RESULTS

Table 1. Patient demographics and treatment complications.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs) and year of Dx of Medulloblastoma</th>
<th>Non-Otologic complications</th>
<th>Pre-CI HA use (yrs)</th>
<th>Pre-CI otologic procedures</th>
<th>Age at CI</th>
<th>Device</th>
<th>Post-CI complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>8 (1989)</td>
<td>CVA</td>
<td>7</td>
<td>Radical tympano-mastoidectomy with blind sac closure</td>
<td>25</td>
<td>Nucleus CI22M</td>
<td>Wound dehiscence treated conservatively*</td>
</tr>
<tr>
<td>2</td>
<td>4 (1999)</td>
<td>Growth and thyroid hormone deficiency</td>
<td>7</td>
<td>Tympanoplasty with OCR, radical tympano-mastoidectomy with blind sac</td>
<td>13</td>
<td>Nucleus CI22M</td>
<td>none</td>
</tr>
<tr>
<td>3</td>
<td>14 (1992)</td>
<td>VPS, Growth and thyroid hormone deficiency</td>
<td>12</td>
<td>Tympanoplasty with OCR, canal plasty, modified radical tympano-mastoidectomy</td>
<td>29</td>
<td>HiRes 90K</td>
<td>Mastoid-cutaneous fistula requiring closure</td>
</tr>
</tbody>
</table>

CI indicates cochlear implantation; CDSM, chronic serous otitis media; CVA, cerebrovascular accident; VPS, ventriculoperitoneal shunt; HA, hearing aid; OCR, ossicular chain reconstruction

*All patients experienced mild neuropsychologic dysfunction manifested as attention deficit and learning disabilities.

**Fourteen days after CI, patient 1 demonstrated minor dehiscence in blind sac closure and was treated with topical (Bacitracin) and oral (Ciprofloxacin) antibiotics for 7 days with complete resolution.

†Patient 3 developed a small post-auricular wound dehiscence with mastoid-cutaneous fistula requiring closure using local anesthesia. Intra-operatively, the fistula tract was excised and closed with a cartilage and perichondrial rotation flap without disruption of the implant. Complete healing was documented after 4 weeks.

Table 2. Cochlear implantation performance data.

<table>
<thead>
<tr>
<th>Patient</th>
<th>CNC-W (%)</th>
<th>CNC-P (%)</th>
<th>HINT-Q (%)</th>
<th>HINT-N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Pre-CI</td>
<td>Post-CI</td>
<td>Pre-CI</td>
<td>Post-CI</td>
</tr>
<tr>
<td>1</td>
<td>8</td>
<td>72</td>
<td>24</td>
<td>67</td>
</tr>
<tr>
<td>2</td>
<td>CNE</td>
<td>CNE</td>
<td>CNE</td>
<td>CNE</td>
</tr>
<tr>
<td>3</td>
<td>0</td>
<td>36</td>
<td>0</td>
<td>64</td>
</tr>
</tbody>
</table>

CI indicates cochlear implantation; CNC-W, consonant-nucleus-consonant words; CNC-P consonant-nucleus-consonant phonemes; HINT-Q, hearing-in-noise test administered in quiet; HINT-N, hearing-in-noise test administered in noise; CNE, could not be evaluated

DISCUSSION

Ototoxicity and Chemotherapy

Cisplatinum ototoxicity is typically characterized by irreversible, bilateral, symmetric, high-frequency SNHL. Ototoxicity can be related to cumulative dose as well as dose intensity, reinforcing the need for audiologic monitoring in these patients. Histopathologic studies of temporal bones treated with cisplatinum demonstrate damage to the inner and outer hair cells, atrophy of the stria vascularis, as well as some degeneration of the spiral ganglion cells and cochlear nerve.

Ototoxicity and Radiation Therapy

Similar histologic findings are seen in temporal bones subjected to radiation therapy. Acute effects include severe effusion, mucosal inflammation and intimal hypertrophy of arteries. SNHL is typically progressive, irreversible and dose-dependent and can manifest long after completion of radiation. While the mechanism of sensorineural damage is not fully understood, ischemic effects on cochlear vessels leading to hair cell and supporting cell death and intra-cochlear fibrosis have all been implicated. Functioning retro-cochlear auditory pathways are a prerequisite for successful auditory rehabilitation with CI and these are subjected to radiation in medulloblastoma patients. Low et al examined the retro-cochlear pathways of nasopharyngeal cancer patients after radiation therapy and showed intact auditory pathways up to 2 years post-treatment. Results of this study support their findings.

CONCLUSIONS

Patients treated for pediatric medulloblastoma with surgical excision and chemoradiation develop otologic sequelae, including profound SNHL, and may require cochlear implantation. Successful management of middle ear and mastoid pathology involves consideration of potential future cochlear implantation. Post-operative performance data supports cochlear implantation in this population.

Chronic Ear Disease

Chronic ear disease following RT is a prevalent side effect and was seen in all three patients. Operative treatment should incorporate the possibility of future cochlear implantation and may include subtotal petrosectomy, obliteration and the creation of a blind sac of the EAC.

Intro-Operative Considerations

Histologic changes lead to softer bone around the otic capsule and facial nerve. Mucosal disease obscuring the round window and cochlear lumen obliteration can compromise electrode insertion. In this series, all patients had full electrode insertion. Poor-wound healing is accompanied by concern for infection and/or extrusion. Careful planning of incisions/flaps, meticulous handling of soft tissue, avoidance flap thinning and judicious use of electrocautery, may minimize post operative wound complications. Keys to effective post-operative management include close follow-up, vigilant PE and aggressive treatment of complications when discovered.

Tumor Surveillance and Cochlear Implants

MRI is used for tumor surveillance of medulloblastoma up to 5 years post treatment. Therefore, CI in this period should be balanced by the need for frequent MRI. Once implanted, magnet removal from the internal device is required for safe imaging and only bipolar electrocautery can be used intraoperatively. Should additional RT be necessary, variable dose concentration due to metallic reflection and shadowing effects must be considered.