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Short Communication

Summary and consensus in 7th International Conference on acoustic neuroma: An update for the management of sporadic acoustic neuromas

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Abstract Sporadic vestibular schwannoma (acoustic neuroma) is a benign tumor arising from cochleovestibular nerve. Nowadays, various specialties and medical centers are treating this disease, and the multidisciplinary collaboration is the trend. In an effort to promote a uniform standard for reporting clinical results, even for treatment indications, the mainly controversies were posed and discussed during the 7th International Conference on acoustic neuroma, and the agreement was summarized by the Committee of this conference. The main symptoms grading and tumor stage should note its name of classification for making them comparable. The goal of the modern managements for vestibular schwannoma is to improve the quality of life with lower mortality, lower morbidity and better neurological function preservation. The experience of surgical team and their preference might be a major factor for the outcome. Because of lacking of long-term follow-up large data after radiotherapy, and with the development of microsurgery, radiotherapy is now less recommended except for recurrent cases or elderly patients.

Introduction

The 7th International Conference on acoustic neuroma was held on April 12—15, 2015 in Shanghai, China. This series conference, where gathers the outstanding experts worldwide, is the most remarkable meeting in the field of acoustic neuroma. The 7th conference was co-hosted by the Xinhua Hospital, People's Liberation Army General Hospital, Tiantan Hospital, and Huashan Hospital. There were more than 700 participants, including 345 foreign attendees from 41 countries, composed of neurosurgeons, neurotologists, radiotherapists, neuro-radiologists, audiologists, plastic surgeons, and basic researchers. After several multidisciplinary discussions, some ancient controversies reached an agreement, and this consensus summarized by the committee of this conference.

Nomenclature

Acoustic neuroma (AN) is also known as vestibular schwannoma, since this benign tumor almost originates from superior or inferior vestibular branch of the cochleovestibular nerve in the internal auditory canal (IAC).[1] Moreover, the tumor is schwannoma in pathology rather than neuroma. The two nomenclatures are both accepted, however, vesctibular schwannoma (VS) is preferable.

Sporadic Vestibular Schwannoma is basically distinct from Neurofibromatosis type 2 (NF2). If it is not specifically noted, VS refers to the sporadic vestibular schwannoma in the context.

Cystic vestibular schwannnoma (CVS) should be distinguish from solid vestibular schwannoma (SVS) because of the their variant clinical, radiological, histopathological features and surgical outcomes.[2—5] CVS can be peripherally located thin-walled tumors, and centrally located thick-walled tumors based on CT or MRI images. CVS frequently presents rapid progression of symptoms with facial nerve involvement.

Standardization of main symptoms grading

The purpose of standardizing the grading of the main symptoms is to unify the description of patients' status, and then to make analyzing management strategy and outcome more precisely. Classically, the AAO-HNS Hearing Classification System,[6] House-Brackmann Facial Nerve Grading System,[7] Tinnitus Handicap Inventory[8] and Dizziness Handicap Inventory[9] are widely accepted and used for VS. But the two latters are in the form of questionnaire which is more complicated, this consensus attempts to classify them into four grades as alternatives (Tables 1 and 2).

Tumor size and stages

Several stage grading systems have been reported according to tumor size.[10—13] Generally, the tumor size should be measured on MRI images, and the maximum diameter (also called tumor diameter) means the one measured in cerebellopontine angle (CPA) along the long axis of tumor. The type of tumor within the IAC should be classified separately. Four commonly used tumor grading are Sterkers classification, House classification, Koos classification and Samii classification (Fig. 1).

Table 1 Tinnitus grading system for acoustic neuromas.

Grade	Descriptions
I	No tinnitus
II	Intermittent or mild tinnitus, can only be heard when the ambient noise is low
III	Persistent or moderate tinnitus, can be heard every day
IV	Persistent and severe tinnitus, interfere with work and sleep

Table 2 Dizziness grading system for acoustic neuromas.

Grade	Descriptions
I	No dizziness or imbalance
II	Occasional and mild dizziness or imbalance
III	Persistent or moderate vertigo or imbalance
IV	Persistent and severe dizziness or imbalance, disturbing daily life

<table>
<tr><th>Tumor size (CPA maximum diameter)</th><th>Sterkers</th><th>House</th><th>Koos</th><th>Samii</th><th>Tumor Description</th></tr>
<tr><td>0 (Intracanalicular)</td><td>Tube type</td><td>Intracanalicular</td><td>Grade Ⅰ</td><td>T1</td><td>Confining to IAC</td></tr>
<tr><td>≤10 mm</td><td rowspan="2">Small</td><td>Grade 1 (Small)</td><td rowspan="3">Grade Ⅱ</td><td>T2</td><td>Surpassing IAC</td></tr>
<tr><td>≤15 mm</td><td rowspan="2">Grade 2 (Medium)</td><td rowspan="2">T3a</td><td rowspan="2">Tumor occupying CPA</td></tr>
<tr><td>≤20 mm</td><td rowspan="2">Mild</td></tr>
<tr><td>≤30 mm</td><td>Grade 3 (Moderately Large)</td><td>Grade Ⅲ</td><td>T3b</td><td>Tumor occupying CPA and contacting the brain stem without compression</td></tr>
<tr><td>≤40 mm</td><td>Large</td><td>Grade 4 (Large)</td><td rowspan="2">Grade Ⅳ</td><td>T4a</td><td>Tumor compressing the brain stem</td></tr>
<tr><td>>40 mm</td><td>Huge</td><td>Grade 5 (Giant)</td><td>T4b</td><td>Severe brain stem displacement and deformation of the fourth ventricle under tumor compression</td></tr>
</table>

Fig. 1 Main grading systems for acoustic neuromas. The classifications on the left side (blue area) are mainly based on tumor size, while those on the right side (green area) are based on the anatomical relationship around the tumor. Koos classification combines the tumor size and anatomical relationship for larger tumors.

As an alternative, rather than using a particular staging systems mentioned above, indicative of intracanalicular type (with size in millimeters) and simple reference to tumor size in the CPA in 10 mm increments might simplify the VS grading and render more consistent tumor size reporting from all centers.

Management goal and strategy

The goal of the modern managements for VS is to improve the quality of life with lower mortality, lower morbidity and better neurological function preservation. VS management is no longer simply limited to surgical resection. The conception of "wait and scan" is accepted worldwide, especially for the small, primary and sporadic tumor.[14–17] Radiotherapy is accepted as mainstream method for the patients with surgical contra-indication.[18,19] The management strategy becomes individual, which mainly depends on the radiological features (cystic or not, tumor size and extension), the biologic feature (tumor growth rate), the severity of symptoms (ipsilateral and contralateral hearing, facial function, other complications), the patient's age, and the general situation and expectations.[17,20–26]

Small tumors with useful hearing

Several long term follow-up studies have found that, during the follow-up of small tumor, the possibility of hearing loss after 5 years was about 70% whether a tumor was increased or not.[27–29] For the small tumor, the hearing preservation probability after surgery is about 60% in experienced institutes if the fundus of IAC free of tumor involvement.[15,30–35] Attending experts in the 7th conference agreed that for younger patients, with grade B or grade A hearing level and free IAC fundus, surgical intervention can be considered earlier. However, for small tumors involving the IAC fundus, the hearing preservation rate is lower than 50% even in experienced institute.[36] Therefore, this consensus proposes that a planned follow-up should be the first choice under this circumstance in consideration of quality of life. However, surgical procedure is reasonable to perform in those patients who are well-informed and willing to take the risk of surgery regarding the situation and options for management.

Small tumors with refractory vertigo or imbalance

In such a situation, regular treatment and observation must go through for 6 months and whether the quality of life of such patient is affected by vertigo or imbalance is determined. If vertigo cannot be alleviated in the short term, surgical intervention should be taken into account.

Small tumors without useful hearing in young people

It has been reported that VS grows slowly. According to a large number of reports, facial paralysis rate after surgery for small tumors with no useful hearing in young people was 10%.[37–41] Therefore, young patients who might be expected to live 25—30 years (or longer) with slow growing tumor and without hearing might be recommended to be followed for at least one year. However, surgical procedure is reasonable to perform in those patients who are well-informed and willing to take the risk of surgery regarding their situation and options for management.

CVS

VS with cystic degeneration or cystic degeneration appears during follow-up often means rapid tumor growth.[15,42,43]

Moreover, this type of VS is less sensitive to radiotherapy has been documented.[44,45] Therefore, the optimal choice of treatment is surgery for these patients and should be performed as soon as possible.

Difference among medical centers

More important, not all centers where manage VS patients achieve comparable results in terms of surgical treatment. The experience of surgical team and their preference might be a major factor. The ratio of post-operative hearing preservation for small tumors can be apparent various among centers, also for the relationship between hearing preservation and internal auditory canal (IAC) fundal involvement. Management strategy should be specific for a patient with VS in any center, in this way the patient could make an informed decision in their particular situation.

Surgical approaches

There are three main surgical approaches, including translabyrinthine approach, retrosigmoid approach, and middle fossa approach. The selection of approach should defer to surgeon's preference and experience. It is generally accepted that the retrosigmoid approach is recommended when hearing preservation is considered. Nowadays, in virtue of endoscopic technique and advanced surgical experience, the extent or/and IAC fundus involvement are no longer the opposition for this approach because some centers achieve excellent hearing outcomes in patients with small tumors with fundus involvement.[46] The translabyrinthine, or enlarged translabyrinthine,[47,48] or modified translabyrinthine approach, as well as in combination with a retrosigmoid exposure,[49] is appropriate to removal of VSs for any size. Because of the endoscope assisted technique in the retrosigmoid approach, the middle fossa approach becomes less selected for hearing preservation, however, this approach is still the main approach for moderate or small tumor in some centers with excellent outcome.[32,50]

Evaluation of tumor resection

Tumor resection only includes total resection, near total resection, subtotal resection, and partial resection. Total resection means no tumor residue. Near-total resection (NTR) was assigned when a small piece of tumor remnant (size was no greater than 25 mm^2 and 2 mm thick, and could not be detected by routine MRI) was intentionally left *in situ* in an effort to preserve neural integrity. Subtotal resection (STR) was used to describe any situation where less than NTR was performed.[51] Partial resection (PR) was defined and used a percentage of the original tumor when Tumor residues greater than $>5\%$.[52]

The size of the residual tumor is indicated by the vertical diameter of each other. Meanwhile, the location of the residual tumor should be documented, for example, residue in IAC, in CPA, on brainstem surface, or on cerebellar surface, etc.

Evaluation and follow-up after radiotherapy

Long-term follow-up is mandatory after tumor radiotherapy which just controls tumor growth. So far, there is a lack of long-term follow-up large data. It has been documented that in longer term follow up after fractionated stereotactic radiotherapy, 30% of tumors continued to grow (defined as at least 15% increase in tumor volume).[53] Furthermore, either the tumor did or did not increase in size after the typical 18 month time frame after radiation in which tumor edema may have occurred.[54,55] Thus, for the young people with VS, radiotherapy is not recommended. Radiotherapy is applied to recurrent cases after surgery or elderly patients. The grade of radiotherapy outcome is shown in Table 3.

Table 3 Radiotherapy outcome for acoustic neuromas.

Grade	Description
1	Tumor control, tumor diameter is reduced by more than 2 mm, and the volume is reduced by more than 10%
2	Tumor stability, tumor diameter reduction is less than 2 mm, and the volume reduction is less than 10%
3	Tumor growth, the tumor does not shrink or tumor size re-increases after shrinking

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