The neurosurgical treatment of glomus jugulare tumors

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Glomus jugulare tumors are discussed with regard to diagnosis and treatment. These tumors, although usually benign, may be fatal because of their location. Therefore, accurate classification by location, size, and the blood supply of the mass is important; use of newer roentgenographic techniques provides key information. Surgical removal is recommended as the treatment of choice. The neurosurgical technique of en bloc excision of glomus jugulare tumors is described and illustrated.

Key Words - glomus jugulare tumor - chemodectoma - nonchromaffin paraganglioma - operative technique

Glomus tumors are relatively newcomers to the medical scene. Even though Valentin had described a ganglion-like formation at the initial portion of the tympanic nerve in 1840, over 100 years passed before Guild accurately described the glomus jugulare. The first tumor of the glomus jugulare was reported only 25 years ago by Rosenwasser. Since that time this tumor has received extensive recognition in the otologic literature. Otologists have classified, divided, and subdivided these lesions by virtue of their location and accessibility to surgical treatment. Many otologists have removed those lesions that were readily accessible and treated others with irradiation. A few otologists have braved the removal of extensive tumors with marked intracranial extension, and occasionally a case report of a combined otologic-neurosurgical venture has been published. Neurosurgeons for the most part, however, have been remiss in ignoring the treatment of this tumor even though it often invades the cranial cavity. Because in our experience this tumor responds erratically, if at all, to irradiation therapy, its neurosurgical treatment needs explanation and clarification.

Classification of Tumor

These tumors originate from a small (0.25 to 0.5 mm) group of cells in the adventitia of the jugular bulb. The glomus jugularis, microscopically, consists of blood vessels of capillary and precapillary caliber and clumps of epitheloid cells which are richly supplied by neural elements from the glossofaryngeal nerve. Similar structures are found associated with the ramus tympanicus of the glossofaryngeal nerve on the cochlear prominence and associated with the ganglion nodosum of the vagus nerve. Most workers agree that tumors arising from these paraganglia are neoplastic adenomas. Although these tumors are usually slow in growing, some of the lesions enlarge rapidly and sometimes metastasize. Some otologists have concluded that the glomus tumor arising at the dome of the jugular bulb is
much more destructive and fast growing than that arising on the cochlear promontory.\textsuperscript{8,18}

Classification of these lesions by location is needed. The small tumor of the middle ear, presenting as a bulging reddish-blue mass seen through the tympanic membrane, is clearly a problem for the otologist. This lesion usually presents symptoms of tinnitus, conductive hearing loss, facial weakness, or bleeding from the ear; it can be totally removed through a transmeatal approach.

Quite different is the very rare tumor arising at the jugular foramen and protruding intracranially. This mass presents symptoms of lower cranial nerve palsies, bruit, and increased intracranial pressure. Neurosurgeons have successfully removed this tumor through a posterior fossa approach\textsuperscript{10} but extension into the temporal bone may be difficult to deal with by that approach.\textsuperscript{3}

Unfortunately, between these two extremes of the spectrum there is a sizable collection of tumors associated with marked destruction of the mastoid process and adjacent structures of the temporal bone. Patients with this type of tumor may present any combination of the aforementioned symptoms. Treatment of these lesions has traditionally been subtotal removal and irradiation. This is the group of patients to whom we would like to call attention.

**Diagnosis**

Radiography, especially some of its newer techniques, is important in the diagnosis of the extent as well as the origin of these tumors. Clouding or erosion of the mastoid process or petrous pyramid can usually be appreciated on plain x-rays of the skull. The anteroposterior or Towne projection of the skull may illustrate bone destruction in the petrous pyramid. A submental vertex view may be helpful in delineating the limits of the bone changes. Mastoid views often elucidate the abnormality. Tomography of the temporal bone is also of great aid in determining the extent of the lesion.

Subtraction angiography, particularly when used in conjunction with selective internal and external carotid and vertebral injections, is absolutely essential in planning an appropriate operative treatment. The selective external carotid arteriogram in Fig. 1 \emph{left} illustrates the significant contribution to the blood supply of these tumors made by branches of this vessel. The subtraction view of the same arteriogram (Fig. 1 \emph{right}) illustrates the feeding vessels much better. It has been our experience that the most significant blood supply to these tumors is via the external carotid and ascending pharyngeal vessels. Subtraction views are important in the capillary as well as arterial phases since the petrous bone can hide the vascular blush indicating the size of the tumor. The extent of the lesion can also be recognized in the anteroposterior views (Fig. 2). Branches of the vertebral artery also contribute blood to these tumors (Fig. 3). Retrograde jugular venography is useful in demonstrating the area of obstruction of the jugular bulb. Sinography of the superior sagittal sinus has
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Fig. 2. Capillary phase of a selective external carotid arteriogram using the subtraction technique to delineate the extent of the lesion.

Fig. 3. Selective vertebral arteriogram illustrating the contribution to the blood supply from this vessel.

helped us delineate the superior level of the obstruction within the sigmoid sinus.

Operative Technique

A reverse “question-mark” incision is made around the ear (Fig. 4 left). The inferior extent of the incision crosses the sternocleidomastoid muscle. The reason for this inferior extension of the incision is so that the external carotid artery and ascending pharyngeal artery can be ligated in the neck prior to the direct attack upon the tumor (Fig. 4 right). The superior extent of the incision is then continued down to the bone.

Fig. 4. Drawings illustrating operative technique. Left: Skin incision. Right: Ligation of the external carotid artery.
The bone just above and behind the ear is exposed by subperiosteal reflection (Fig. 5 left). In cases where the middle ear has been filled with tumor, the external meatus is divided and the skin of the external canal closed.

Multiple burr holes are placed above and below the level of the lateral sinus and the craniectomy is completed with a rongeur. It is important to expose the junction of the superior petrosal and lateral sinuses where they form the sigmoid sinus. When possible the sigmoid sinus proximal to this point is ligated rather than both the superior petrosal and lateral sinuses. To make this surgical judgment, preoperative sinography is of great help. The craniectomy is continued anteriorly and inferiorly into the petrous bone until the tumor is recognized (Fig. 5 right). A high-speed drill can then be used to remove additional bone as the tumor and the sigmoid sinus are followed inferiorly. Great

Fig. 5. Left: Exposure of the bone. Right: Extent of craniectomy with exposure of the tumor invading the temporal bone.

Fig. 6. Left: Ligation of the sigmoid sinus. Right: Ligation of the internal jugular vein and exposure of the entire posterolateral surface of the tumor.
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care is necessary to avoid injuring the facial nerve which is always exposed in the course of this bone removal. Usually the patients are deaf on the side of the lesion; if not, removal of the bone medially can open the semicircular canals and cochlea, resulting in deafness. The bone is usually removed from the internal auditory meatus medially to the jugular foramen inferiorly.

Small dural flaps are elevated anteriorly and posteriorly to the sinus just above the tumor (Fig. 6 left). Since these dural incisions flank the sigmoid rather than the transverse sinus, they both lie below the tentorium. The sinus is ligated with 00 silk and opened longitudinally below the ligature. Additional bone is removed as necessary. No tumor should be left within the mastoid air cells. The internal jugular vein is followed inferiorly through its exit from the skull. The vein then passes just in front of the transverse process of C-1; this transverse process is exposed subperiosteally and removed. The relationship of the vertebral artery to this bone landmark must be kept in mind.

The jugular vein is ligated at this level (Fig. 6 right). In doing this, care must be taken to avoid injuring the seventh, ninth, tenth, and twelfth nerves and the internal carotid artery. Magnification is helpful in preserving the facial nerve within the temporal bone. The vein is opened longitudinally above the level of the ligature, and tumor removal is completed. The entire vein may be reflected superiorly, as the intima of the vein will usually delineate the tumor margin. Often it is necessary to excise the entire jugular bulb as well as its surrounding dura. The tumor is dissected from the petrous pyramid and excised en bloc. There is a cavity within the petrous pyramid after the tumor has been excised. Obviously in this situation a dural graft is needed to close this sizable defect. In our experience the isobutyl cyanoacrylate tissue adhesive has been extremely helpful in bonding the margin of the graft medially along the clivus where the dura is always very thinned out. Laterally the dural graft can be sutured in place.

Extensive tumors have been followed medially to the cavernous sinus. When a large tumor has been removed from the petrous pyramid, the dead space can be obliterated by use of sternocleidomastoid muscle. Often a large section of this muscle can be rotated into the cavity with its blood supply still attached. The wound is then approximated in layers without drainage.

Conclusions

Because the glomus jugulare tumor is uncommon, our experience remains limited. Based on the surgical management of seven glomus tumors, however, we have reached the following conclusions:

1. This tumor is accessible to complete neurosurgical removal.
2. Bleeding from the lesion can be controlled with proximal ligation of the external carotid and ascending pharyngeal arteries.
3. Small lesions can readily be excised from within the lumen of the jugular bulb.
4. In removing larger tumors, the facial nerve, semicircular canals, cochlea, and internal carotid and vertebral arteries are structures of which one must be cognizant and which can be avoided.

References

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