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GLOMUS TUMORS —— A REVIEW OF 8 CASES WITH INTRODUCTION OF A MODIFIED COMBINED INTRA-AND EXTRACRANIAL APPROACH

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Eight cases of glomus tumors hospitalized in our department from 1982 through 1995 were reviewed. It comprised of five glomus tympanic tumors and three glomus jugular tumors. Discussion was centered on it's contemporary diagnosis, classification and surgical treatment with introduction of a modified combined intra-and extracranial surgical approach to resect the extensive glomus tumors.

Key words: Glomus tumor, Surgical approach.

Glomus tumors are also referred to as chemodectomas or nonchromaffin paragangliomas. Although benign histologically, glomus tumors can grow aggressively and , by nature of their location can cause multiple cranial nerve abnormalities, displace or erode ossicles and even the bony carotid canal, damage inner ear structures, or extend into the posterior cranial fossa.[1] Glomus tumors have so far posed a diagnostic and therapeutic challenge to otolaryngologists and neurosurgeons. We think it interesting to present our experience with 8 cases of glomus tumors admitted from 1982 to 1995.

MATERIALS AND METHODS

Clinical Features

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The clinical features of eight cases with pertinent signs and symptoms were summarized in Table 1. Both sex were equally distributed. The age ranged from 16 to 52 years with a mean of 33 years. The average duration of symptoms at the time of referral was 7.3 years (ranging from 0.25 to 20 years). Seven of the eight cases had pulsatile tinnitus as well as hearing loss (5 cases with conductive hearing loss, 2 cases with mixed hearing loss). Six patients complained of intermittent aural bleeding with estimated blood loss of 2 ml to 20 ml. Two cases presented with facial nerve palsies and three with IXth 、 Xth 、 XIth 、 XIIth cranial nerve palsies.

Radiographic Features

Of four cases with plain X-ray films of jugular foramen, only two showed dilated jugular foremen. Tumor size and extent were determined from preoperative contrast-enhanced computerized tomography (CT) with bone windows, magnetic resonance (MR) imaging, and digital subtraction angiography (DSA) if available as shown in Table 1. Of five cases evaluated with CT, two were diagnosed as glomus tumors, one as malignant tumors of middle ear involving the inner auditory canal, and extending into middle and the posterior cranial fossa, one as neoplasm of uncertain nature and one as normal. All the four cases with preoperative MR were diagnosed as glomus tumors. Preoperative DSA and selective

intraarterial embolo-zation therewith were performed in two cases, where the highly vascular space occupying lesions were shown to be lobular and situated at the jugular foramen with their blood supply deriving from the ascending pharyngeal, internal maxillary and vertebral arteries.

Table 1. Clinical features at referral of 8 cases with glomus tumor

Case No.	Age (yr.) sex	Duration of symptoms (yr.)	Tinnitus	Hearing loss	Aural bleed	VIIth palsy	IXth–Xth palsy	X-ray	CT	MR	DSA	Operative approaches
1	48M	13	yes	yes	yes	yes	yes		anor	anor	anor	combined
2	36F	20	yes	yes	yes	no	no	nor		anor		tympanotomy
3	14F	0.5	no	no	no	no	yes		anor			Shapiro
4	26M	1	yes	yes	yes	no	no	anor				tympanotomy
5	46F	13	yes	yes	yes	no	no	nor	nor			no
6	52M	10	yes	yes	yes	yes	no					mastoidetomy
7	16M	0.5	yes	yes	no	no	yes		anor	anor	anor	combined
8	26F	0.25	yes	yes	yes	no	no	anor	anor	anor		mastoidetomy

nor: normal; anor: abnormal

Preoperative Diagnosis

The presumptive diagnoses at first visit in our clinic were glomus tumors in six cases, granular tissues of external auditory canal (possibly due to otitis midia or external) in one case, and facial neurilemoma of middle ear in another case. Among four cases preoperatively transmeatally biopesied, only one was confirmed histopathologically as glomus tumor, in the remaining three cases only blood clot and/or granular tissues were revealed.

Classification

The classification have been proposed by other authors.[2,3] Four types of glomus tumor were categorized based on the information provided by the imaging study regarding their size and different anatomical involvements in our cases, i.e.:

I: Tumor limited to the tympanic space

II: Tumor not only filling the tympanic space, but also extending into the external auditory canal, attic and mastoid and involving the facial nerve

III: Tumor locating at the jugular foramen and extending into the middle ear and external auditory canal

IV: Tumor originating from the jugular foramen but extending massively into both middle and posterior cranial fossa including the internal auditory canal, petrous apex, cavernous sinus, clivus etc.

Surgical Approaches

Surgical treatments were performed on all cases except one who refused resolutely to accept the operation. Depending on the tumor classification, different surgical approaches were adopted. Tumors were removed via tympanotomy in two cases who were classified as having type I and via radical mastoidectomy in another two cases who were classified as having type II. One type III tumor was operated on using Shapiro approach. For the remaining two cases with type IV tumor, we used a modified combined intra-and extracranial approach, of which a brief description is given as follows:

Under general anesthesia, a long Y-shaped pre- and postauricular incision was made (Figure 1), so as to transect the external auditory meatus and create a big triangular superiorly basedflap with full exposure of the temporal as well as the upper cervical area (Figure 2). The sterno-cleida-mastoid muscle was freed and retracted inferiority and posteriorly and divided at the level of mastoid tip. As a result such structures as diagastric muscles, jugular vein, internal and external carotid arteries and IX、X、XI、XII cranial nerves could be clearly identified and the external carotid artery could be easily ligated. Parotidectomy was done after raising of the parotid

flap and identification of the facial nerve at the stylomastoid foramen. The next step was to complete a mastoidectomy with removal of the bony external auditory canal, tympanic membrance, and middle ear contents if there is any. The facial nerve was then mobilized out of its fallopian canal and translocated anteriorly. Sigmoid sinus bony plate was extensively drilled away to fully expose the sinus and to put a ligature on its proximal portion. The internal jugular vein was also ligated from the neck region. Petrosal segment of the internal carotid artery was intentionally exposed to avoid damage. In order to achieve better exposure of the tumor mass that concealed itself within the tympanum, the floor of bony external auditory canal as well as the tympanum were adequately removed with drill. Now we turned to the temporal craniotomy which was routinely done by taking out a rather quadrilateral bony flap after elevation of the temporal muscle. While elevating and pushing away the dura of the middle cranial fossa anteromedially, we were able to identify and ligate the superior petrosal sinus so as to greatly facilitate the resection of the lateral third of the petrosal bone, thus allowing access to the entire petrous apex. The petrous segment of the internal carotidartery was further uncovered proximally up to its entrance into the petrous bone and distally into the posteroinferior aspect of the cavernous sinus. At this point the tumor could be worked on by a combined dissection from both above in the tympanum and below in the jugular foramen area using the standard microsurgical technique. An enbloc resection of the tumor could be achieved as long as the internal carotid artery had not been invaded. However, resection and replacement with vascular graft of the internal carotid artery which has been occluded by tumor requires a second stage operation. The transposed facial nerve could be easily relocated and the dural defect was repaired with temporal fascial graft. The incision was sutured in layers with insertion of a suction drain. The most common post operative complication was facial weakness secondary to the facial nerve manipulation.

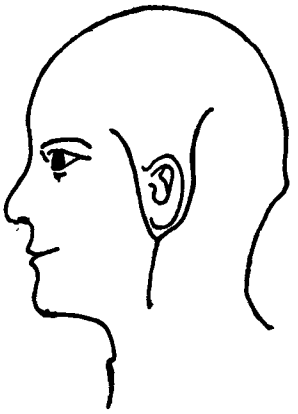

Fig. 1. Y-shaped incision

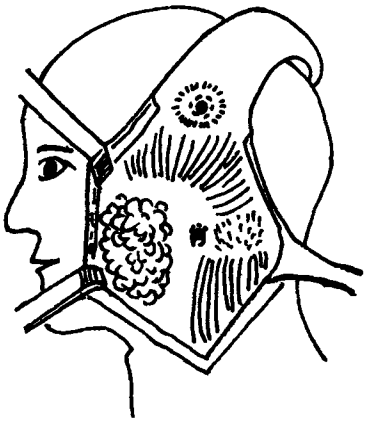

Fig. 2. Full exposure of the temporal and the upper cervicalarea

RESULTS

Complete microscopic resection was obtained in all of the seven cases. The immediate postoperative complications included aspiration pneumonia in two cases which was rapidly controlled with antibiotic therapy, otorrhea in all seven eases which was completely resolved one month after operation, and CSF otorrhea in two other cases that persisted until three months later. The use of preoperative antibiotic prophylaxis helped to prevent the infection. None of the cases develped any wound infection nor meningitis.

At a median follow-up period of five years, none of the operated seven cases has had a recurrence or progression of the disease. The only case who was not operated on was died one year later. Two cases who required mobilization and anterior translocation

of the facial nerve developed transient postoperative facial nerve palsy, but the facial function returned to normal six months after operation. The sequelae with VIIth nerve palsy in two cases and with IXth、Xth、XIth、XIIth cranial nerve palsies in three other cases remain unchanged so far.

DISCUSSION

Glomus tumor which arises from chemoreceptor bodies in the wall of the jugular bulb and along the Jacobson's nerve is a vascular tumor. Based on its point of origin and the extent of its involvement in the temporal bone, the tumor is classified into two types; i.e. glomus tympanic tumor originating from the chemoreceptor body along the Jacobson's nerve and glomus jugular tumor originating from the wall of the jugular bulb. Although histologically benign, glomus tumors are well known for their aggressive behavior. Both types may involve external auditory canal, attic and mastoid and cause impairment of the facial nerve. Besides the glomus jugular tumor may further involve petrous apex, cavernous sinus anteriorly invade internal auditory canal posteriorly and even extend deep to the clivus and foramen magnum resulting in multiplecranial nerve deficits.[4–6] As mentioned above, there are five glomus tympanum and three glomus jugular tumors in our series.

For those with typical and full-klown symptoms and signs, it is usually not difficult to make the diagnosis clinically. However, when a patient of glomus tympanic tumors presents only with intermittent or persistent pulsatile tinnitus, mild hearing loss and some granulation tissue-like mass in the external auditory canal with little bleeding, it may be misdiagnosed as granulation tissue resulting from otitis media or/and external, cholesterol granuloma, malignant tumor of the middle ear and idiopathic hemotympanum etc. In our series, seven patients who complained pulsatile tinnitus and hearing loss at their initial visits were all so misdiagnosed. Therefore, one should be adequately cautious whenever facing such a case. In the presence of unilateral characteristic tinnitus, pressure with a pneumatic otoscope may accentuate the pulsation and vigorous pressure can cause the pulsation to cease. This procedure may provide some hints to the diagnosis. On the other side, patient with glomus jugular tumors who presents with multiple (9–12th) cranial nerve palsies accompanied with or without tunnitus and deafness may be easily misdiagnosed as suffering from the tumors of cerebellopontine angle such as acoustic neurilemoma, cholesteatoma etc. Under such circumstances in addition to careful history taking, physical examination, audiological study and vestibular function test etc., the updated imaging studies including CT and MR have been shown to be very helpful in making the diagnosis. Moreover MR is especially helpful in determing the tumor size and extent of involvement and allows the surgeon to select the most appropriate surgical approach. In the four cases who had been MR imaged preoperatively, the diagnoses suggested and their sizes and the extent of involvement determined by the MR were all confirmed by the operative findings as well as the post operative histopathology. In contrast, the accurate rate of diagnosis for CT was rather lower in our experience. Only in two of the five cases scanned with CT the diagnosis of glomus tumors were suggested. Preoperative DSA can accurately determine the bloodfeeders of the tumor with simultaneous intraarterial embolization, thus remarkable decreasing the intraoperative blood loss.

Operative resection has undeniably had an important role in the treatment of glomus tumors. Depending on the size and involving extent of the tumor, classification had been proposed and so have been the various surgical approaches. As stated above the classification we proposed was based on the precise information mainly provided by the updated imaging studies regarding the size and the regions of involvement, we feel that such classification seems more helpful in deciding the surgical strategy and the different approaches for different case, and more closely reflect the degree of difficulty involved in the tumor resection. For instance, type I tumor can be removed with ease by tympanotomy, type II tumor by radical mastoidectomy, the removal of type III tumor requires Shapiro approach, while type IV tumor can be en bloc resected only via a modified combined intra-and-extra cranial approach. With the recent advances in diagnostic and interventional radiology, skull-base surgery and microsurgical techniques, vascular reconstruction and anastomosis ect., there seems no major obstacle any longer that can not be overcome in this type of surgery. The goal of the surgery is to render the case disease-free while minimizing the permanent disability related to the injury of the cranial nerves, brain stem, and major arteries. In order to

achieve this goal, it is equally important to make close collaboration with neurosurgeon and anesthesiologist and to set up an excellent team work among them, just as we have successfully done in our cases. The operative mortality in our small series is zero and the incidence of morbidity is low with the most common postoperative complication being the temporary facial weakness secondary to facial nerve manipulation.

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