32 Tympano-Jugular Paragangliomas and Skull Base Surgery

Sanna Mario • Falcioni Maurizio • Flanagan Sean • Yusuke Takata • De Donato Giuseppe

TJP are rare lesions with an estimated incidence of 1 in 300,000. However they constitute the second commonest tumour of the temporal bone following vestibular schwannoma, and the commonest tumour affecting the jugular foramen.

TJPs are normally sporadic, but may be associated with one of the 4 familial paraganglioma syndromes related to mutations in the succinate dehydrogenase gene. A far higher rate of multicentricity is found with these syndromes.

While usually described as exhibiting slow growth, these tumors can show an unpredictable growth pattern, so that some of them require surgery while others may be maintained under radiological surveillance.

Treatment of TJP has always represented a difficult task for the skull base surgeon, at least until the standardization of the infratemporal fossa type A approach by Ugo Fisch in 1978[1]. Since then ongoing improvements in microsurgical and anesthesiological techniques, as well as the progressive development of preoperative interventional radiological techniques such as preoperative embolization[2] and internal carotid artery (ICA) preoperative treatment[2–4] has allowed the management of lesions previously considered inoperable.

However in spite of these terrific improvements, management of TJPs remains a demanding task.

Pathology

Paragangliomas are classified as a neuroendocrine neoplasms with neural differentiation. They arise from the paraganglion system, aggregations of cells found throughout the body associated with vascular and neuronal adventitia.

Structurally the paraganglia are 1–1.5mm in length and found along the course of Jacobson's nerve in the inferior tympanic canaliculus or the promontory, Arnold's nerve in the mastoid canaliculus and in the adventitia of the jugular bulb.

Histology of the paragangliomas reveals that tumour tissue replicates the normal structure of the paraganglia. Two cell types are present: chief cells, or type I cells, with granules containing catecholamines, and schwann like satellite cells, or type II cells.

A classic Zellballen (cell ball) configuration is noted, with abundant blood vessels.

Clinically significant rates of catecholamine secretion is found in only 1–2% of TJP.

Less than 4% of TJP are found to be malignant; importantly there are no histologic critieria for malignancy.

Genetics

Whilst the majority of TJP are sporadic it has been the investigation of familial types that has revealed genetic abnormalities in these and sporadic cases[5]. Genetic mutations have been identified in around 25% of the patients affected by head and neck paragangliomas, with the risk decreasing progressively with advancing age.

Familial paragangliomas are inherited in an autosomal pattern. The genetic defect involves mutations in SDH, a mitochondrial enzyme complex playing a role in the Krebs cycle and intracellular oxygen sensing and signaling[6].

Mutations of the SDH subunits are responsible for paragangliomas in paragangliomas syndromes (PGL).

Multicentricity has been reported to occur in sporadic cases, but at a higher rate in familiarly inherited tumours[7].

Whilst the reported rates vary considerably it is paramaount that the possibility of multicentricity is considered, due to the obvious fact that compound and or bilateral cranial nerve palsies inevitably leads to significant morbidity.

There is mounting evidence of some genetic changes increasing the risk of malignant change.

Clinical History

While pulsatile tinnitus usually affects the majority of the

patients, the other symptoms are a direct consequence of the specific growth pattern. In particular hearing loss depends on the invasion of the middle and inner ear, while lower cranial nerve deficits usually develop as a consequence of the progressive invasion of the jugular foramen. Nerve deficit induced by tumor growth is generally very slow, allowing a progressive compensation so that the patient is sometimes unaware of the deficit itself.

Clinical Examination

In the majority of the cases a red mass is visible on otoscopic examination. It can be a retrotympanic mass for lesions expanding into the middle ear cleft, while tumors invading the tympanic bone from below can show the classic aspect of the "rising sun". These tumours can also extend through the tympanic membrane and be confused with an inflammatory polyp. Obviously any vascular mass seen on otoscopy, the margins not seen in it's entirety, involves the jugular bulb until proven otherwise.

A full cranial nerve examination is essential including upper aero-digestive tract endoscopy and careful palpation of the neck.

Routine screening for urinary catecholamines is recommended despite the low rates of secreting tumours, and genetic testing if advisable.

Pathophysiology

The precise site of origin of TJP is often difficult to establish, and is academic unless it is confined to the middle ear cleft without erosion of the jugular plate.

With progression, TJPs most frequently follow a path of least resistance, through air cell tracts to involve the intrapetrous carotid canal, into the neck along the carotid sheath and or within the jugular vein and, in later stages, intracranially. This is however accompanied by an often underestimated degree of bony erosion. The tympanic bone is widely involved early, with extension in all directions.

Lower cranial nerve involvement occurs later and is usually related to invasion through the medial wall of the jugular bulb. The facial nerve lies in close proximity to the jugular bulb in its vertical segment and is also at risk. The otic capsule is relatively spared but extension to the inner ear can occur, as well as invasion of the occipital condyle, the XII cranial nerve and vertebral artery. Further anteromedial spread can involve the cavernous sinus, with significant intra-dural involvement usually found in the advanced stages of disease.

Tumours arising in younger patients tend to behave in a more aggressive fashion, with more advanced disease at presentation and have higher recurrence rates following treatment.

Diagnostic Work-Up

The diagnosis and assessment of TJPs, like most skull base pathology is based on radiologic information, not on histopatholgy from biopsy specimens. To differentiate between different pathologies directly affects the management options, surgical approach and prognosis, even for lesions affecting the same anatomical compartment. This situation is particularly hightlighted in lesions of the jugular foramen.

CT and MRI are mandatory and complementary in the evaluation of any jugular foramen lesion. In fact while MRI with gadolinium allows a better understanding of the tumor extension as well as neck and intracranial involvement, CT permits better assessment of bony erosion.

Low to intermediate T1 signal and relatively high T2 signal are typical for paragangliomas. A classic "salt and pepper" pattern in TJPs greater than 2cm is seen on T1 after gadolinium infusion, due to the presence of intratumoral vasculature appearing as flow voids. Dural invasion is not always easy to detect, because often the dura is infiltrated and pushed medially, without true invasion of the posterior fossa. Sagittal sections allow to appreciate the intra- and extracranial extension of the lesion **(Fig. 32.1)**.

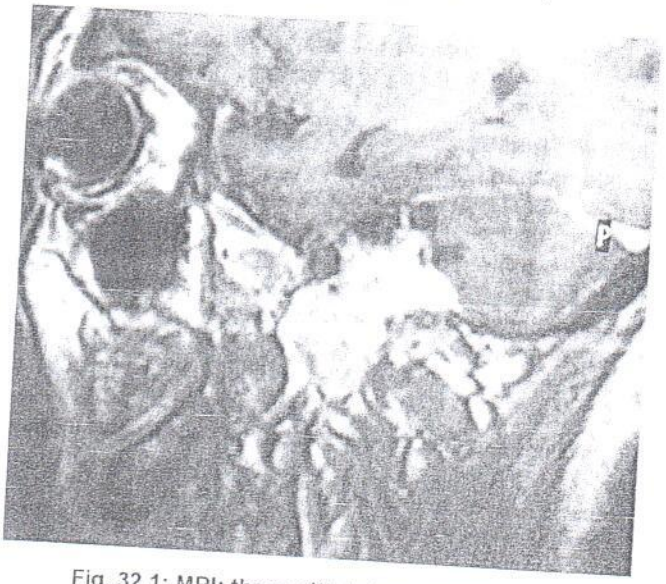


Fig. 32.1: MRI; the sagittal view allows a clear visualization of the intra- extracranial extent of the lesion.

On CT TJPs show characteristic irregular bony erosion (moth eaten bone) **(Fig. 32.2)**. Extension through the jugular plate from either direction occurs early, with involvement of the hypotympanum and jugular bulb. Early changes are represented as an indistinct lateral margin of the jugular foramen, followed by erosion of the caroticojugular crest and jugular spine. The extent of involvement of the infralabyrinthine cell tract and carotid canal is used to classify the lesion according to the Fisch classification[8 libro Fisch]. As mentioned, the degree of bony involvement is often difficult to assess. This is especially

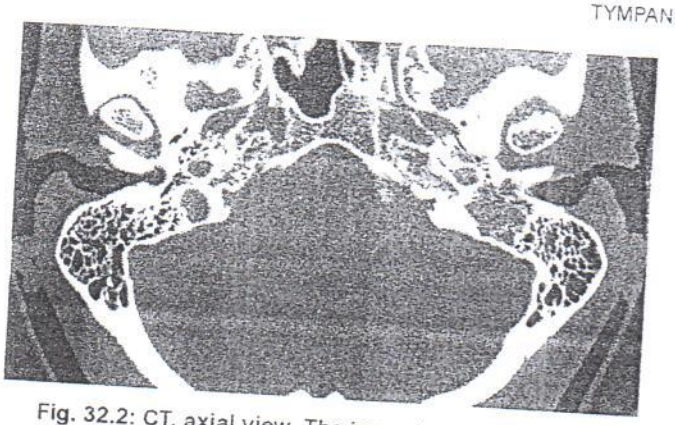


Fig. 32.2: CT, axial view. The irregular erosion of the jugular foramen represents a peculiarity of paragangliomas.

problematic in upstaging early lesions, necessitating the conversion of a transmastoid to an infratemporal type A approach, as well as in extensive involvement of the petrous apex, clivus anteriorly, and occipital condyle and hyoglossal canal posteriorly.

Two classification systems are in common use, that of Fisch and that of Glasscock-Jackson[9]. In terms of describing the involvement of the internal carotid artery (ICA), the most critical aspect in planning the surgical approach, it is Fisch's system that we recommend be used. There is also a close correlation between the C class and the likelihood of intra-cranial extension.

In the majority of cases CT and MRI allow a clear differentiation from other lesions involving the jugular foramen area, mainly jugular foramen schwannomas and meningiomas[10,11] biblio Dedo e Bacciu. Schwannomas characteristically reveal smooth enlargement of the jugular foramen, with early sparing of the lateral wall. While menigiomas can produce an irregular margin of the jugular foramem, the degree of bony erosion is significantly less and are often associated with hyperostosis. Meningiomas may also show intratumural calcification and a dural tail while intratumoral cysts are more frequent in schwannomas.

It is also essential to assess the venous and arterial status bilaterally. In contrast to other lesions of the jugular foramen, TJPs often infiltrate and invade the jugular bulb, so large collaterals often have developed, and the status of the contralateral venous is drainage paramount. The current use of magnetic resonance angiography (MRA) and venous phase CT normally provides adequate information.

Four vessel digital angiography with embolisation is integral in the management of TJP. It can also aid in the diagnosis in difficult cases. Rare lesions such as chondrosarcomas and endolymphatic sac tumors with inferior extension should also be considered. TJPs show a coarse blush, greater than that of a meningioma, and rapid diffusion on venous phase due to rapidly draining veins. The primary blood supply is from the ascending

Fig. 32.3: on digital angiography tympano-jugular paragangliomas show a characteristic "blush".

pharyngeal artery, gaining additional supply from both external and internal carotid systems with tumour growth **(Fig. 32.3)**.

Screening for co-existent lesions is also very important. MRI scanning of the entire neck is required, with investigation of the abdomen if there are any signs of catecholamine excess or family history.

Treatment Options and Decision Making

Often patient factors are just as important as the extent of pathology when formulating a management plan. The following parameters should be carefully taken into consideration:

- Patient's age, general medical condition, life expectancy
- Lower cranial nerve function;
- Facial nerve function;
- Patency of the contralateral sigmoid sinus and jugular bulb;
- Tumor size, with particular regard to intracranial extension and carotid artery involvement.
- Presence of other ipsilateral or contralateral lesions

The available options for treatment of TJP are: radiological surveillance, conventional radiotherapy, stereotactic radiotherapy and microsurgery. If required different options may be used together.

Radiological surveillance is based on the general belief that TJPs are benign, slow-growing lesions. Unfortunately the course of at least a subgroup of large tumors, usually affecting younger patients, seems completely different[12,13] and presently this policy may be recommended in selected cases, mainly in elderly patients with intact lower cranial nerve function.

Conventional radiotherapy has no direct effect on the tumoral cells, producing only fibrosis of the vessels supplying the tumor[14–16]. Recurrences are also documented[13]; potentially life-treatening complications such as osteoradionecrosis and induction of malignancy have also a consideration[17,18].

There are still no large series treated with stereotactic radiotherapy to confirm its effectiveness in the long-term. It is important to stress that majority of the recurrences become apparent at least 5 years after surgical removal. Stereotactic radiotherapy is mainly suggested for small TJPs confined to the jugular bulb[19] and is not indicated in large lesions with extensive bone infiltration, where its effects are questionable and the risks of osteoradionecrosis are comparable to those of conventional radiotherapy.

For these reasons microsurgery remains the preferred treatment modality in the majority of the cases, at least in patients with life expectancy longer than 20 years.

The conundrum that exists between the radicalness of resection and neural preservation is never more sharply defined as in the treatment of TJP. Ultimately this decision is one based upon judgement gained from experience and the experience of others.

Planned partial removal has[20] progressively gained consensus among the skull base community. However indications for a planned partial removal should be accurately evaluated; the temporary benefit of avoiding facial nerve (FN) weakness, hearing loss or lower cranial nerves palsies should be balanced against the possibility that, because of the infiltrating nature of TJPs, tumoral remnants may spread along the dural surfaces into cancellous bone, reaching areas were they become unresectable. In fact large lesions affecting young patients usually show a particular aggressive pattern, with high recurrence rate.

Another option proposed has been to allow the pathology to slowly paralyse the lower cranial nerves, especially in cases where the likelihood of neural preservation is low. Compensation tends to occur pre-operatively and radical resection can precede without the concern of neural conservation. As in much of skull base surgery personal experience, judgement and philosophy colour ones management decisions.

Special Situations

In the case of advanced age or poor medical status there is rarely an indication for radical surgical removal. Radiological follow-up often represents the best treatment in these cases. Occasionally resection of the middle ear and mastoid components with a blind sac closure provides excellent control in elderly patients with troublesome otorrhagia.

Compensation following acute compound lower cranial nerve palsies is particularly difficult in elderly patient and normal lower cranial nerve function should be considered a contraindication for surgery in patients older than 60 years.

The likelihood of preserving the function of the LCNs is mainly related to tumor invasion of the medial wall of the jugular bulb. Preoperative judgment on the integrity of the medial wall of the bulb is not always feasible, with an indirect sign represented by the presence of intradural tumoral invasion. In fact transdural growth usually occurs initially at the level of the jugular foramen.

FN function is another point to be considered during pre-treatment evaluation. The presence of a facial deficit essentially dictates that the nerve will need to be resected and grafted, with the results directly related to the duration of pre-operative facial dysfunction.[Falcio,21]. The converse however is not always true, with a number of cases with normal pre-operative facial function revealing extensive neural invasion.

Surgical treatment for TJPs inevitably means sacrifice of the ipsilateral jugular bulb. This is usually occluded by the presence of the tumor and some compensation has already taken place, often through the contralateral bulb. However in rare occasions, the ipsilateral bulb may be still patent or the compensation has occurred through collaterals, such as the posterior condylar vein. In the presence of an hypoplastic contralateral venous system, sacrifice of the ipsilateral jugular bulb is risky, because it may lead to intracranial hypertension or more rarely venous infarction of the temporal lobe, occasionally limiting treatment options.

Subtotal removal is indicated for tumors reaching inaccessible areas; remnants in such areas showing growth are better managed through stereotactic radiotherapy.

The possibility of multiple and contralateral lesions should also be considered, especially in the young patient with a family history or who has tested positive to genetic screening. Treatment selection in these cases should be carefully tailored according with the specific situation of each patient; of course a more conservative treatment is

usually considered in the presence of bilateral risk to lower cranial nerves.

The most common synchronous lesion with a TJP is a carotid body paraganglioma. Strong consideration can be given to treatment of both lesions at the same surgery, especially if on the same side.

Preoperative Treatment

All patients scheduled for surgery must undergo an angiographic evaluation with embolization. This is a standardized procedure that has significantly reduced the intraoperative blood loss and increased the rate of total tumor removal. The procedure is to be planned not more than 72 hours before the surgery because after this time the feeding vessels start to be recanalized.

Involvement of the ICA is common in TJP. Indications for interventional radiological preoperative treatment of the ICA include:

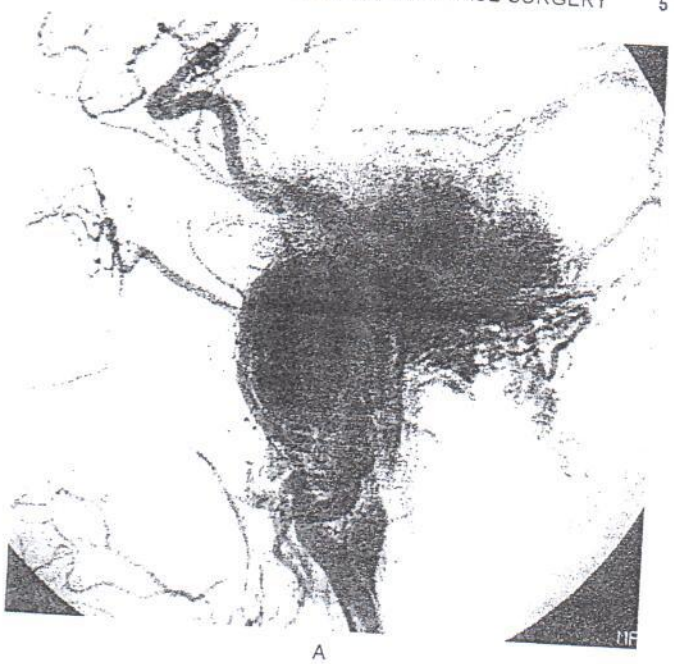
1. Engulfment by the tumor for more than 270 degree as shown by MRI;
2. Evidence of stenosis of the arterial lumen or irregularities of the arterial walls on the angiography;
3. cCass C3 and C4 TJPs;
4. Extensive blood supply from branches of the ICA;
5. Recurrent or post-radiation cases.

Engulfment of the artery for more than 270 degrees, even if limited to the vertical segment, requires at least mobilization of the artery from its bony canal, in order to completely remove the infiltrated canal and surrounding bone. However the anterior and medial surfaces of the artery are not easy to control, placing the artery at risk.

Stenosis of the arterial lumen or irregularities of the arterial walls on the arteriography are strongly suggestive of arterial wall infiltration **(Fig. 32.4A, B)**. Subadventitial dissection is mandatory in order to achieve gross total removal. However this surgical manoeuvre results in a high risk of arterial tear in the absence of adequate preoperative measures.

Class C3 and C4 tumors, according to the Fisch classification, extend far from the carotid genu, toward the petrous apex. Drilling of the carotid canal and dissection of the arterial walls in these areas is particularly demanding with limited ability to control the distal ICA.

Tumors with massive involvement of the ICA often receive a significant blood supply from small vessels originating from the ICA itself, but usually angulation and small size of these feeding vessels precludes selective microcatheterization and embolization. However maintenance of this blood supply produces an increase in the intraoperative bleeding, making much more difficult the already dangerous surgical steps in close proximity to the ICA. In such a situations preoperative management of the ICA facilitates radical tumor removal, increases devascularisation of the pericarotid tumor and reduces the risk of ICA laceration.



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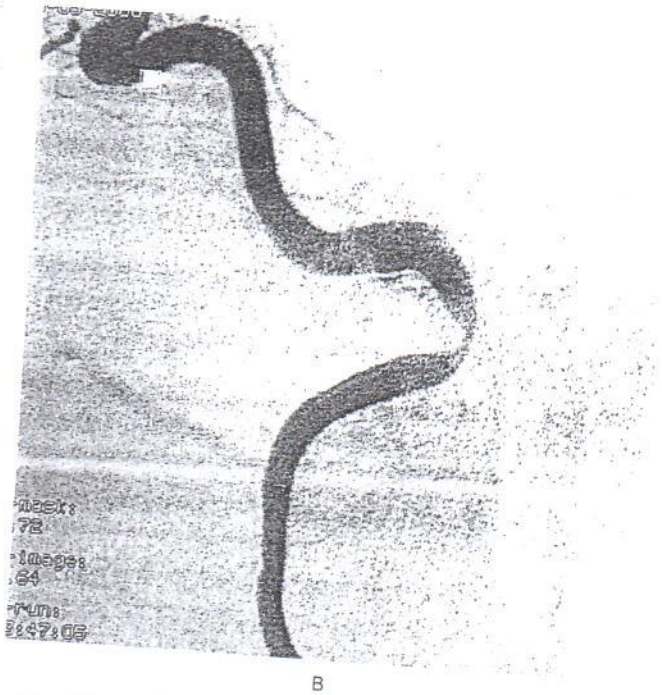


B

Fig. 32.4: huge lesions (in this case a combination of a tympano-jugular and a vagal paraganglioma) often produce significant involvement of the carotid artery. This may be suspected on MRI scans (A) and confirmed on angiography (B) (in this case an arterial stenosis is present at the level of the junction between the 2 lesions).

Options for preoperative radiological ICA treatment include:

- Preoperative balloon occlusion (PBO)

- PBO with carotid by-pass
- Reinforcement with stent (RWS)

If preoperative angiography shows adequate collateral circulation through the circle of Willis, a balloon test occlusion of the ICA with clinical and angiographic tolerance, followed by permanent balloon occlusion[22] **(Figs. 32.5 & 32.6)**, is considered the simplest and most effective procedure. However, this procedure is not risk free, and, although rare, complications can be profound[3]. In addition, it cannot be used in cases of inadequate collateral circulation.

The bypass of the ICA[23] has been considered the only option in cases of inadequate collateral circulation, but, regardless of the technique used, this is a major procedure and carries a relatively high risk of stenosis, thrombo-embolism, and occlusion[24,25] The recent introduction of preoperative stenting **(Figs. 32.7 & 32.8A, B)** has made more aggressive carotid dissection feasible, and has greatly reduced the need for permanent balloon occlusion. In addition, when the tumor infiltrates the wall of the artery, it may derive abundant blood supply directly from the ICA; in such situations positioning of the stent also contributes to preoperative devascularization of the lesion. Stenting is performed 4 to 6 weeks before the planned date of the

Fig. 32.5: postinterventional X-ray allows for confirmation of the correct position of the 3 balloons into the carotid artery.

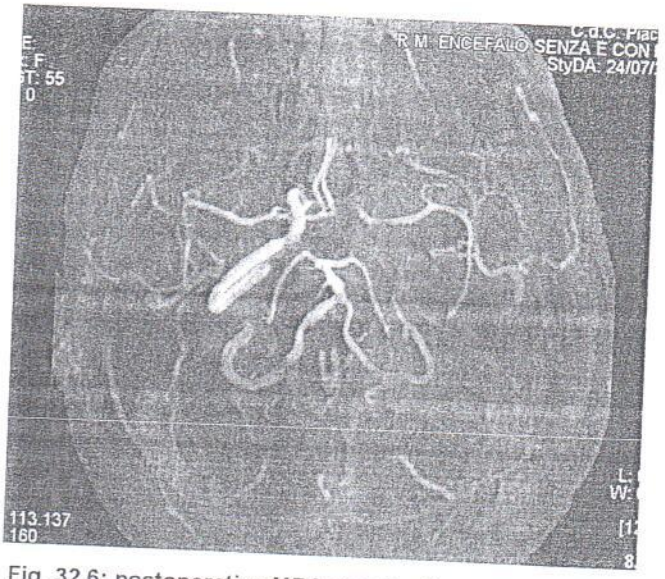


Fig. 32.6: postoperative MRA showing the absence of the internal carotid artery removed with the tumor after preoperative occlusion.

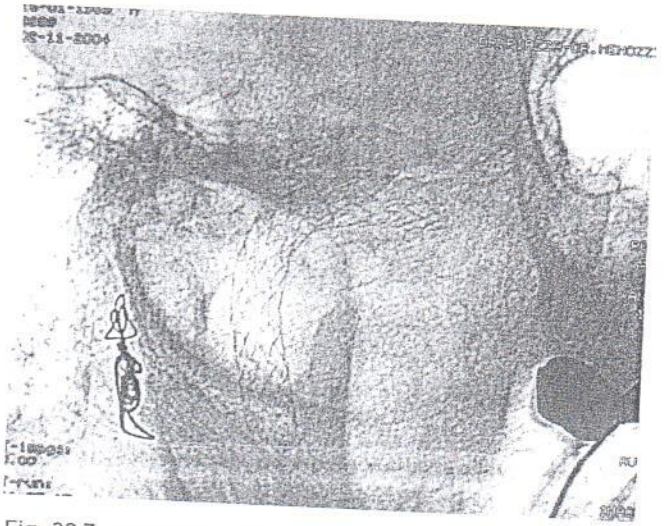


Fig. 32.7: postinterventional X-ray allows for confirmation of the correct position of the stent into the carotid artery.

operation to allow a stabilized neointimal lining to form on the luminal surface of the stent. The risk of stent thrombosis is still present and pre- and poststenting antiplatelet therapy should be continued for life.

Surgical Management

The IFT type A is the cornerstone of TJP surgery[1]. The morbidity associated with a IFTA approach includes:

- Maximal conductive hearing loss
- FN dysfunction: 70% of patients undergoing permanent anterior re-routing regain HB I or II within 1 year post-operatively.

While an ideal goal of any approach is to minimise associated morbidity, only selected TJP are amenable to more restricted exposure. Essentially this is limited to few

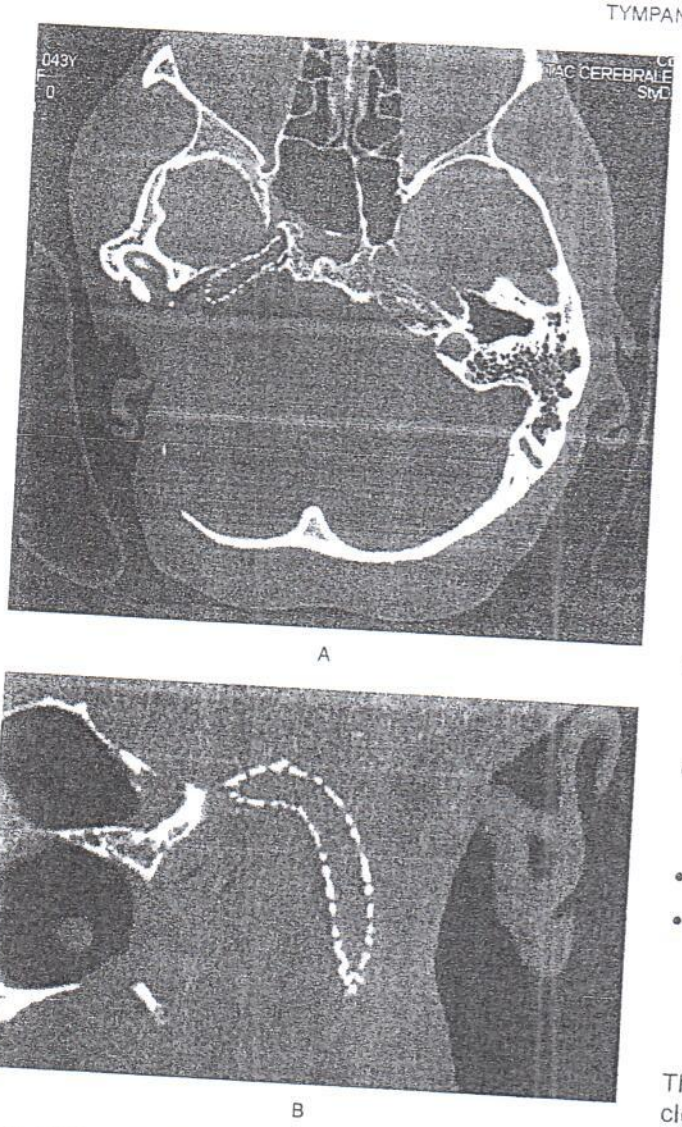


Fig. 32.8: postoperative CT showing the stent inside the internal carotid artery; (A) the horizontal portion on axial view; (B) a longer segment of intratemporal stent on oblique reconstruction.

C1 tumours with predominantly posterior extension that can be approached without anterior re-routing of the FN and sacrifice of the external auditory canal.

While the IFTA is associated with a degree of morbidity it provides wide un-interupted access to jugular foramen and intra-petrous carotid artery.

Based on the IFTA approach various extensions are dictated by the location of the lesion. The standard extension we use is transjugular paracondylar, which allows additional postero-inferior access to the jugular fossa, widening the exposure and facilitates venous and neural control. The widened angle also affords better access to the petrous apex, medial to the carotid artery. Occasionally this can be further widened by a trans-condylar and trans-tubercular extension. Very rarely a far lateral is employed with exposure of the vertebral artery.

The use of a translabyrinthine extension is occasionally required for otic capsule involvement. Modified trans-cochlear or combination with the IFT type B approaches are rarely required to access petrous apex and clival involvement.

Intradural Tumour

The combination of an infratemporal approach type A, due to the extensive neck dissection, and a contemporary large dural opening exposes the patient to a significant risk of CSF leak through the neck incision. This is due to the difficulty of reinforcing the area of primary dural closure, secondary to extensive removal of the skull base. A staged resection of the intradural component minimises this risk. There are 4 important points to be made regarding the secondary approach.

- The approach is performed through the same route without any neck extension
- The FN should be permanently anteriorly mobilised during the 1st stage in order to be far from the surgical field during the 2nd stage.
- The second stage should be conducted within 3–4 months due to contraction of the access and ***pseudo-encephalocele*** formation.
- New embolization is indicated.
- Preservation of at least some bony anatomy during the first stage facilitates orientation.

Surgical Highlights IFTA with Para-Condylar Extension

The operation starts with a sub-total petrosectomy and closure of the external auditory canal. The tympanic membrane and ossicles are removed with the exception of the stapes footplate. The sigmoid sinus is skeletonized and prepared for closure.

The neurovascular bundle is identified in the neck **(Fig. 32.9)**. Frozen section for parajugular nodes are taken to identify the rare malignant cases. If positive a modified neck dissection is accomplished. The sternocleidomastoid muscle from the mastoid tip are detached and the muscle is posteriorly reflected. The posterior belly of the digastric muscle is sectioned and anteriorly reflected and the styloid process is divided in order to expose the skull base. The FN is then identified at the exit from the stylomastoid foramen and followed to it's bifurcation. A subtotal petrosectomy permits exposure of the entire course of the facial nerve from the geniculate ganglion to the parotid, as well as minimising the risk of recurrence in this area.

The nerve is then decompressed **(Fig. 32.10A)**, displaced anteriorly **(Fig. 32.10B)** and allocated a new groove drilled into the antero-superior wall of the external auditory canal and secured with fibrin glue. Another groove for the terminal portion is created in the parotid gland. Displacement of the FN represents the key-point of the surgery

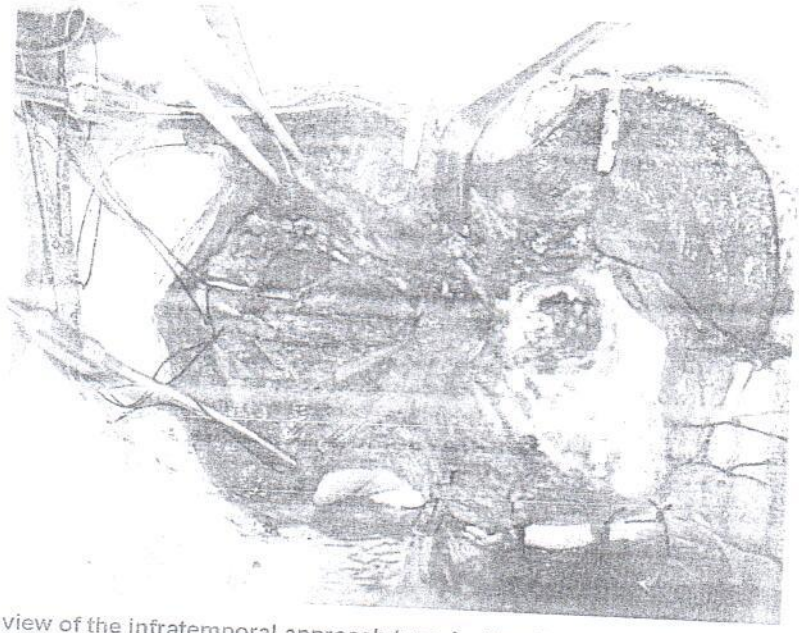
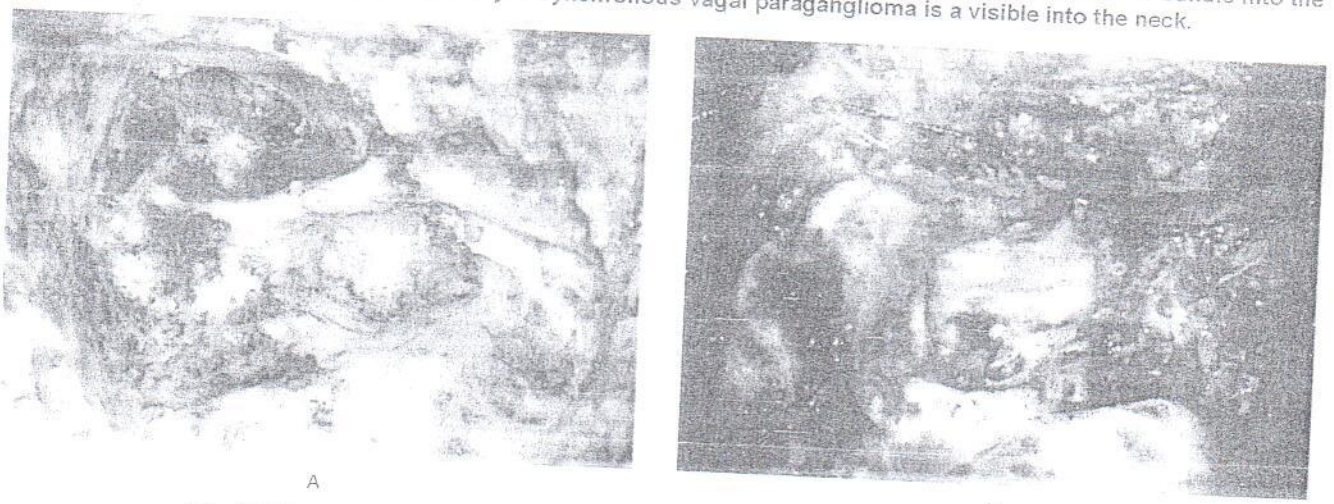


Fig. 32.9: intraoperative view of the infratemporal approach type A after dissection of the neurovascular bundle into the neck and subtotal petrosectomy. A synchronous vagal paraganglioma is a visible into the neck.



A B

Fig. 32.10: after carefull skeletonization (A) the facial nerve is anteriorly rerouted (B).

because it allows unobstructed control of the jugular foramen and the carotid canal. Tips to be adopted to correctly perform the anterior rerouting include complete bony decompression of the geniculate ganglion area sharp dissection of nerve from the 3rd portion of the fallopian canal.

The anatomical integrity of the facial nerve can be spared in the majority of the cases. In the presence of epineurial involvement the nerve should first be re-routed. The involved epineurium is then dissected to reduce the risk of nerve interruption during the rerouting of a fragile nerve deprived of its epineurium. In other cases the invaded segment of the nerve needs to be resected and grafted. Tension free interposition grafting using the greater auricular nerve is the commonest technique.

The head of the mandible is then anteriorly mobilised.

The area of the jugular foramen is now widely exposed. Further bone is removed postero-inferiorly which represents the bone of the jugular process of the occipital bone, the postero-lateral third of the occipital condyle, and the jugular tubercle, can also be removed to extend exposure. The hypoglossal canal lies a few millimetres deep to the removed bone.

The sigmoid sinus is occluded at the level of the transition into the lateral sinus, by means of extra-luminal packing, and the jugular vein is ligated into the neck. The vein is then transected and the dissection is developed in an inferior to superior direction.

The jugular bulb is opened and tumour dissection performed in a piecemeal fashion using bipolar forceps.

The critical point is represented by the invasion of the

medial wall of the jugular bulb; in the presence of such infiltration radical surgery requires removal of the entire bulb, with an extremely high possibility of damage to the lower cranial nerves. On the contrary if the medial wall of the bulb can be preserved so too can neural function. Care must be taken when packing branches of the inferior petrosal sinus. These can be multiple and usually enter between the IX and X,XI nerves.

The final stage of surgery involves management of carotid artery involvement.

The artery may require the following types of treatment, depending on degree of involvement

- Simple decompression in presence of bony infiltration of the carotid canal;
- Decompression with partial mobilization of the artery in presence of bony infiltration anterior to the vertical portion;
- Subadventitial dissection when a infiltration is predicted;
- Artery removal (after a preoperative occlusion).

Naturally in presence of significant involvement, surgery is preceded by radiological treatment as discussed. Preoperative stent insertion allows the skull base surgeon to perform an aggressive ICA dissection with a significant reduction of the surgical risks **(Fig. 32.11)**. With presence of an intraluminal stent the surgeon is usually able to establish a cleavage plane on the lateral surface of the stent, allowing total removal of the involved portion of the carotid wall, minimising the risk of inadvertent rupture. This is particularly true when working at the level of the carotid genu and/or the horizontal segment of the artery; in this area in fact the surgical room and the mobility of the artery are reduced, and the direct control of the antero-medial wall is particularly demanding. The surgeon is now able to achieve dissections previously considered too risky. It is important to start this dissection proximally where it is easier to find the correct cleavage plane.

Special Situations: Intradural Extension

Intradural extension usually starts at the level of the jugular foramen. As already mentioned our policy is to remove the intradural lesion in a single stage only in presence of small extension (< 2 cm in diameter). In the case of a larger intradural tumoral component its removal is postponed to a 2nd stage to be performed after 3–4 months **(Fig. 32.12A, B)**. Usually the second stage is performed through the same route without any extension to the neck **(Fig. 32.13A, B)**.

SURGICAL CONTROVERSIES

A less aggressive treatment as been advocated by some Authors, especially regarding the possibility of approaching remove C1 and C2 TJPs without the need to mobilise the FN and remove the external and middle ear, using the fallopian bridge technique[26]. The use of this conservative approach to manage a highly vascular tumour limits control of the area of origin of the lesion and of the ICA. As a consequence this approach will likely produce a higher rate of recurrence.

While the goal should be to preserve neural function when possible, skull base surgeons must also be aware of the extreme aggressiveness of these tumours. An initial conservative surgery may lead to extension of the lesion to areas where it becomes very difficult to be managed. Conductive hearing loss and temporary FN palsy with a high percentage of recovery to grade I or II in around 70% of cases (65.8% in our series, ranging from 51% and 95%

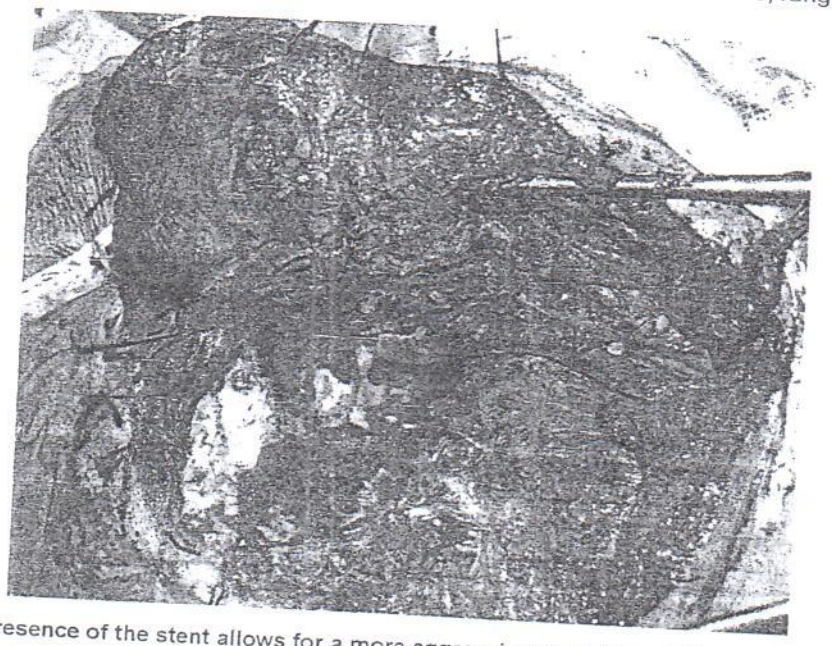
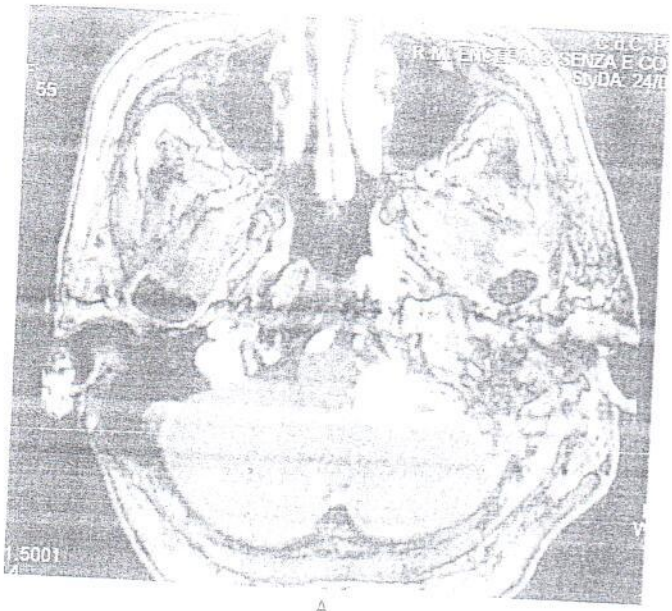
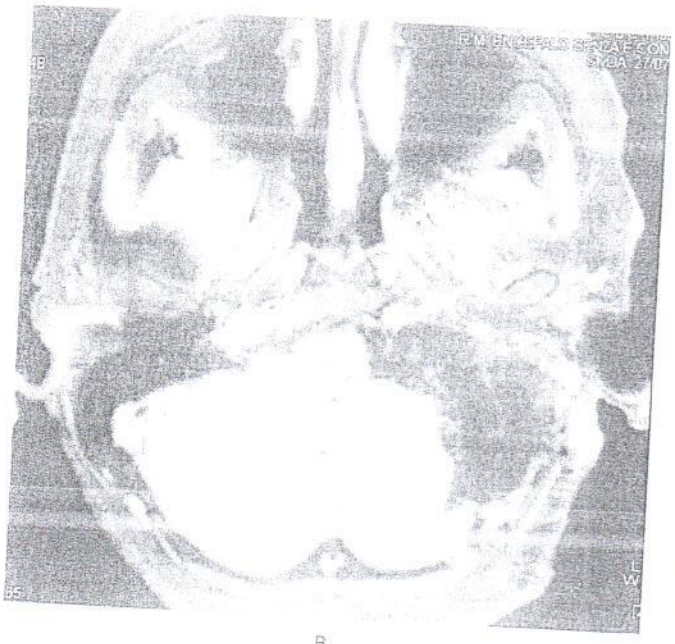


Fig. 32.11: the presence of the stent allows for a more aggressive dissection of the internal carotid artery.



A



B

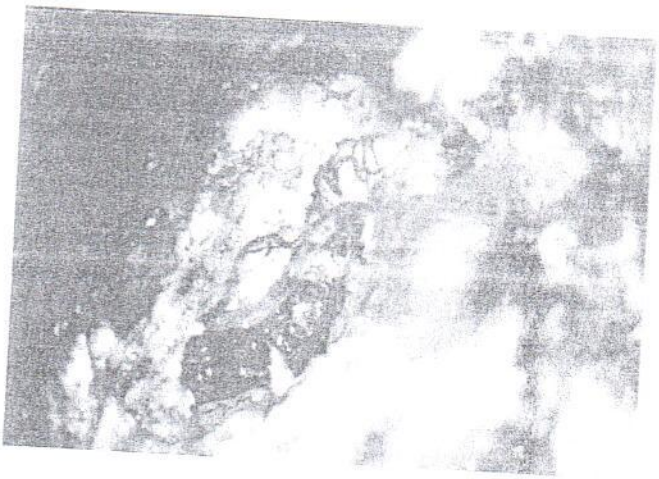
Fig. 32.12: (A) postoperative MRI after the 1st operation of a staged procedure; the intradural component has been left in place; (B) postoperative MRI of same case after the 2nd stage, showing the total removal of the intradural remnant.

in literature)[27-30] indicate that the increased safety of ICA exposure and lower rates of recurrence do not come at too high a price. Recurrence in the presence of a facial nerve that was not re-routed during the first surgery portends an extremely high risk of complete facial palsy.

POSTOPERATIVE CARE

The need for prolonged postoperative care and rehabilitation after TJP surgery is mainly dependent on the functionality of the lower cranial nerves. Acute loss of the IX and X cranial nerves is not well tolerated by patients,

A



B

Fig. 32.13: intraoperative view of a 2nd stage; (A) the dura has been opened and the dissection started from the anterior border of the lesion; (B) final view after total tumor removal.

especially in association with a XII cranial nerve palsy. Even if there is a preoperative deficit, complete section of the nerves can produce a decline in function, due to the sacrifice of a few fibers still maintaining some muscular tone.

The majority of the patients are able to start oral alimentation after a few days; however aspiration with secondary pneumonia represents a significant risk for these patients. Intensive speech and swallowing rehabilitation is strongly indicated in all patients, with thyroplasty and pharyngoplasty employed when indicated. The risk of tracheostomy or prolonged percutaneous gastrostomy is now extremely low.

Management of temporary facial paresis is also necessary, with corneal protection a priority.

With the use of a blind sac closure and staging of the removal of intracranial pathology the rates of CSF leak are very low. If present, lumbar drainage followed by surgical exploration is occasionally required.

Follow-up

TJPs are infiltrative lesions with a high risks of recurrence, even in case of aggressive surgery. It is important to reinforce that even those units with extensive experience with these lesions report recurrence rates in the long term of 5–10%[31].

A prolonged postoperative follow-up is then necessary, taking into consideration that the majority of the recurrences are detected at a mean of 7 years from the surgery[27]This is also due to the fact that following a large surgical removal as in the IFT type A and the consequent large amount of postoperative scar tissue, postoperative scans are usually difficult to be evaluated, and recurrences are usually obvious only when already large. An MRI study is indicated at 1, 3, 5, 7 and 10 years from the surgery, with angiography reserved for doubtful cases.

The theoretical risk of implantation cholesteatoma must also be considered when evaluating follow up scans, and the possibility of developing further paragangliomas.

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