Bilateral Acoustic Tumors: Its Clinical Features and Surgical Treatment

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Summary

The authors have experienced 18 cases of bilateral acoustic tumors. They were found to occur most often in relatively young subjects and often to be associated with von Recklinghausen's disease. In spite of the existence of bilateral tumors, the patients frequently retained unilateral auditory function. Visual disturbance due to secondary optic atrophy was observed occasionally.

The mortality rate of operation was fairly high. The cause of death was mostly brainstem damage induced by simultaneous removal of large bilateral tumors. In many of the surviving patients, auditory function was sacrificed in spite of fair postoperative course.

The most advisable stratagem for surgery is thought to be as follows.
1) Bilateral large tumors should be submitted to surgery in two sessions.
2) The maintenance of residual hearing function should be considered superior to radical removal of tumors.
3) Decompressive craniectomy alone is not effective

Key words: Bilateral acoustic tumors, clinical features, surgical management

Among 369 cases of acoustic tumors admitted to the Department of Neurosurgery, University of Tokyo, 18 cases (4.9%) had bilateral tumors. These patients were frequently associated with von Recklinghausen's disease and had characteristic clinical features that were different from unilateral ones. Surgical treatment of such patients is difficult because of the postoperative morbidity, including total deafness, loss of labyrinthine function and/or bilateral facial paralysis. When bilateral tumors are large and compress the brain stem from both sides, postoperative death due to brain stem damage may ensue. We present the case histories of 5 patients in this group that will illustrate the characteristic clinical features and the difficult problems involved in surgical management.

Report of Cases

Pertinent clinical features in these 18 cases are tabulated in Tables 1 and 2. Operations were performed in 15 out of 18 cases using a suboccipital transmeatal approach except for one case in which operation was carried out via the middle fossa route (Table 3). Bilateral total removal at one session was carried out in 7 cases. In 4 cases, suboccipital decompression

Table 1 Initial symptoms of bilateral acoustic tumors

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>14</td>
</tr>
<tr>
<td>Headache</td>
<td>3</td>
</tr>
<tr>
<td>Vertigo</td>
<td>2</td>
</tr>
<tr>
<td>Gait disturbance</td>
<td>2</td>
</tr>
<tr>
<td>Visual disturbance</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2 Neurology on admission

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>18</td>
</tr>
<tr>
<td>(Bilat. deaf)</td>
<td>(8)</td>
</tr>
<tr>
<td>Choked disc</td>
<td>14</td>
</tr>
<tr>
<td>(Visual disturbance)</td>
<td>(12)</td>
</tr>
<tr>
<td>Cerebellar sign</td>
<td>14</td>
</tr>
<tr>
<td>(Bilat. involvement)</td>
<td>(12)</td>
</tr>
<tr>
<td>Other cranial nerves sign</td>
<td>16</td>
</tr>
<tr>
<td>(Bilat. involvement)</td>
<td>(10)</td>
</tr>
</tbody>
</table>
with or without shunting was performed. Recently, a two-stage procedure was employed in 2 cases and tumors were unilaterally extirpated in another 2 cases.

Postoperative mortality was experienced in 4 cases, due to brainstem damage in 2 cases and associated complications in the remaining 2 cases.

**Case 1**
A 25-year-old female was admitted with the complaints of hearing loss, staggering gait and swallowing difficulty for two years’ duration. Neurological and clinical diagnosis of bilateral acoustic tumors was made. Bilateral cerebellopontine angle exploration at one operation was carried out. An egg-sized tumor on the left side and a small one of 1 cm in diameter on the right side were totally extirpated. She withstood the operation well and postoperatively had no facial weakness, although her hearing function was completely lost.

**Case 2**
When a 33-year-old female was admitted, she was totally deaf and blind. She had large acoustic tumors on both sides and they were radically extirpated at one operation. Postoperatively tracheostomy was performed and recovery of consciousness was slow, although, for the next few days, her consciousness was clear. On the fifth postoperative day, she bled from the gastro-intestinal tract and died in a state of shock.

**Case 3**
A 30-year-old female with neurofibromatosis had, in addition to bilateral acoustic tumors, spinal cord tumors in the cervical and thoracic regions. The right-sided acoustic tumor was removed first because it was the side with the lessened hearing acuity. After about four weeks, spinal cord tumors were extirpated. Postoperatively her general course was good, although right facial palsy and bilateral motor weakness of the limbs remained.

**Case 4**
A 23-year-old male was one of twins with von Recklinghausen’s disease. On admission, he had bilateral choked disc, cerebellar ataxia and bilateral palsy of the 5th to 8th cranial nerves. CT scan examination revealed bilateral giant tumors in the cerebellopontine angles. As the first procedure, suboccipital decompressive craniectomy with V-P shunt was performed, but his general condition worsened and he became bed-ridden. As apneic attacks began to occur several times, it was decided to operate on the tumors directly.

Both tumors were excised subcapsularly in two sessions. Postoperatively the patient’s respiratory state became stable, but his general condition remained unchanged.

**Case 5**
A 19-year-old female with neurofibromatosis was admitted with complaints of hearing loss predominant on the left side and ataxic gait. Neuroradiological evaluation with CT scan
showed extremely large tumors occupying both the cerebellopontine angles (Fig. 1). The huge tumor on the left side was removed in two sessions separated by an interval of three weeks. A part of the tumor was resected by a suboccipital approach and the remaining portion was totally removed via the middle fossa route. After two months, the right-sided tumor was partially removed.

One year later, as the tumor on the right side had grown to a huge size, subcapsular removal was performed via the middle fossa route (Fig. 2). Postoperatively she was generally well, her hearing function remained and she returned to a normal social life.

Discussion

Clinical features
The average age of the patients with bilateral acoustic tumors was 27.6±8.8 (mean±SD) years on admission. This is in marked contrast with unilateral acoustic tumors which usually arise in middle age.6,8)

The typical stigmata of von Recklinghausen's disease, such as café au lait spots or neurofibromatosis,1,6,8) were observed in 15 cases (83.3%). Sixteen patients had associated lesions of the central nervous system, which consisted of spinal cord tumors in 9 cases, neurinomatosis of the lower cranial nerves in 4 cases, meningiomas in 2 cases and a cerebral arteriovenous malformation in one case.

Hereditary predisposition is also one of the characteristic features of bilateral acoustic tumors.2,3,6,7,8) The classic report of Gardner and Frazier,3) in which a family of five generations with bilateral deafness in 38 members, is well known. Moyes7) also reported familial bilateral acoustic tumors affecting 14 members from four generations. In our cases, hereditary factor was noted in 6 cases and 5 of them were associated with von Recklinghausen's disease.

Among 18 cases, only 8 cases had bilateral hearing loss. Auditory function in at least one ear was kept almost normal in one third of the cases (Fig. 3).
Visual disturbance was not infrequently observed and some patients became blind due to secondary optic atrophy.

Clinical features of bilateral acoustic tumors can be summarized as follows: 1) onset at a relatively young age; 2) often associated with von Recklinghausen's disease; 3) familial predisposition; 4) unilateral hearing often retained; and 5) visual disturbance occasionally.

Surgical treatment
Eleven of 15 surgical cases were submitted to surgery before 1975 and were operated on without the present stratagem of surgical management. Among these 11 cases, bilateral tumors were totally excised in one session in 7 cases. When bilateral tumors were huge and compressed the brainstem from both sides, the postoperative course was extremely poor. The cause of death was brainstem damage due to surgical maneuvers.

When one or both of bilateral tumors were small and did not compress the brainstem, the postoperative course was uneventful. However, 4 of these 5 cases lost residual hearing function.

In 4 cases, decompressive craniectomy was performed. Postoperatively 2 patients died because of associated complications and the other 2 patients showed no neurological improvement.

Since 1976, we have performed the operation in two sessions when bilateral tumors are large and compress the brainstem, and better results have been obtained.

Furthermore we have taken care to maintain the patients' residual hearing function. In this respect, subcapsular removal or only unilateral resection on the side without residual hearing were employed on some occasions.

Our present stratagem for surgery of bilateral acoustic tumors is as follows:
1) Bilateral large tumors should be submitted to surgery in two sessions.
2) The maintenance of residual hearing function should be considered important. Subcapsular removal or unilateral removal may be advisable in some cases.
3) Decompressive craniectomy alone is not effective.

Acknowledgments
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References
1) Davidoff, L. M. and Martin, J.: Hereditary


