or retroauricolar transcervical approach



FN RESULTS

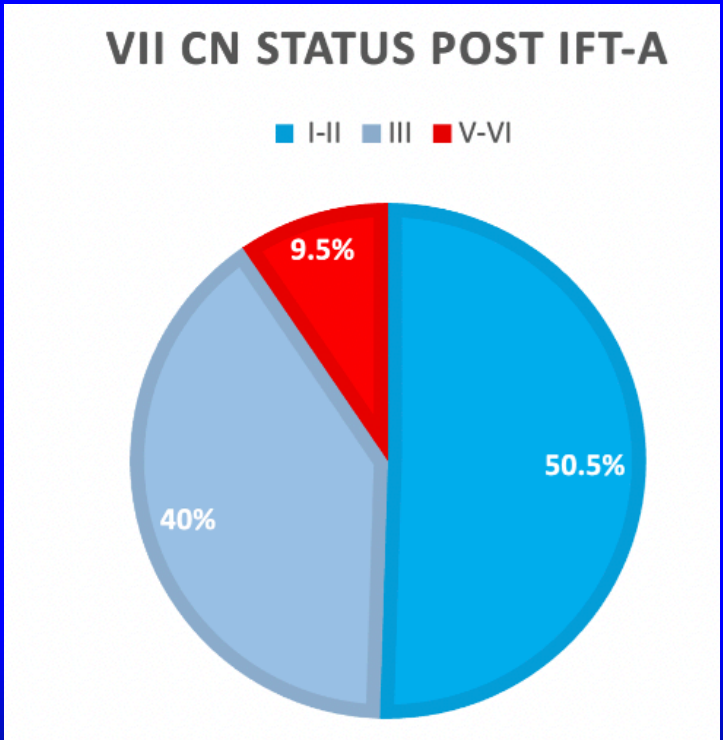
Original Study

OTOLOGY & NEUROTOLOGY

Facial Nerve Outcomes After Infratemporal Fossa Approach Type A With Anterior Rerouting for Class C-D Tympanojugular Paragangliomas Removal: Analysis From a Quaternary Referral Center

Yi Sun[a,b], Giuseppe Fancello[b,c], Melcol Hailu Yilala[b,d], Francesca Gaino[a,f], Virginia Fancello[b,g,h], Janina Roxana Becherescu[i], Lorenzo Lauda[b], Jianming Yang[a,*], Mario Sanna[b]

Results of FN rerouting

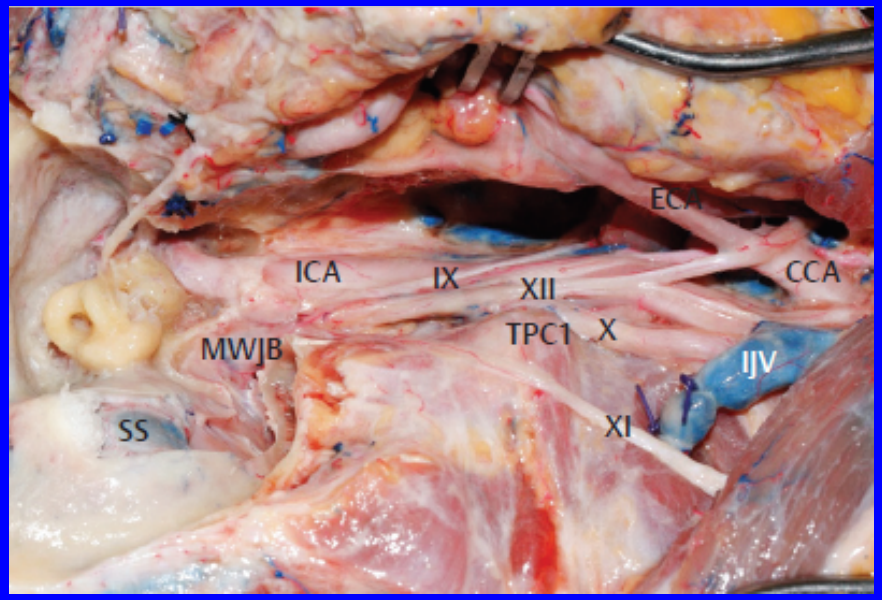


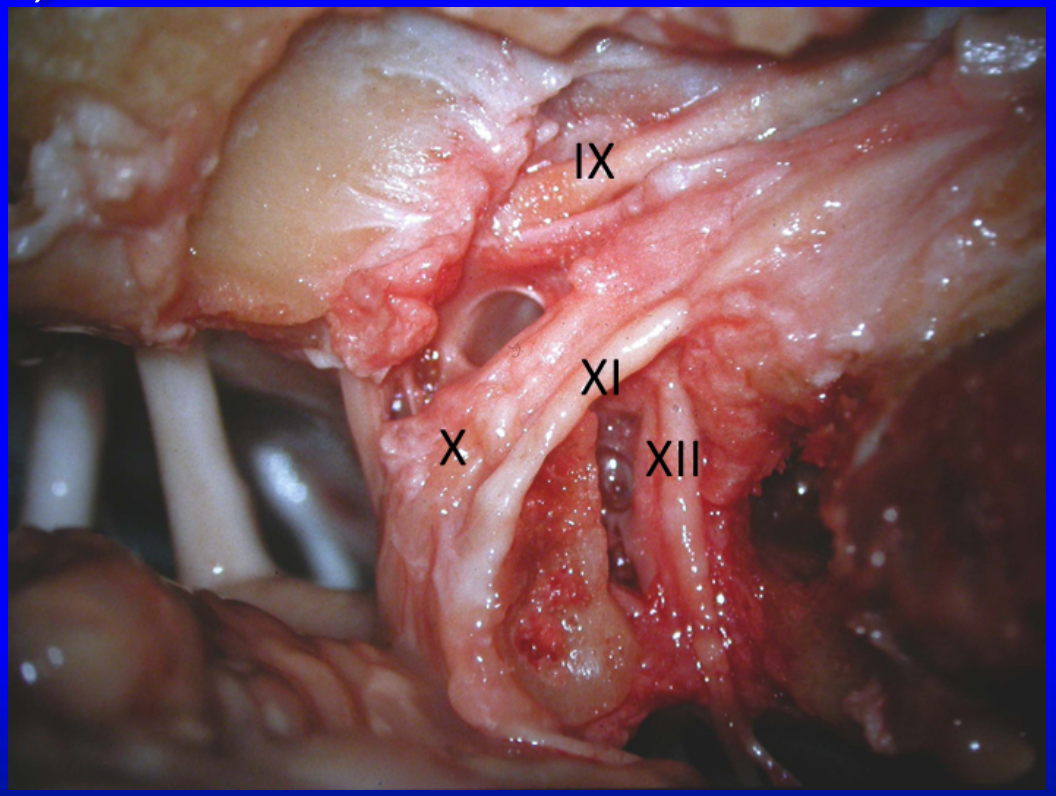
PRESERVATION OF THE MEDIAL WALL OF THE JB

MARIO SANNA FOUNDATION ONLUS

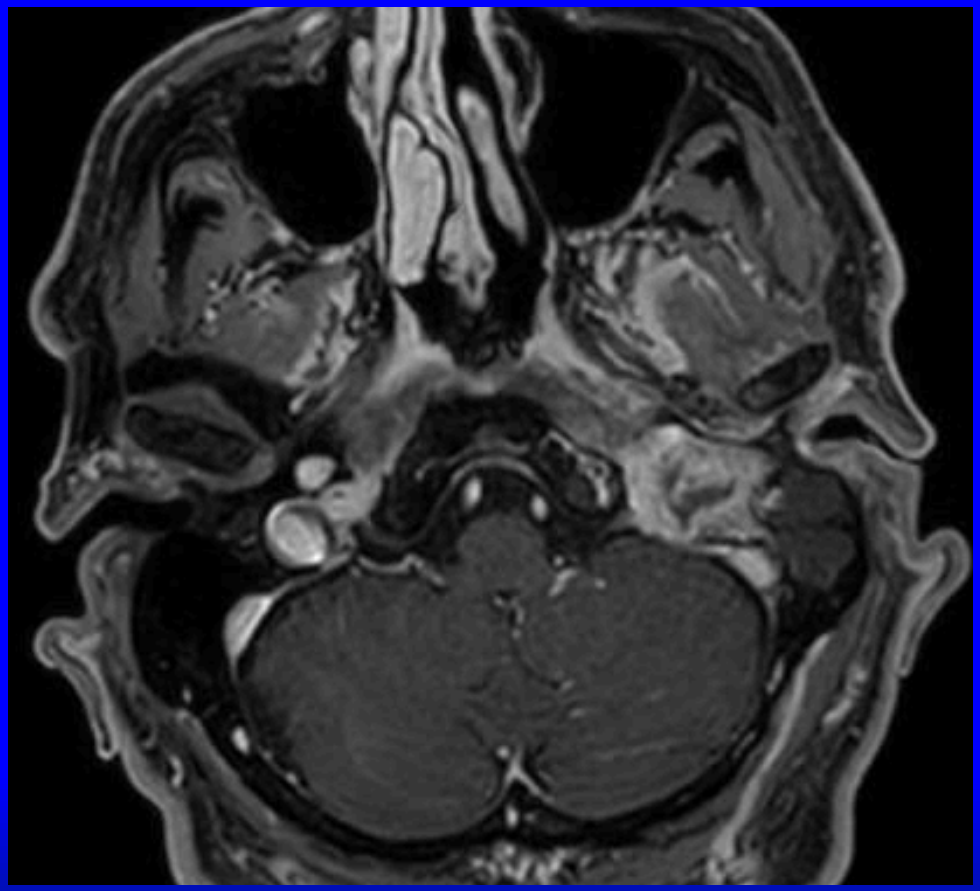
GRUPPO OTOLOGICO 1983

It allows anatomical and functional preservation of the Lower Cranial Nerves in early stages TJPs (C1-C2)





PRESERVATION OF THE MEDIAL WALL OF THE JB

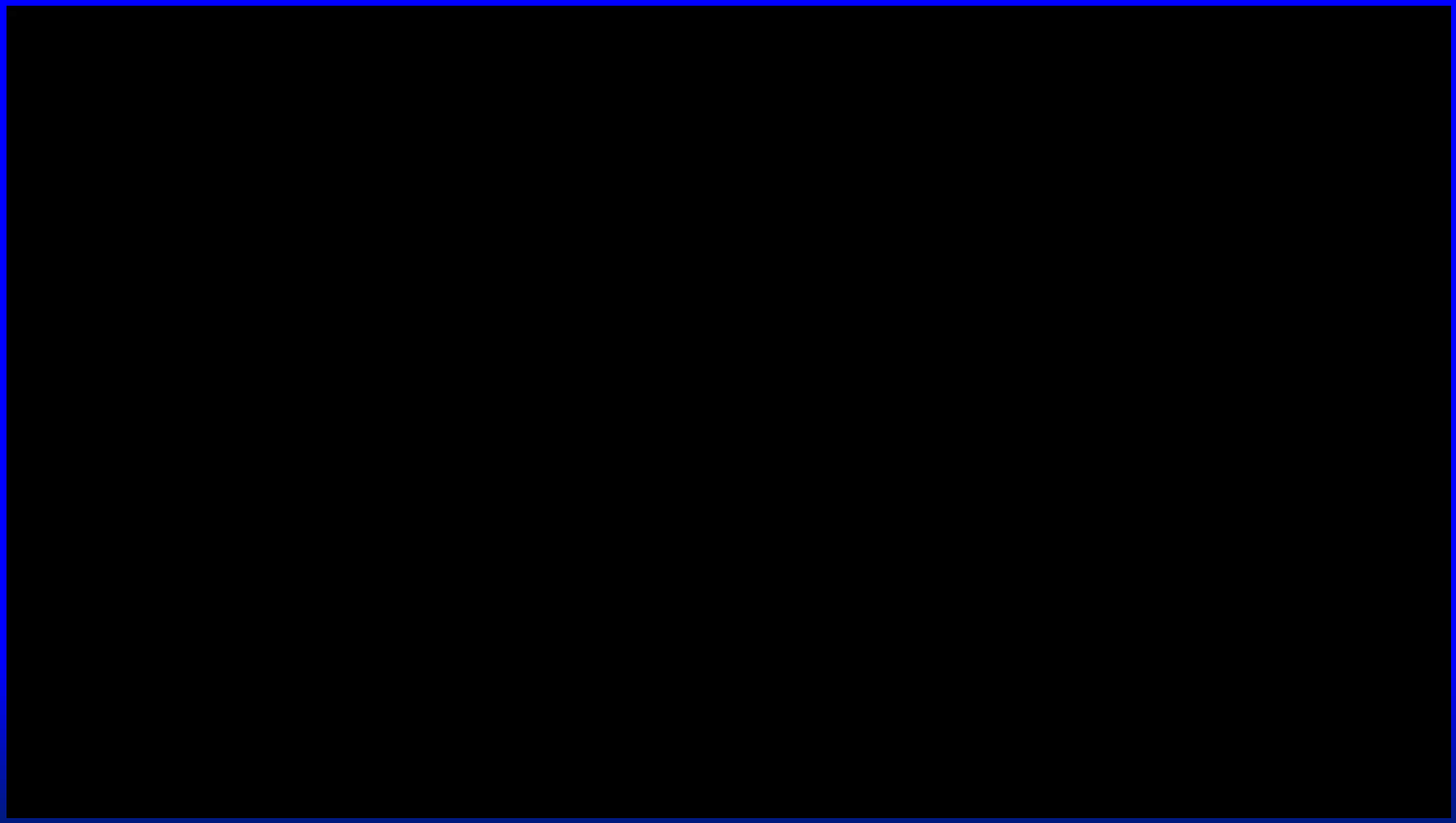


Delayed surgery due to pandemic COVID-19

Tumor growth

Ongoing facial palsy (grade VI HB)

PRESERVATION OF THE MEDIAL WALL OF THE JB



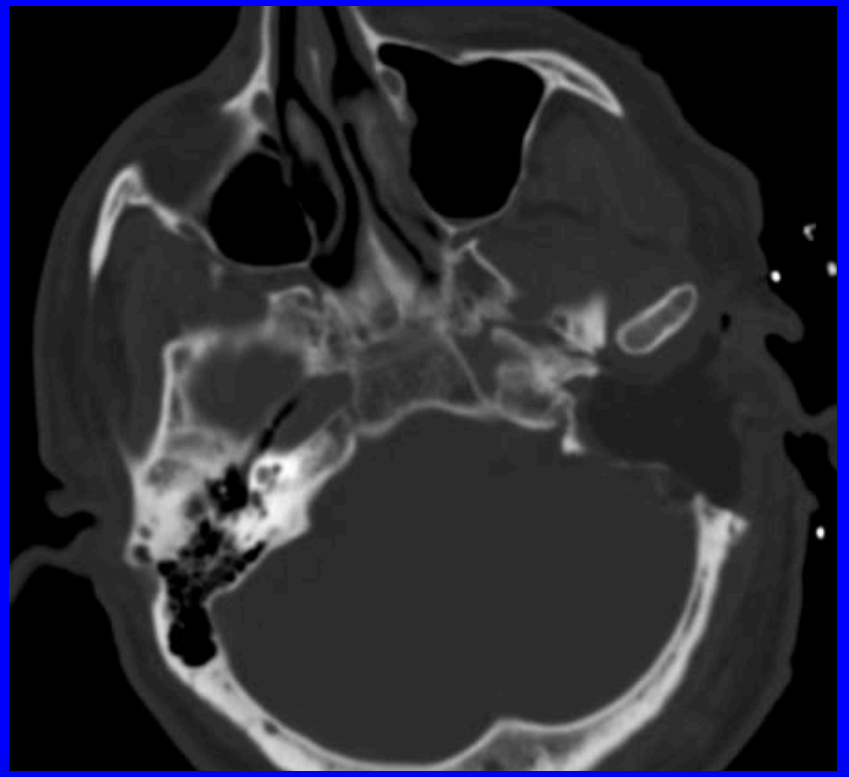
PRESERVATION OF THE MEDIAL WALL OF THE JB

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

POST-OP

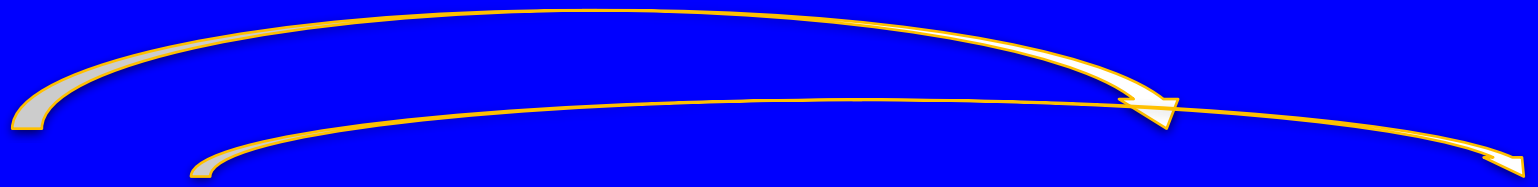
No LCNs impairment



LCN RESULTS

Pre-op (normal LCNs)

Post-op (normal LCNs)

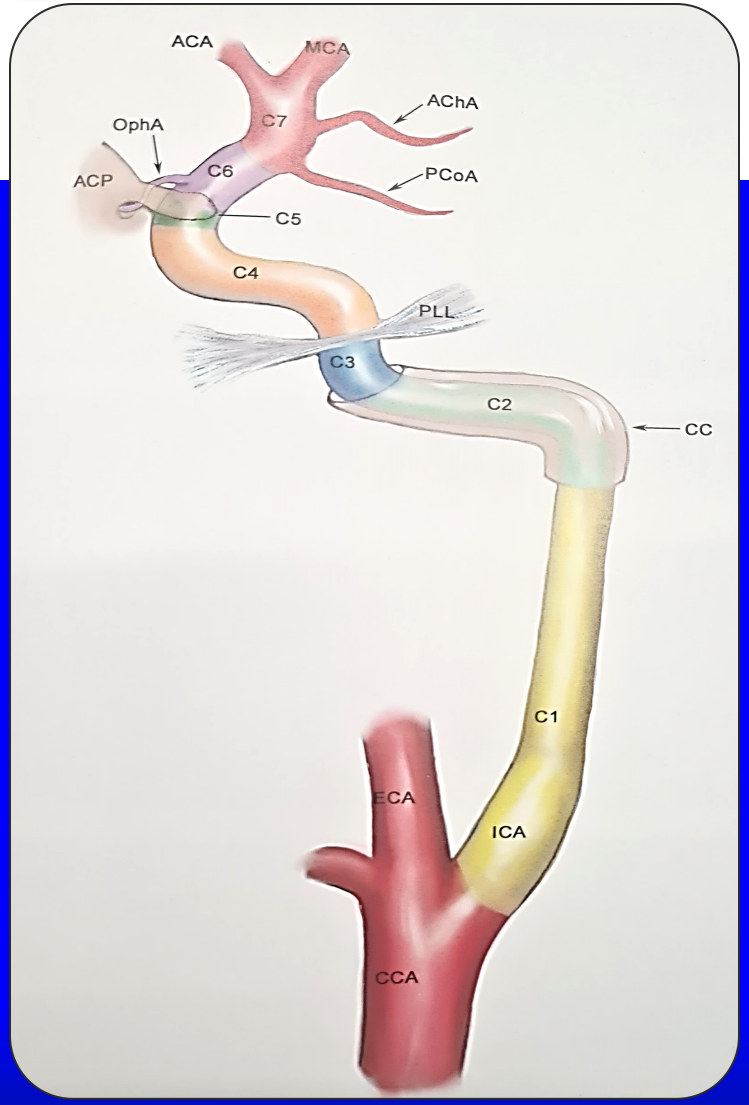


INTERNAL CAROTID ARTERY MANAGEMENT

Tumors of the lateral skull base mainly affect the first three segments

- Cervical segment (C1)
- Petrous segment (C2)
- Lacerum segment (C3)

Management of these segments is fundamental before tumor removal



STENTING OF THE ICA

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

INDICATIONS

- ICA engulfment (> 270°)
- ICA stenosis or irregularities
- Extensive blood supply from ICA
- Recurrent cases
- Insufficient collateral blood supply
- Single ipsilateral ICA

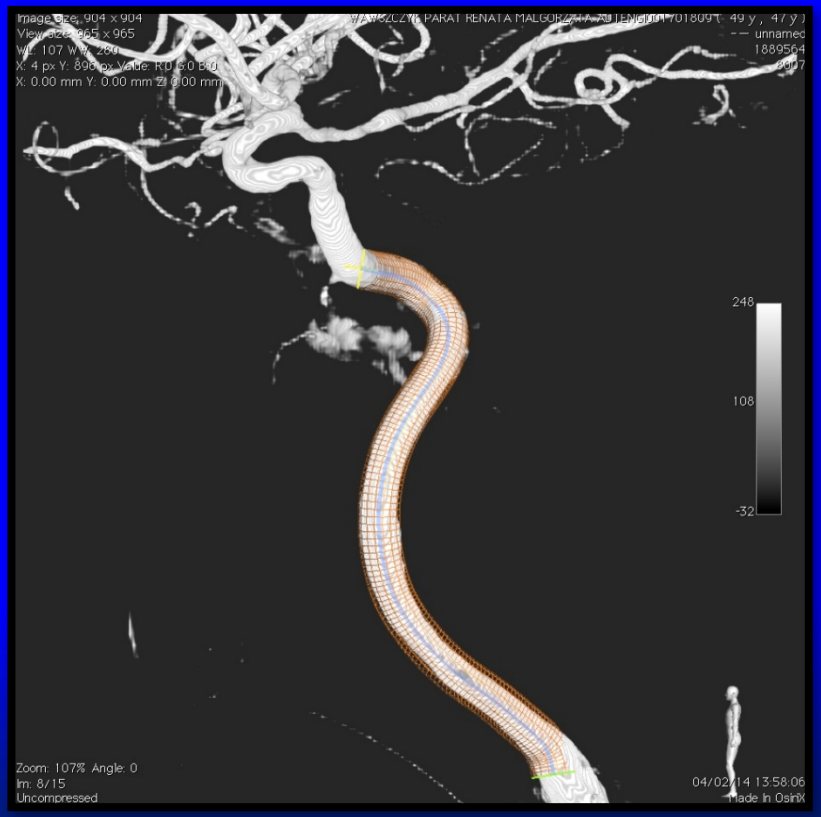
Has emerged as the 1st choice in C3-C4 TJPs and Shamblin 3 CBP and VP

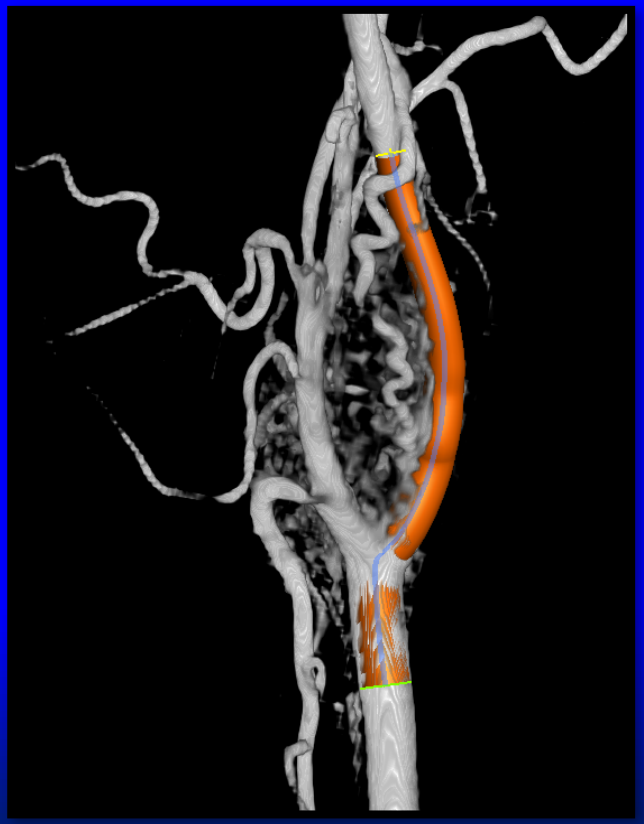
STENTING OF THE ICA

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

- Digital study before using stent to study feasibility of stenting





STENTING OF THE ICA

MARIO SANNA FOUNDATION ONLUS

GRUPPO OTOLOGICO 1983

ADVANTAGES

- Avoids theoretical short & long term complications of balloon occlusion
- Allows to operate 'inoperable' cases like SINGLE IPSILATERAL ICA

Audiol Neurootol. 2013;18(6):345-52. doi: 10.1159/000354158. Epub 2013 Oct 4.

Preoperative protective stenting of the internal carotid artery in the management of complex head and neck paragangliomas: long-term results.

Piazza P[1], Di Lella F, Bacciu A, Di Trapani G, Ait Mimoune H, Sanna M.

Open Journal of Clinical & Medical Case Reports

Volume 9 (2023)
Issue 12

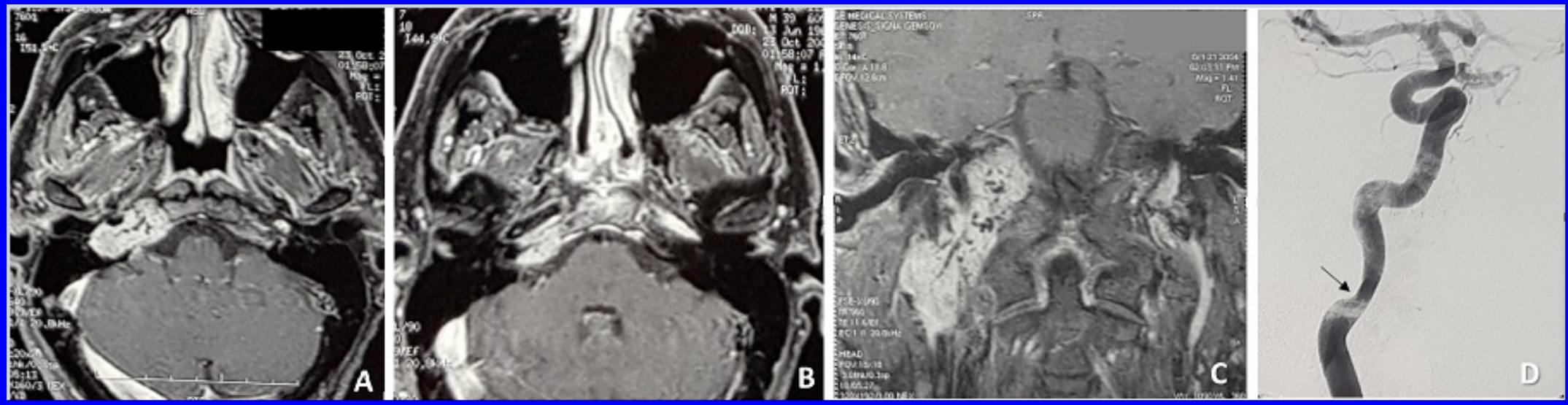
Case Report **ISSN: 2379-1039**

Long term outcomes of stenting of a single internal carotid artery in complex head and neck paragangliomas

Gianluca Piras*; Golda Grinblat; Lorenzo Lauda; Antonio Caruso; Dikran Mardighian; Mario Sanna

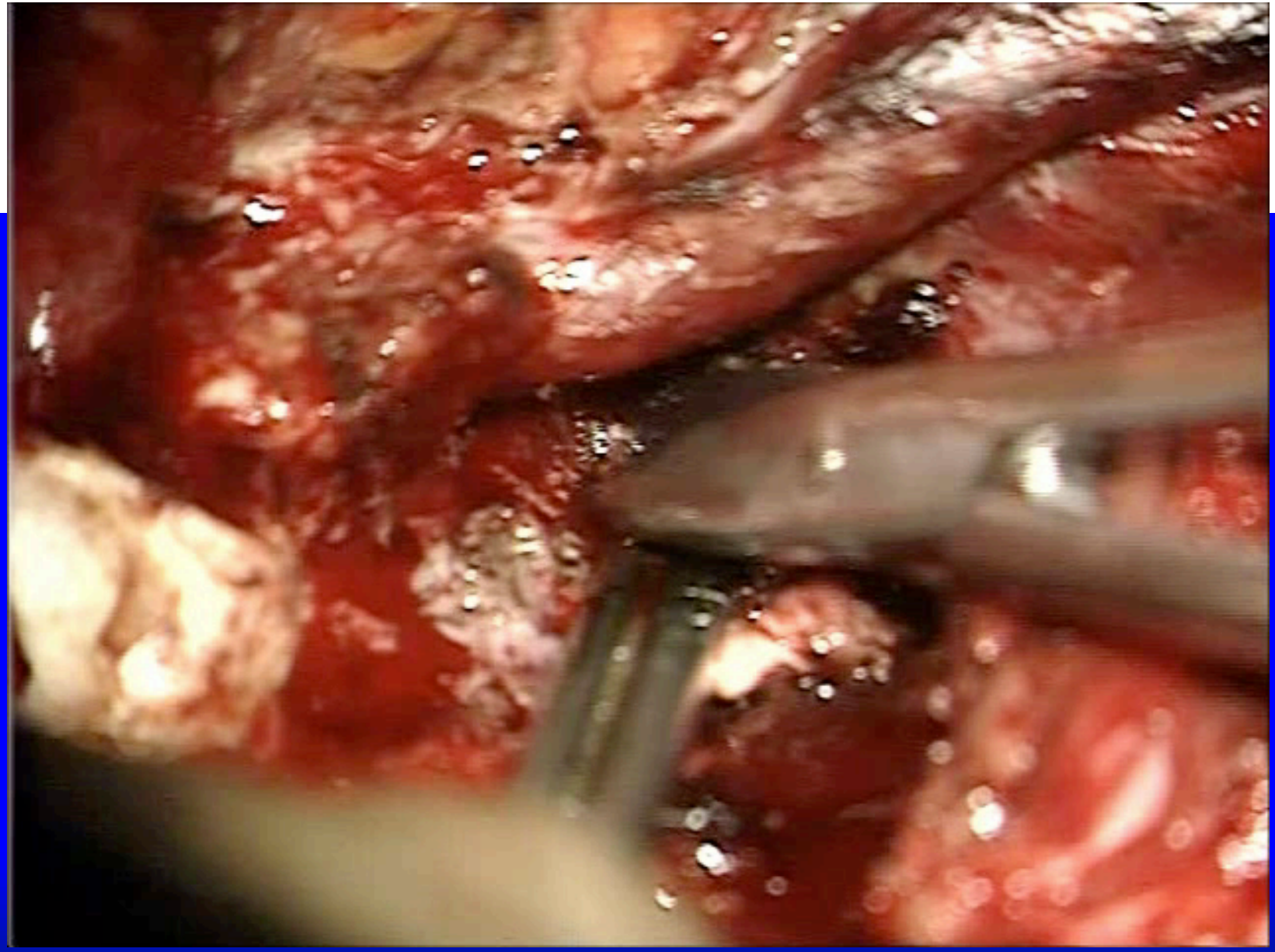
CASE: C3 TJP + VP ON A SINGLE ICA

Pre-op



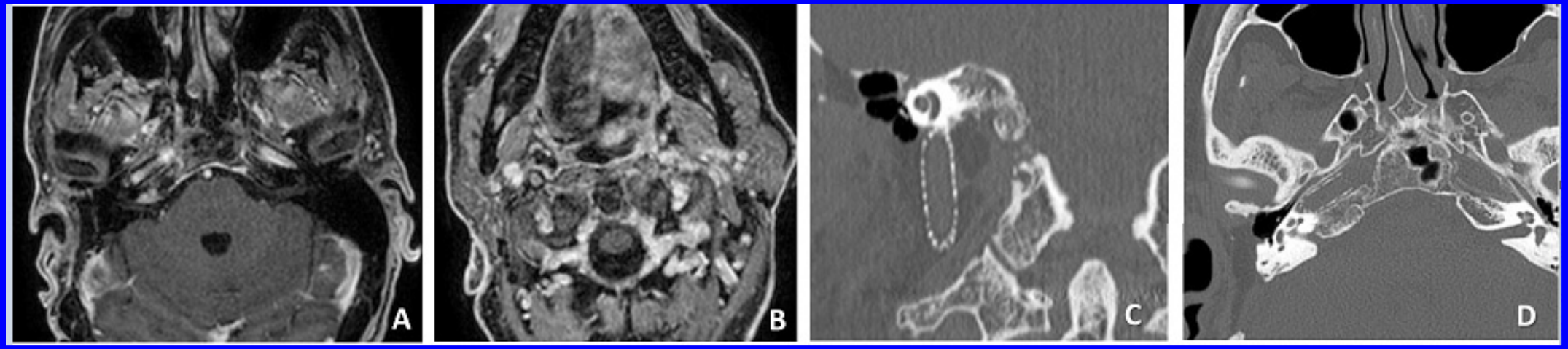
Previous left CBP removal with closure of the common ICA (vascular surgeon)

Planned IFTA (single stage with pre-operative stenting of the ICA) for the right TJP



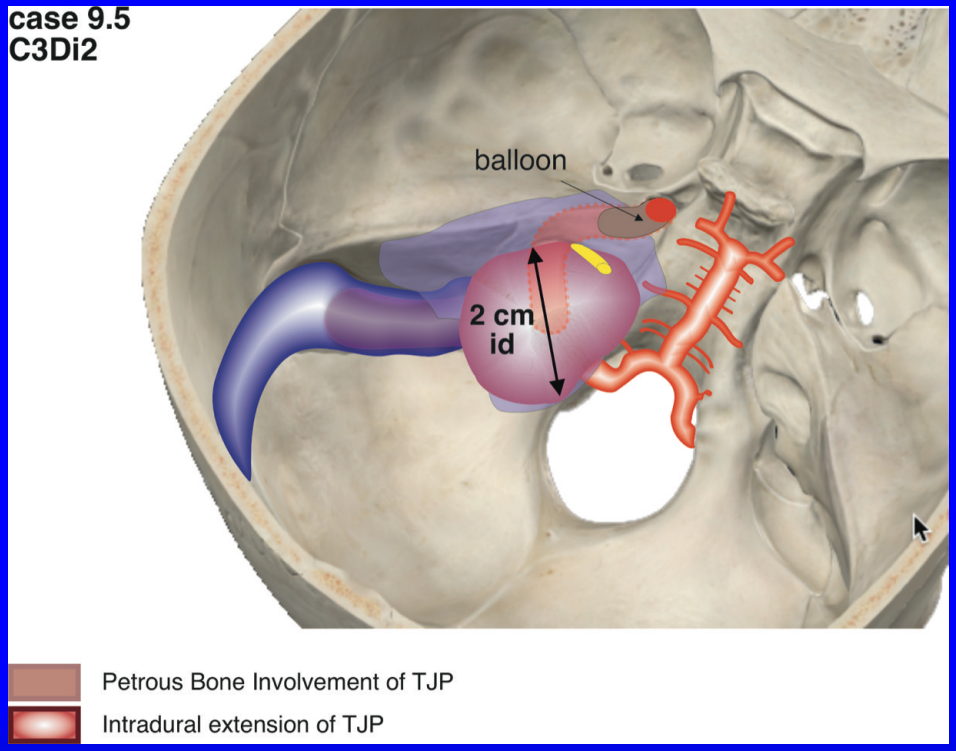
CASE: C3 TJP + VP ON A SINGLE ICA

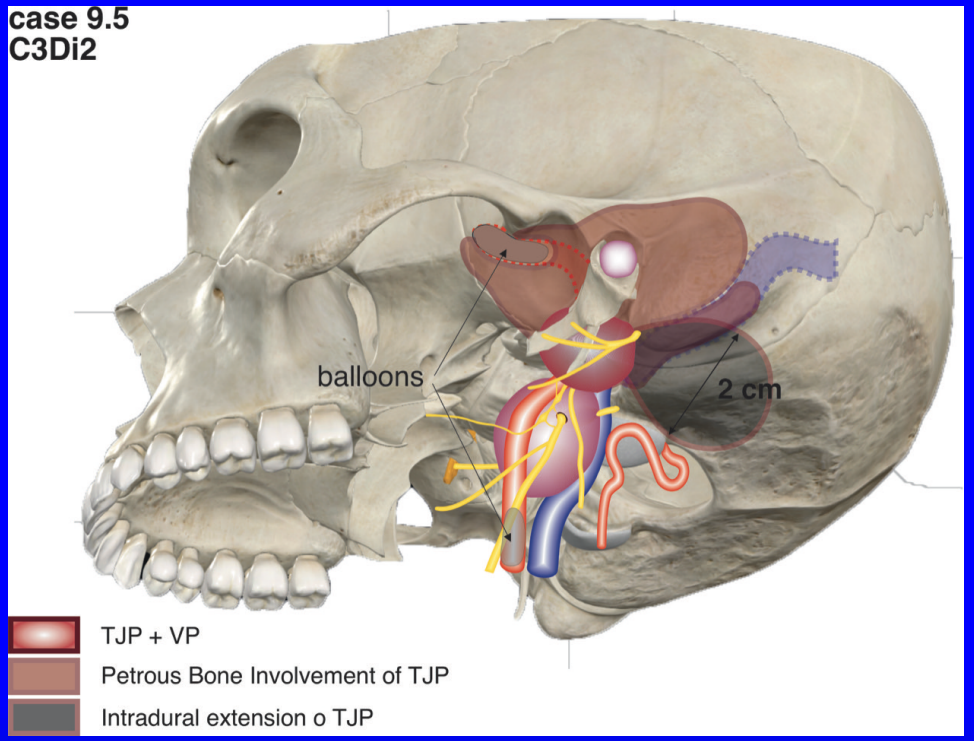
Post-op (11 yrs)



CLINICAL CASES

CASE 1
CLASS C3Di2 TJP + VP + CBT

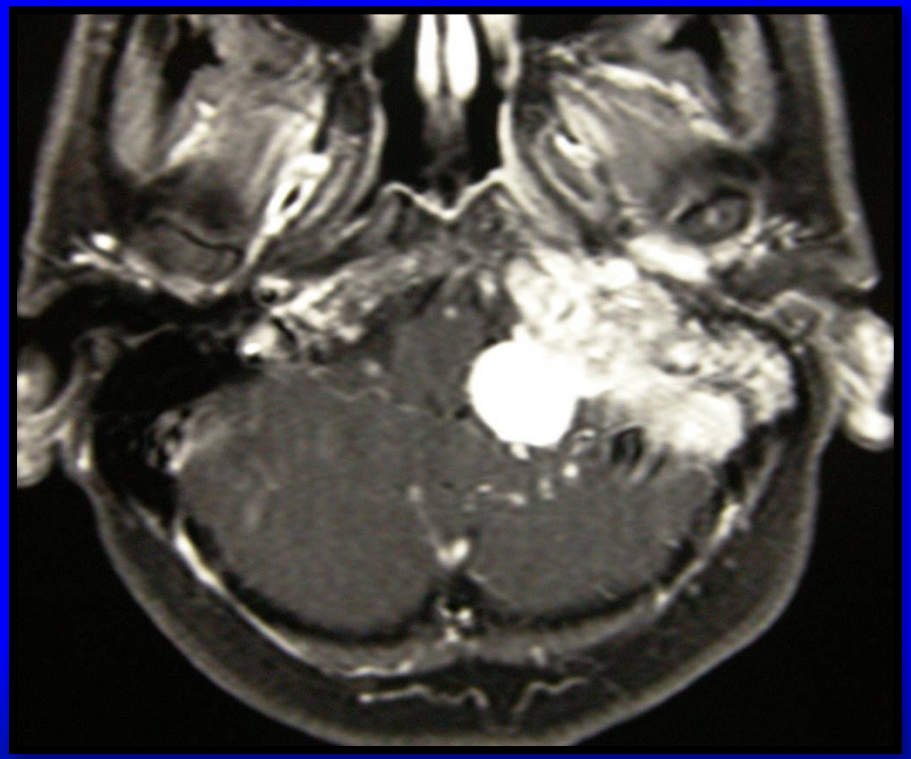


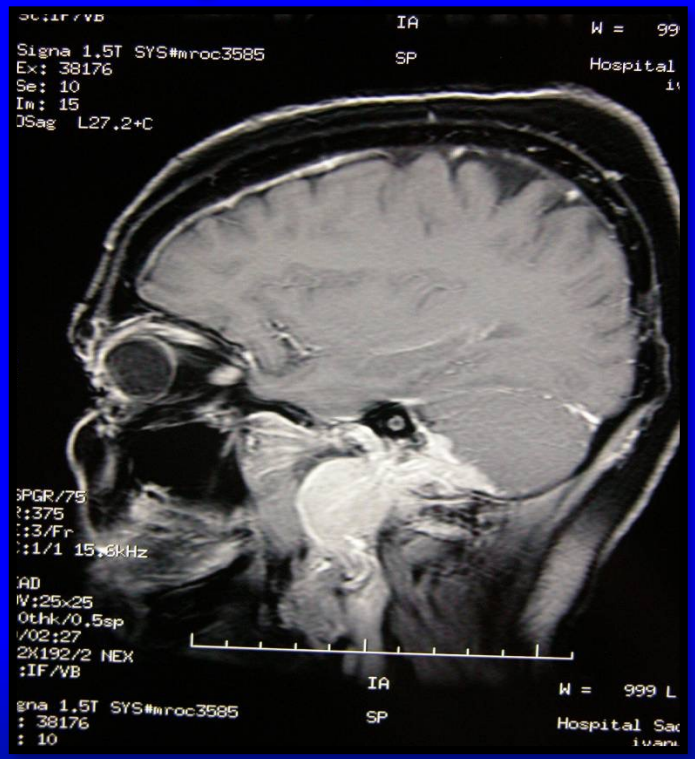


CASE 1
CLASS C3Di2 TJP +VP + CBT

52 yrs woman

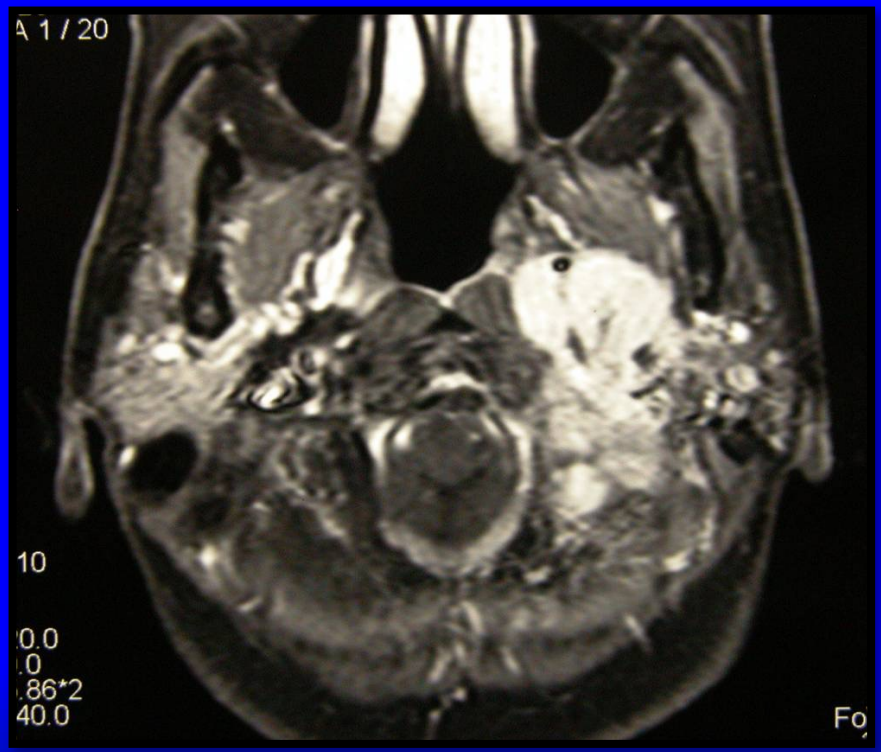
4-year history of dysphonia, progressive hearing loss, pulsatile tinnitus





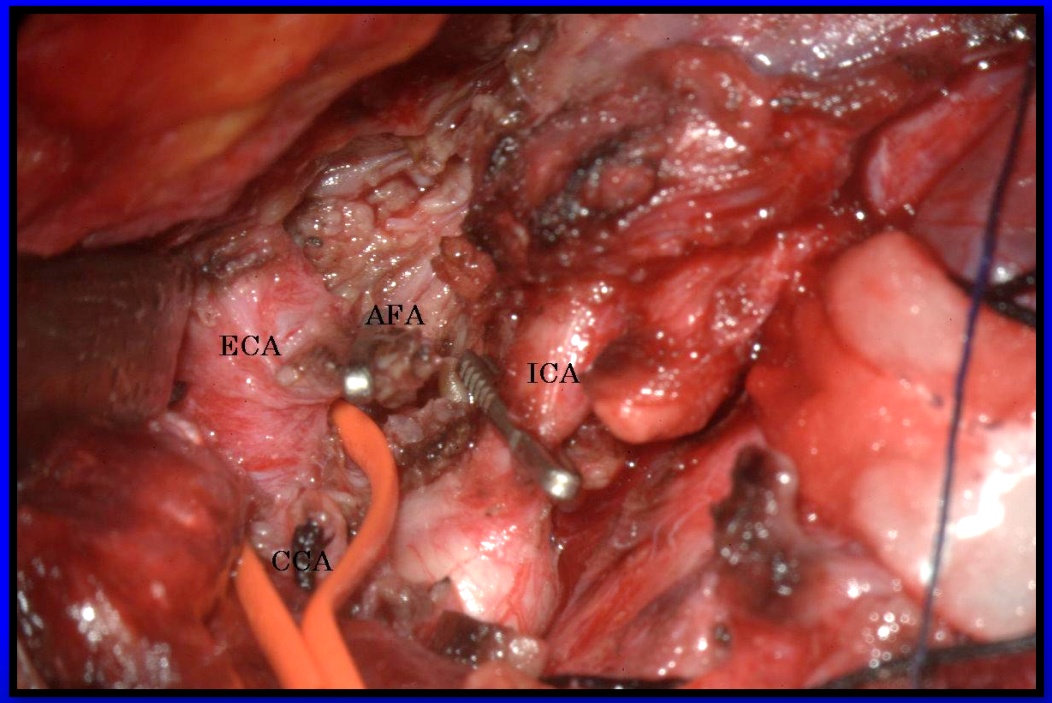
CASE 1
CLASS C3Di2 TJP +VP + CBT

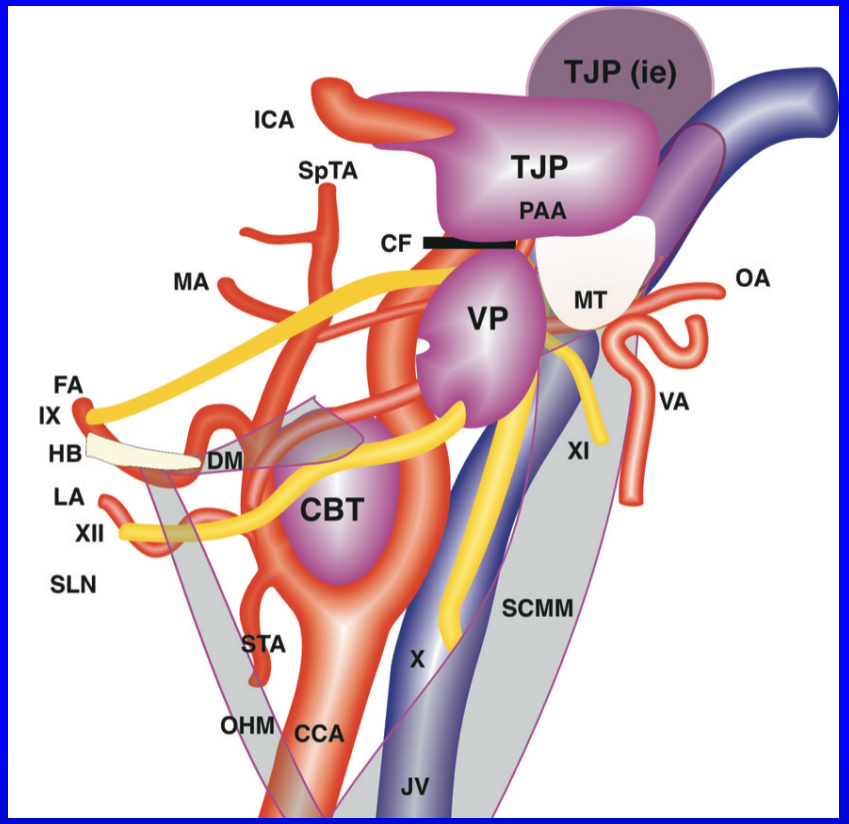
- Severe stenosis of the ascending portion of the petrous ICA



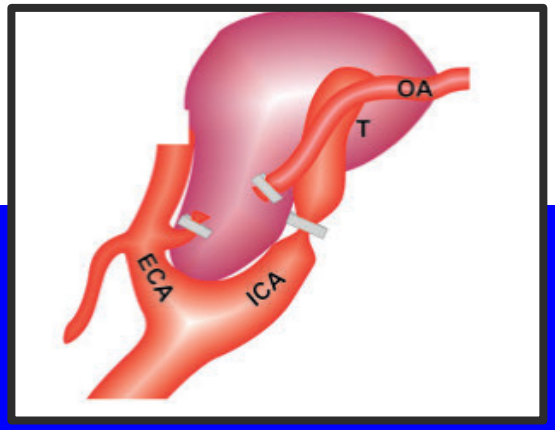
CASE 1
CLASS C3Di2 TJP +VP + CBT

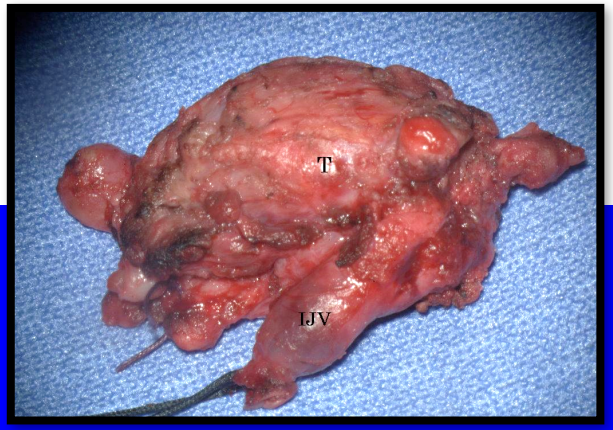
- I st stage: ITFA Type A after PBO



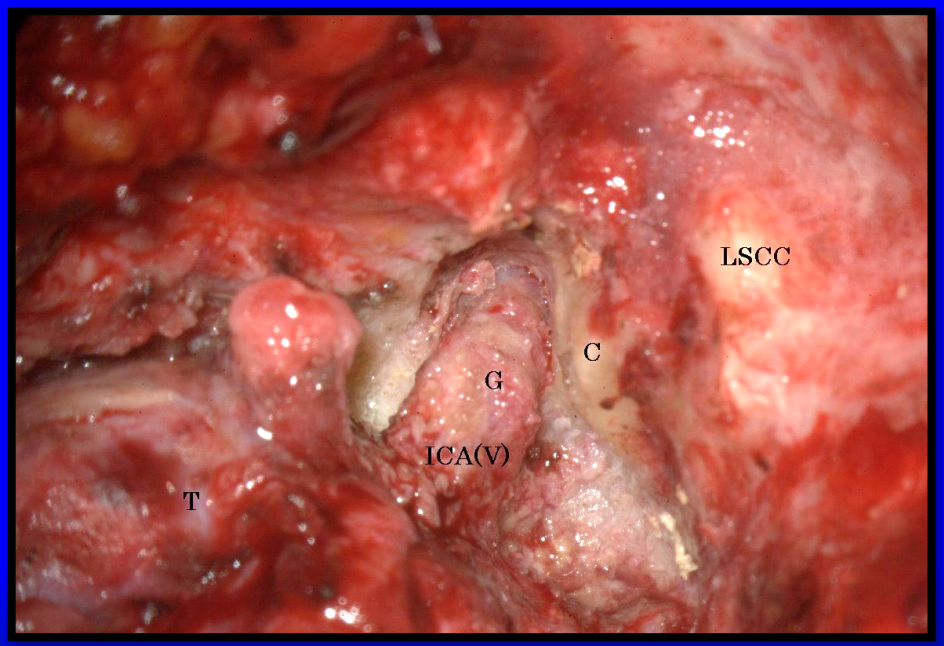


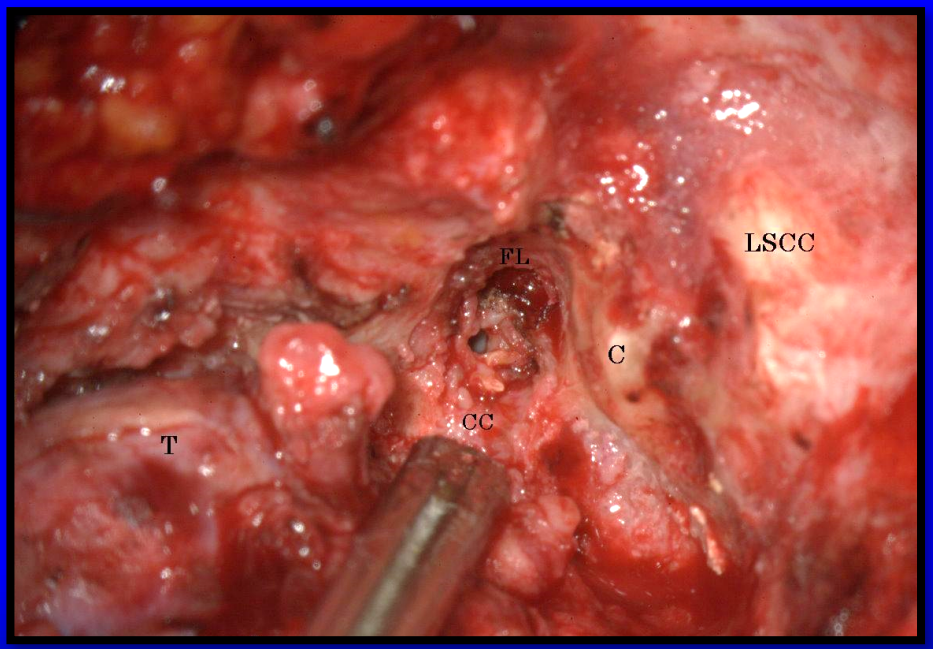
CASE 1
CLASS C3Di2 TJP +VP + CBT





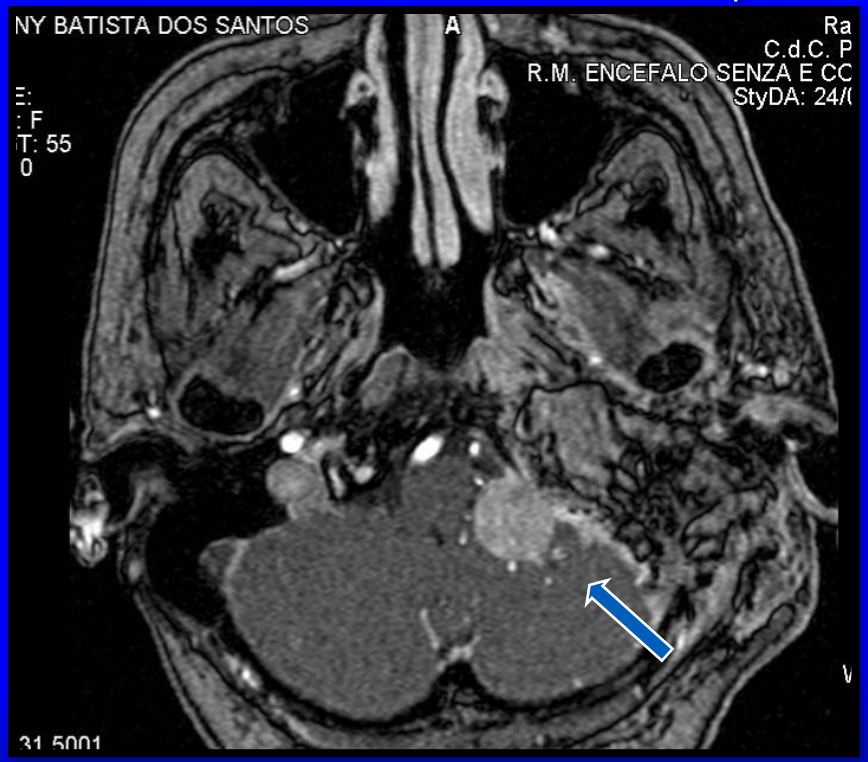
- I st stage: ITFA Type A after PBO

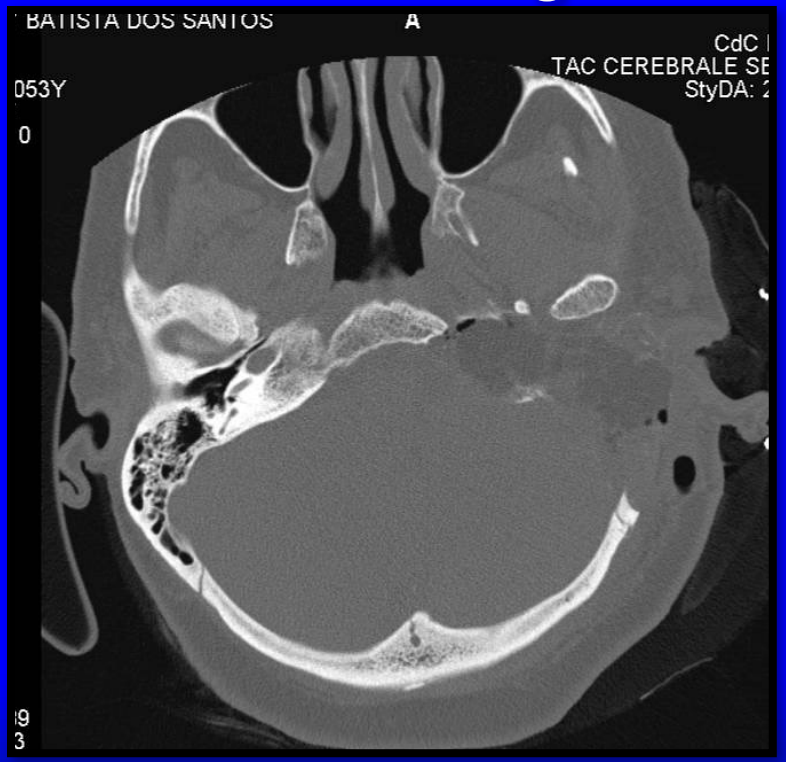




CASE 1
CLASS C3Di2 TJP +VP + CBT

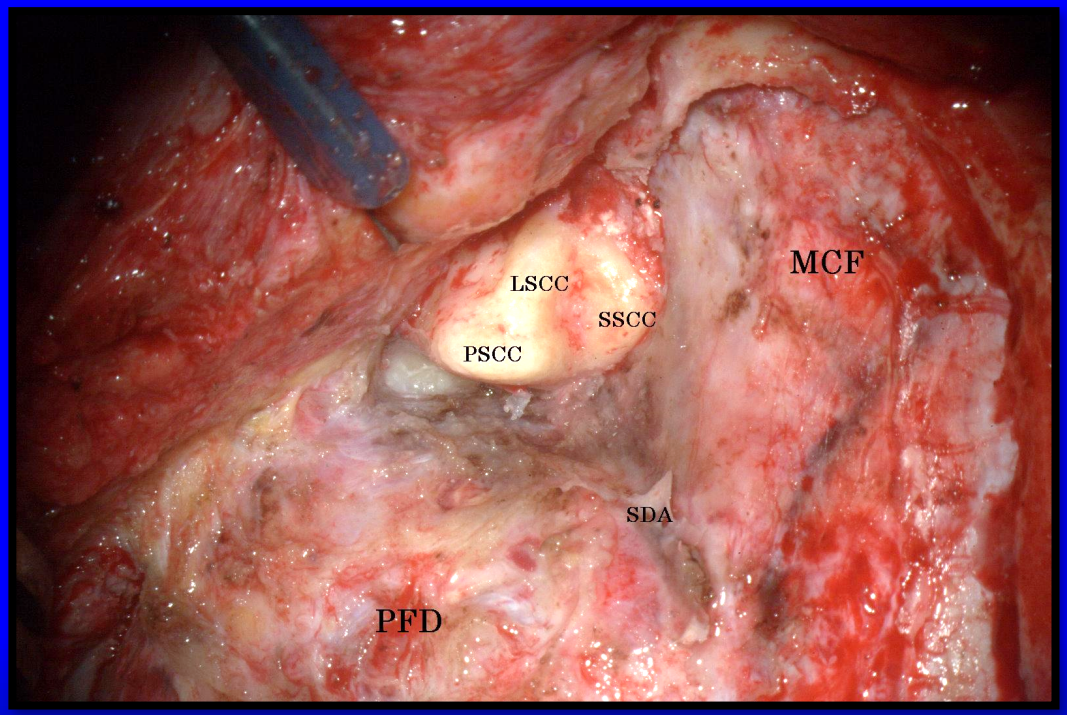
- Intradural component in situ after Ist stage

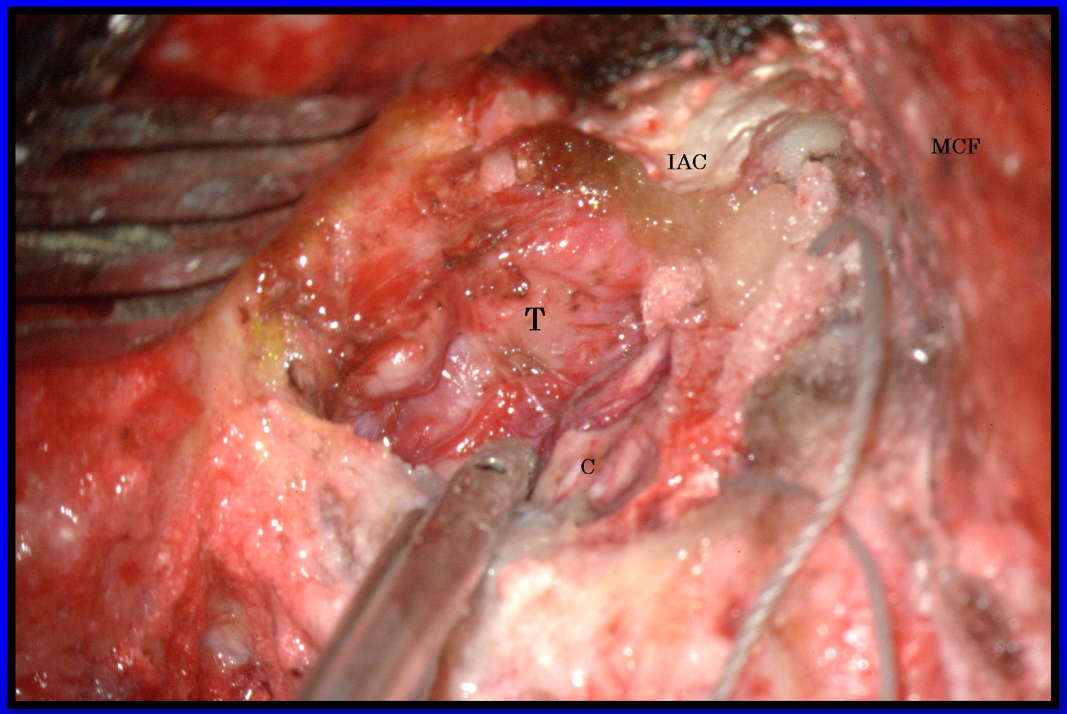




CASE 1
CLASS C3Di2 TJP +VP + CBT

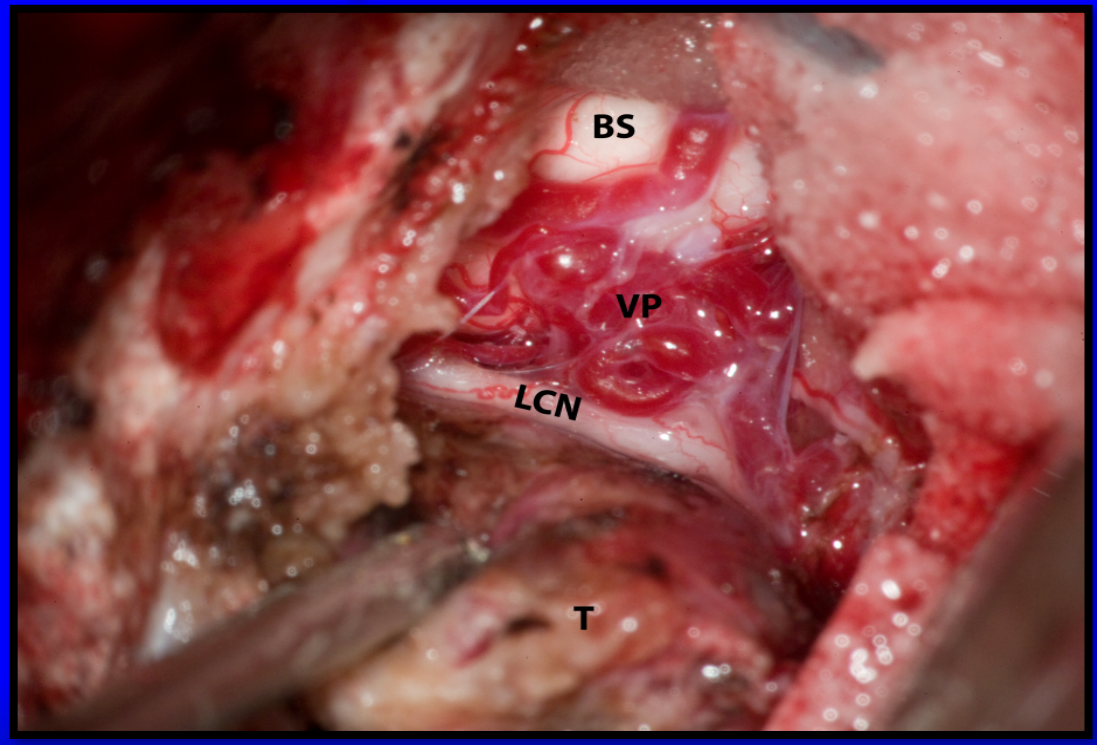
- 2nd stage : TRANSLABYRINTHINE APPROACH

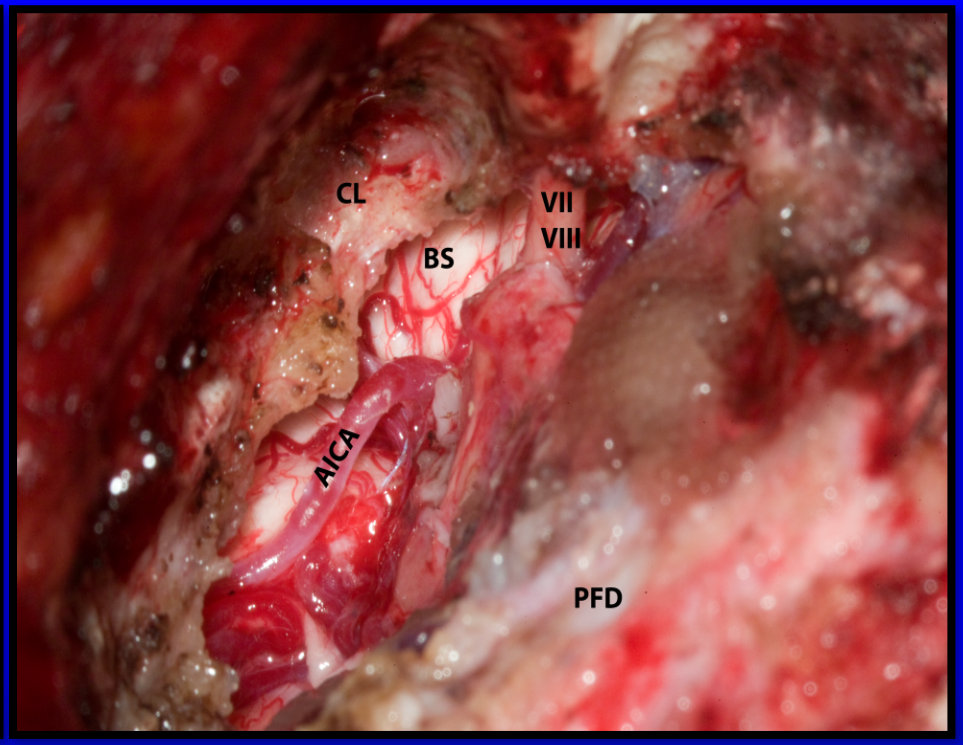




CASE 1
CLASS C3Di2 TJP +VP + CBT

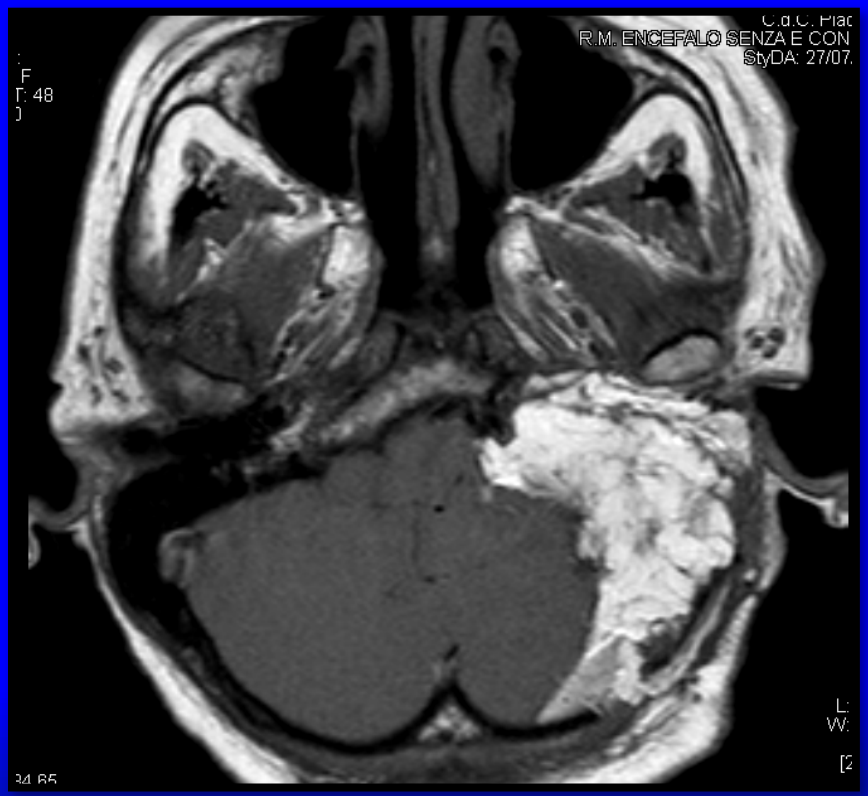
- 2nd stage : TRANSLABYRINTHINE APPROACH total tumor removal



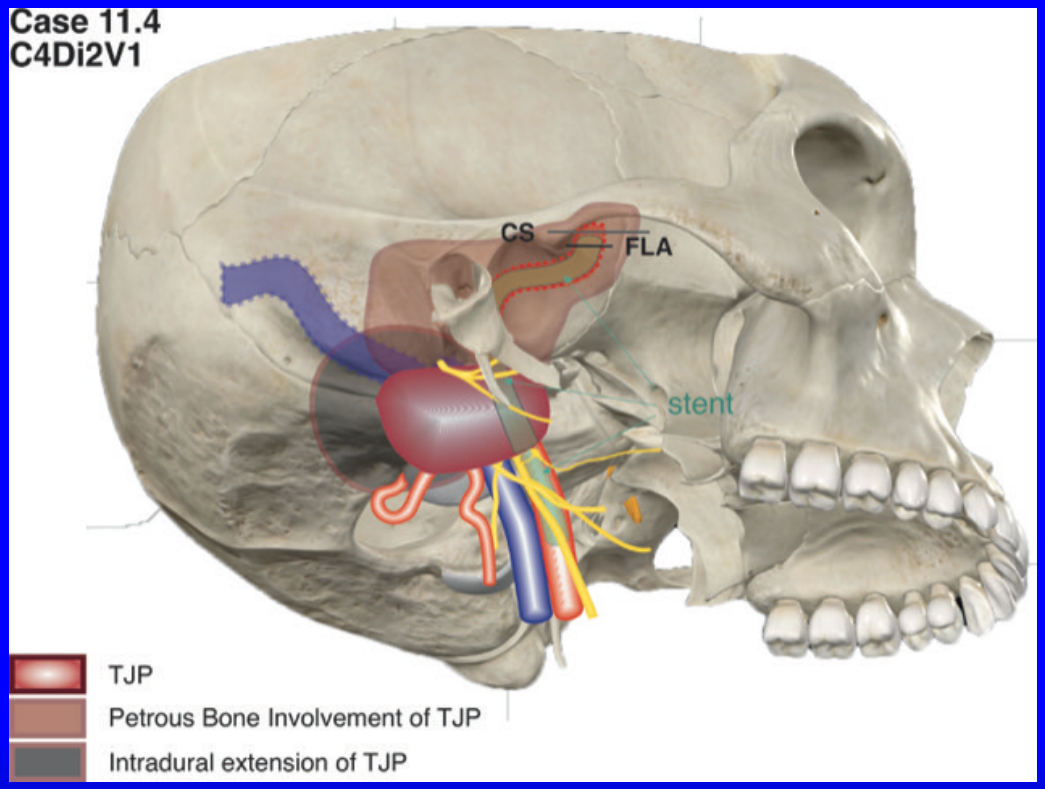


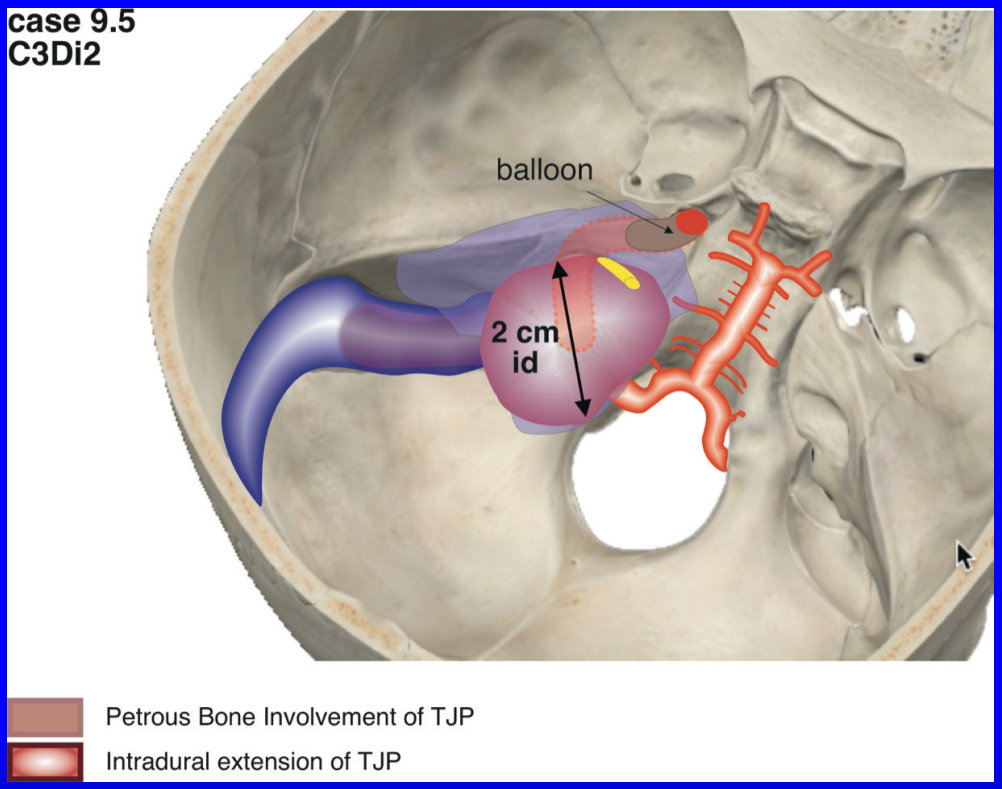
CASE 1
CLASS C3Di2 TJP +VP + CBT

- Post-op



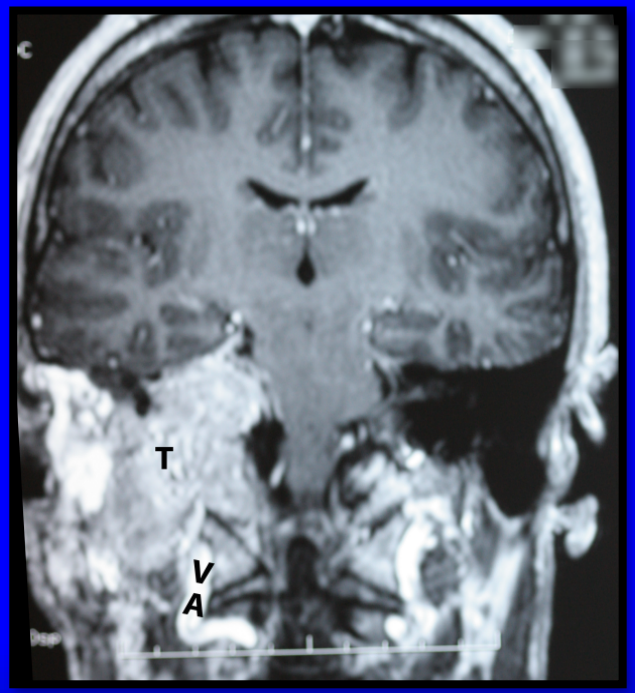
CASE 2: CLASS C3Di2Ve TJP

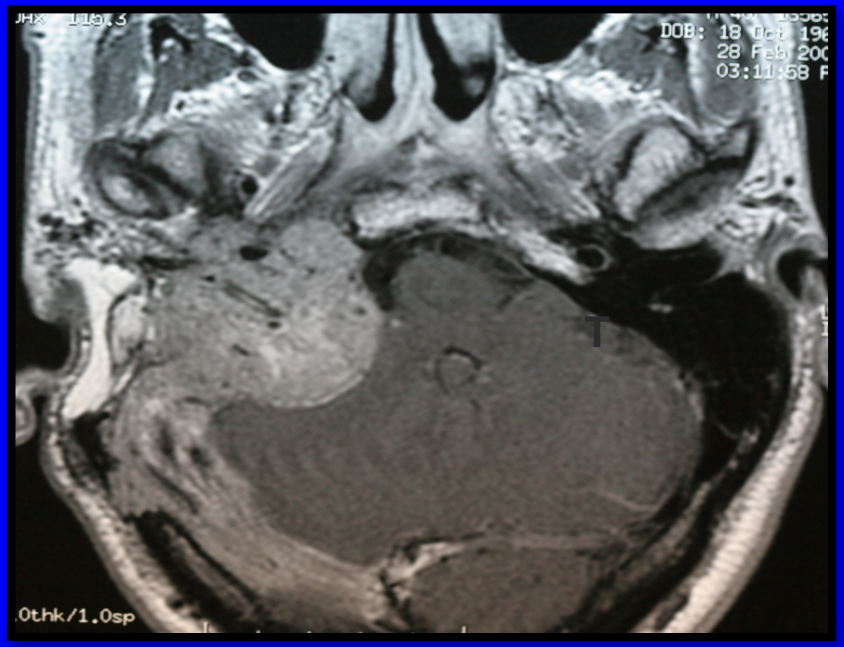


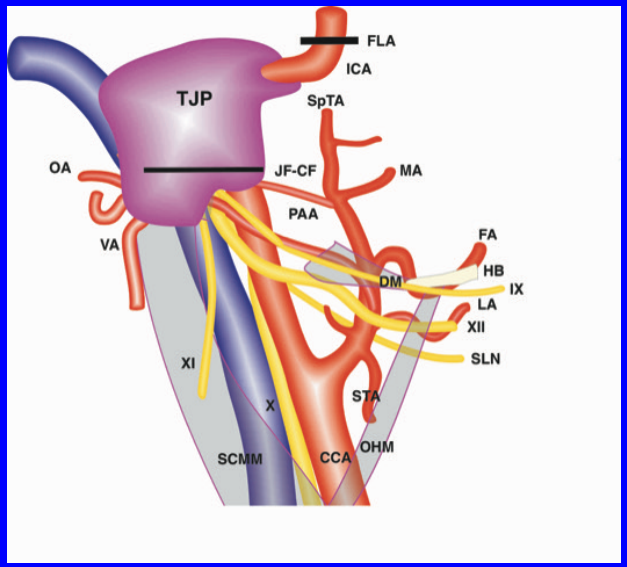


CASE 2: CLASS C3Di2Ve TJP

50 yrs old, 3 prior surgeries elsewhere ICA + Vertebral artery involvement

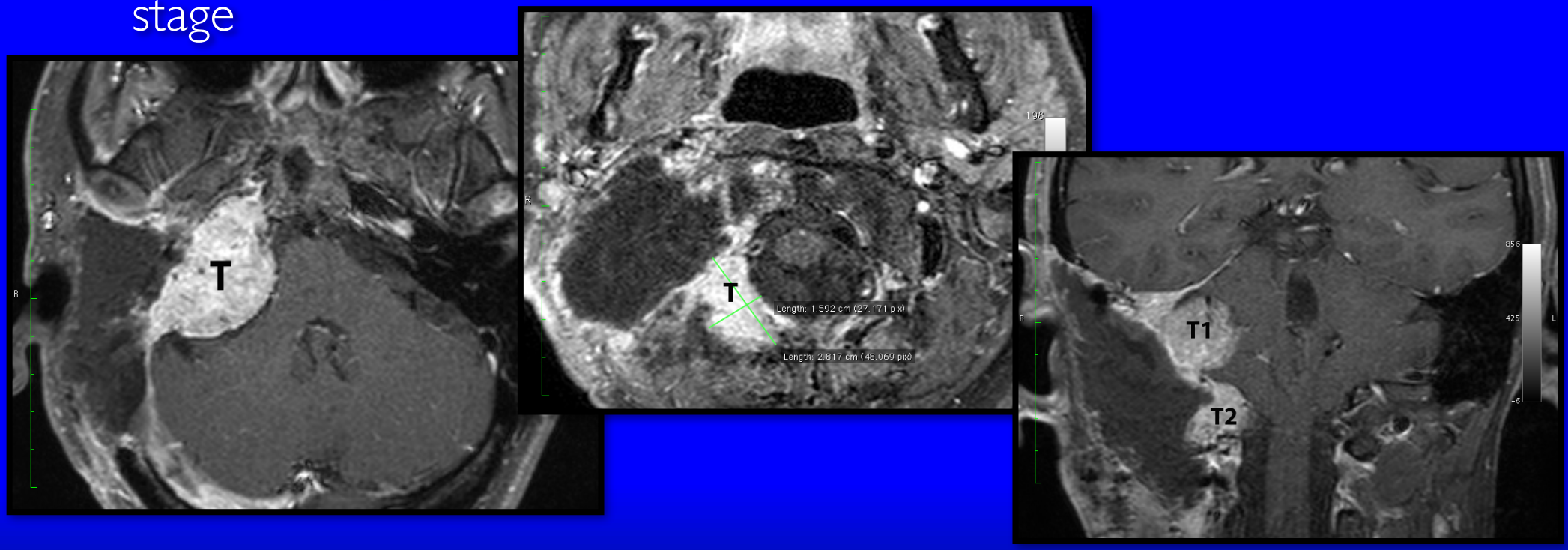






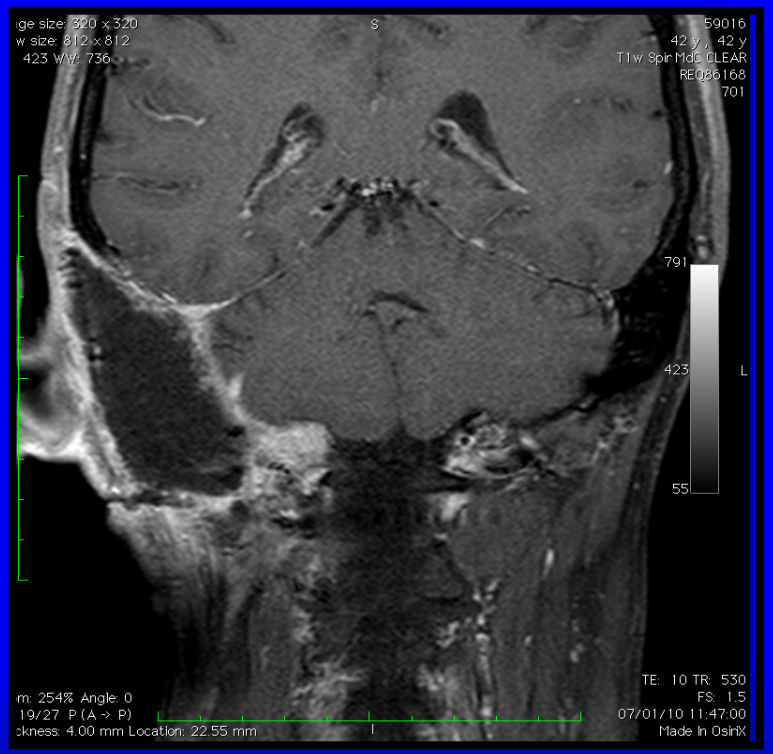
CASE 2: CLASS C3Di2Ve TJP

After 1st stage ITFA type A: intradural component left behind for 2nd stage



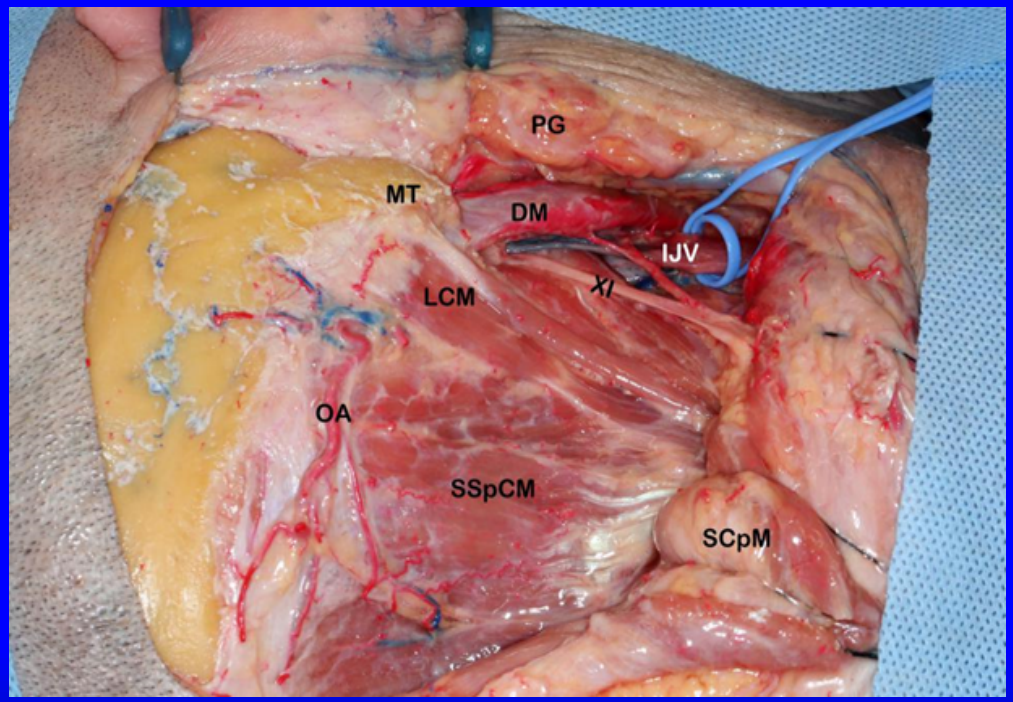
CASE 2: CLASS C3Di2Ve TJP

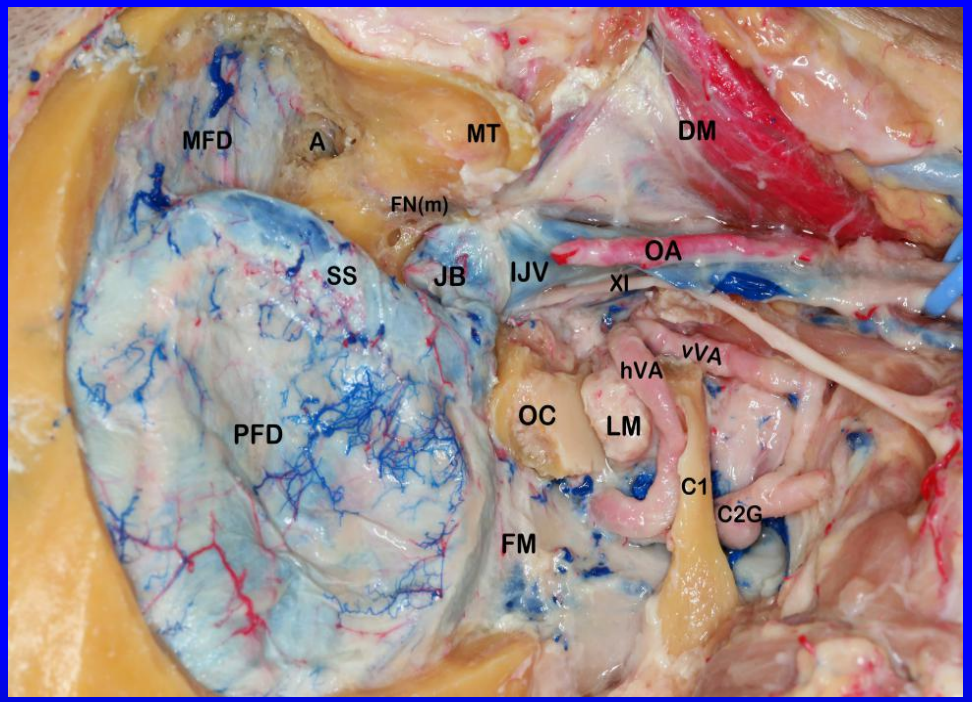
After 2nd stage ITFA type A: still some residual tumor near the foramen magnum



CASE 2: CLASS C3Di2Ve TJP

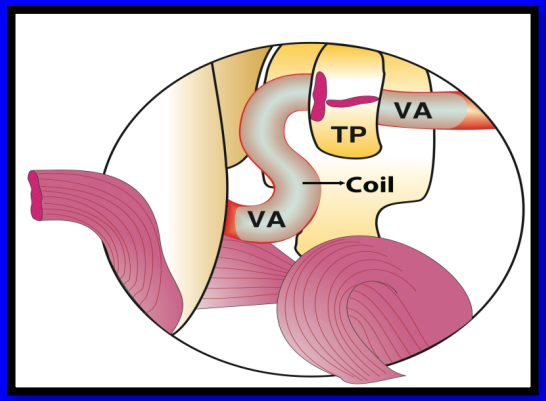
Extreme Lateral Transcondylar Approach to remove the last part of the tumor

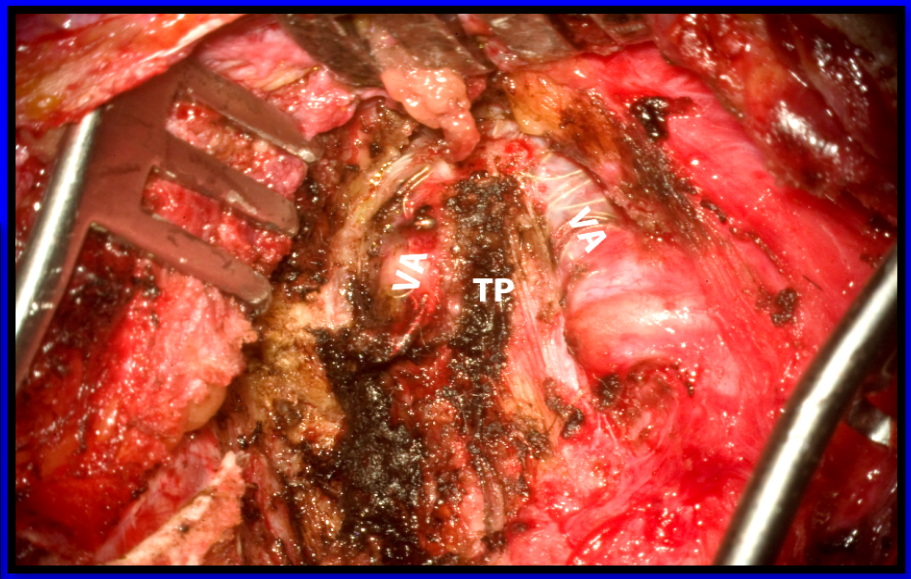




CASE 2: CLASS C3Di2Ve TJP

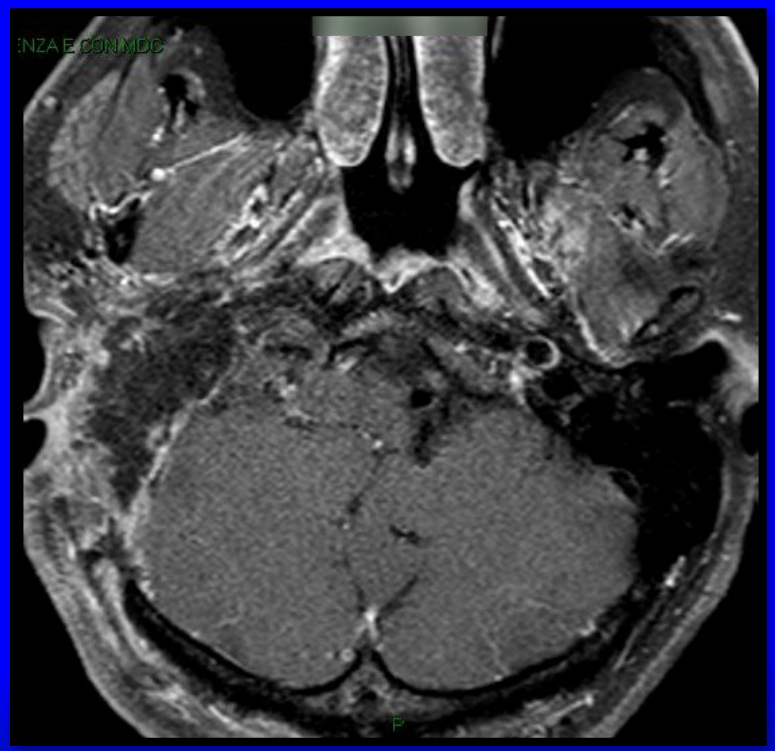
Extreme Lateral Transcondylar Approach after PBO of the Vertebral artery

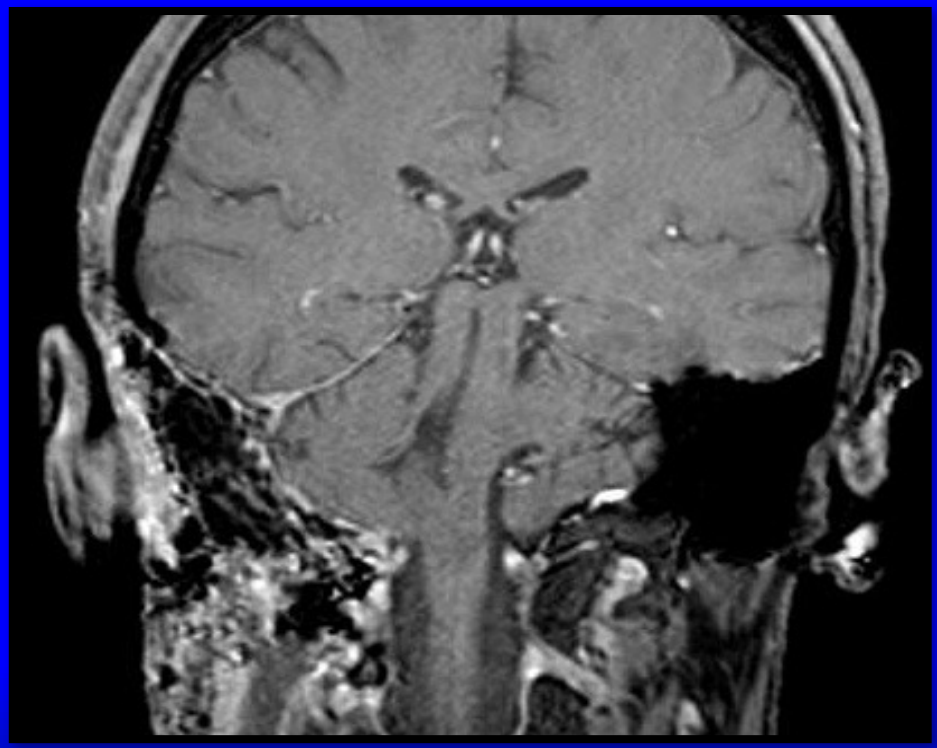




CASE 2: CLASS C3Di2Ve TJP

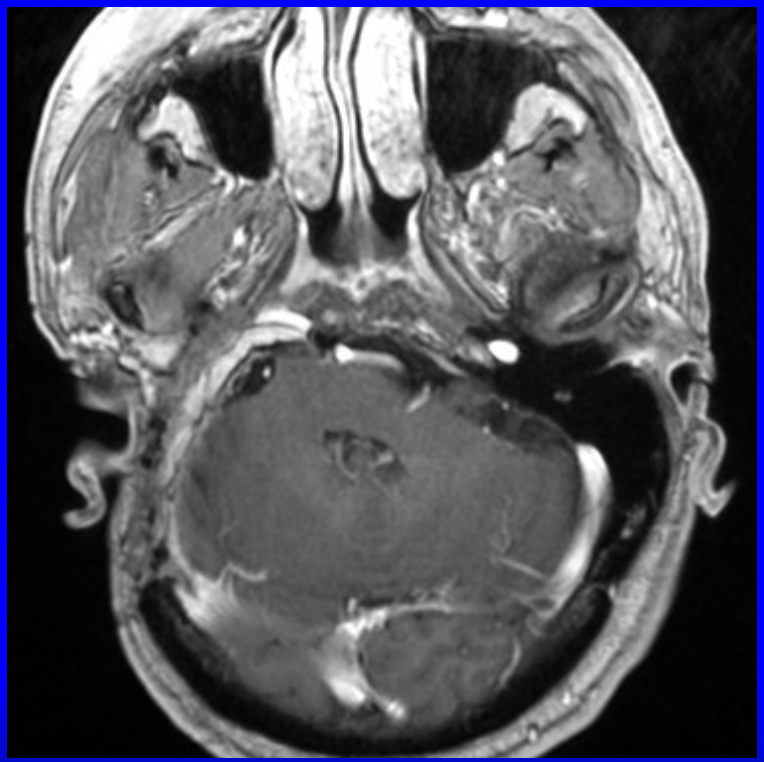
Total tumor removal

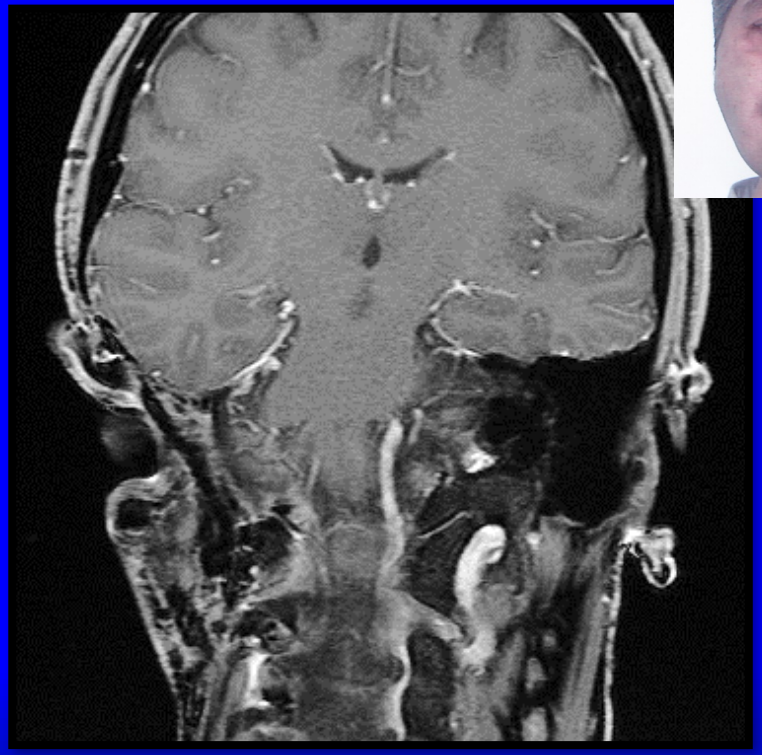




CASE 2: CLASS C3Di2Ve TJP

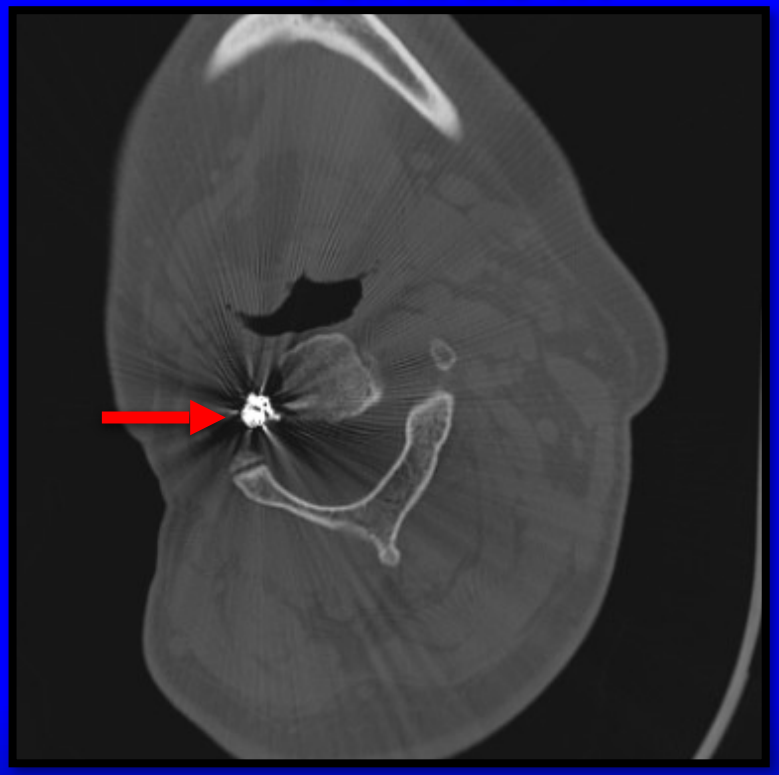
MRI after 10 years shows total tumor clearance



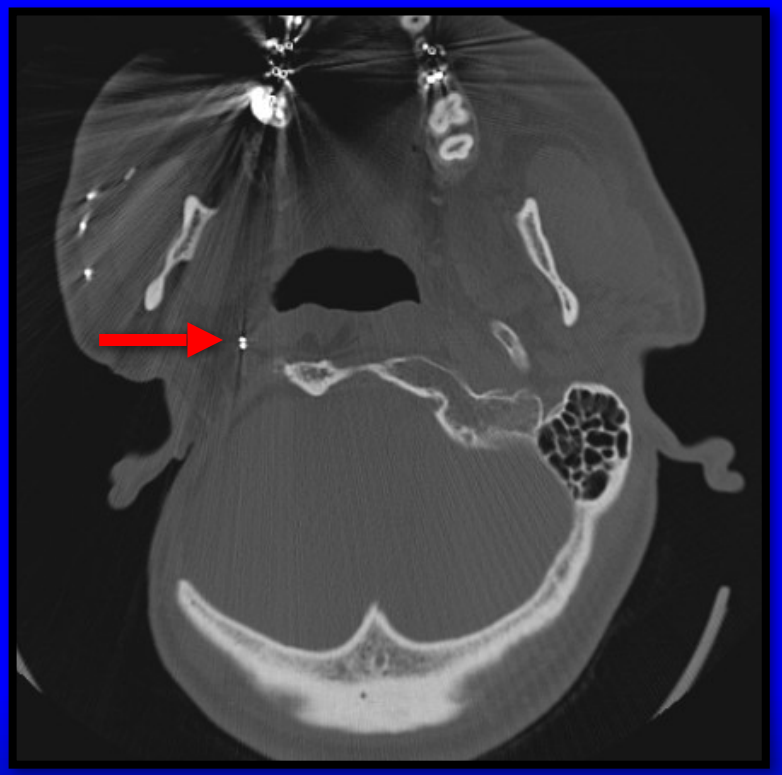


CASE 2: CLASS C3Di2Ve TJP

Coil in the VA



Coil in the ICA



OUR EXPERIENCE

Otology & Neurotology
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Management of Jugular Paragangliomas: The Gruppo Otologico Experience

Mario Sanna, Yogesh Jain, Giuseppe De Donato, Rohit, Lorenzo Lauda, and Abdelkader Taibah

Gruppo Otologico, Piacenza-Rome, Italy

ORIGINAL ARTICLE

Strategies and long-term outcomes in the surgical management of tympanojugular paragangliomas

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HEAD AND NECK PARAGANGLIOMAS

Gruppo Otologico Experience

HEAD AND NECK PARAGANGLIOMAS (1983-2025)	
Class A/B	230
CBT + VP	50
Class C / D	250
Class C / D (wait and scan)	105
TOTAL	635

COMPLICATIONS IN CLASS C/D TJPS

Complication	Number of patients	%
Mortality	0	0
Stroke	1	0.3
ICA injuries	3 (1 short stent)	1.2
CSF leak	9 (3 surgery)	3.6
Tracheostomy	1 (temporary)	0.3
PEG	4 (temporary)	1.6
Vocal cord medialization	9	3.6

THE GRUPPO OTOLOGICO EXPERIENCE

STENTING OF THE ICA

- 52 cases
- No complications
- No blow out
- No events on long term follow-up

CONCLUSIONS

GRUPPO OTOLOGICO

TAKE HOME MESSAGE

- TUMOR CLASSIFICATION
  &
- PRE-OP ASSESMENT

….are as important as the surgery itself!

1

TAKE HOME MESSAGE

- ITFA and its VARIANTS are the workhorse of tbps
- FACIAL NERVE RE-ROUTING is necessary in large tumors & the price to pay for total removal
- LCNs preservation is possible in the majority of Class C1-C2 cases

TAKE HOME MESSAGE

- With preoperative radiological ICA TREATMENT (STENT/ BALLOON) total removal is feasible and should be attempted in the majority of the cases

3

TAKE HOME MESSAGE

- HIGH SUCCESS RATE in skull base paragangliomas (even complex cases) by:
- Thorough knowledge of anatomy / radiology / surgery

+

- Logical decision making

4


WELCOME TO
GRUPPO OTOLOGICO

Second International Congress
on the Management of

HEAD & NECK PARAGANGLIOMAS

23-25 September 2026
Cattolica University Congress Center Piacenza, Italy

Live Surgery free of charge 21-22 September
Headquarters of the Gruppo Otologico

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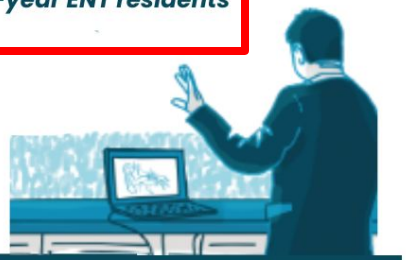
100 free registrations for final-year ENT residents



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SAVE THE DATE!!!

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