Chapter 23: Glomus and other tumours of the ear

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Glomus jugulare

The glomus jugulare is a collection of ganglionic tissue within the temporal bone in close relation with the jugular bulb. The first description of this tissue were probably by Valentin (1840), who described his 'ganglia tympanica', and Krause (1878), his 'glandula tympanica'. Both writers described ganglionic-like tissue, but the credit for recognizing the histological relationship to the carotid body goes to Guild in 1941. He originally called the structure the glomus jugularis, but in a later report (Guild, 1953) he accepted the terminology glomus jugulare accorded by Lattes and Waltner (1949). Lattes also suggested that the generic term for these structures in the body should be non-chromaffin paraganglia (any associated tumour being called a non-chromaffin paraganglioma).

The paraganglia are cells derived from the neural crest and are found widely distributed in the autonomic nervous system, the usual sites being the carotid, ciliary and vagal bodies, along the aorta and its main branches, in the glomus jugulare complex, in the bladder, in the para-adrenal area, and most notably the adrenal medulla. The paraganglia of the adrenal medulla secrete adrenaline and noradrenaline and histologically they stain chromaffin positive, hence the term 'chromaffin paraganglia'. Paraganglia having a negative chromaffin reaction are termed 'non-chromaffin'; they do not normally secrete hormones. The nerve supply of the latter is mainly sensory and, although the carotid body has been shown physiologically to be a chemoreceptor, responding to changes in blood pH and oxygen tension, the glomus jugulare has never been shown to be a physiologically active chemoreceptor.

Guild's (1953) anatomical studies were based on 88 temporal bones, in which he found an average of three glomus bodies in each bone. They were usually found in close relationship with either the tympanic branch of the glossopharyngeal nerve or the auricular branch of the vagus nerve; both nerves had an equal distribution of glomus bodies. The bodies were supplied with non-medullated sensory fibres from the adjacent nerve and, in most cases, the blood supply was from the ascending pharyngeal artery. Apart from their close relationship with the two nerves, their anatomical position was very variable, but 50% could be found in the adventitia of the jugular bulb and 25% in the mucosa of the promontory. Histologically, they were similar to the carotid body with epithelioid cells interspersed in a highly vascular stroma of capillary and precapillary vessels. The proportion of cells to vessels was variable and Guild recognized two groups, the cellular glomus bodies and the vascular glomus bodies, with a slight preponderance of the former. Their size was variable, but they tended to be ovoid in shape with a long diameter of 0.5 mm, equally distributed between the two ears in both sexes and found more commonly in the middle age group.

Glomus jugulare tumours

There are several reports of vascular tumours in the ear over the latter part of the eighteenth and the early part of the nineteenth century (Simpson and Dallachy, 1958). In
particular, Lubbers (1937) reported a case of metastatic carotid body tumour in the ear, with a contralateral carotid body tumour.

Rosenwasser (1945) was the first surgeon to recognize the relationship between these tumours and the normal glomus jugulare. In 1942, he removed a vascular tumour from the middle ear and mastoid and, on histological examination, found it to be very similar to the carotid body, but he could find no other primary tumours in the neck, and called it a carotid-body-like tumour. In his 1945 paper, Rosenwasser proposed that the tumour arose from the glomus jugularis described by Guild. Since that time there have been a variety of names attached to the tumour in an attempt to indicate its pathological origin.

Winship, Klopp and Jenkins (1948) first used the term 'glomus jugulare', and Latters and Waltner (1949) proposed that the tumours were called non-chromaffin paragangliomata. Mulligan (1950) introduced the general term 'chemodectoma' for the carotid body and glomus jugulare tumours, based on their common histological appearances and probable origin from chemoreceptor tissue. Boyd, Level and Griffith (1959) objected to this term as the glomus jugulare has no demonstrable chemoreceptor function.

Current usage suggests that these tumours should be considered as non-chromaffin paragangliomata. The most common term used is 'glomus tumours', and the terms 'glomus tympanicus' and 'glomus jugulare tumours' are used primarily for the clinical description of a particular tumour.

**Pathology**

Histological examination of the glomus jugulare tumour shows a similar appearance to the normal glomus jugulare; cytologically they are not very active with only rare mitotic figures, and they usually have a well-defined thin fibrous capsule. Clinically, however, they can be locally invasive and destructive of bone and facial nerve. In the author's series, they showed a great propensity to infiltrate through the mastoid air-cell system.

**Sex and age incidence**

The glomus tumour, in contradistinction to the glomus body, shows a predominance in females, but both tend to be more common in the middle age group.

**Endocrine activity**

They are usually considered to be non-chromaffin paragangliomata with no endocrine function, but there has been an increasing number of reports of vasoactive tumours (Duke et al., 1964; Matishak, Symon and Cheesman, 1987), and clinically it is important to look for evidence of endocrine activity by urinary assay of the metabolites dopamine and 3-methoxy-4-hydroxymandelic acid (vanillylmandelic acid).
**Multicentricity**

Glomus tumours are sometimes multicentric presenting in both ears (Winship and Louzan, 1951), or in conjunction with other parangangliomata, the carotid body commonly being the second site (Spector et al, 1975).

**Metastases**

The glomus jugulare is generally considered to be of low malignancy, mainly causing problems because of its site in the complex anatomy of the skull base. However, there are well documented cases of malignant glomus jugulare tumours, with both nodal and distant metastases; fortunately the incidence is very rare (Brown, 1967).

**Natural history and presentation**

The slow growth of these tumours means that the diagnosis is often missed until the tumour is very extensive. Alford and Guilford (1962) found the average delay to diagnosis was 6 years from the original symptoms, the extremes being 42 years and 2 weeks. The first symptoms noted generally follow middle ear involvement, and are often ignored. Pulsatile tinnitus and conductive deafness are, equally, the commonest presenting symptoms. A red mass (the rising sun behind the drum) on routine examination is not uncommon, but quite a high proportion do not present until cranial nerve palsy occurs. Some 30% of cases, in most series, present with facial palsy; the pareses, resulting from involvement of the nerves of the jugular foramen, often do not cause sufficient symptoms in most cases to warrant presentation. Table 23.1 shows the clinical findings in the author’s series of 21 cases and is similar to most other reports. Otalgia and aural bleeding are other fairly common symptoms.

**Table 23.1. Presenting clinical features in 21 patients with temporal region glomus tumours**

<table>
<thead>
<tr>
<th>Symptoms and signs</th>
<th>Cases</th>
<th>No of cases</th>
<th>Percentage</th>
</tr>
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<tbody>
<tr>
<td>Hearing loss</td>
<td>18</td>
<td>86</td>
<td></td>
</tr>
<tr>
<td>Middle ear mass</td>
<td>15</td>
<td>71</td>
<td></td>
</tr>
<tr>
<td>Tinnitus (pulsatile)</td>
<td>14 (5)</td>
<td>67 (24)</td>
<td></td>
</tr>
<tr>
<td>Cranial nerve deficits</td>
<td>11</td>
<td>52</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>1</td>
<td>5</td>
<td></td>
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<tr>
<td>VI</td>
<td>1</td>
<td>5</td>
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<tr>
<td>VII</td>
<td>8</td>
<td>38</td>
<td></td>
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<tr>
<td>IX</td>
<td>6</td>
<td>29</td>
<td></td>
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<tr>
<td>X</td>
<td>7</td>
<td>33</td>
<td></td>
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<tr>
<td>XI</td>
<td>5</td>
<td>24</td>
<td></td>
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<tr>
<td>XII</td>
<td>7</td>
<td>33</td>
<td></td>
</tr>
<tr>
<td>Unsteady, dizzy, vertigo</td>
<td>5</td>
<td>24</td>
<td></td>
</tr>
<tr>
<td>Otorrhoea</td>
<td>2</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Endocrine syndrome</td>
<td>1</td>
<td>5</td>
<td></td>
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</tbody>
</table>
Classification

There have been many attempts at classification. Lundgren (1949) used a basic division into glomus tympanicus tumours arising from the promontory, and glomus jugulare tumours arising from the jugular bulb. This had clinical value in planning the surgical approach for small tumours. Bickerstaff and Howell (1953) used a symptomatic classification, but this had little value, either for management or prognosis. Alford and Guilford (1962) reviewed the world's literature and proposed a staging system for their own cases. The staging was determined by the degree of spread shown radiologically and symptomatically; they then used the staging to indicate various combinations of surgery or radiotherapy.

The most widely used classification was devised by Oldring and Fisch (1979). They proposed four types, A, B, C, D, based on site and size:

- **Type A:** tumours localized to the middle ear cleft (glomus tympanicus tumours)
- **Type B:** tympanomastoid tumours with no destruction of bone in the infralabyrinthine compartment of the temporal bone
- **Type C:** tumours invading the infralabyrinthine region and extending towards the petrous apex with destruction of infralabyrinthine compartment of temporal bone
- **Type D:** tumours with intracranial extension.

This classification, although anatomically based, was primarily differentiated by the surgical approaches used. In 1982, Fisch further subdivided types C and D to cope with the different surgical problems encountered. While the original classification into four types has considerable clinical value, Fisch’s further subdivision is of limited value unless one follows his surgical approach exactly.

Investigation

Those cases presenting to the otologist usually have a red mass behind the tympanic membrane and this should indicate the probable diagnosis. The two other conditions that mimic this appearance are the high jugular bulb or an aberrant carotid artery. The red drum of otitis media should be obvious from the history. More extensive spread involving the external auditory meatus may appear to be a squamous cell carcinoma, which can often bleed profusely. Those cases presenting to the neurologist often cause considerable diagnostic problems, as neuromata of the last four cranial nerves have a common symptomatology.

The first step in investigation entails a careful clinical examination. Observation of the drum under the microscope will frequently show pulsations of the mass, which will be soft and often blanch on palpation. The hearing loss is generally conductive. Neurological assessment of the cranial nerves is very important, and often gives considerable information regarding the extent of the tumour.

Radiological

The main investigation must be radiological and, with the advances in imaging over the last decade, a very detailed assessment of the tumour can be made. Although
computerized tomographic (CT) scanning and angiography are the main techniques utilized, mention must be made of plain X-rays, tomography and venography.

For type A and B tumours, plain mastoid X-rays will show only clouding of the middle ear and mastoid air cells, although special views of the jugular foramen may show unilateral enlargement. Type C tumours may show bone erosion on plain films, but the jagged erosion of the normally well corticated jugular bulb is best seen on polytomography. The absence of the normal crest of bone between the carotid canal and jugular fossa on lateral tomography is virtually diagnostic of a glomus jugulare tumour (Phelps’ sign).

Retrograde venography by catheterization of the internal jugular vein was popular to differentiate between a glomus tympanicus and glomus jugulare, the latter showing as a filling defect. However, with modern CT scanning this invasive technique is no longer necessary.

High resolution CT scanning is the technique of choice for establishing the diagnosis in those cases presenting with a red mass behind the drum (the rising sun sign). The first step is a conventional axial scan; this will show whether the jugular bulb is enlarged and, if there is any erosion of the cortex, a glomus jugulare tumour is likely. Smooth well-corticated enlargement is generally due to the abnormally high jugular bulb. If the jugular fossa has normal dimensions, coronal CT scanning is necessary (the coronal cuts obtained by scanning with the patient’s head in the submentovertical position are necessary as the coronal computer reconstructions do not give adequate detail). If on coronal cuts the carotid canal is normal, the mass must be a glomus tympanicus. An aberrant carotid artery is demonstrated by a more laterally placed carotid canal often with a deficient bony wall. With the more extensive glomus tumours, the erosion of the infralabyrinthine part of the temporal bone and intracranial spread are clearly demonstrated.

In the unlikely event of doubt as to the correct diagnosis still existing after CT scanning, angiography will demonstrate virtually all glomus tumours apart from the very early glomus tympanicus tumour which may not be obvious even with selective angiography of the ascending pharyngeal artery. If the requirement is to establish the diagnosis, digital subtraction angiography using intravenous contrast injection will often confirm. Arteriography is best reserved for the preoperative detailed assessment of the extent of the tumour and indication of the arterial blood supply. If the arteriography is accompanied by preoperative embolization, it should not be performed more than 8 days prior to the planned surgery, otherwise collateral vessels may open, thereby restoring the tumour's vascularity.

Endocrine

Prior to any surgery, it is important to exclude the secretion of any vasoactive hormones by the tumour, particularly in those cases with an elevated blood pressure. A 24-hour urine collection will demonstrate any raised vanillylmandelic acid levels (normal level up to 7 mg/24 hours). Direct biopsy of the tumour should not be necessary if the suggested radiological assessment is used. Biopsy of an obvious tumour presenting in the external auditory meatus is occasionally performed to exclude a squamous cell carcinoma; in such cases bleeding will occur, but it is rarely severe and always stops with a (BIPP) pack.
**Management**

After many years of controversy, it is now possible to propose a series of reasonable therapeutic options in the management of glomus jugulare tumours. In the past, the main arguments have been over the value and place of radiotherapy. Initially it was used when surgical resection was deemed impossible or, more often, when the operation had to be curtailed because of unpredicted technical problems in the hands of a surgeon unfamiliar with this particular tumour. Subsequently, it was used as the sole form of treatment in many centres, and presently tends to be used mainly as an adjunct to surgery.

The main types of surgical procedure described over the years have often reflected the interests of general otology. In the 1940s and 1950s, open techniques were usually performed. In the 1960s the intact canal wall procedures were popular, and since 1970 increasingly more sophisticated skull base procedures have been described.

The use of other modalities of treatment have been suggested, but none have found wide acceptance. Tumour reduction by diathermy was initially popular and, with the evolution of cryotherapy, the cryoprobe was suggested, but in both cases it was found impossible to treat any tumours apart from the smallest glomus tympanicus. Intra-arterial embolization under radiological control is useful for intracranial vascular malformations, but when used for glomus tumours the duration of tumour reduction is very limited. It has no place as a permanent treatment, but is excellent as a preoperative adjunct to surgical resection.

The current treatment options for glomus tumours may be summarized as follows:

1. no active treatment and continued observation
2. primary radiotherapy
3. surgical resection with planned adjunctive radiotherapy
4. surgical resection.

**No treatment**

Glomus tumours are extremely slow growing and may have a long natural history. They usually present in the middle age groups and, where general health is good, treatment is definitely indicated in view of the patient's expected lifespan. Some patients do not present until the latter part of their sixth or seventh decades and, providing that repeat CT scans do not show very extensive spread or rapid growth, and the patient's symptoms are minimal, no treatment is indicated apart from explanation and reassurance.

**Radiotherapy**

There have been several detailed studies on the effects of radiotherapy monitored clinically and radiologically, and several workers have also looked at the histopathological changes in irradiated tumours (Capps, 1952; Silverstone, 1973; Spector, Maisel and Ogura, 1974). Unfortunately, there is too much variation in both method and dose of radiation to draw definite conclusions. Most clinicians agree that radiotherapy is rarely curative, but it does have some effect on slowing tumour growth. Rosenwasser (1968) made the profound generalization 'that the inherent tendency of the glomus jugulare tumours to slow growth may
be more important in determining its radiocurability than is its actual responsiveness to irradiation'. Clinically, following irradiation, visible tumour often shrinks and bleeding generally ceases, and although tinnitus and vertigo may improve, the deafness and other cranial nerve palsies persist. Repeat angiography shows little change in either vascularity or in the extent of the tumour, apart from intracranial extensions which often regress.

Histopathological examination of irradiated tumour shortly after radiotherapy may not give the true picture, as radiation fibrosis generally develops 6-12 months later. Most advocates of radiotherapy stress that improvement may take several years to become obvious. Cytologically, the epithelioid or chief cells show very little change apart from being broken up into nests of tumour surrounded by sheets of fibrous tissue. Most of the radiation effects appear to involve the stroma with changes typical of endarteritis obliterans, and this in some cases causes thrombosis of some areas of the tumour.

Many of the documented complications of radiation, such as cerebral necrosis and radionecrosis of the temporal bone, can probably be attributed to the use of older methods of radiation such as orthovoltage (Jackson and Koshiba, 1974). Today, better tumour localization and megavoltage irradiation should result in fewer complications. The usual practice is to deliver 4000-5000 cGy (rads) over a 3-4 week period.

Most clinicians agree that an elderly or infirm patient with a symptomatic, growing tumour should be treated solely with radiotherapy. In the absence of an experienced surgical team, radiotherapy will probably cause less problems than surgery with types C and D tumours, but it must be remembered that 40% of tumours may continue to grow after initial radiation control (Spector et al, 1974). Rosenwasser, reviewing the results of treatment in 1969, came to the conclusion that surgery, if possible, was the method of choice in the management of glomus jugulare tumours. Despite the advances in skull base surgery over the last decade, it is extremely difficult to eradicate those extensive tumours that invade the petrous apex around the internal carotid artery. If such cases have limited neurological deficit on presentation, surgery may well increase the neurological deficit, and in such cases subradical surgery with postoperative radiotherapy is probably the method of choice. In this respect, the use of the newer interstitial radioactive implants, such as iodine-125 with its long half-life, may well be the ideal form of treatment in the future.

**Surgery**

The objectives of surgery are total resection of the tumour where possible, and this should ideally be achieved without increasing the patient's neurological deficit. In certain cases, improvement of the hearing may also be achieved.

Most of the frightening complications described in the past have resulted from inadequate appreciation of the extent of the tumour preoperatively, and from inadequate exposure of the tumour at the time of surgery.

Type A tumours, or glomus tympanicus tumours, can usually be approached via the external auditory meatus.
Type B tumours can often be encompassed by a combined approach (intact canal wall) procedure.

Type C tumours need some form of skull base approach utilizing an upper cervical dissection and transmastoid approach.

Type d tumours require a skull base approach and posterior fossa craniotomy, some surgeons preferring to perform the resection in two stages.

**Surgical technique**

The various techniques available are well described in the textbooks of operative surgery, but considerable experience is also required for the surgery to be performed safely.

Various methods of reducing the tumours' vascularity have been suggested. Spector et al (1974) favoured preoperative irradiation which reduced vascularity and, by inducing stromal fibrosis, permitted easier tumour dissection. Other surgeons including the author have favoured the use of preoperative embolization. This requires the help of an experienced neuroradiologist as there is about a 1% chance of inadvertent embolization of the internal carotid artery system leading to a stroke. Selective angiography of the individual vessels supplying the tumour is performed and embolization achieved either with gelfoam or lyophilized dural fragments. The procedure should be performed some 4-8 days prior to planned resection, and a light general anaesthetic is used.

The highest quality of anaesthetic help is required for this type of surgery. Careful work-up should have excluded any production of vasoactive hormones by the tumour. Profound hypotension controlled by intra-arterial monitoring reduces bleeding to an acceptable level, and dissection with gauze soaked in 1:1000 adrenaline is also useful.

**Transmeatal approach**

The very small glomus tympanicus tumours can be removed by simple tympanotomy if all their borders can be visualized and additional exposure can be obtained by dissecting the malleus handle free of the drum.

Additional exposure can also be obtained by lowering the inferior annulus in the hypotympanic approach described by Shambaugh (1955); in 1967, Farrior described further extension of this approach by removing the mastoid tip and mobilizing the vertical portion of the facial nerve.

**Extended facial recess approach**

With the advent of combined approach mastoidectomy for cholesteatoma, it became clear that large type A and moderate-sized type B glomus tumours could be removed with an intact canal wall procedure instead of by the traditional radical cavity. House (1968) combined the intact canal wall procedure with a neck approach to the jugular bulb, but did not transpose the facial nerve. Glasscock, Harris and Newsome (1974) further developed this technique.
By extending the facial recess inferiorly, reasonably good access is obtained to the hypotympanum, particularly if the chorda tympani is sacrificed. Even better exposure is obtained by skeletonizing, in turn, the vertical portion of the facial nerve, the sigmoid sinus, and the posterior semicircular canal. With these structures clearly identified, the tympanic recess can be opened widely medial to the vertical portion of the facial nerve and this gives adequate exposure for type B tumours. In such cases, total tumour removal can be accomplished with restoration of hearing.

**Infratemporal fossa approach (lateral approach)**

The essential features of this approach are: (a) the resection of the jugular bulb after ligating the internal jugular vein in the neck, and packing off the sigmoid sinus superiorly, and (b) the anterior transposition of the facial nerve to allow direct access to the jugular bulb region. Such an approach was attempted by Capps in 1955, but the stormy postoperative period caused him to recommend radiotherapy for such extensive tumours. The approach was successfully used by Shapiro and Neues (1964) and Gejrot (1965). Gejrot, in particular, emphasized the importance of preserving the medial wall of the sinus if possible, thus protecting the neural compartment of the jugular bulb and maintaining intact the dura of the posterior fossa. Since that time, many surgeons have utilized this basic approach but especial credit must be given to Fisch (Fisch, Fagan and Valvanis, 1984) who extended the approach for a variety of lesions in the lateral skull base.

The basic steps of Fisch's infratemporal fossa approach are as follows: a postaural incision is extended both superiorly and inferiorly into the neck. The facial flap and pinna are raised and reflected anteriorly. The cartilaginous meatus is transected and closed off as a blind-ending sac. The parotid region is dissected to mobilize the peripheral branches of the facial nerve, and the nerves and vessels of the upper neck are carefully mobilized up to the skull base. Control ligatures are placed around the internal jugular vein and internal carotid artery, but not tied at this stage. A complete mastoidectomy (subtotal petrosectomy) is performed removing all the air cells, the posterior meatal wall, the drum, malleus and incus. The outer wall of the hypotympanum is drilled away and the facial nerve skeletonized along both its horizontal and vertical portions. It is dissected free from the canal and permanently transposed anteriorly.

If, during mobilization of the facial nerve, it becomes apparent that it has been invaded by the tumour, the involved section is resected and a cable-graft using sural nerve is placed between the cut ends.

The sigmoid sinus is ligated in the region of the sinodural angle and the internal jugular vein is ligated in the neck. The tumour is then mobilized, first peripherally then centrally. If possible the medial wall of the sinus is preserved, but if infiltrated with tumour it is resected along with the nerves of the medial compartment. At this stage bleeding occurs from the inferior petrosal sinus where it enters the jugular bulb medially. It is controlled by packing the lumen with Surgicel gauze. A plane of cleavage can often be found between the tumour and the internal carotid artery, otherwise the tumour on the wall of the carotid is controlled with judicious diathermy. At the completion of the procedure, any dural defect is repaired with fascia, the eustachian tube is closed off with bone wax, and the whole cavity filled with a free fat graft.
The anterior transposition of the facial nerve may occasionally be achieved without immediate loss of function. It is more usual to develop a temporary paresis which recovers in 2-3 months with a satisfactory final result in 85% of cases.

Intracranial extensions of greater than 2 cm are best managed, according to Fisch, by a second-stage procedure to reduce the chances of cerebrospinal fluid leak and meningitis.

**Posterolateral approach**

Cheesman and Symon (1987) have recently described a modification to the infratemporal fossa (or lateral) approach of Fisch. They have termed it the 'posterolateral approach' and with it have been able to achieve total resection of most type C and D tumours without transposition of the facial nerve. They originally managed type D tumours by combining a posterior fossa craniotomy with a 'Fisch-type' infratemporal fossa approach at the same operation, and were fortunate not to suffer the same complications as seen by Fisch. Their wide posterolateral exposure allows preservation of the posterior meatal wall and does not require transposition of the facial nerve in most cases. Radical mastoidectomy and transposition of the facial nerve was only necessary in those cases with extensive tumour around the internal carotid artery. In such cases the necessary medial dissection of the skull base often resulted in increased neurological deficit, particularly when the intracranial extension reached anterior to the internal auditory meatus, and they currently feel postoperative irradiation is a useful adjunct in such cases, enabling a more conservative resection. However, long-term follow-up will be necessary to validate their views.

**Management of secretory glomus jugulare tumours**

There has been an increasing number of functionally active parangangliomata reported (Matishak, Symon and Cheesman, 1987), of which nine have been functional glomus jugular tumours. Many of these cases present as phaeochromocytomata and their localization often creates a diagnostic problems. The management of the blood pressure, however, creates even greater problems for the anaesthetist. The initial hypertension often requires both alpha- and beta-blockade and, following embolization and surgical resection, the loss of vasoconstrictor tone results in circulatory collapse, needing correction with massive intravenous infusion, and the use of an anti-gravity suit to the lower body to increase venous return.

To anticipate these potential problems, it is prudent to perform a routine vanillylmandelic acid estimation of a 24-hour urine collection preoperatively.

**Postoperative neurological complications**

The slow development of nerve palsies preoperatively generally allows adequate compensation, and few patients on presentation have any aspiration or swallowing problems. However, if the vagus and glossopharyngeal nerves are further damaged at surgery, there is often an acute swallowing problem postoperatively. Fortunately, most patients rapidly adapt to the unilateral paresis and generally return to normal function after 1-2 weeks. Nasogastric feeding is used in the intervening period and occasionally temporary tracheostomy is necessary. Hoarseness also generally improves with time, but if it persists for more than 6 months the unilateral paralysed cord can be corrected with an intralaryngeal Teflon injection.
The temporary facial palsy following anterior transposition has already been discussed. When
the patient has a facial palsy on presentation, nerve grafting is invariably needed and the
results depend on the duration of the preoperative facial palsy. Palsies of greater than one
year's duration often achieve poor results, and adjunctive facial rehabilitation is often
necessary.

Infratemporal fossa route to lateral skull base

Fisch, in particular, has developed the infratemporal fossa approach as a route to the
lateral skull base. He has described a type B approach to the region of the clivus, and a type
C approach to the posterior aspect of the maxillary antrum, parasellar region, nasopharynx and
sphenoid sinus.

He uses the type B approach primarily for a chordoma of the clivus, cholesteatoma
of the petrous apex, and meningiomata. The operation is an anterior extension of his glomus
approach (type A) and he gains further access by removing bone of the posterior zygomatic
arch and occasionally the head of the mandible. He then works forward along the eustachian
tube, dividing the mandibular branch of the trigeminal nerve and middle meningeal artery to
gain access to the clivus anterior to the carotid artery. His type C approach is basically the
same, except that the zygoma is divided more anteriorly and he continues the dissection more
medially by removing the pterygoid plates, and also divides the nerves of the pterygopalatine
fossa to enter the nasopharynx and sphenoid region. He uses this latter approach for the
removal of some large juvenile angiofibromata, adenoid cystic carcinoma, and squamous cell
carcinoma of the nasopharynx, which extend laterally into the infratemporal fossa.

The author has used both of these approaches in the past for access to the lateral skull
base, but now favours the lateral craniofacial approach (Cheesman and Symon, 1987) to this
region via the temporal fossa and floor of the middle fossa, which leaves the ear and facial
nerve intact. However, in surgery, the description of any particular technique merely increases
the number of possible approaches to solve any particular problem. The way forward in skull
base surgery is to consider all the possibilities for each case, and to provide an individual
solution for each. This often entails the combined approach by both otolaryngologist and
neurosurgeon; if they can work well together the patient benefits.

Epithelial tumours

The only benign epithelial tumour found in the middle ear is the rare adenoma and this
can usually be excised by the standard combined approach mastoidectomy with preservation
of hearing. The common malignant tumour is the squamous cell carcinoma and this is dealt
with in Chapter 22, the essence of treatment being a combination of radiotherapy and radical
surgery. The other malignant epithelial tumours are adenocarcinoma and adenoid cystic
carcinoma, both being relatively radioresistant, and initial radical surgery offers the best
chance of cure. Radiotherapy postoperatively may have some effect on residual disease.

Mesenchymal tumours

The paragangliomata have already been discussed in detail. Schwannomata of the
nerves of the jugular foramen, in particular of the glossopharyngeal nerve, present in a similar
fashion, but are avascular and produce symptoms by expansion. Radiological examination confirms an avascular tumour. Surgical resection by the posterolateral approach is the treatment of choice, but as the tumour is so closely applied to the other nerves of the jugular foramen, additional neurological deficit may result. In view of this, simple decompression in the elderly patient is sometimes more sensible. A schwannoma may occur on the facial nerve. Providing the facial palsy has been of short duration, simple resection and repair by cable-graft can produce excellent results. However, if the facial palsy has been present for more than one year, the results of grafting are less satisfactory, and a hypoglossofacial anastomosis for rehabilitation is more beneficial.

Primary sarcomata of the temporal bone are rare, the most common type being the embryonal rhabdomyosarcoma. This is the most common childhood malignancy; unfortunately it is sometimes initially missed, being reported on pathologically as granulation tissue, but the rapid growth and extensive destruction should make the diagnosis obvious. The more recent use of combination chemotherapy and radiotherapy (Raney et al, 1983) has produced spectacular improvements in prognosis and, although these must remain the main modalities, adjunctive surgical debulking occasionally can be beneficial.

**Secondary tumours**

The temporal bone may occasionally be the site of metastatic tumour, usually the hypernephroma. More commonly, it is involved by direct spread from tumours of the parotid and nasopharynx. In most cases, palliative radiotherapy is indicated, unless the primary tumour can be controlled in which case the metastasis can occasionally be encompassed by radical surgery.

Primary meningioma of the middle ear has been reported, but it is usually associated with an *en-plaque* meningioma of the middle fossa not demonstrated radiologically. A good clearance of the tumour from the middle ear can be achieved by a middle fossa approach, but extensive *en-plaque* involvement of the middle fossa dura is left as its rate of growth is extremely slow.

**Granulomata of the temporal bone**

Wegener's granulomatosis may occasionally present in the middle ear and, if the general condition of the patient is good, may present a diagnostic problem with histological reports of granulation tissue only. Management is discussed in Volume 4, but see also Chapters 15 and 17.

Eosinophilic granuloma or histiocytosis X is a rare condition of unknown aetiology, typified by single or multiple osteolytic lesions with granulomatous replacement. The granulomata are composed of histiocytes and eosinophils. Three distinct clinical variants are recognized: eosinophilic granuloma, Hand-Schüller-Christian disease, and Letterer-Siwe disease.

Eosinophilic granuloma is a disease of young adults with a male predominance. The granulomata present in the middle ear and meatus often complicated with secondary infection. The osteolytic lesions seen on X-ray are generally assumed to be cholesteatoma
preoperatively, but at surgery the granulomatous replacement of bone is obvious, and confirmed by histological examination. Local resection followed by low dose radiotherapy is generally curative.

Hand-Schüller-Christian disease was originally described as a triad of skull base granulation, exophthalmos and diabetes insipidus. It is a more severe form of the disease with multifocal granulomata. It is usually a disease of childhood and there is accompanying systemic upset with recurrent respiratory infection, hepatosplenomegaly, lymphadenopathy and, frequently, diabetes insipidus if the sella is involved. Low dose chemotherapy has been used to control the condition, but it tends to be a chronic disease with a mortality of 10-20%. Letterer-Siwe disease is a fulminating condition in children under 3 years old. The diffuse granulomatous deposits replace the bone marrow, and skin deposits are frequent. Death generally follows intercurrent infection and bleeding diathesis. (See also Chapter 15.)