Jugular foramen schwannomas

ANDREW H. KAYE, F.R.A.C.S., JOSEPH F. HAHN, M.D., SAM E. KINNEY, M.D., RUSSELL W. HARDY, JR., M.D., AND JANET W. BAY, M.D.

Departments of Neurosurgery and Otolaryngology, Cleveland Clinic Foundation, Cleveland, Ohio

Thirteen patients with schwannomas of the jugular foramen were operated on at the Cleveland Clinic between 1974 and 1983. The authors’ experience in managing these rare tumors is presented. Three major growth patterns of jugular foramen schwannoma were seen, and it is postulated that the position of the tumor depends on its point of origin from the nerves as they pass through the pars nervosa of the jugular foramen. The more distal lesions will expand inferiorly out of the base of the skull, and the more proximal lesions will enlarge into the posterior fossa. Tumors in the mid region will tend to expand primarily into bone. The schwannoma was primarily intracranial in six patients. In five patients the tumor expanded the bone at the base of the skull, with only a small intracranial component, and in two patients the tumor was primarily extracranial, with a small extension into the bone or posterior fossa. The presentation of the patients varied according to the tumor growth pattern. Deafness, vertigo, and ataxia were present in all patients with a major intracranial component, and in most of these there were only minimal deficits of the jugular foramen nerves. By contrast, lower cranial nerve involvement, including hoarseness and weakness of the trapezius and sternocleidomastoid muscles, occurred in patients in whom the tumor was primarily within the bone or extracranial. Three of the five patients with the major component of the schwannoma within the bone also had deafness. Symptomatic history was longest in those with tumor mainly involving the bone at the base of the skull, and shortest in patients with entirely extracranial tumor. Surgical resection was accomplished with a joint neurosurgical-otological approach, usually combining a posterior fossa exploration with either a translabyrinthine transcochlear or infralabyrinthine procedure. The exact nature of the operation depended upon the presence of intracranial tumor and on the extent of bone or extracranial involvement. Total excision was performed in all cases. There was no operative mortality, and surgery resulted in loss of function of the ninth, 10th, and 11th cranial nerves in most patients. The major postoperative morbidity consisted of swallowing difficulties and sputum aspiration.

KEY WORDS • jugular foramen • schwannoma • cranial nerve tumor

Schwannomas of the jugular foramen are rare tumors. Maniglia, et al.,10 reviewed the literature in 1979 and found 56 reported cases. Although Pluchino, et al.,14 reported a personal series of 12 patients with jugular foramen schwannomas who presented to the Milan Neurological Institute between 1946 and 1971,14 most reports have been of either single cases3,4,10,11,13 or of only two8,15 or three1,6,9 patients.

Thirteen patients with schwannomas of the jugular foramen have been operated on at the Cleveland Clinic Foundation over the 9-year period from 1974 to 1983. This report describes our experience in managing these rare tumors.

Description of Tumor Type

Three growth patterns of jugular foramen schwannomas were seen in our series. It has been postulated that the position of the tumor depends on its point of origin from the nerves as they pass through the pars nervosa of the jugular foramen.9 The more distal lesions will expand inferiorly out of the base of the skull, whereas the more proximal lesions will enlarge into the posterior fossa. Tumors arising in the mid region will tend to expand primarily into the bone. In this series, the tumors that were primarily intracranial with only a small extension into bone are called Type A; those of which the main mass was within bone, with or without an intracranial component, are considered Type B; and those that were primarily extracranial with only a minor extension into bone or into the posterior fossa are designated Type C. The presentation of the tumor and the surgical management of the patients varied according to the tumor type.

Clinical Material and Methods

Summary of Cases

Table 1 summarizes the clinical data on the 13 patients with jugular foramen schwannomas who were included in this series. There were five men and eight...
The patients' ages at the time of admission ranged from 20 to 58 years, with a mean of 43 years. The presenting features are shown in Table 2, and the initial symptoms in Table 3. Deafness, vertigo, and ataxia were all present to some degree at the time of admission in all patients with Type A tumor, but there were no significant deficits of the nerves of the jugular foramen in any of these patients. Involvement of the lower cranial nerves occurred earlier in patients in whom the tumor was primarily within the bone or extracranial (Types B and C, respectively), so that these tumors often had elements of the jugular foramen syndrome (Vernet's syndrome) or Jackson's syndrome as described by Svien, et al. However, in common with those authors, we are not certain that taste in the posterior third of the tongue was reliably tested in some patients, and it is probable that sensation in the pharynx and larynx was not studied in all cases. Thus, although possibly some patients might be erroneously listed as having no ninth cranial nerve defect, this has not been of any serious practical significance.

The deafness in Type A patients was sensorineural, whereas in Type B patients it was either sensorineural, conductive, or mixed (one patient each). The duration of symptoms was shortest in those patients with extracranial tumors (Type C, mean 7.5 months), and longest in those whose tumor was primarily within the bone (Type B, 3.6 years).

The preoperative diagnosis in each tumor group is shown in Table 4. The most common diagnosis before surgery was glomus jugulare tumor, although those tumors with a major intracranial extension (Type A) were most frequently misdiagnosed as being acoustic neuromas.

### Radiological Investigations

Table 5 shows the radiological investigations used in this series in each tumor group and the number of

* For description of tumor types see text.
† PFC = posterior fossa craniotomy; TLTC = translabyrinthine transcochlear approach; and IL = infralabyrinthine approach. Some of the operations were performed in several stages.
‡ Patient presented to our institution after previous biopsy of tumor through the ear.
§ Facial weakness in this patient developed 1 year after initial subtotal excision of the tumor by PFC. A two-stage operation was performed at that time.
abnormal results found. It is of note that, although the plain skull films revealed abnormalities in all the cases of Type B tumor, they were helpful in only one of the Type A and one of the Type C tumors. Tomography confirmed the plain x-ray findings. In three patients with Type A tumors, the tomography performed was too limited to show abnormalities around the jugular foramen.

Cerebral angiography was used in all cases (including digital subtraction angiography in one case). One Type B tumor exhibited marked vascularity and one Type A tumor moderate vascularity; in all other cases the tumor was avascular. Computerized tomography (CT) scanning was of considerable help in evaluating the intracranial extension of the tumor and, more recently, sector CT scanning has been invaluable in showing the extent of the tumor, not only intracranially but throughout the bone and in extracranial locations as well.

**Surgical Considerations**

Two patients with Type B tumors presented following a previous operation at another institution, which consisted in each case of a biopsy of the tumor through the ear. Both procedures resulted in facial nerve palsies requiring subsequent facial nerve grafts, one with total success and one with only partial return of function.

Surgery was performed in all cases by a joint neurosurgical-otological approach, usually combining a posterior fossa exploration with either a translabyrinthine transcoclelear, or infralabyrinthine procedure. The exact nature of the operation depended upon the presence of intracranial tumor and the extent of involvement in bone and in extracranial areas. Total excision was achieved in every case.

Preoperative radiological evaluation has allowed accurate diagnosis of the extent of these lesions, and is essential in determining the approach that should be used to remove the entire lesion. Besides assessing the extent of intracranial involvement, the most important aspect of planning of the surgical approach is to determine the superior limit of the lesion. This is best done by the use of high-resolution CT scanning. If the patient's internal auditory canal is intact, and in particular

### TABLE 2

**Presenting clinical features in patients from each tumor group**

<table>
<thead>
<tr>
<th>Clinical Feature</th>
<th>Type of Tumor</th>
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<tbody>
<tr>
<td></td>
<td>Type A</td>
</tr>
<tr>
<td>deafness</td>
<td>6</td>
</tr>
<tr>
<td>vertigo</td>
<td>6</td>
</tr>
<tr>
<td>ataxia</td>
<td>6</td>
</tr>
<tr>
<td>facial weakness</td>
<td></td>
</tr>
<tr>
<td>hoarseness</td>
<td></td>
</tr>
<tr>
<td>sternocleidomastoid &amp; trapezius weakness</td>
<td></td>
</tr>
<tr>
<td>swallowing difficulty</td>
<td></td>
</tr>
<tr>
<td>tinnitus</td>
<td></td>
</tr>
<tr>
<td>Babinski response</td>
<td></td>
</tr>
<tr>
<td>diminished taste in posterior 1/3 of tongue</td>
<td></td>
</tr>
<tr>
<td>glosal atrophy</td>
<td></td>
</tr>
<tr>
<td>total cases</td>
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</tbody>
</table>

### TABLE 3

**Initial symptoms in patients from each tumor group**

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Type of Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Type A</td>
</tr>
<tr>
<td>deafness</td>
<td>5</td>
</tr>
<tr>
<td>vertigo</td>
<td>1</td>
</tr>
<tr>
<td>hoarseness</td>
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</table>

### TABLE 4

**Preoperative diagnosis correlated with tumor type**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Type of Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Type A</td>
</tr>
<tr>
<td>acoustic neuroma</td>
<td></td>
</tr>
<tr>
<td>epidermoid tumor</td>
<td></td>
</tr>
<tr>
<td>glomus jugulare tumor</td>
<td></td>
</tr>
<tr>
<td>chordoma</td>
<td></td>
</tr>
<tr>
<td>jugular foramen schwannoma</td>
<td></td>
</tr>
</tbody>
</table>

### TABLE 5

**Radiological investigations performed in 13 cases of jugular foramen schwannoma***

| Investigation            | Type A | | Type B | | Type C |
|--------------------------|--------| |--------| |--------|
|                         | No. Performed | No. Abnormal | No. Performed | No. Abnormal | No. Performed | No. Abnormal |
| plain x-ray films        | 6      | 1 | 5      | 5      | 2      | 1      |
| tomography               | 4‡     | 1 | 5      | 5      | 1      | 1      |
| cerebral angiography     | 6‡‡    | 6 | 5      | 2      | 2      | 2      |
| pneumoencephalography    | 3      | 3 | 2      | 1      |        |        |
| CT scanning              | 5      | 5 | 5      | 5      | 2      | 1      |
| sector CT scanning       | 1      | 1 | 2      | 2      | 1      | |

* For a description of tumor types see text. CT = computerized tomography.
‡ Three of these patients had tomography in too limited an area.
‡‡ One of these patients underwent intra-arterial digital subtraction angiography.
if there is normal hearing without evidence of sensorineural hearing loss or vestibular dysfunction, an infralabyrinthine approach to the skull base would be indicated. However, if radiological studies show that the tumor involves the internal auditory canal, or in particular if it has extended superior to the internal auditory canal to the middle fossa tegmen, then the superior limits of the tumor cannot be reached by an infralabyrinthine procedure and a transcocchlear translabyrinthine procedure is indicated. These otological approaches to the areas of involved bone and extracranial extension have been described by Fisch and Pillsbury, and a brief description of each follows.

**Operative Techniques**

**Infralabyrinthine Approach.** The surgical procedure for the infralabyrinthine approach to the jugular foramen involves first identifying the internal carotid artery, external carotid artery, internal jugular vein, and ninth, 10th, 11th, and 12th cranial nerves in the neck and following them to the base of the skull. This entails separating the digastric muscles from the digastric groove and detaching the sternomastoid muscle from the mastoid.

The otological approach to the superior aspect of the jugular foramen is then carried out. A complete mastoidectomy is performed, and the facial nerve is identified just inferior to the horizontal semicircular canal and is followed down to the stylomastoid foramen. All bone lateral to the facial nerve around the inferior aspects of the external auditory canal is removed anterior to the point of the temporomandibular joint (Fig. 1). After complete decompression of the nerve, it is mobilized and gently elevated out of the stylomastoid foramen up to the level of the horizontal semicircular canal, and is then retracted anteriorly around the inferior portion of the external auditory canal.

Next, the lateral and sigmoid sinuses in the mastoid are followed down inferiorly to the level of the jugular bulb, and the carotid artery is completely exposed up to the level of the eustachian tube. The posterior aspect of the dissection is followed back to the posterior fossa dura and, after the superior plane of the tumor has been developed, the mass can then be delivered out of the skull base.

**Translabyrinthine Transcochlear Approach.** With the translabyrinthine transcocchlear approach to the petrous apex, the vessels and nerves in the neck are identified and dissected as for an infralabyrinthine exposure. Then a complete radical mastoidectomy is performed, with removal of the internal auditory canal, including the anterior portion of the external auditory canal. The facial nerve is mobilized in the parotid gland, including elevation of the nerve off the deep lobe of the parotid in all three branches. The nerve is then completely decompressed from the stylomastoid foramen around to the geniculate ganglion and is relocated in a new canal into the parotid gland (Fig. 2).

A complete labyrinthectomy can then be performed, exposing the entire internal auditory canal. By removal of the cochlea, the extent of the dissection can be carried anterior to the carotid artery at the level of the cavernous sinus, thereby exposing the middle fossa tegmen of the petrous apex at the level of the clivus.

The translabyrinthine transcocchlear approach allows the surgeon to obtain superior access to the tumor up to the level of the middle fossa tegmen and permits him to explore anteriorly and medially along the skull base to the foramen spinosum and foramen ovale. Although limited tumor extensions into the posterior fossa can be removed by this approach, craniotomy is necessary for removal of any significant intracranial involvement.

**Summary of Operations**

Table 6 lists the operations performed for each type of tumor.

<table>
<thead>
<tr>
<th>Operative Procedure</th>
<th>Type of Tumor</th>
</tr>
</thead>
<tbody>
<tr>
<td>posterior fossa craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>posterior fossa craniotomy and translabyrinthine</td>
<td>1</td>
</tr>
<tr>
<td>transcocchlear approach</td>
<td>3</td>
</tr>
<tr>
<td>posterior fossa craniotomy and infralabyrinthine</td>
<td></td>
</tr>
<tr>
<td>transcocchlear approach</td>
<td>1</td>
</tr>
<tr>
<td>translabyrinthine transcocchlear approach</td>
<td></td>
</tr>
<tr>
<td>infralabyrinthine approach</td>
<td></td>
</tr>
</tbody>
</table>

*For a description of tumor types see text.
† This patient (Case 7) had a previous posterior fossa craniotomy and subtotal excision of the tumor, which rapidly recurred.
Jugular foramen schwannomas

Type A Tumors. All patients with Type A tumor had a suboccipital (posterior fossa) craniotomy. In addition, one patient had translabyrinthine transcochlear excision of a small bone extension. The suboccipital craniotomy was performed in a sitting position in four patients and in the park-bench position in two. It was possible to positively identify the tumor arising from the ninth cranial nerve in three patients. In the other three patients, although the tumor arose from the nerves of the jugular foramen, the exact identity of the nerve of origin could not be ascertained with certainty.

Type B Tumors. Two patients with Type B tumor (Cases 5 and 9) had had a previous biopsy of the tumor through the ear. Four patients had a suboccipital craniotomy and otological procedure. One of these patients (Case 7) had a suboccipital craniotomy with subtotal excision of the tumor after presenting with lower cranial nerve involvement. One year later she developed a facial palsy, and repeat investigation showed that the tumor extended from the auditory canal to the clivus, with a 3-cm posterior fossa extension. A two-stage operation was then performed. With a combined translabyrinthine suboccipital approach, some tumor tissue was removed from the posterior fossa up to the level of the petrous apex. During the second stage, the remaining tumor was removed from the petrous apex through a translabyrinthine transcochlear approach to the petrous apex. The facial nerve had been destroyed from the stylomastoid foramen to the internal auditory canal and could not be grafted.

One patient with a Type B tumor (Case 8) had minimal intracranial involvement and underwent total excision via a translabyrinthine transcochlear approach in two stages. The facial nerve was permanently rerouted anterior to the geniculate ganglion. The labyrinth and cochlea were removed. The jugular vein and carotid artery were exposed in the neck, and the tumor was followed into the base of the skull up to the region of the mid clivus. The surgical defect was closed with a free abdominal fat graft and overclosing of the external auditory canal as described by Fisch and Pillsbury.

In the four patients with Type B tumors who underwent a suboccipital craniotomy, the tumor was extradural in three and the other patient had a dural graft with fascia lata.

Type C Tumors. One patient with a Type C tumor (Case 3) underwent total excision with an infralabyrinthine otological procedure. The other patient (Case 10), who had tumor extension into the posterior fossa, had a combined posterior fossa craniotomy and infralabyrinthine approach.

Postoperative Results

All patients had a total macroscopic tumor removal, and there has been no recurrence in any case since the time of each patient's last operation. One patient (Case 7), who had subtotal excision via a craniotomy as an initial procedure, had rapid regrowth of the tumor. Following the last operation there has been no evidence of recurrence.

All patients experienced some degree of swallowing difficulty and sputum retention postoperatively, albeit only very mild in three. All but one patient had ninth, 10th, and 11th cranial nerve dysfunction after opera-

FIG. 2. Diagrams showing a translabyrinthine transcochlear mastoidectomy on the right side. In each illustration, the anterior portion of the skull is to the right of the figure. Left: The extent of the radical mastoidectomy is demonstrated. The facial nerve has been mobilized in the parotid gland, and a complete bone decompression performed from the stylomastoid foramen to the geniculate ganglion. The facial nerve has been elevated out of its bone canal, rerouted, and relocated both in a new bone canal going anteriorly from the geniculate ganglion and also in a soft-tissue passage in the substance of parotid gland. Right: The jugular foramen, terminal sigmoid sinus, proximal internal jugular vein, and internal carotid artery are exposed in the skull base. The condyle of the mandible has been removed to allow access to the infratemporal fossa. The middle meningeal artery arising from the maxillary artery and the third division of the trigeminal nerve are seen at the skull base. The facial nerve has been mobilized (as shown left). The lower cranial nerves have been omitted for simplification.
tion, and one patient (Case 12) had only ninth and 10th cranial nerve palsy. Also, all patients had some degree of hoarseness postoperatively; in five it was severe enough to warrant injection of Teflon into the vocal cord.

Three patients developed transient facial nerve weakness postoperatively. In one case this followed a translabyrinthine transcochlear procedure in which the facial nerve was rerouted, in one it occurred after an infralabyrinthine resection, and in the third it followed resection of a large Type A tumor. As mentioned previously, two of the patients (Cases 5 and 9) had presented with facial nerve palsy following biopsy of the tumor, and another patient (Case 7) developed a permanent facial palsy 1 year after an incomplete resection.

Deafness was reversed in one patient (Case 12) who had a Type A tumor. One patient (Case 5) had a transient collection of cerebrospinal fluid (CSF) under the skin incision postoperatively. There was no operative mortality in the series, and all patients are at present alive and well.

Pathology
In two of the six cases of Type A tumors, a macroscopic cystic component was found at surgery (see Fig. 3). However, this was not noted in any of the other tumors. The pathology in all cases was that of a schwannoma. Most tumors, including all in the Type A group, had mixed Antoni A and Antoni B components, and there was no significant difference in the pathology type between the three groups.

Illustrative Case Reports
Case 12 (Type A Tumor)
This 40-year-old man presented 4 years after the onset of vertigo and 3 years after he became deaf. For the previous six months, he had been unsteady when walking. On examination, he had fine nystagmus on left lateral gaze, a diminished left corneal reflex, left sensorineural deafness, and mild gait ataxia. A CT scan (Fig. 3) showed a left cerebellopontine angle tumor, and plain x-ray films of the skull showed no significant bone erosion.

A posterior fossa craniotomy was performed in the park-bench position, and a partially cystic tumor was found to be arising from the ninth cranial nerve, extending up to the level of the fifth cranial nerve. The seventh and eighth cranial nerves were easily peeled off the upper part of the capsule of the tumor. There was only minimal extension into bone, and a complete excision was performed, leaving the 10th and 11th cranial nerves intact.

Save for minor swallowing difficulties, sputum retention, and a slight degree of hoarseness, the patient made an uneventful recovery and was discharged home 8 days postoperatively. At the time of follow-up examination 3 months later, his deafness had partially resolved.

Comment
This patient had a Type A tumor in which the nerve of origin was easily identified. Although the tumor arose from the ninth cranial nerve and compressed the 10th and 11th cranial nerves, the patient did not present with any features of lower cranial nerve dysfunction, as was typical of Type A cases.

Case 11 (Type B Tumor)
This 53-year-old woman presented following 3 years of tinnitus and hoarseness and 6 months of intermittent vertigo. On examination she had right glossal atrophy and paresis of the right true vocal cord, with good palatal movement. There was moderate bilateral sensorineural hearing loss with moderate reduction of word discrimination on the right and good word discrimination on the left. Tympanometry was consistent with normal middle-ear function. The CT scans (Fig. 4) demonstrated a destructive lesion in and around the jugular foramen with intracranial posterior fossa extension (Fig. 5), and angiography showed the mass to be avascular.

To achieve complete removal, a two-stage operation was planned. At the first stage, a posterior fossa crani-
Jugular foramen schwannomas

*Left Pair:* Part of the extent of bone involvement.  
*Right Pair:* Posterior fossa extension.

Otorrhaphy was performed in the park-bench position. The posterior fossa tumor extension was entirely extradural and, after incision of the dura, this portion was easily removed. At the second stage, the area of bone involvement was removed through an infralabyrinthine approach. The postoperative course after each operation was uneventful, except for minor swallowing problems and sputum aspiration.

Comment

This patient had a Type B tumor, which was most easily removed by a staged neurosurgical-otological procedure. As in four of the five cases of Type B tumor, this patient had evidence of lower cranial nerve dysfunction at the time of presentation.

Case 10 (Type C Tumor)

This 50-year-old man was seen 12 months before surgery when he presented with a complaint of slight hoarseness. Six months later an otolaryngologist diagnosed a right true vocal cord paralysis, and from that time he had gradual onset of difficulty in swallowing and right shoulder weakness. Examination at the time of hospital admission showed paralysis of the right ninth, 10th, and 11th cranial nerves, a mild high-tone sensorineural hearing loss on audiography, and normal electronystagmograms. Skull x-ray films, polytome x-ray films, CT scans, and bilateral carotid and vertebral angiograms showed a relatively avascular tumor at the base of the skull in the area of the jugular foramen. The tumor extended up to the level of the cochlea and the internal auditory canal; it did not involve the petrous apex, and extended only slightly into the posterior fossa. The tumor extended approximately 3 cm into the neck (Figs. 6 and 7). There was no blood flow in the sigmoid sinus, and the internal carotid artery was occluded at its origin.

A combined neurosurgical-otological approach to the tumor was planned. The facial nerve was rerouted up to the external auditory canal, with preservation of the external auditory canal and middle ear structures. A suboccipital craniectomy was performed and, with extradural elevation of the cerebellum, the superior limit of the tumor could be mobilized. The carotid artery was identified in the neck and was followed superiorly to accomplish total tumor removal. Postoperative hoarseness and swallowing difficulties were markedly improved after an injection of Teflon into the vocal cord.

Comment

Vocal cord paralysis was the presenting symptom in both patients with Type C jugular foramen lesions. This patient's tumor apparently began in the most distal portion of the jugular canal pars nervosa. The pattern of growth was mostly inferior, below the base of the skull. Suboccipital craniectomy with extradural exploration of the posterior fossa allowed access to the superior extension of the tumor, with preservation of auditory, vestibular, and facial nerve function.
A. H. Kaye, et al.

FIG. 6. Case 10. Computerized tomography scans with contrast enhancement in a patient with a Type C tumor. Tumor can be seen extending through the jugular foramen.

Discussion

Jugular foramen schwannomas are rare tumors. Although there have been sporadic reports of these tumors, there has been little discussion on their various growth patterns and how these patterns can influence the symptomatology of the patient. Reports have emphasized either an intracranial location of the tumor or tumor growth into bone or extracranial areas, and have rarely mentioned that this tumor may grow in three basic directions, thereby producing quite distinct clinical presentations and management problems.

Three major growth patterns of jugular foramen schwannomas were seen in this series, and it is postulated that the position of the tumor depends on its point of origin from the nerves as they pass through the pars nervosa of the foramen jugulare. The jugular foramen is actually a canal that courses anteriorly, inferiorly, and laterally so that the more distal lesions will expand inferiorly out of the base of the skull and the more proximal lesion will enlarge into the posterior fossa. Tumors arising in the mid region will tend to expand primarily into bone. The presentation and surgical management of the patients vary according to the type of tumor.

Patients with Type A tumors presented with symptoms of a posterior fossa mass without deficits of the jugular foramen nerves. All patients presented with a history of deafness, vertigo, and ataxia. The deafness, which was sensorineural, was the first symptom in five of the patients. Although at operation the seventh cranial nerve was markedly distorted by the tumor in five cases, in only one of these was there any preoperative facial weakness. This presentation, mimicking that of acoustic neuroma, has been described by others. In the series of Pluchino et al., of 12 intracranial neuromas, all patients had hearing loss and four were ataxic; however, seven also had raised intracranial pressure at presentation, which was not seen in this series or in others. Reversible hearing loss (Case 12) has been documented before, and a diminished corneal reflex (Case 12) and Babinski response (Case 6) have also been noted in larger tumors. Tumors with their primary growth in either bone or extracranial locations (Types B and C, respectively) have received less attention, and by contrast all but one in this series had evidence of lower cranial nerve dysfunction without significant posterior fossa signs. It is assumed that, in Type A tumors, the posterior fossa allows a relatively capacious environment in which the tumor can expand, and so pressure on the lower cranial nerves is minimal. However, those tumors expanding into bone or the upper cervical region meet some resistance, and so jugular foramen cranial nerve dysfunction is present early. This is further borne out by the fact that, in those patients with Type

FIG. 7. Diagram of a coronal section of the skull base showing the extent of tumor involvement in Case 10. The tumor extends into the neck, and the superior extension reaches the cochlea and internal auditory canal. This represents a typical Type C jugular foramen schwannoma.
Jugular foramen schwannomas

A tumors, the first symptom in five of them was due to pressure on the eighth cranial nerve, which lay some distance from the jugular foramen. Jugular foramen syndromes accompanying foramen jugulare schwannomas have also been described, and some features of Vernet's and Jackson's syndromes were seen in most of the Type B and all of the Type C cases in this series.

Radiological investigation has in the past included plain x-ray films, tomography, pneumoencephalography, angiography, and nuclear scanning. High-quality CT scanning (including sector CT scanning of intracranial tumors, there was erosion in all of the cases. In the series of Pluchino, et al., of intracranial tumors, there was erosion in all of the cases studied with special plain x-ray projections and/or tomography. Di Chiro, et al., stated that jugular foramen schwannomas cause enlargement of the jugular foramen involving predominantly the pars nervosa, with smooth indistinct sclerotic margins. In contrast, glomus jugulare tumors caused irregular enlargement of the jugular foramen, with indistinct margins. While this holds true for Type A and Type C tumors in this series, it was not the case for Type B neuromas, which were associated with considerable bone destruction. Angiography (either the conventional procedure or digital subtraction angiography) may help differentiate the schwannoma from a glomus jugulare tumor in Type B and C cases, since the glomus tumor is highly vascular while the neuroma has variable, often slight, vascularity.

Although surgery of Type A tumors is relatively straightforward, being just a modification of acoustic schwannoma surgery, those tumors that have a major extension either into bone or extracranially present particular technical difficulties. That complete excision is paramount for these lesions is borne out by our Case 7, in which the patient had rapid regrowth of tumor after a primary subtotal excision. Complete excision has best been achieved by a combined neurosurgical-otological procedure, with the particular type of operation depending upon the position of the tumor.

Of particular importance is the prevention of a CSF leak when the tumor extends from the posterior fossa either into the bone or extracranially. This can be averted by using a free fat graft to fill the extradural defect, as well as meticulous dural closure with fascia if necessary.

The postoperative problems of sputum retention and aspiration have been troublesome. Even though the vagus nerve was spared in most cases, some degree of sputum retention and aspiration invariably occurred postoperatively, as it has in other series. This was particularly serious in two patients, but all responded to intensive physiotherapy.

Pluchino, et al., reported a 16.1% operative mortality rate in their series and a 9.1% rate in their review of the literature. There were no operative deaths in this series, but, as most of our patients were operated on since publication of the report of Pluchino, et al., we have had the benefit of considerable technical advances, such as the operating microscope. In addition, a knowledge of the growth patterns and improved radiology aided in the preoperative diagnosis and planning of the surgical approach in these tumors. By using a combined neurosurgical-otological approach, it was possible to achieve a complete and safe excision of the tumor.

References


Manuscript received May 23, 1983. Accepted in final form November 3, 1983.
Address reprint requests to: Andrew H. Kaye, F.R.A.C.S., Department of Neurosurgery, Royal Melbourne Hospital, Grattan Street, Parkville, Victoria 3050, Australia.