Aberrant Internal Carotid Artery in the Temporal Bone

Imaging Findings and Management

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Objectives: To describe the clinical and radiological features of the vascular anomaly aberrant internal carotid artery (ICA) in the temporal bone and to discuss management strategies.

Design: Retrospective study.

Patients: Sixteen cases of aberrant ICA were diagnosed between 1982 and 2003.

Results: Of 16 cases, 11 were recognized by imaging assessment, 4 were recognized during middle ear surgery, and 1 was recognized clinically. Among the 11 cases, 8 malformations were diagnosed because of otologic symptoms related to the abnormal ICA or chronic otitis, while the other 3 were identified incidentally because of an underlying accompanying disease. In 4 cases, the diagnosis was made during surgery related to chronic otitis media (n = 2) or conductive hearing loss (n = 2). In these 4 cases, massive bleeding resulted from surgical injury to the vessel. Packing the external auditory canal and the middle ear first controlled the bleeding. Endovascular procedure was required in 2 cases to exclude an aneurysm or to control bleeding but was followed by anterior cerebral stroke in 1 case. The aberrant ICA could be identified on computed tomographic scan by the following features: intratympanic mass, enlarged inferior tympanic canalculus, absence of the vertical segment of the ICA canal, and absence of bone covering the tympanic portion of the ICA. Conventional angiography was mandatory when intervention was planned to control bleeding or aneurysm.

Conclusions: This study highlights that aberrant ICA has to be identified before any middle ear surgery because misdiagnosis may lead to dramatic surgical complications, whereas diagnosis with computed tomographic scan is easy. Bleeding is a minor complication compared with the putative neurologic deficit due to endovascular occlusion.

roradiology departments. The mean ± SD age of patients was 38.6 ± 15.0 years, and the population consisted of 6 men and 10 women. Of 16 cases, 11 were diagnosed by imaging assessment, 4 during middle ear surgery, and 1 by clinical tympanic examination (Figure 1 and Table).

Of the 11 cases of imaging diagnosis, 3 were incidental imaging diagnoses, with the CT scan being performed because of another disorder, and 8 cases were diagnosed because of otologic disorders. Patients with incidental diagnosis (n=3) did not complain of otologic symptoms. One incidental diagnosis was made during CT scan of sinuses, and 2 were made during conventional angiography for subarachnoid hemorrhage (n=1) and carotid artery stenosis (n=1). The otologic disorders (n=8) were chronic otitis media with conductive hearing loss (n=2) and suspicion of glomus tumor because of pulsatile tinnitus, conductive hearing loss, and anteroinferior red mass in the retrotympanic space on otoscopy (n=6). The first 2 cases were assessed by CT scan and the other 6 cases by CT scan, MR imaging, and/or arteriography. A careful tympanoplasty was performed without bleeding complication in 1 case.

For the 4 cases set during middle ear surgery, 2 patients presented with chronic otitis media, 1 presented with conductive hearing loss with suspected otosclerosis, and 1 presented with a retrotympanic mass. Middle ear surgery (tympanoplasty or middle ear exploration) revealed an anterior red mass. Unaware of the type of the mass, surgeons performed either a puncture with a needle or a biopsy with forceps, which led to massive bleeding (n=4). Hemorrhage was immediately controlled by packing the external auditory canal and middle ear. Arteriography was performed as soon as possible after the surgical injury, which confirmed the diagnosis of aberrant ICA (n=2) and allowed an endovascular procedure (n=2). Computed tomographic scan was performed secondarily to confirm the vascular malformation and in the third case revealed contact between aberrant ICA and incus and a persistent stapedial artery, which may explain the conductive hearing loss.

The aberrant ICA was associated with persistent stapedial artery in 4 cases and with contralateral carotid hypoplasia in 1 case. Persistent stapedial artery was evidenced by CT scan but not by arteriography.

**IMAGING ASSESSMENT**

Imaging assessment included angiotomography (n=1), MR angiography (n=2), arteriography (n=10), and/or temporal bone CT scan (n=14). The following CT scan features were seen: an anterior retrotympanic mass, an

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**Table. Clinical Characteristics of Patients**

<table>
<thead>
<tr>
<th>Case No./Sex/Age, y</th>
<th>Otologic Signs</th>
<th>Otoscopy Finding</th>
<th>Diagnosis</th>
<th>Carotid Injury</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/54</td>
<td>Conductive hearing loss</td>
<td>Chronic otitis</td>
<td>CT scan</td>
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<td>None</td>
</tr>
<tr>
<td>2/F/27</td>
<td>Conductive hearing loss and pulsatile tinnitus</td>
<td>Retrotympanic red mass</td>
<td>CT scan, MRI, and MRA</td>
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<td>None</td>
</tr>
<tr>
<td>3/M/24</td>
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<td>Cholesteatoma</td>
<td>Preoperative CT scan</td>
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<td>None</td>
</tr>
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<td>4/M/22</td>
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<td>None</td>
<td>Incidental diagnosis on sinuses</td>
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<td>None</td>
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<tr>
<td>5/F/37</td>
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<td>None</td>
<td>Intraoperative FNAs</td>
<td>Puncture with a needle</td>
<td>None</td>
</tr>
<tr>
<td>6/F/34</td>
<td>Conductive hearing loss</td>
<td>Chronic otitis</td>
<td>Intraoperative FNAs</td>
<td>By biopsy forceps</td>
<td>ICA occlusion</td>
</tr>
<tr>
<td>7/F/42</td>
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<td>Retrotympanic red mass</td>
<td>Otoscopy and angiography</td>
<td>None</td>
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<tr>
<td>8/M/74</td>
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<td>None</td>
<td>Incidental diagnosis with arteriography</td>
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<td>None</td>
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<td>Puncture with a needle</td>
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<td>Intraoperative diagnosis</td>
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<tr>
<td>12/F/56</td>
<td>None</td>
<td>Retrotympanic red mass</td>
<td>Intraoperative FNAs</td>
<td>Puncture with a needle</td>
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<td>CT scan</td>
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<td>Retrotympanic red mass</td>
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<tr>
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<td>Otoscopy</td>
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<td>Retrotympanic mass</td>
<td>CT scan</td>
<td>None</td>
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</tr>
</tbody>
</table>

Abbreviations: CT, computed tomographic; ICA, internal carotid artery; MRA, magnetic resonance angiography; MRI, magnetic resonance imaging.
enlarged inferior tympanic canaliculus, absence of the vertical segment of the ICA canal, and absence of bone covering the tympanic portion of the ICA (Figure 2 and Figure 3). Findings from arteriography and MR angiography showed a reduced diameter of the tympanic ICA. In a frontal view, the vertical segment of the carotid artery was lateral to a line drawn vertically through the vestibule (light round cavity) (Figure 4 and Figure 5).

MANAGEMENT

In the absence of hemorrhage, aberrant ICA was neither managed with surgery nor with endovascular procedure, even in the case of pulsatile tinnitus or conductive hearing loss. On the contrary, interventional procedures were mandatory when surgical injury occurred. The first-line procedure consisted of packing the external auditory canal and the middle ear. Then, arteriography with endovascular procedure was required in 2 cases of persistent bleeding. In the first case, aberrant ICA was excluded with 2 detachable balloons positioned distally and proximally to the damaged area, which successfully controlled bleeding. However, when the patient awoke, he had a contralateral hemiparesis. In the second case, arteriography revealed an aneurysm of the ICA protruding in the middle ear. Exclusion of the ICA was not allowed because the anterior communicans artery was not functioning well. Thus, endovascular procedure consisted of filling the aneurysm with detachable platinum coils through a stent positioned in the ICA (Figure 6).

COMMENT

This study shows that aberrant ICA in the middle ear is primarily asymptomatic. Misdiagnosis of the anomaly may lead to serious morbidity because of bleeding or vascular occlusion. Computed tomographic scan has become the standard for diagnosis, while arteriography is required for the management of bleeding complication.
Normally, the ICA enters the petrous bone medial to the styloid process via the carotid canal. The initial vertical segment is anterior to the cochlea and separated from the tympanic cavity by a thin plate of bone (mean thickness, 0.24 mm). The ICA then turns anteriorly to lie inferior and posteromedial to the Eustachian tube, traverses the foramen lacerum, and enters the medial cranial fossa.

Anomalies of the ICA include arterial displacement and aneurysms. Several hypotheses have been formulated concerning the genesis of aberrant ICA. Congenital anomaly described by Lasjaunias and colleagues is the most likely. They hypothesized the alternate blood flow theory (Figure 7): the C1 portion of the ICA involutes owing to the persistence of the pharyngeal artery system, and as a consequence, an anomalous course develops with blood flowing via the ascending pharyngeal artery to the enlarged inferior tympanic artery with retrograde flow through the caroticotympanic vessels into the horizontal segment of the ICA. This theory can explain the radiological features with enlargement of the inferior tympanic canalculus, the presence of a tissular mass found in the anterior hypotympanum, and the absence of the vertical portion of the ICA. Alternatively, the second congenital theory is that persistence of embryological vessels (ie, the stapedial artery) pulls the ICA into the middle ear. We could not support this theory because persistence of stapedial artery was found in only 4 cases. Moreover, this theory cannot account for the enlargement of the inferior tympanic canalculus. A less likely hypothesis is related to disappearance of the bony plate between the ICA and the middle ear. Here again, this cannot account for the enlarged inferior tympanic canalculus.

Clinical diagnosis of aberrant ICA is difficult because symptoms and signs are often nonspecific or absent. Otologists should be aware that symptoms and signs such as pulsatile tinnitus, conductive hearing loss, and a pulsatile retrotympanic mass in the anteroinferior part may be related to an aberrant ICA. Clinical presentation could be regarded as otosclerosis, glomus tumor, or other vascular malformation. However, the retrotympanic mass due to aberrant ICA looks different from a glomus tumor: anterior, pulsatile, and white or rosy. Results from audiometric evaluation may be normal or show a conductive hearing loss owing to persistent stapedial artery, malleus or incus blockage, or associated middle ear disease. Thus, aberrant ICA, which is asymptomatic most of the time, will be diagnosed during middle ear surgery. To avoid surgical injury due
to misdiagnosis, CT scan of the temporal bone should ideally be performed before any middle ear surgery. However, if, in the absence of a CT scan, a retrotympanic mass with aberrant ICA characteristics is observed during middle ear surgery, a CT scan should be performed prior to puncture.

On axial CT scan, aberrant ICA is identified by (1) an ICA that runs adjacent to the jugular bulb, in a posterior position and with a reduced diameter; (2) an enhancing mass in the hypotympanum; (3) a deficient bony plate along the tympanic portion of the ICA; and

![Figure 4](image4.jpg) Figure 4. Oblique magnetic resonance angiography. The arrow shows the aberrant internal carotid artery with a reduced diameter.

![Figure 5](image5.jpg) Figure 5. Coronal arteriography. The arrow shows the aberrant internal carotid artery (left) with a reduced diameter.

![Figure 6](image6.jpg) Figure 6. False aneurysm endovascular procedure. A, False aneurysm (arrow) before endovascular procedure on arteriography; B, postendovascular procedure, with exclusion of the false aneurysm (arrow); and C, coils expulsion (arrow) through the tympanic membrane a few months later.

![Figure 7](image7.jpg) Figure 7. Pathogenesis of the aberrant internal carotid artery (ICA): the alternate blood flow theory. The C1 portion of the ICA involutes owing to the persistence of the pharyngeal artery system, and as a consequence, an anomalous course develops with blood flowing via the ascending pharyngeal artery to the enlarged inferior tympanic artery with retrograde flow through the caroticotympanic vessels into the horizontal segment of the ICA. Figure adapted from Som and Curtin.25
(4) absence of the vertical segment of the carotid canal. The main features at angiography are (1) a reduced diameter of the tympanic ICA and (2), in a frontal view, the vertical segment of the ICA is lateral to a line drawn vertically through the vestibule (light round cavity).

Knowledge of this rare entity is essential to any otorhinolaryngologist who undertakes myringotomy and middle ear surgery. Accidental injury after myringotomy or in case of misdiagnosis with a glomus tumor may have disastrous consequences. Bleeding will be initially controlled by packing the ear canal and applying a pressure bandage. If bleeding recurs after packing, angiography with embolization is required because ICA occlusion by surgical methods may induce anterior circulation stroke. The choice of the embolization technique depends on the ability to occlude the ICA. Therefore, a carotid test occlusion is performed to appreciate whether intracranial anastomoses are present and functional enough to supply the occluded vessel. The occlusion test is achieved by temporary inflation of a balloon in the ICA with simultaneous injection of the contra-lateral ICA and dominant vertebral artery. The occlusion is well tolerated if the delay in the venography between both hemispheres is less than 2 seconds. This allows definite occlusion of the ICA with detachable balloons and/or coils. If the occlusion test is not tolerated, selective embolization of the pseudoaneurysm is achieved with platinum coils. In a case report, Alexander et al.23 suggest an alternative procedure using a covered stent. This procedure allows carotid artery patency, and if it proves to have long-term durability and safety, it may supplant carotid occlusion.

In case of an asymptomatic and proven aberrant ICA, most authors recommend a conservative approach,1,2,3,18 as opposed to Ruggles and Reed,24 who advocated surgery to relieve the patient of troublesome symptoms (tinnitus and hearing loss) and to prevent possible destruction of the middle ear structures and formation of an aneurysm. They recommended separation of the ICA from the middle ear space by covering the vessels with fascia and then compressing it into the promontory defect and covering it with a bone graft. However, this may further compromise the blood flow through these already narrowed vessels, with the risk of consecutive neurological disorders. This could be disastrous if this aberrant ICA is associated with a contralateral carotid agenesis, as reported in 1 case in our series. Finally, in this series, aberrant carotid artery, without surgical injury, was not followed by complications. We thus recommend a conservative approach in case of aberrant ICA without bleeding complications.

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References