Giant Cholesteatoma: Recommendations for Follow-up

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ABSTRACT

This report presents the management of five patients who presented with giant recurrent or residual cholesteatoma after periods of 2 to 50 years. Their case histories are highly diverse, but all provide evidence of the need for long-term follow-up.

KEYWORDS: Cholesteatoma, middle ear, skull base, surgery

Chronic otitis media with cholesteatoma is a potentially serious disease. It can lead to life-threatening intracranial complications such as meningitis, encephalitis, or brain abscess. Follow-up is generally considered necessary after surgical treatment for cholesteatoma. Recurrent or residual cholesteatoma can present in several ways that depend on its location. The presence of recurrent or residual disease is not always obvious to the clinician.

In the literature, the incidence of both recurrence and residual disease varies greatly and seems to increase over time. Few studies have prolonged follow-up.¹⁻⁶ Although most recurrences develop within the first 5 years, some reports

describe recurrence up to 14 years after the initial surgery.^{7,8}

To our knowledge, no guidelines for follow-up in adults have been proposed, which is in contrast to that of children.^{2,3,9} It is difficult, if not impossible, to generalize for each patient because of a range of clinical considerations pertinent to each one. Nonetheless, follow-up after surgery for cholesteatoma is essential if the presence of recurrent or residual disease is to be found in time.

This article presents five cases that demonstrate the need for meticulous and prolonged follow-up. They illustrate that the risk of recurrence persists for a very long time after initial treatment

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for cholesteatoma and probably never disappears completely.

CASE REPORTS

Patient 1

A 75-year-old woman presented to the Department of Otorhinolaryngology and Head and Neck Surgery, Radboud University Nijmegen Medical Centre, the Netherlands with a 4-month history of unsteadiness on her feet. This was associated with left-sided hearing loss and tinnitus. She did not have otalgia, otorrhea, or headache. Fifty years previously she had undergone surgery on that ear through a retroauricular approach. There were no operative records. Her eardrums appeared normal, and other than a hearing loss, there were no other neurological deficits. Pure-tone audiometry showed an asymmetrical sensorineural hearing loss of 42-dB hearing level (HL) on the left side and 27-dB HL on the right side (average of thresholds at 0.5, 1.0, and 2.0 kHz).

A computed tomography (CT) scan revealed a large, destructive, left-sided, temporal bone lesion. On axial T2-weighted magnetic resonance (MR) images, the lesion had irregular signal intensity (Fig. 1). Surgical exploration was undertaken through a retroauricular incision, and the mass, a cholesteatoma, was immediately apparent. The dura of both the middle and posterior fossa was exposed, and the cholesteatoma extended inferiorly onto the sternocleidomastoid muscle. The cholesteatoma was removed completely, and the resultant cavity was obliterated with abdominal fat. The surgery was uncomplicated. Histological examination confirmed the diagnosis of cholesteatoma.

Patient 2

A 28-year-old man presented with meningism, complaining of headache and neck stiffness. He

Figure 1 Magnetic resonance axial T2-weighted image of patient 1 with a giant lesion of the left temporal bone, with no obvious invasion of the parenchyma of the brain.

had undergone a radical, left-sided mastoidectomy when 4 years old. Revision surgery had been undertaken on two occasions when he was 14 years and 26 years old. There had been regular follow-up but without any imaging whatsoever. Throughout this period, his left ear had discharged. He had no residual hearing on that side. His right ear was entirely normal and he did not have vertigo. On examination, his left-sided mastoid cavity contained granulation tissue and pus. Facial nerve function was normal. Pure-tone audiometry confirmed that he had total loss of hearing on the left side with normal hearing on the right side. The axial CT images showed massive destruction of the temporal bone on the left side (Fig. 2). MR imaging revealed that the mass extended into the petrous apex and the posterior fossa (Figs. 3 and 4).

An extended retroauricular approach was performed to achieve sufficient access to the cholesteatoma. The vertical and horizontal part of the carotid artery was covered with cholesteatoma, which extended into the clivus. Abdominal fat was used to obliterate the cavity. The surgery was uncomplicated, and there were no neurological or otologic sequelae.

Figure 2 Computed tomography scan of patient 2 showing massive bone destruction of the skull base on the left side.

Figure 4 Magnetic resonance axial T1-weighted image of patient 2, showing a mass of the left skull base, isointense to gray matter.

Figure 3 Magnetic resonance axial T2-weighted image of patient 2 with a hyperintense lesion of the left skull base.

Patient 3

An 81-year-old woman commented at a routine outpatient clinic appointment for a left mastoid cavity review that she had experienced vertigo for some time and had noticed increased right-sided hearing loss and tinnitus. In early childhood, she had suffered from a right-sided, postauricular abscess, but no surgical intervention had been considered. At the age of 40 years, she underwent a left-sided radical mastoidectomy. No data could be obtained about that operation. At the age of 65 years, she developed a right-sided facial paralysis, which was thought to be idiopathic in nature, Bell's palsy. On examination, she had a dry and clean radical mastoidectomy cavity on the left side and a normal right ear. Her facial paralysis on the right side had not recovered at all, a grade VI paralysis according to the House-Brackmann classification.¹⁰ Pure-tone audiometry showed profound deafness on the right side. On the left side, the side of the radical mastoidectomy, a conductive hearing loss

was superimposed on age-related sensorineural hearing loss. MR and CT images revealed a large mass in the right temporal bone (Figs. 5 and 6). The mass extended into the middle and posterior fossa.

Surgery was performed through a retroauricular approach. A cholesteatoma immediately protruded through the incision after elevation of the retroauricular periosteal flap. It had eroded the cortex of the temporal squama as well as the bony margins of the middle and posterior fossa. It extended into the root of the zygoma. After removal of the cholesteatoma, the cavity was filled with a free muscle graft. There were no postoperative complications.

Patient 4

A 70-year-old woman was admitted unconscious to the neurology department. She had a history of ear surgery on the right side 14 years previously. Since the operation, she had been deaf in that ear. Four

Figure 5 Magnetic resonance axial T2-weighted image of patient 3 showing a large mass in the temporal bone on the right side, hyperintense to gray matter.

Figure 6 Coronal computed tomography scan image of patient 3 showing massive bone destruction on the right side.

years later, she developed a right-sided facial palsy. No other information was available. Her conscious state precluded any other information being obtained. Clinical examination was also limited, and the only conclusions were that she had a right-sided VII, VIII, and IX deficits. A CT scan showed a large destructive lesion in the skull base arising from the right temporal bone that extended into the sella and sphenoid sinus (Fig. 7). Multiple brain abscesses were present. Biopsy confirmed the suspected diagnosis of cholesteatoma. To remove the cholesteatoma, a retroauricular incision was extended into the neck. Behind the sigmoid sinus, an opening already existed from which

Figure 7 Axial computed tomography scan image of patient 4 showing the lesion on the right side, extending onto the sella and the sphenoidal sinus.

cholesteatoma protruded. Large amounts of cholesteatoma were removed. The mass extended from the medial fossa to the foramen magnum, with near-total destruction of the temporal bone. Her postoperative course was complicated by epilepsy, septicemia, and cardiovascular instability. Because of airway-management problems, a tracheotomy was performed. After a period of rehabilitation that lasted several months, she was discharged to a nursing home with a slight hemiplegia in addition to her facial palsy and hearing loss.

Patient 5

A 76-year-old man presented with symptoms of chronic otitis media and retro-ocular pain on the left side. At the age of 72 years, he had undergone a left-sided radical mastoidectomy for cholesteatoma. He was type 2 diabetic managed by oral hypoglycemic agents. On examination, he had a left-sided VI and VII palsy, consistent with Gradenigo's syndrome. A CT scan revealed a large, left-sided, destructive lesion that occupied almost one-third of his skull base and extended into his neck (Fig. 8). After surgical removal of the mass, reexploration was needed for a persistent cerebrospinal fluid leak. After closure of the leak, the patient was discharged. One month after discharge, this patient presented with intractable sepsis and died. At autopsy, the extension of the residual cholesteatoma was clear. More than one-third of the skull base was covered with cholesteatoma, in combination with extensive destruction and osteomyelitis of the skull base (Fig. 9). There was involvement of the clivus, the dorsum of the sella, the greater wing of sphenoid bone, and temporal bone. The brain stem showed signs of compression.

DISCUSSION

Giant cholesteatoma of the skull base is not common, and as the presentation of residual or recurrent

Figure 8 Axial computed tomography scan image of patient 5 revealing a giant lesion, covering almost one-third of the skull base on the left side.

cholesteatoma is even more rare. But this report illustrates that it is indeed a realistic possibility in patients who have been lost to follow-up, or in patients for whom follow-up ended after several years.

In cases 2 and 5, the giant cholesteatoma had developed over a period of just 2 and 4 years, respectively. This is within the generally accepted "normal" range of follow-up. In these cases, the extension and aggressiveness of the cholesteatoma is

Figure 9 Photograph taken of the skull base of patient 5 during autopsy. The extensive destruction of the skull base with the remains of the cholesteatoma is visible (left side).

impressive, considering that so few years had passed since surgery.

Cases 1 and 4 are somewhat different than cases 2 and 5 in that the diagnosis was made many years after the patients had their last otologic surgery. They were diagnosed with giant cholesteatoma 50 and 14 years, respectively, after their primary surgery. This illustrates that the manifestation of recurrent or residual disease is not limited by time. A literature review showed that recurrence of cholesteatoma in operated ears ranges from 6 to 27% and not as low as some assume (5 to 10%).¹¹ Moreover, it has been demonstrated that the recurrence rate is higher with a longer observation period.^{3,5} Tos and Lau described an increase of 2.3% in recurrence for every 3 years of follow-up.⁵ In children, the increase in recurrence rate is even higher with prolonged follow-up.³ In some more recent reports concerning revision surgery for chronic otitis media, a recurrence rate of 14% was noted after 5 years of follow-up.⁶ O'Leary and Veldman reported a recurrence rate of 8% in revision surgery for adults, with a maximum follow-up duration of 15 years.¹² This is considerably lower than the expected number based on percentages from older research. Although the percentages mentioned here might vary, the recurrence or residual rate is still considerable. Moreover, it seems to be higher with prolonged follow-up.

Patient number 2 had regular clinical follow-up, but no imaging was undertaken during this period. We have no information about the follow-up of patient number 5. We presume that follow-up had been done regularly after the radical mastoidectomy on his left side, but probably without imaging. Patient number 3 was diagnosed with Bell's palsy and did not have any surgery on her right ear, where the giant cholesteatoma was localized. When her facial paralysis was concluded to be Bell's palsy, no imaging of the temporal bones was conducted. This omission can be seen as questionable, because her medical history suggested a possible otologic cause. For patient number 2, no imaging was done in the brief 2-year follow-up period even though he suffered from chronic otor-

rhea. These cases emphasize the value of applying the imaging techniques that are currently available. It is highly likely that in all cases the imaging techniques would have revealed the presence of the cholesteatoma at a much earlier stage.

CT scanning is most frequently used for evaluating the air spaces and osseous components of the temporal bone or middle ear.¹³ It is valuable in the diagnostic workup for patients with chronic otitis media and possible cholesteatoma. When a soft tissue mass is present in an operated ear, it is much more difficult to assess the radiological criteria for residual or recurrent cholesteatoma. The criterion of bone erosion with a soft tissue mass, visible on CT scanning, is unreliable after surgery. Therefore, attempts have been made to use MR imaging as a reliable diagnostic tool in a postoperative middle ear. In a report from 1999, Vanden Abeele et al concluded that a second-look procedure could not be substituted by MR imaging.¹⁴ Recent developments, however, show promising results in the value of MR imaging for the postoperative follow-up of middle ear status, particularly with the use of delayed contrast-enhanced T1-weighted images or diffusion-weighted MR imaging (DWI).^{15,16} However, in a report by Vercruysse et al, inclusion of patients with a planned second-stage surgery (without a clinical suspicion of recurrent cholesteatoma, $n = 45$) resulted in a lower sensitivity for detection of recurrent or residual disease with echoplanar DWI, compared with patients with a clinical diagnosis of cholesteatoma. They stated that due to the smaller size of the residual cholesteatoma, air-bone artifacts of echoplanar DWI and technical limitations, this imaging technique in combination with standard MR imaging cannot replace a second look.¹⁷ With the use of spin-echo DWI, some of these limitations can hopefully be overcome. In a small number of patients, results indicate that smaller cholesteatomas can be detected.^{18,19}

So there is still debate on whether imaging is appropriate for the postoperative follow-up of cholesteatoma and, if so, on which technique is most suitable. At present (planned second-stage) surgery cannot be replaced by any kind of imaging.

Standardization of follow-up on an international basis would be ideal. Consensus in both the length of the follow-up and in the use of imaging techniques would provide an excellent basis for comparison of results worldwide. In the five patients, giant recurrent or residual cholesteatomas had developed after a period of insufficient or absent follow-up. These five case histories underline the need for (prolonged) follow-up, but they are not sufficient to serve as a basis of a well-defined guideline and protocol for follow-up in all cholesteatoma patients. We suggest, however, that follow-up duration should not be limited in time (or at least be longer than 5 years) and that imaging techniques play a role in the detection or confirmation of recurrent or residual cholesteatoma. There is no doubt that CT and MR imaging shortly after primary surgery of cholesteatomas are valuable, as they provide a basis for future imaging studies.

CONCLUSION

This article reports on five highly diverse giant cases of recurrent or residual cholesteatoma after 2 to 50 years. These cases illustrate the need for prolonged follow-up after surgery for otitis media cholesteatomatosa. It is difficult to outline a uniform strategy for follow-up that would be applicable to all patients. Developments in imaging techniques show promising results, but at present surgery remains the most reliable method for detecting recurrent or residual cholesteatoma. Shortly after primary surgery for cholesteatoma, CT and MR imaging are valuable in providing a baseline for further follow-up studies.

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