

## Cochlear fistula found in a naturally healed tympanic cavity

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### Abstract

A cochlear fistula found in a naturally healed mastoid cavity is reported. The patient is a 53-year-old Japanese woman who was complaining of unsteadiness. She had a long history of otorrhoea in childhood. Her tympanic and mastoid cavities were widely open and were covered by thin epithelium. The posterior wall of the ear canal was missing. She had no hearing in the ear but responded to electrical promontory testing. Exploratory surgery was indicated at which fistulae of the basal turn of the cochlea and the horizontal semicircular canal were found. These fistulae were sealed by pieces of bone and muscle.

### Introduction

Labyrinthine fistula is one of the well-known conditions with which an extensive cholesteatoma may present. Fistulae most commonly involve the horizontal semicircular canal, but a fistula into the cochlea is rare. The authors describe a case of an

extensive opening of the cochlear bony shell in a naturally healed mastoid cavity.

### Case report

A 53-year-old Japanese woman complained of unsteadiness since January 1990. She also had occasional pain in the right postauricular region. Extensive destruction of the ear canal was noted by her otolaryngologist and she was referred to the Department of Otolaryngology, Hiroshima University on February 3, 1990. She had suffered from hearing loss and otorrhoea in the right ear during childhood, but had not been treated. Her past history included bronchial asthma and rheumatoid arthritis.

On examination, the posterior wall of the external ear canal was absent resulting in a large space from the mastoid through the tympanic cavity. The entire space was covered by thin, dry epithelium; no air space was visible, nor were the ossicles.

Pure tone audiometry revealed total deafness on the right side, and 17.5 dB (in average) in the left ear. On vestibular testing no gaze or spontaneous nystagmus were seen. In the righting reflex tests, Mann's test was normal, yet Romberg's test



FIG. 1

Schueller x-ray. Note normal finding in the left ear (top) and a large radio-lucent area in the right mastoid portion (bottom).

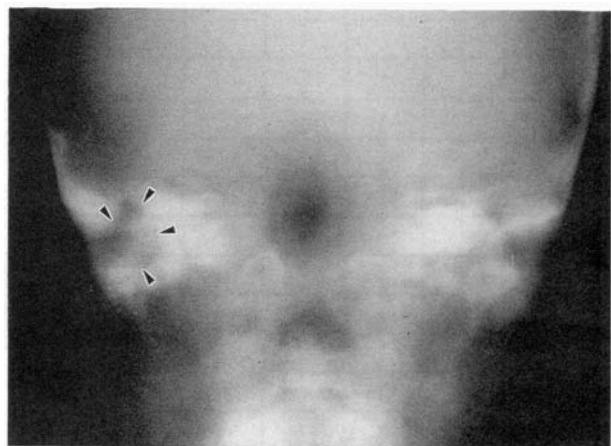


FIG. 2

The tomogram in the frontal section, showing a large cavity from the right ear canal through the antrum (arrows).

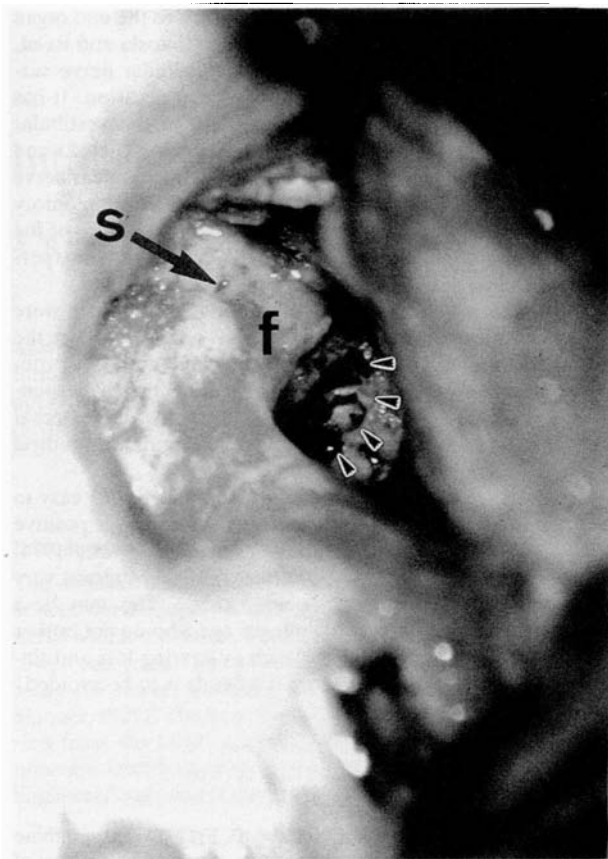


FIG. 3

A view through the surgical microscope. The cochlear fistula is shown by the arrows. f: exposed facial canal, s: fistula of the lateral semicircular canal.

was poor when the eyes were closed. Goniometry was within the normal limit. On the stepping test, she showed no lateral deviation, but tended to shift forward. The fistula sign was negative on both positive and negative pressure. A caloric response could not be elicited from the right ear. The optokinetic nystagmus and eye tracking test results were normal. Electrical promontory test was performed using House's cochlear nerve tester. Sound sensation described as "buzzing" was obtained on stimulation with currents of 1.2 volts AC at 30, 60, 90 and 120 Hz.

X-ray (Schueller's view) and tomography showed a large area of bone destruction in the mastoid. No mass shadow was observed (Figs. 1 & 2).

The patient had been on several kinds of antivertiginous drugs for three weeks without effect. She was therefore scheduled for an exploratory surgery on April 3, 1990.

#### Operative findings

Under general anaesthesia, the posterior part of the mastoid was approached via a postauricular incision. The skin covering the mastoid cavity was elevated and reflected forward. This exposed the smooth bony surface of the mastoid cavity, antrum and the dome of the lateral semicircular canal. The posterior wall of the ear canal had been destroyed leaving a low hump. A small fistula was found in the lateral semicircular canal. The epithelium continued from the mastoid through into the tympanic cavity. The tympanic scutum was also absent and epithelium covered the entire portion of the tympanic cavity. All of these findings were exactly as one named after a radical mastoidectomy. The epithelium of the tympanic portion of the facial nerve was elevated; the bony wall of the facial canal was missing for two-thirds of the horizontal portion. No ossicles

were found. The oval window was occluded by bone. The round window could not be seen, but the upper rim of the niche could be identified. A very small hole was found below this rim. The most noteworthy finding was widespread destruction of the cochlear bony shell, from the upper rim of the round window through the oval window (Fig 3). Inside the bony defect, the lateral wall of the cochlear duct and the osseous spiral lamina could be seen (Fig. 4). No fluid escaped from the defect. A small piece of bone was placed in the semicircular canal fistula. A piece of muscle was inserted into the cochlear defect and was covered by several pieces of bone. The elevated skin was replaced in the tympanic and the mastoid cavities. Two pieces of the temporalis fascia were used to seal defects of the skin flap inadvertently made during the procedure.

The post-operative course was uneventful. The replaced skin flap was viable and the cavity dried up in seven weeks.

CT scans taken after surgery show the cochlear fistula to be sealed by soft tissue (Fig. 5).

#### Discussion

A number of studies have been done on fistulae of the semicircular canal caused by cholesteatoma. Of special interest to surgeons is how to treat a labyrinthine fistula, when the inner ear has good function. In many cases total removal of the matrix from the semicircular canal fistula does not jeopardize cochlear function (Palva *et al.*, 1971; Gacek, 1974) and sometimes even improves the hearing (Sheehy and Brackmann, 1979). However, when treating an only hearing ear or a large fistula, the surgeon may choose to leave the matrix on the fistula untouched. According to Gacek (1974), when the fistula is larger than 2 mm, the matrix is adherent to the membranous canal and therefore, it is more risky to attempt its removal. A fistula into the cochlear part, on the other hand, gives a poorer

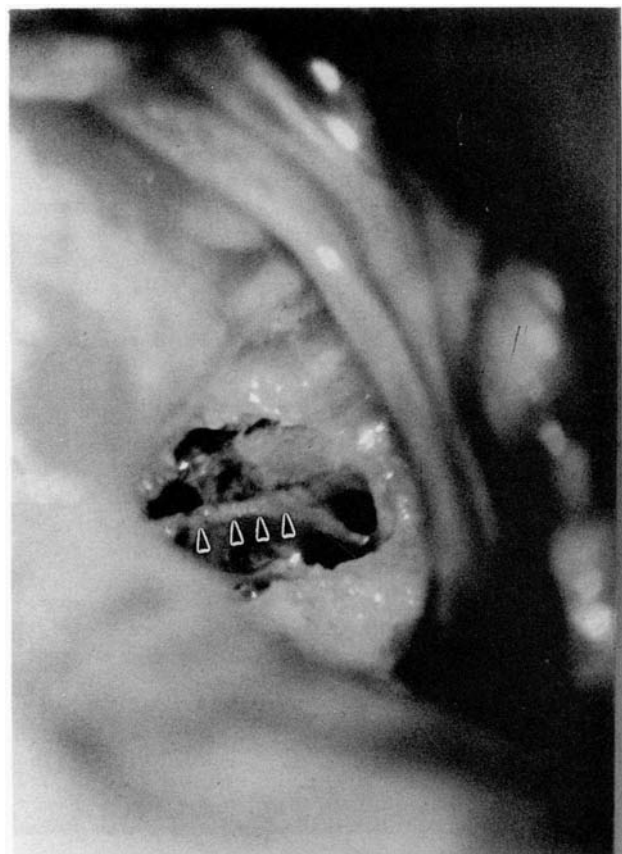


FIG. 4

High magnification view of the cochlear fistula. The arrows indicate the osseous spiral lamina.

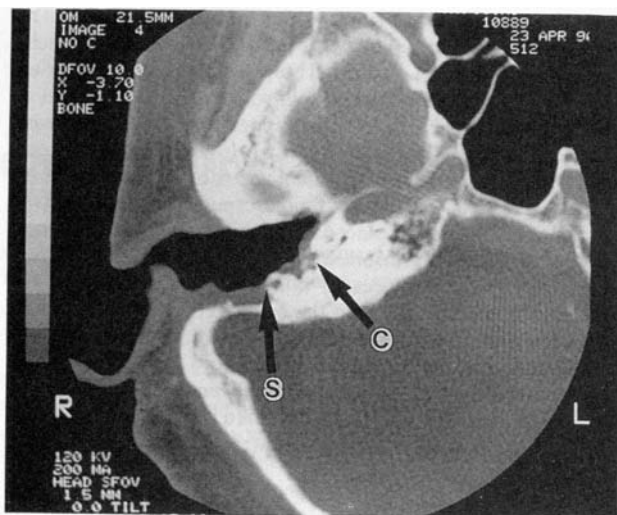


FIG. 5

Postoperative CT scan, showing the sealed fistulas of the cochlea (c) and the lateral semicircular canal (s).

result (Palva *et al.*, 1971; Abramson *et al.*, 1974; Gacek, 1974; Sheehy and Brackmann, 1979). In the present case, both semicircular canal and cochlear fistulae could be handled without special care, because the patient had no auditory or vestibular function.

A fistula of the cochlea is a rare occurrence. Its incidence in middle ear disease is 5 out of 168 by Gacek (1974), one out of 110 by Edwards (1964), two out of 830 by Palva *et al.* (1971) and one out of 212 by Abramson *et al.* (1974). The proportion of cochlear fistulae found in relation to labyrinthine fistulae is low. Ritter (1984) reported two cochlear fistulae in 50 labyrinthine fistulae.

In most cases, cochlear fistulae are located on the promontory area between the oval and the round windows (Edwards, 1964; Abramson *et al.*, 1974; Gacek, 1974). A cholesteatoma originating in the posterosuperior quadrant has more chance to affect the cochlea. This type of cholesteatoma is less common compared with that arising in the pars flaccida. This is one of the reasons that a cochlear fistula is rare. On the other hand, the vestibular organ, especially the horizontal semicircular canal may well be involved by either type of cholesteatoma.

The majority of the cochlear fistulae occur in active and infected cholesteatoma (Abramson, *et al.*, 1974; Gacek, 1974; Sheehy and Brackmann, 1979). Because of the functional vulnerability of the cochlea as compared to the vestibular organ, cochlear fistulae lead to severe sensorineural hearing loss (Sheehy and Brackmann, 1979). However, Inafuku *et al.* (1984) reported a case of a cochlear fistula with residual hearing. They stated that because the cholesteatoma matrix was dry and free from suppurative infection, the cochlear function had been preserved. Presumably, in the present case the infection was so active, that the cochlear and vestibular functions totally deteriorated, resulting in a dead ear.

A noteworthy finding is that our particular case responded to electrical stimulation to the cochlea. The frequencies used for the testing ranged from 30 to 120 Hz. This indicates that significant numbers of cochlear neurons remained. It has been

reported that nerves often tolerate the insult to the end organ and survive (Spoendlin and Gacek, 1963; Ylikoski and Belal, 1981). Even after labyrinthectomy the vestibular nerve survives, thus causing recurrence of vertigo on occasion. It has also been reported that after axotomy of the vestibular nerve, the dendrites persist in the end organ (Ylikoski and Belal, 1981). The same finding was noted in the cochlear nerve by Spoendlin and Gacek (1963). The results of promontory testing in the present case also suggest some resistance of the cochlear nerve. Most probably the vestibular neurons also persisted although a caloric response could not be elicited.

The whole process of how the middle ear structures were destroyed and led to the cochlear fistula is unknown. Since the patient was suffering from otorrhoea in childhood, it is probable that cholesteatoma associated with infection was responsible for both the fistulae and the destruction of the external bony meatal wall creating large self-cleansing natural radical cavity.

As Ritter (1984) states, a labyrinthine fistula is not easy to detect pre-operatively. The fistula test is not always positive (65%–78.5% in published series, Abramson *et al.*, 1974; Sheehy and Brackmann, 1979; Ritter, 1984). Symptoms vary from person to person and are not specific. This may be a special problem in children of younger age who do not bother much about cochlear symptoms such as hearing loss and tinnitus. Early surgery is mandatory if a fistula is to be avoided.

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