Combined approaches for resection of extensive glomus jugulare tumors

A review of 12 cases

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Complete resection with conservation of cranial nerves is the primary goal of contemporary surgery for glomus jugulare tumors. This publication reports the value of combined surgical approaches in achieving this goal in 12 patients with extensive tumors. Eleven of these tumors were classified as Fisch Class C and/or D, while eight were categorized as Jackson-Glasscock Grade III or IV. Intracranial (intradural) extension was present in 10 patients; four patients had tumor extension into the clivus and two into the cavernous sinus. The petrous internal carotid artery (ICA) was involved in eight and the vertebral artery (VA) in one.

Subtemporal-infratemporal, retrosigmoid, and/or extreme lateral transcondylar approaches were added to the usual transtemporal-infratemporal approach. This improved the exposure, provided early control of the petrous ICA, and facilitated tumor removal from the clivus, cavernous sinus, posterior fossa, and foramen magnum, allowing a single-stage resection in eight patients. Ten patients had a complete microscopic resection with no mortality. The facial nerve was preserved in nine cases, with tumor involvement requiring nerve resection followed by grafting in the remaining three. Mobilization of the facial nerve was avoided in five cases; of these, three had intact function and two had House-Brackmann Grade III function on follow-up review. Only one patient had a mild persistent swallowing difficulty. The ICA was preserved in 10 patients and resected in two, while the VA required reconstruction in one case. There were no instances of stroke, and blood transfusions were required in five patients who had tumors with nonembolizable ICA or VA feeders. While complete resection provides the best possibility for cure, the important role of adjuvant radiation therapy in cases with residual tumor is discussed. The importance of degrees of brain-stem compression and vascular encasement is emphasized in classifying the more extensive tumors.

Key Words • glomus jugulare tumor • grading systems • paraganglioma • surgical approach • facial nerve function

With the advances made recently in diagnostic and interventional radiology, skull-base approaches, and microsurgical techniques, the prevailing modality for the management of extensive glomus jugulare tumors is complete surgical excision. Although involvement of the internal carotid artery (ICA) and intracranial extension are no longer deterrents to surgery, they may threaten serious complications and, moreover, preservation of cranial nerves still remains a challenge. For more extensive tumors with intracranial extension, there is also the added risk of injuring the brain stem and intracranial vessels. Extension of the tumor into the adjacent clivus, cavernous sinus, and foramen magnum adds further challenges. Therefore, the goal of contemporary surgery is to render the patient disease-free while minimizing permanent disability relating to the cranial nerves, brain stem, and major arteries.

We present our surgical experience with 12 cases of extensive glomus jugulare tumors and describe the combined approaches used. In most cases, a subtemporal-infratemporal approach was added to the usual transtemporal-infratemporal approach, with a few lesions requiring the addition of a retrosigmoid cranio-
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### TABLE 1
Clinical features at referral of 12 patients with extensive glomus jugulare tumors

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Duration of Symptoms (yrs)</th>
<th>Prior Treatment</th>
<th>Cranial Nerve Signs &amp; Symptoms*</th>
<th>Gait Disturbance</th>
<th>Headaches</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>22, F</td>
<td>none</td>
<td>no - conductive</td>
<td>VI Hearing Loss</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>2</td>
<td>36, M</td>
<td>5 ops &amp; radiotherapy</td>
<td>yes - conductive</td>
<td>no</td>
<td>yes</td>
<td>yes</td>
</tr>
<tr>
<td>3</td>
<td>42, F</td>
<td>none</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>4</td>
<td>39, F</td>
<td>2 ops</td>
<td>no - conductive</td>
<td>yes</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>5</td>
<td>58, F</td>
<td>1 op</td>
<td>no - conductive</td>
<td>yes</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>6</td>
<td>60, F</td>
<td>1 op</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>7</td>
<td>68, F</td>
<td>none</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>8</td>
<td>49, M</td>
<td>1 op</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>9</td>
<td>28, M</td>
<td>none</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>10</td>
<td>66, M</td>
<td>none</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>11</td>
<td>30, M</td>
<td>none</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
<tr>
<td>12</td>
<td>40, M</td>
<td>2 ops</td>
<td>no - conductive</td>
<td>no</td>
<td>no</td>
<td>no</td>
</tr>
</tbody>
</table>
* Roman numerals denote cranial nerves. = normal; \( \downarrow \) = paresis; \( \downarrow \downarrow \) = complete paralysis.

### Clinical Material and Methods

Twenty-eight patients were registered with a diagnosis of paraganglioma in the Cranial Base Tumor Registry at the University of Pittsburgh Medical Center. Of these, 12 patients were identified who had undergone surgery for extensive glomus jugulare tumors.

### Clinical Features

The clinical features of the 12 patients, with pertinent signs and symptoms, are summarized in Table 1. Six had undergone previous operations at this or other institutions and were referred for significant regrowth of residual disease; the other six were newly diagnosed patients referred for primary treatment. There were six men and six women, with a mean age of 45.7 years (range 22 to 68 years). The average duration of symptoms at the time of referral was slightly greater than 3 years (range 1 to 12 years). All 12 patients had hearing difficulty; 11 had a conductive hearing loss only and one had a sensorineural deficit. Of the four patients presenting with swallowing difficulty, two became symptomatic after previous operations. Only one patient had a history of aspiration pneumonia, while two others suffered from occasional coughing spells with aspiration identified on postcontrast swallowing tests. Table 2 shows the preoperative facial nerve function according to the House-Brackmann facial nerve grading system. The facial palsy noted in four patients occurred after previous surgery. Aside from hearing loss, the preponderance of signs and symptoms were related to the lower cranial nerves; the majority of these were in patients who had had previous operations. Three patients presented with ataxia related to brain-stem compression. None of the patients suffered from labile hypertension or were suspected of harboring endocrine-active tumors.

### Radiographic Features

Tumor size and extent were determined from preoperative contrast-enhanced computerized tomography (CT) with bone windows, magnetic resonance (MR) imaging, and four-vessel cerebral angiography.
The size of intracranial extension, degree of brain-stem compression and vascular encasement, and regions of extension are shown for each patient in Table 3. The brain stem was compressed in three cases and had tumor abutment in another three. The ICA was involved in eight patients: four with partial encasement, three with narrowing from total encasement, and one with complete occlusion. The vertebral artery (VA) was encased and narrowed in one patient (Case 2). Another patient (Case 9) had an occipital dural arteriovenous malformation in addition to the tumor. Four patients had clival involvement by the tumor, two of whom had cavernous sinus extension. The Jenkins and Fisch classification system for delineating the extent of glomus tumors (Table 4) was applied and 11 tumors were categorized as Class C and/or D and one as Class B; by the Jackson-Glascoek classification system (Table 5), seven tumors were categorized as Grade IV, one as Grade III, and four as Grade II.

Preoperative Protocol

In all patients, collateral cerebral blood flow (CBF) reserve was assessed to prepare for possible intraoperative temporary or permanent occlusion of the ICA and/or VA. At our institution, this is done by clinical evaluation during a 15-minute balloon test occlusion of the ICA; if the results were satisfactory, this was followed by a balloon-occluded stable xenon-enhanced CT study of the CBF. In one patient whose ICA was already occluded, a balloon test occlusion of the ipsilateral tumor-encased VA was performed. None of the patients failed the test occlusion, and CBF remained bilaterally symmetrical at less than 35 cc/min/100 gm during temporary occlusion.

Superselective catheterization techniques were used to embolize feeders from the ascending pharyngeal, occipital, internal maxillary, or (in one case) the vertebral and anterior inferior cerebellar arteries. Embolization was attempted in all patients except one (Case 5), in whom the feeders were too small and originated mostly from the ICA.

Surgical Approaches

In all 12 patients, standard anesthetic techniques were used to facilitate cranial nerve stimulation and monitoring and also to record electroencephalographic tracings and somatosensory evoked potentials when necessary. The various combinations of surgical approaches, the management of vascular involve-

### TABLE 3

*Extent of tumor and surgical procedures*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Radiographic Findings</th>
<th>Surgical Approaches</th>
<th>ICA &amp; VA Status</th>
<th>Resection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>2</td>
<td>+++ (H) (45 mm)</td>
<td>O</td>
<td>–</td>
<td>incomplete</td>
</tr>
<tr>
<td>3</td>
<td>++ (8 mm)</td>
<td>–</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>4</td>
<td>++ (10 mm)</td>
<td>N</td>
<td>+</td>
<td>complete</td>
</tr>
<tr>
<td>5</td>
<td>+ (8 mm)</td>
<td>PE</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>6</td>
<td>+++ (10 mm)</td>
<td>–</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>7</td>
<td>++ (18 mm)</td>
<td>PE</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>8</td>
<td>–</td>
<td>PE</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>9</td>
<td>+++ (22 mm)</td>
<td>PE</td>
<td>–</td>
<td>incomplete</td>
</tr>
<tr>
<td>10</td>
<td>+ (7 mm)</td>
<td>–</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>11</td>
<td>+ (12 mm)</td>
<td>N</td>
<td>–</td>
<td>complete</td>
</tr>
<tr>
<td>12</td>
<td>+ (9 mm)</td>
<td>N</td>
<td>–</td>
<td>complete</td>
</tr>
</tbody>
</table>

*Intracranial extension: – = none; + = present without brain-stem involvement; ++ = brain-stem abutment; +++ = brain-stem compression; H = hydrocephalus.

†ICA = internal carotid artery; VA = vertebral artery; CS = cavernous sinus; FM = foramen magnum; ITF = infratemporal fossa; – = not involved; + = involved; N = narrowed, totally encased; O = occluded; PE = partially encased.
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**TABLE 4**

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>tumors limited to the middle ear cleft (tympanicum)</td>
</tr>
<tr>
<td>B</td>
<td>tumors limited to the tympanomastoid area with no bone destruction in the infralabyrinthine compartment of the temporal bone</td>
</tr>
<tr>
<td>C</td>
<td>tumors involving the infralabyrinthine compartment with extension into the petrous apex</td>
</tr>
<tr>
<td>D</td>
<td>tumors with intracranial extension</td>
</tr>
</tbody>
</table>

* See Reference 46.

**TABLE 5**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>small tumor involving the jugular bulb, middle ear, and mastoid</td>
</tr>
<tr>
<td>II</td>
<td>tumor extending under internal auditory canal; might have intracranial extension</td>
</tr>
<tr>
<td>III</td>
<td>tumor extending into the petrous apex; might have intracranial extension</td>
</tr>
<tr>
<td>IV</td>
<td>tumor extending beyond petrous apex into the clivus or infratemporal fossa; might have intracranial extension</td>
</tr>
</tbody>
</table>

* See Reference 43.

ment, and the completeness of resection are shown in Table 3.

**Trans temporal-Infratemporal Approach.** In all cases, the initial approach performed by our otology colleagues was transtemporal/infratemporal, as described by Fisch. A curvilinear incision is made behind the ear, extending forward superiorly (for temporal craniotomy) and inferriorly through a midcervical crease to expose the great vessels and lower cranial nerves. The facial nerve is exposed from its exit at the stylomastoid foramen to the pes anserinus in the parotid gland (Fig. 1 left). A radical mastoidectomy is performed to expose the sigmoid sinus and bony fallopian (facial) canal, after transection and blind-sac closure of the external ear canal and forward reflection of the pinna with the scalp flap (Fig. 1 center). In order to avoid a cerebrospinal fluid (CSF) leak, closure of the external auditory canal was considered a necessity in all cases in which there was intradural extension of the tumor. The entire bony external ear canal is then removed with the remaining skin of the canal and the tympanic membrane. Tumor involving the middle ear can be removed through this exposure (as in all but one of our cases).

**FIG. 1.** Cadaver dissection photographs of the transtemporal-infratemporal approach. *Left:* Initial retroauricular incision with detachment of the sternocleidomastoid muscle (SCM) from the mastoid process (M). Inferior neck dissection showing the internal jugular vein (IJV), lower cranial nerves (LCNs), and facial nerve (FN) from its exit at the stylomastoid foramen into the parotid gland (P). *Center:* Mastoidectomy exposing the sigmoid sinus (small black arrow) and mastoid segment (asterisk) of the facial nerve (FN). Blind-sac closure (BS) after transection of the external ear canal (arrowhead). The parotid gland (P), sternocleidomastoid muscle (SCM), lower cranial nerves (LCNs), and internal jugular vein (large black arrow) are shown. *Right:* Radical mastoidectomy further exposing the mastoid facial nerve (black arrow) and posterior bend of the petrous internal carotid artery (asterisk). The internal jugular vein (white arrow), lower cranial nerves (LCNs), mandibular condyle (C), zygomatic arch (Z), and parotid gland (P) are also seen.
As can be observed from Fig. 1 center and right, any tumor extending more anteriorly, specifically along the petrous ICA and more medially toward the clivus, would require anterior transposition of the facial nerve from the fallopian canal. Both proximal and distal control of the great vessels (sigmoid sinus, internal jugular vein, and ICA) is key during this operation. While proximal ICA control in the neck is easily provided by the transtemporal-infratemporal approach, distal (horizontal petrous ICA) control may be difficult without directly injuring or stretching the facial nerve if it is kept in its canal (Fig. 1 right). Moreover, even translocation of the facial nerve anteriorly to gain access to the petrous ICA still would not provide distal control prior to tumor dissection from the artery. Anterior transposition of the facial nerve distal to the geniculate ganglion was carried out in three cases and the vertical mastoid segment alone was mobilized in one. Due to significant tumor encasement, three patients required facial nerve resection and reconstruction with a greater auricular nerve interposition graft. To avoid postoperative facial paresis, in five patients operated on recently the nerve was left in place in the intact fallopian canal, since the subtemporal-infratemporal approach was added to provide the necessary anteromedial exposure.

Subtemporal-Infratemporal Approach. A preauricular subtemporal-infratemporal approach was added whenever the tumor involved the ICA and/or extended anteromedially into the clivus and cavernous sinus. Briefly, this involves elevation of the temporalis muscle (TM), temporal craniotomy, and removal of the zygomatic arch (Z) with the glenoid fossa (large arrow). Also shown are the mandibular condyle (C), parotid gland (P), facial nerve (parotid FN) and mastoid (asterisk) segments, the sternocleidomastoid muscle (SCM), and the sigmoid sinus (small arrows). Center: Combined approaches showing the entire petrous internal carotid artery (ICA[P]), the transected mandibular nerve (V3), mastoid segment of the facial nerve (asterisk), sigmoid sinus (small arrows), internal jugular vein (large arrow), parotid gland (P), temporal muscle (TM), and sternocleidomastoid muscle (SCM). Right: Anterior reflection (large arrow) of the petrous internal carotid artery (ICA[P]) exposing the petrous apex and clivus (arrowhead). The sigmoid sinus (small arrow), internal jugular vein (medium arrow), mastoid segment of the facial nerve (asterisk), and parotid gland (P) are also shown.
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Xenon-CT studies of CBF after the occlusion had occurred showed no decrease or asymmetry in blood flow, and the ICA was thus resected during a second-stage operation without placing a bypass vein graft.

**Tumor Resection Through Combined Approaches.** With the great vessels under better control, tumor resection can proceed through the wide superior (subtemporal-infratemporal) and inferior (trans temporal-infratemporal) exposures working anterior and posterior to the facial nerve without having to transpose or stretch it (Fig. 2 center). After ligation of the sigmoid sinus and the internal jugular vein above and below the tumor, the latter is dissected with the jugular bulb from the pars nervosa (glossopharyngeal, vagus, and accessory nerves) of the jugular foramen. Bleeding, often brisk, from the inferior petrosal sinus at its entrance into the jugular bulb is easily stopped with Surgicel packing. Involvement by the tumor necessitated resection of the glossopharyngeal nerve in five patients, the vagus nerve in four, and the accessory nerve in one.

Once totally free of tumor from the peristeum of the bony canal, reflection of the petrous ICA (horizontal genu and segments) laterally allows access to the entire petrous apex and upper and middle clivus (Fig. 2 right). The eustachian tube, opened during this exposure, should be packed with fat and/or muscle and closed with suture in order to prevent a CSF leak, especially when intradural exposure is likely. Tumor is then removed from these regions through the subtemporal-infratemporal approach. This was done in four of our 12 cases; two patients had small posterior cavernous sinus extensions that were extradurally dissected through this approach.

**Intracranial Extension.** Six patients with large intracranial tumor extension (> 10 mm) required the addition of a retrosigmoid craniectomy and posterior fossa exploration for tumor removal. In these cases, the C-shaped scalp incision is curved back far enough behind the ear to allow for the craniectomy. Intradural tumor dissection from the brain stem, cranial nerves, and major arteries is performed using the appropriate microsurgical techniques for preserving these structures. Tumor encasement of the nerve required sectioning of the hypoglossal nerve in three patients. One patient with a tumor-encased VA (Case 2) underwent primary VA resection and reconstruction.

Two patients required a second-stage extreme lateral transcondylar approach to resect tumor extending into the foramen magnum and up to C-1. For this approach, the suboccipital (retrosigmoid) craniectomy is extended laterally and inferiorly, removing portions of the occipital condyle and lateral mass of C-1 so as to expose dura up to 1 cm in front of the dural entrance of the vertebral artery (Fig. 3). This approach provides excellent proximal control of the VA and a direct view of the tumor-brain stem interface to facilitate dissection.

Various combinations of the approaches were used in 10 of the 12 cases, allowing single-stage resections to be accomplished in eight. The approaches provided adequate anterior, lateral, and posterior exposures of the tumor (Fig. 4), with complete control of the ICA, sigmoid sinus, and internal jugular vein.

**Closure**

Dural closure with pericranial or fascia lata grafts and the addition of a temporal muscle flap and fat to cover the defect was the usual practice, especially in patients with intradural intracranial tumor extension. In two patients who had undergone multiple procedures (one with postoperative radiation therapy) and large surgical defects, the plastic surgeons performed closure with a free flap of revascularized rectus abdominus muscle.

Intraoperative blood loss required transfusion in only five of the 11 patients with preoperative embolization. These five patients underwent external carotid artery embolization; however, additional significant feeders from the ICA could not be embolized.

**Illustrative Case**

**Case 2**

This 36-year-old man presented after five previous operations and radiation therapy with regrowth of tumor into the posterior fossa, clivus, posterior cavernous...
sinus, and infratemporal fossa (Fig. 5). He also exhibited hydrocephalus from an obstructed fourth ventricle, and presented with a persistent right facial palsy due to his prior operations.

**Examination.** Preoperative angiography demonstrated feeding vessels couring to the tumor from retrograde filling of an occluded ICA and also from the anterior inferior cerebellar artery; both of these arteries were successfully embolized. Angiography and MR imaging confirmed narrowing of the VA due to tumor encasement. Balloon test occlusion of the VA was tolerated, and xenon-CT CBF studies showed a flow of more than 35 cc/min/100 gm during temporary occlusion. The CBF increased significantly after administration of Diamox (acetazolamide).

**Operations.** The patient underwent a two-stage procedure and placement of a ventriculoperitoneal shunt between stages that was prompted by CSF otorrhea secondary to hydrocephalus. The first stage entailed a trans-temporal approach with a preauricular subtemporal-infratemporal approach and attempted resection of tumor; however, there was failure to distally occlude the ICA. At the second stage, retrosigmoid and extreme lateral transcondylar approaches were combined with the previous approaches to successfully remove the tumor. Although there was significant compression of the brain stem, the tumor had not invaded the pia mater. The encased VA was transected during tumor resection, and primarily reanastomosed.

**Postoperative Course.** A postoperative vertebral angiogram showed normal patency of the vessel. A segment of the tumor-encased facial nerve was resected and grafted with sural nerve. Dural closure was accomplished using pericranial and fascia lata grafts, fibrin glue, and a covering of fat from the thigh. The entire defect was then covered with a revascularized rectus abdominus muscle free flap. Minimal residual tumor, suspected in the midclival area and intentionally left in the sphenoid sinus in order to avoid CSF leak, remained without regrowth at the 3-year follow-up evaluation. The patient has resumed his previous occupation with minimal hoarseness and no swallowing difficulty.

**Results**

Complete microscopic resection was obtained in 10 of the 12 patients. Besides Case 2, one other patient with incomplete resection underwent surgery early in this series and had residual tumor in the medial clivus. This patient (Case 9) had radiation therapy postoperatively and remains without progression of disease 6 years after surgery. At a median follow-up period of 44 months, none of the 12 patients has had a recurrence or progression of disease. Eleven of the 12 patients are independent (Karnofsky score > 70), eight of whom are working (median follow-up period 44 months). One patient who was mildly retarded since 1 year of age remains so postoperatively.

Postoperative facial nerve function is summarized in Tables 2 and 6. All five patients not requiring mobilization of the facial nerve had return of facial function: three were normal and two were in House-Brackmann Grade III on follow-up evaluation. All four patients with mobilization of the facial nerve exhibited significant (Grade VI) postoperative facial paralysis; at 1 year postoperatively, none of these had improved to normal (one to Grade II, two to Grade III, and one remaining in Grade VI). Due to extensive tumor involvement, three patients required segmental grafting, two of whom had improved facial nerve function. The third patient remained with complete facial paralysis 1 year after surgery.

Although the external auditory canal was closed in all cases, sensorineural hearing was maintained in ev-
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Complications

The three patients with brain-stem distortion had worsened ataxia postoperatively. This resolved within 6 months in one patient and improved to independent walking in the other two over a 2-year period. Postoperatively, hemiparesis developed in one patient (Case 2) with severe brain-stem distortion and hydrocephalus; however, this completely resolved within 2 years, at which time the patient could walk independently.

Nine of 10 patients with a swallowing difficulty postoperatively eventually improved without requiring a permanent gastrostomy or feeding tube. None suffered from aspiration or pneumonia. In five patients with a decreased gag reflex preoperatively, temporary (<4 weeks postoperatively) tracheostomy and gastrostomy were used. Patients with preoperative glossopthenic and vagus nerve deficits had a tendency toward earlier improvement in swallowing than those who had no preoperative deficit. Postoperative hoarseness in nine patients was temporary (<1 year) and only four required injection of Teflon into the vocal cords.

Otorrhea developed after the first-stage operation in one patient (Case 2) and resolved after placement of a ventriculoperitoneal shunt for hydrocephalus. Three patients developed a large neck effusion, which promptly resolved after placement of a lumbar drain for a few days. All of these four patients had undergone intradural tumor removal. There was no incidence of wound dehiscence or meningitis. Two patients developed benign intracranial hypertension within weeks after their operation and required lumbo-peritoneal shunt placement. There were no deaths or ischemic complications in this series.

Classification

Although histologically benign, glomus jugulare tumors are well known for their aggressive behavior, involving the middle ear structures, jugular bulb and vein, sigmoid sinus, petrous carotid artery, clivus, cavernous sinus, and cranial nerves, and extending intracranially and/or into the infratemporal fossa. The ever-changing classifications are likely a reflection of the limited radiographic modalities available in the past which are essential in evaluating these very “anatomically involved” tumors.

Not surprisingly, due to its temporal bone location and early involvement of the glossopthenic and vagus nerves and the middle ear, otolaryngologists have had the opportunity to make most of the advances in the diagnosis, classification, and treatment of these tumors. The classifications proposed by Jenkins and Fisch and Jackson, et al., are based on precise information regarding size and regions of involvement as noted on contrast-enhanced CT and CT with bone windows (Tables 4 and 5). Such classifications cer-

Table 6

<table>
<thead>
<tr>
<th>Facial Nerve Manipulation</th>
<th>No. of Cases</th>
<th>Normal</th>
<th>Grade ≤ III</th>
<th>Grade &gt; III</th>
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<tr>
<td>not mobilized</td>
<td>5</td>
<td>3</td>
<td>2</td>
<td>0</td>
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<tr>
<td>mobilized</td>
<td>4</td>
<td>0</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>severed &amp; grafted</td>
<td>3</td>
<td>0</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

* House-Brackmann facial nerve grading system.

Fig. 5. Gadolinium-enhanced magnetic resonance images, preoperative axial and coronal images (upper) and postoperative axial images (lower).
tainly help in deciding the surgical strategy and approaches utilized in individual cases and often indicate the degree of difficulty of tumor resection.

Additional information, lacking in these classifications, relates to brain-stem compression and degree of vascular involvement. The degree of brain-stem compression certainly affects the neurological status of the patients and, as in all three of our patients with this complication, clearly is implicated in their postoperative neurological deficits. The increased difficulty of resection is intuitive whenever there is brain-stem compression, as was our experience with these three patients. One patient with severe distortion of the brain stem had obstructive hydrocephalus that aggravated a CSF leak and required a shunting procedure. Major arterial encasement by tumor certainly deserves close scrutiny prior to surgical management. The degree of encasement not only may imply dissectability of the tumor from the vessel, but should alert the surgeon to prepare for temporary occlusion if injury to the vessel should occur. Repair of the vessel with temporary occlusion was required in our three patients with total encasement of the ICA while, in the four patients with only partial encasement, dissection of the tumor was easier. These patients certainly are at greater risk for stroke complications, although we were fortunate. For similar reasons, verteobasilar involvement should also be carefully scrutinized preoperatively. Involvement of the cavernous sinus, clivus and foramen magnum, and upper spine requires the additional approaches that were used in several of our patients. The location of tumor not only dictates the surgical approaches utilized, but determines the resectability of the tumor as in two of our cases with residual tumor in the upper clivus.

It appears that for the more extensive tumors with brain-stem distortion and vascular involvement, perhaps a classification based on regions of tumor involvement and the degrees of vascular encasement and brain-stem distortion may be more useful (Table 7). This would certainly fulfill the purpose of the existing classification and provide the essential additional information outlined above.

**Combined Surgical Approaches.** Since the initial description of glomus jugulare tumors by Rosenwasser,71 both neurosurgeons1,2,4,7,57,68,84 and otolaryngologists1,3,13,19,82,83,86, working independently, often met with failure to remove the tumor in its entirety due to either involvement of the temporal bone or intracranial extension, respectively. Forty years ago, Seiffert first described a surgical exploration of this tumor extending into the lumen of the jugular bulb and producing a jugular bulb syndrome.7,54 Surgical approaches developed since then have the common goal of total tumor removal by exposing and excising the jugular bulb with relative hemostasis. To do this, Capps52 initially proposed mobilization of the mastoid segment of the facial nerve. Shapiro and Neues57 and Gejrot27 added an inferior neck dissection and rerouting of the facial nerve, allowing control of the extracranial great vessels at the skull base and improved access to the jugular bulb. After House and Glasscock40 reported improved transtemporal techniques in 1968, Glasscock, et al.,79 and Gardner, et al.,24 independently described removal of larger tumors. However, this still fell short of resecting the intracranial extension and tumor located more anteriorly. Fisch22 pioneered the transtemporal-infratemporal approaches with facial nerve transposition. Additional variations of these approaches to remove significantly more extensive tumors have been reported by others.25,30,45 Neurosurgeons often used the suboccipital approach solely to remove intracranial tumors1,2 until Portmann19 described removing large tumors with intracranial extension in two stages, emphasizing the need for a combined and neurosurgical participation in such situations. Since then, several others have popularized combined suboccipital and transtemporal approaches for single-stage resection of these lesions.1,26,38,45,49

Sekhar, et al.,75 previously described the subtemporal-infratemporal approach to expose the petrous ICA and resect extracranial tumors of the petrous apex and upper and middle clivus.74,76 With ICA displacement. For these extensive glomus neoplasms, this added approach proved quite useful when removing tumor from these areas and in providing a "rostral" exposure giving good distal control of the tumor-involved ICA without having to transpose the facial nerve. Prior reports have shown that, in a majority (58%) of the cases with incomplete resection, the tumor involves the anterior carotid canal (petrous), cavernous sinus, and clivus.44 In our series, the combined approaches allowed complete resection in all but two of such cases, with the residual tumor being in the upper midline clivus, an area that can occasionally prove difficult to reach. One of these two patients had prior radiation therapy and both had multiple operations, and these factors certainly rendered the dissection more difficult. No tumor was left in the petrous apex, anterior to the ICA, or in the cavernous sinus.

Extension into the brain stem also resulted in partial resections in prior reports.44 The use of the combined retrosigmoid or the extreme lateral transcondylar approach solved the problem. The latter approach also gave better access to the lower clivus and anterior fo-

<p>| TABLE 7 |
| Classification of extensive glomus jugulare tumors |</p>
<table>
<thead>
<tr>
<th>Regions of Tumor Involvement</th>
<th>Vascular Enencasement</th>
<th>Brain-Stem Compression</th>
</tr>
</thead>
<tbody>
<tr>
<td>petrous bone</td>
<td>none</td>
<td>none</td>
</tr>
<tr>
<td>clivus</td>
<td>partially encased</td>
<td>mild indentation</td>
</tr>
<tr>
<td>cavernous sinus</td>
<td>encased/narrowed</td>
<td>severe compression</td>
</tr>
<tr>
<td>infratemporal fossa</td>
<td>occluded</td>
<td>or invasion (with or without hydrocephalus)</td>
</tr>
<tr>
<td>posterior fossa</td>
<td></td>
<td></td>
</tr>
<tr>
<td>foramen magnum/</td>
<td></td>
<td></td>
</tr>
<tr>
<td>upper cervical spine</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Encasement of the internal carotid artery, vertebral artery, or basilar artery.

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ramen magnum region with a direct view of the tumor-brain stem interface in addition to providing the necessary control of the VA and verteobasilar junction.

Facial Nerve Preservation. While transposition of the facial nerve may permanently compromise normal postoperative function, leaving it in its canal often led to an incomplete resection.\textsuperscript{20,47,58} Glasscock, et al.,\textsuperscript{20,43,45} also proposed the idea of limiting facial nerve mobilization; however, they continued to advocate the need for its anterior transposition or temporary severance for extreme anterior exposure and to achieve distal ICA control. In the series reported by Jackson, et al.,\textsuperscript{41} the facial nerve was preserved in 62% of patients with Fisch Class C tumors and 80% of those with Class D tumors. By the Jackson-Glasscock classification system,\textsuperscript{43} 63% of Grade III and 57% of Grade IV tumors had facial nerve preservation. Leonetti, et al.,\textsuperscript{52} reported on the use of a modified technique for anterior facial nerve mobilization, with some improvement in its functional outcome. Although we used this modification, our results seem to favor keeping the nerve in its canal if oncologically feasible (Table 6). The added subtemporal-infratemporal approach appears to facilitate tumor removal more anteriorly without having to transpose the facial nerve. The nerve was preserved in six of the eight patients with anteriorly located tumors involving the ICA. Scarring from prior surgery and tumor infiltration required nerve severance in the remaining two patients. These factors appeared to predict poor facial nerve outcome. As alluded to by Jackson, et al.,\textsuperscript{41} and supported by our series, early diagnosis (when the tumor is smaller) and complete initial resection would be the most ideal procedure for facial nerve function.

Functional Recovery. In order to prevent a CSF leak in patients with large tumors with intracranial extension, closure of the external auditory canal with permanent conductive hearing loss is acceptable morbidity.\textsuperscript{22,25,42} Most of these patients present with hearing loss anyhow. Although both Farrior\textsuperscript{20} and Maniglie, et al.,\textsuperscript{55} described an inferior mastoidectomy-anterior hypotympanic approach that preserves hearing, this approach is not suited for tumors that are anteromedial to the ICA or extend intracranially.

Contemporary surgery has invalidated the claim that resection of these lesions results in long-term disability, especially regarding lower cranial nerve function.\textsuperscript{14,66} Although temporary tracheostomies and enteral feeding tubes may be required in a few cases, compensatory return of function is the rule rather than the exception. Patients with preoperative swallowing dysfunction tend to recover more rapidly than those in whom the glossopharyngeal and vagal nerves were functioning normally. The mechanisms underlying this compensation are not clear. Gelfoam or Teflon injection of the vocal cords is certainly a useful option in those patients who have no recovery of vocal cord function. Although none of the patients in this series required it, cricopharyngeal myotomy may be performed in patients with long-term swallowing difficulty and those who are at risk of aspiration.

Cerebrospinal Fluid Leak

The risk of CSF leakage is especially increased in patients who have intradural tumor and have undergone previous operations or radiotherapy. With proper closure of the external auditory canal, eustachian tube, and dura, this complication can be avoided. Regional muscle flaps or revascularized free flaps should be used in addition to lumbar spinal drainage with larger defects or questionable wounds.\textsuperscript{48} One of our cases emphasized to us that preoperative hydrocephalus must be treated prior to tumor resection. Postoperative benign intracranial hypertension, although rare,\textsuperscript{11} may certainly aggravate CSF leakage despite meticulous closure. It is of interest to note that, in both patients who developed postoperative benign intracranial hypertension, either a dominant but only partially occluded sigmoid sinus or a major draining mastoid emissary vein was ligated. In both cases, cerebellar and temporal lobe swelling was noted during the operation. Particular attention may be necessary in patients who undergo ligation of major draining veins and develop venous congestion with brain swelling intraoperatively. Preoperative angiographic assessment of venous drainage and intraoperative measurement of venous pressures directly from the sigmoid sinus during temporary occlusion may be useful in preventing or anticipating venous congestion and intracranial hypertension.

Adjuvant Therapy

Preoperative Embolization. The extreme vascularity of the tumor discouraged many earlier surgical attempts, as exemplified by Parkinson\textsuperscript{68} in his description of an operation on a 17-year-old patient as “the most vivid vascular experience we have ever encountered, requiring 6 hours and 17 bottles of blood.” Earlier attempts with external carotid artery embolization often met with failure due to “opening up” of anastomotic channels from the vertebral, internal carotid, opposite external carotid, ascending, and deep cervical arteries.\textsuperscript{37,50,65} For this reason, embolization should be performed only as a presurgical procedure and not as primary therapy. Although potential risks exist, current selective superselective catheterization techniques allow embolization of more specific feeders to the tumors.\textsuperscript{67,68} As a result, operative time and blood loss have been reduced in most cases.\textsuperscript{62} Although most of the blood supply to glomus jugulare tumors comes from the external carotid artery, there are a few cases that involve ICA and VA feeders. Such intracranial feeders can also be embolized, as in Case 2 reported above; however, if embolization of these feeders is not feasible, operative blood loss will remain a problem.

Radiotherapy. Radiotherapy has undeniably had an important role in the treatment of glomus jugulare tumors. Several reports have shown tumor growth control with radiation therapy.\textsuperscript{3,15,23,25,31,64,79} It is also clear that these tumors are not curable by irradiation and regression of the tumor is never achieved. Long-term studies (10 to 20 years) demonstrate control rates of 70% to 85% with several recurrences.\textsuperscript{65,78,82} Postirradiation tumor cell survival has also been shown,\textsuperscript{10,30,89} including
continued secretion of catecholamine. The only patient in our series with prior radiotherapy had a massive regrowth of his tumor. Therefore, while radiation therapy may render a glomus tumor “stable,” it is not curative, and in the younger patients with an additional life expectancy of at least 20 years, surgical extirpation of the tumor provides the best modality for cure. With this treatment most patients, although suffering from acute operative deficits, recover substantially, are disease-free, and are very functional. Radiation therapy should continue to play a role in treating patients who are clinically disabled or elderly and those who have unresectable residual tumor.

Conclusions

The need for a multidisciplinary team approach to the overall management of extensive glomus jugulare tumors is well established. As part of this team, neurosurgeons must often deal with intracranial extension and with vascular and cranial nerve involvement. We recommend, where necessary, the addition of subtemporal-infratemporal, retromastoid, and/or extreme lateral approaches to the known transtemporal-infratemporal approach for the resection of extensive glomus jugulare tumors. This strategy, in most cases, achieves the goal of complete resection with minimal risks of vascular complications and acceptable cranial nerve morbidity and good overall functional outcome.

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