

Sudden Deafness Due to Intralabyrinthine Haemorrhage: A Possible Rare Late Complication of Head and Neck Irradiation

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Abstract

Introduction: Radiation injury resulting in sudden, late onset sensorineural hearing loss is a recognised complication in patients who have received head and neck irradiation. We describe the magnetic resonance imaging (MRI) of the internal acoustic canal (IAC) of 3 such patients and postulate a cause for these findings. **Clinical Picture:** A total of 63 patients were referred for MRI IAC for sudden-onset sensorineural hearing loss. Of these patients, only 5 patients had abnormal MRI finding in the affected ear and the remaining patients had normal studies. Two patients had acoustic neuromas. Three patients demonstrated high T1-weighted signal in the labyrinths of the affected ears and had past histories of head and neck irradiation. The MRI findings and medical records of these 3 patients were reviewed and described in this case series. **Outcome:** High labyrinthine signal on unenhanced T1-weighted images in the symptomatic ear of these patients was observed, suggesting the possibility of haemorrhage. In the patient who had a history of brain tumour, susceptibility artifacts were also seen in the right hemipons on the gradient-echo images, indicating the presence of paramagnetic substances from previous therapy. **Conclusion:** We postulate that labyrinthine haemorrhage is a rare, late complication of head and neck irradiation, resulting in sudden sensorineural hearing loss.

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Introduction

The auditory apparatus and vestibular part of the inner ear are often included in the radiation field treatment of patients with head and neck tumours.¹⁻⁵ Radiation injury resulting in late-onset sensorineural hearing loss is a recognised complication, but the underlying pathological processes are poorly understood.⁴ Magnetic resonance imaging (MRI) of the brain and internal acoustic canal (IAC) has become an important modality in the diagnosis of various diseases that may be responsible for sudden deafness. In this case series, we report the finding of labyrinthine high signal on unenhanced T1-weighted MRI of the IAC in patients who presented with sudden onset sensorineural hearing loss 5 to 20 years following head and neck irradiation. We postulate that these findings are due to labyrinthine haemorrhage as a rare, delayed complication of head and neck irradiation.

Clinical Picture

Between December 2000 and June 2003, 632 patients had MRI IAC in our department. Of these, 63 patients were

referred for sudden-onset sensorineural hearing loss. Of this group, only 5 patients had abnormal MRI IAC findings. These 5 patients comprised 2 patients who were found to have acoustic neuromas and 3 patients who demonstrated high signal in the labyrinth of the symptomatic side on the T1-weighted images. The remaining 58 patients had normal studies and their symptoms were presumed to be due to a viral or ischaemic labyrinthitis. It is not known if any of the patients with “normal studies” had previous head and neck irradiation.

The medical records of 3 patients who demonstrated high T1-weighted signal changes in the labyrinths of the symptomatic ear on MRI were reviewed. MRI of the IAC was performed at our institution using a 1.5-T MRI system (Siemens Vision, Erlangen, Germany) and a phase-array head coil. Axial oblique T1-weighted fast spin echo sequences of the temporal bone were obtained with the following parameters: TR 400 ms, TE 8 ms, 2 mm, 0 gap, 168 x 256, 90 degree flip angle, 4 NEX, FOV 180 mm. Coronal oblique T1-weighted fast spin echo images were obtained with the following parameters: TR 400 ms, TE 8

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Fig. 1a. Unenhanced T1-weighted axial image showing high signal from the left cochlea (white arrow).

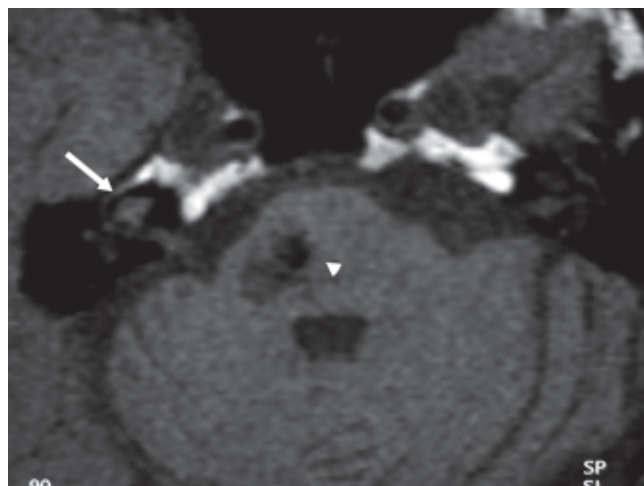


Fig. 2a. Unenhanced T1-weighted axial image showing high signal from the right cochlea (white arrow). There is also a hypointense area in the right hemipons (white arrowhead).

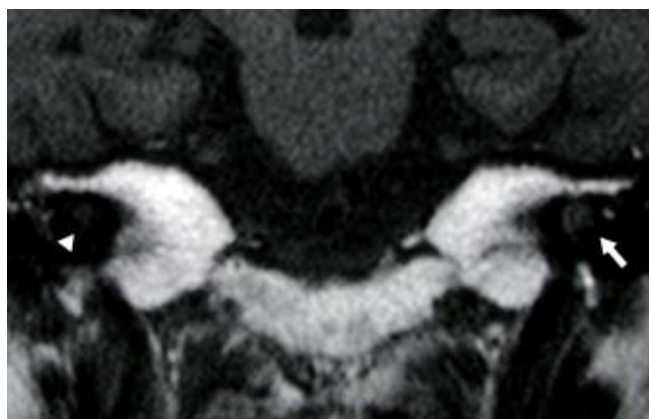


Fig. 1b. Unenhanced T1-weighted coronal images showing high signal from the left cochlea (white arrow). The right cochlea shows normal signal, which is isointense to the cerebrospinal fluid (white arrowhead).

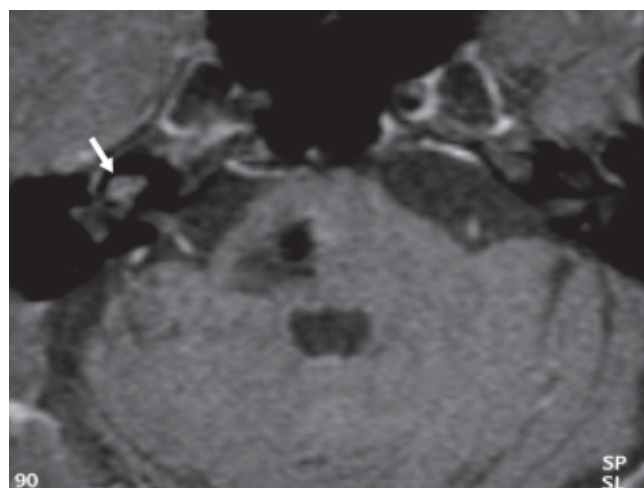


Fig. 2b. Contrast-enhanced T1-weighted axial image at the same level showing no appreciable increase in the high signal in the right cochlea (white arrow).

ms 2 mm, 10% gap, 168 x 256, 90 degree flip angle, 4 NEX, FOV 180 mm. Post-intravenous gadolinium (10 mL gadopentetate dimeglumine, Omniscan, 0.5 mmol/mL, Guerbet, Aulnay-sous-Bois France), and enhanced axial and coronal oblique images were obtained with the same parameters.

Case Reports

There were 2 male and 1 female patients whose ages ranged from 48 to 61 years. Two patients had radiotherapy for nasopharyngeal carcinoma (NPC) and the third patient had radiotherapy for a brain tumour. The individual case reports illustrate the pathologies detected in the MRI of the IAC studies.

Case 1

This 56-year-old Chinese lady presented with the sudden onset of sensorineural hearing loss in the left ear

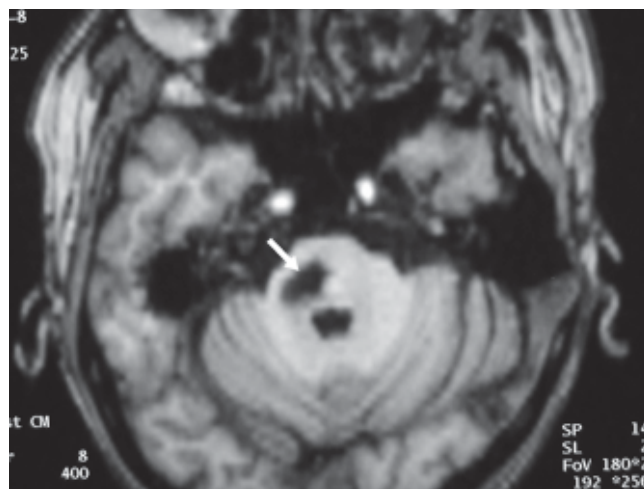


Fig. 2c. Gradient-echo image of the brain showing susceptibility artifact or the right hemipons suggestive of deposition of paramagnetic substance from treatment of the previous tumour (white arrow).

unaccompanied by vertigo. She had a history of stage 1 NPC (T2NOM0) for which she had received a radiation dosage of 70 Gy, 5 years prior to her presentation. She had remained free from disease recurrence in her subsequent clinical assessments. Her physical examination was unremarkable, with the absence of dermatologic, otologic or neurological findings. Audiogram was not performed in this patient. Laboratory evaluation consisting of full blood count (FBC), electrolytes, thyroid function test (TFT) and erythrocyte sedimentation rates (ESR) were all within normal range. MRI showed elevated signal intensity in the left cochlea in the unenhanced T1-weighted images (Figs. 1a and 1b). High-signal intensity was also observed in the post-contrast images. She was treated conservatively and did not recover from her hearing loss.

Case 2

This 48-year-old Chinese man presented with a 4-day history of right sensorineural hearing loss of sudden onset. He had a history of a brainstem tumour that had been treated with radiotherapy 20 years prior to his symptoms. The record of the radiation dose administered was not available. Physical and laboratory evaluation were otherwise normal. The audiograms revealed profound right sensorineural hearing loss. The MRI of the IAC showed right cochlea hyperintensity on the unenhanced and enhanced T1-weighted images (Figs. 2a and 2b). There were also susceptibility artifacts in the right hemipons on both the T1- and T2-weighted sequences, which demonstrated “blooming” in the gradient-echo sequence (Fig. 2c). This indicates the presence of paramagnetic substances such as haemosiderin or calcification, suggesting previous radiotherapy treatment for a pontine tumour. The patient was given a course of prednisolone at 1 mg/kg, but he did not recover his hearing loss.

Case 3

This 61-year-old Chinese man had been diagnosed with stage 1 NPC (T2NOM0) 7 years ago, for which he had received a radiation dosage of 72 Gy and he had since remained recurrence-free. He presented with sudden onset of sensorineural hearing loss in the left ear associated with vertigo and tinnitus. Physical examination was unremarkable, as were the laboratory investigations, which included FBC, TFT, ESR and electrolytes. Audiometric evaluation revealed profound left sensorineural hearing loss. The MRI also showed high signal intensity in the left cochlea in the unenhanced and enhanced T1-weighted sequences. The patient was treated conservatively and defaulted on subsequent clinical follow-up appointments.

Discussion

Radiotherapy is an integral component in the treatment

of various head and neck tumours. Continued refinements in radiation treatment strategies have reduced the radiation exposure to sensitive and vital structures in the central nervous system and visual apparatus. However, otologic structures are often included in the treatment field and may be injured, resulting in hearing loss.¹⁻⁵ Studies have shown that patients with NPC, in particular, are at high risk of radiation exposure to the ear.⁶ In brain tumour patients who have received radiotherapy, late hearing loss has been observed with fraction doses of less than 2 Gy and total doses of less than 60 Gy.⁴

The onset of sensorineural hearing loss in patients who have received radiation to the head and neck is varied and in cases of NPC, the incidence ranges from 3% to 54%.¹ Hearing loss may be progressive or sudden and may occur as early as 3 months after irradiation or many years later.^{2-5,7} A proportion of patients suffer post-irradiation sudden deafness (PISD), defined as the sudden onset of deafness in patients who have previously received irradiation of the temporal bone.⁷ In a large retrospective study of patients who had radiotherapy for NPC, Young et al⁷ found that PISD was a rare, late complication of radiotherapy in 15 patients, occurring between 4 and 23 years after treatment with a mean interval of 12 years, comparable to the 5 to 20 years observed in our patients. Profound or total sudden hearing loss was common and high-frequency hearing loss was frequent. Other authors have also made similar observations in patients who have had irradiation for NPC and brain tumours.¹⁻⁵ The clinical findings in these patients were ascribed to a variety of causes, including radiation necrosis of the temporal lobe, VIII nerve damage, tumour infiltration, otomastoiditis, endarteritis obliterans of the stria vascularis and damage to the organ of Corti.^{1-5,7,8} In most of these studies, however, the patients were evaluated with tympanometry and audiometry and the few that had magnetic resonance (MR) evaluation, mainly described temporal lobe necrosis, tumour infiltration and radiation otomastoiditis as causes for the hearing loss.^{7,8}

MRI has emerged as a powerful tool in the evaluation of endocochlear diseases that cause sudden sensorineural hearing loss.⁹⁻¹¹ Weissman et al¹² found high intralabyrinthine signal on unenhanced T1-weighted images of 2 patients with sudden deafness. The authors attributed the findings to small amounts of haemorrhage or elevated concentrations of protein macromolecules within the labyrinth. This theory was supported by Shinohara et al¹³ in a study of 5 patients with sudden sensorineural hearing loss. Mild contrast enhancement on T1-weighted images is frequently observed, although it may be difficult to appreciate the increase in signal intensity after intravenous contrast when the pre-contrast T1-weighted images already show high signal.^{12,13}

Intralabyrinthine haemorrhage is a known but uncommon cause of sudden sensorineural hearing loss. It has been attributed to coagulopathy,^{14,15} non-steroidal anti-inflammatory drugs,⁹ trauma or labyrinthitis^{11,12} and is uncommon in healthy individuals with no predisposing factors.¹² To our knowledge, the MR imaging features of labyrinthine haemorrhage associated with sudden sensorineural hearing loss as a delayed complication of head and neck irradiation have not been described. The MRI IAC studies of all our patients showed high signal in the labyrinth of the symptomatic ear on the unenhanced T1-weighted images. One of our patients had MRI features of old haemorrhage in the right hemipons, possibly from a previous brainstem tumour that had been treated with radiotherapy.

There are many studies in the literature describing the pathological changes in irradiated animal ears, which include haemorrhage, lymphatic oedema with compression of the endolymph disruption of the organ of Corti in the cochlea and inflammation.¹⁶⁻¹⁸ These are mostly acute processes that were observed in experimental animals sacrificed soon after irradiation and there are few studies that describe the pathological alterations responsible for delayed sensorineural hearing loss. In one such study, Bohne et al¹⁹ found degeneration of sensory and supporting cells and loss of the VIII nerve fibres in the organ of Corti, 2 years after the experimental animals had been irradiated. Specifically, delayed labyrinthine haemorrhage has not been described in both animal and human subjects after head and neck irradiation. However, delayed *intracerebral haemorrhage* has been observed in patients who have received radiation dosage between 18 and 60 Gy for the treatment of brain tumours. This phenomenon has been attributed to radiation vasculopathy.²⁰⁻²⁴ Capillaries, in particular, are the most radiosensitive component of the vasculature.²⁴ It has been shown that cranial irradiation disrupts the tight junctions between vascular endothelial cells resulting in the destruction of the blood-brain barrier.²² Radiation-induced microvascular injury also results in focal alterations in the fibrinolytic enzyme system, causing thrombosis and occlusion. In addition, capillary teleangiectasis with proliferation of small friable blood vessels occur and may result in haemorrhage.²⁰⁻²⁴ Why these haemorrhages occur in a delayed fashion is poorly understood. One theory is that radiation injury has been known to require a few cell cycles to manifest and endothelial cells have long turnover times, in the range of months to years.²¹

The blood-labyrinth barrier is similar to the blood-brain barrier. It is also characterised by endothelial tight junctions, tightly regulated pinocytotic vesicle transport and negatively charged basal lamina.²⁵ In addition, the stria vascularis is a

multilayered epithelium comprising many capillaries. It is therefore conceivable that the same radiation vasculopathy that causes delayed haemorrhage in the brain may also occur in the inner ear in patients who have had head and neck irradiation. We hypothesise that disruptions in the blood-labyrinth barrier, teleangiectasis and small vessel proliferation of the stria vascularis may result in labyrinthine haemorrhage.

Our patients had normal platelet levels and clotting profiles. They had no clinical findings suggestive of recent viral infection and they had not consumed ototoxic drugs. The MR studies did not demonstrate any evidence of tumour recurrence. This made other causes of sensorineural hearing loss, in the setting of head and neck malignancy, such as drug toxicity, infective labyrinthitis and tumour infiltration, unlikely. In case 2, the patient's symptoms are unlikely to be due to the post-irradiation haemorrhagic or calcific changes in the pons. If this was the cause, we would expect the patient to have bilateral sensorineural hearing loss, worse on the contralateral side, rather than ipsilateral hearing loss because two-thirds of the nerve fibres arising from the cochlear nucleus in the pons at the level of the inferior cerebellar peduncle cross to the contralateral side. The only common factor in all our patients was a history of previous head and neck irradiation. Although no pathological proof was available in this study, we postulate that the high signal in the labyrinths noted on the unenhanced T1-weighted MRI images of the IAC was due to haemorrhage occurring as a result of previous head and neck irradiation.

In conclusion, we describe the MR imaging findings in 3 patients who presented with sudden sensorineural hearing loss 5 to 20 years after radiotherapy. The finding of high signal in the labyrinths of our patients on unenhanced T1-weighted images, in the absence of other causes, led us to postulate that labyrinthine haemorrhage may be a late sequelae of head and neck irradiation, despite the lack of histological proof. Although the recognition of this finding does not impact clinical management in terms of active intervention, it is nevertheless useful information for the clinician, who has to explain and reassure the patient as to the cause of his frightening symptoms. Therefore both the clinician and the radiologist should be aware of this differential diagnosis in a patient with sudden deafness and a history of head and neck irradiation.

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