Cancer Imaging (2008) **8,** 191–198
DOI: 10.1102/1470-7330.2008.0031

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REVIEW ARTICLE

Imaging of carcinoma of the external auditory canal: a pictorial essay

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Date accepted for publication 17 September 2008

Abstract

Carcinoma of the external auditory canal presents a challenge in management, largely due to limited experience in treating this rare disease and the lack of a universally accepted staging system. Prognosis is most dependent on the extent of local disease at presentation, while resection margin status is also a strong determinant of survival in post-operative patients. The intent of this pictorial essay is to review the pattern of tumour spread and highlight the value of imaging, particularly magnetic resonance imaging in pre-operative tumour mapping.

Keywords: *Carcinoma; external auditory canal; computed tomography; magnetic resonance imaging.*

Introduction

Carcinoma of the external auditory canal (EAC) is a rare malignancy. Treatment planning is challenging because the accumulated experience of any particular individual or institution in managing this tumour is rather limited. This problem is further compounded by the absence of a universally accepted tumour staging system, making treatment outcome comparison difficult. It is now known that the single most important factor determining survival is the local tumour extent at presentation[1]. It is therefore crucial to establish the disease extent in order to optimize treatment options. The patterns of tumour spread are well described in the surgical literature[2,3]. Computed tomography (CT) can demonstrate bony erosion with exquisite detail but magnetic resonance (MR) imaging is preferred in the delineation of tumour spread along vascular channels, neural pathways, intracranial extension and involvement of the extra-temporal soft tissues.

Radiological features

On temporal bone CT, early carcinoma of the EAC commonly presents as a soft tissue mass within the EAC. As the disease progresses, aggressive underlying bony destructive changes are demonstrated. As the tumour infiltrates and spreads deep into the surrounding tissue, it is best shown as a heterogeneously enhancing lesion on contrast-enhanced MR imaging.

Carcinoma of the EAC most commonly spreads through the floor of the EAC into the soft tissue below the temporal bone[4–6]. This may be related to the presence of the fissures of Santorini along the inferior aspect of the cartilaginous EAC. Less frequently, the tumour erodes posteriorly into the mastoid bone or anteriorly through the tympanic portion of the EAC to invade the temporomandibular joint (Fig. 1)[7]. On the other hand, superior extension into the middle cranial fossa and medial extension into the middle ear are rare[4–6,8].

Medial extension into the middle ear cavity is of special interest because of the resultant poor prognosis[8]. This negative prognostic indicator is related to the subsequent disease spread beyond the middle ear making surgical excision increasingly difficult. Tumour in the middle ear can readily extend superiorly through the thin tegmen tympani into the middle cranial fossa (Fig. 2). However, the presence of temporal lobe oedema

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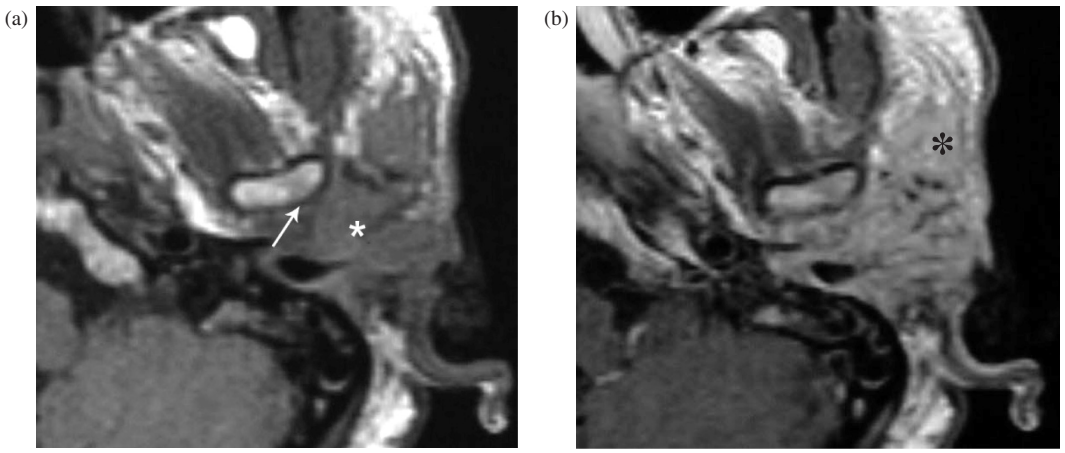

Figure 1 **A 64-year-old female with EAC carcinoma involving the temporomandibular joint. (a) Axial T1-weighted MR image shows intermediate signal tumour in the left EAC (asterisk) extending into the temporomandibular joint and displacing the mandibular condyle (arrow) anteriorly. (b) Axial contrast-enhanced MR image shows tumour enhancement. Note the tumour extension into the left parotid region (asterisk).**

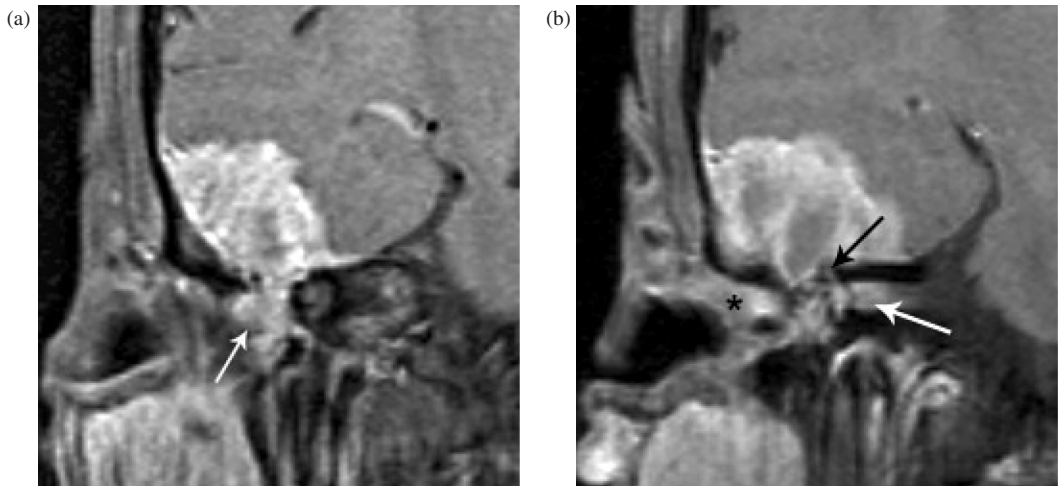

Figure 2 **A 58-year-old male with right EAC carcinoma. (a) Coronal contrast-enhanced MR image shows tumour in the right middle ear (arrow) with extension through the tegmen tympani into the middle cranial fossa. (b) Coronal contrast-enhanced MR image (posterior to (a)) shows tumour in the right EAC (asterisk) with middle ear and intracranial extension. Note enhancement in the labyrinth (black arrow) and the internal auditory canal (white arrow).**

associated with tumour extension into the middle cranial fossa does not necessarily indicate cerebral parenchymal infiltration.

From the middle ear cavity, tumour can spread medially to erode the inner ear structures. Tumour may gain access into the labyrinth through the lateral semicircular canal, oval or round windows. Malignant infiltration often extends further proximally along the vestibular or cochlear nerves into the internal acoustic canal (Fig. 2). The resistant cortical bone of the inner ear, especially the otic capsule, often limits the amount of gross bone destruction but aggressive tumours will eventually involve the cochlea (Fig. 3).

In contrast to the difficulty of invading cortical bone, tumour can readily spread from the middle ear posteriorly through the aditus into the mastoid antrum (Fig. 4). Further posterior spread leads to extension into the posterior cranial fossa and involvement of the sigmoid sinus (Fig. 5).

Tumour erosion through the floor of the middle ear anteriorly results in the involvement of the carotid artery while inferior extension through the floor posteriorly involves the jugular foramen (Fig. 6). Cancers with extensive inferior spread involving the jugular bulb and internal carotid artery exclude the surgical options of lateral temporal bone resection and subtotal

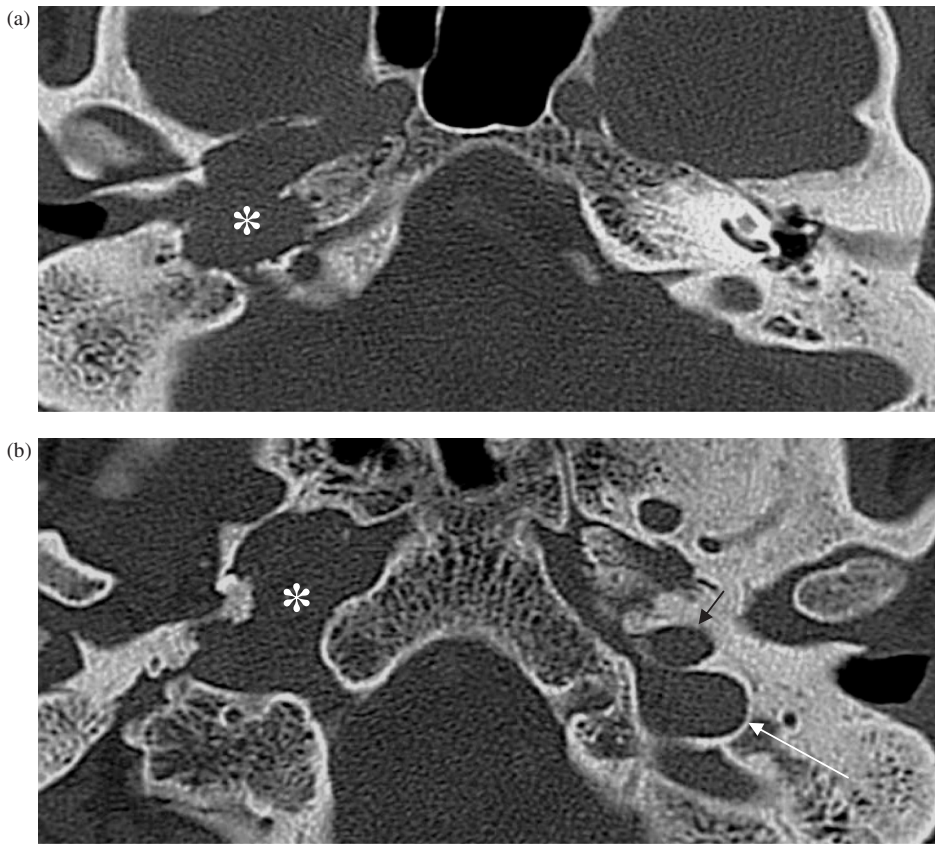

Figure 3 **(a) Axial CT shows a right EAC tumour eroding the otic capsule and destructing the cochlea (asterisk). Note the normal left cochlea for comparison. (b) Axial CT (inferior to Fig. 4a) shows inferior spread of the right EAC tumour involving the internal carotid artery and jugular bulb (asterisk). Note the normal left carotid foramen (black arrow) and jugular foramen (white arrow) for comparison.**

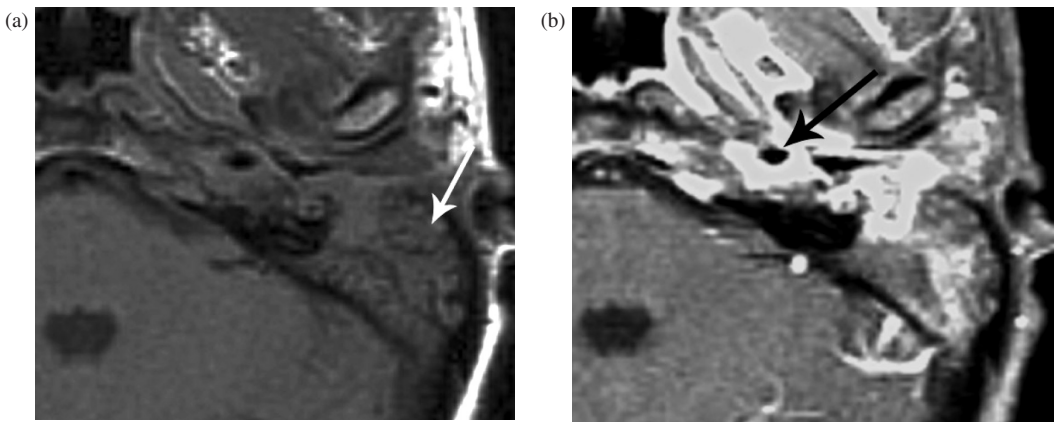

Figure 4 **(a) Axial T1-weighted MR image shows an intermediate signal intensity tumour in the left middle ear cavity with extension posteriorly into the mastoid (arrow). (b) Axial contrast-enhanced MR image shows heterogeneous tumour enhancement. Note tumour surrounding the left petrous internal carotid artery (arrow).**

temporal bone resection, and are often unresectable (Figs. 6 and 7).

Tumour in the hypotympanum often extends anteromedially along the Eustachian tube. This manner of spread can be diagnosed by the identification of soft tissues anterior and parallel to the petrous segment of the internal carotid artery (Fig. 8). When this manner of spread occurs, tumour comes into close proximity with the foramen ovale and the mandibular nerve in the central skull base. Proximal perineural

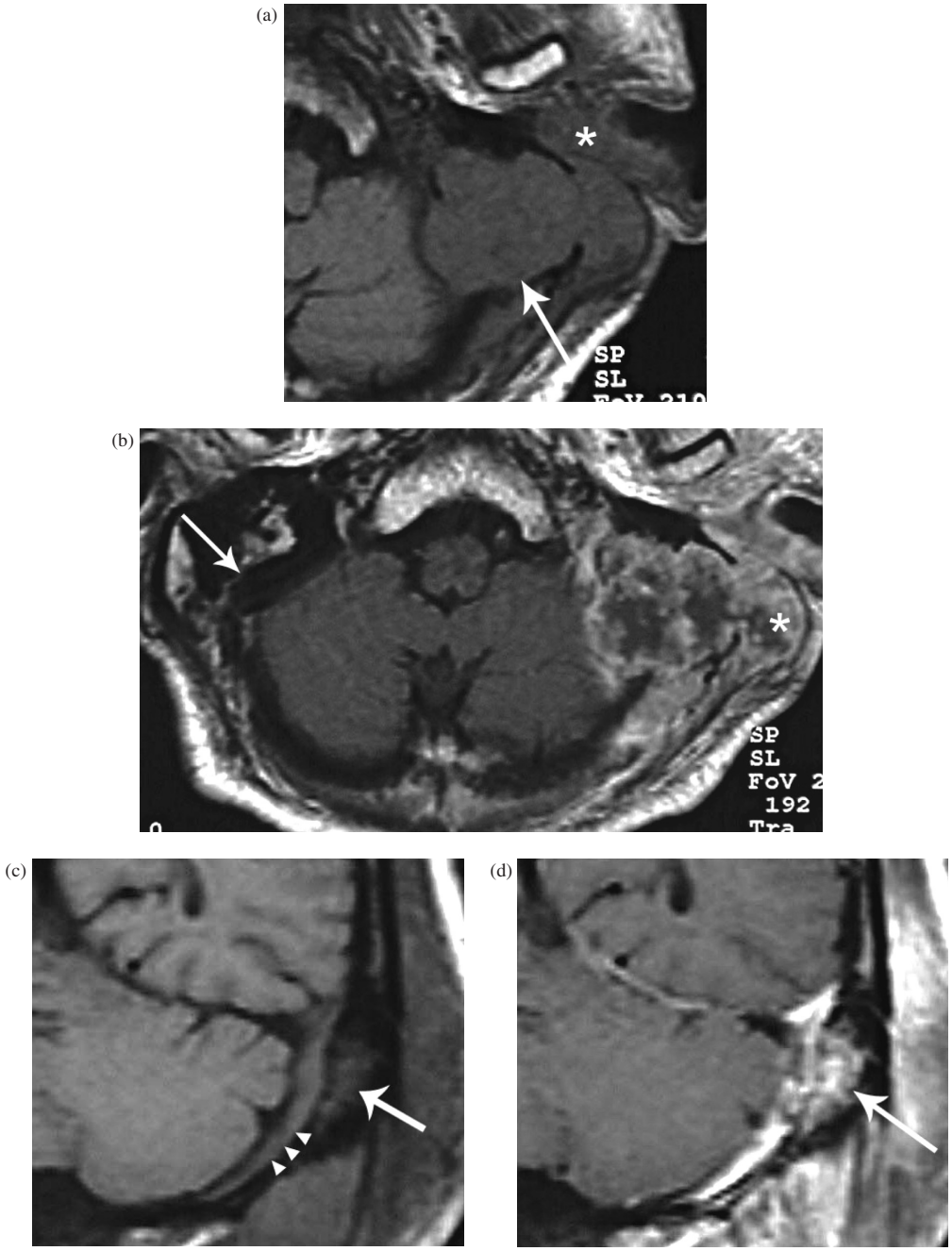

Figure 5 **A 55-year-old female with a left EAC tumour extending into the posterior cranial fossa. (a) Axial T1-weighted MR image shows intermediate signal intensity tumour in the left EAC (asterisk) with extension into the posterior cranial fossa (arrow). (b) Axial contrast-enhanced MR image shows heterogeneous contrast enhancement in the tumour. There is obliteration of the left sigmoid sinus (asterisk). Note normal signal void in the right sigmoid sinus for comparison (arrow). (c) Coronal T1-weighted MR image shows intermediate signal intensity tumour in the posterior aspect of the left mastoid bone (arrow) and along the dural venous sinus (arrowheads). (d) Coronal contrast-enhanced image shows enhancement of the tumour (arrow).**

spread along the mandibular nerve leads to intracranial extension. At this stage, signal changes in the muscles of mastication may be seen, indicating the onset of denervation atrophy. Tumour extending medially along the Eustachian tube may eventually reach the nasopharynx (Fig. 8). The superior portion of the parapharyngeal space is adjacent to the Eustachian tube. The parapharyngeal space is a well-recognized route for the spread of inflammatory and malignant disease[9]. Tumour may therefore extend into the parapharyngeal space and spread into the neck with relative ease[10].

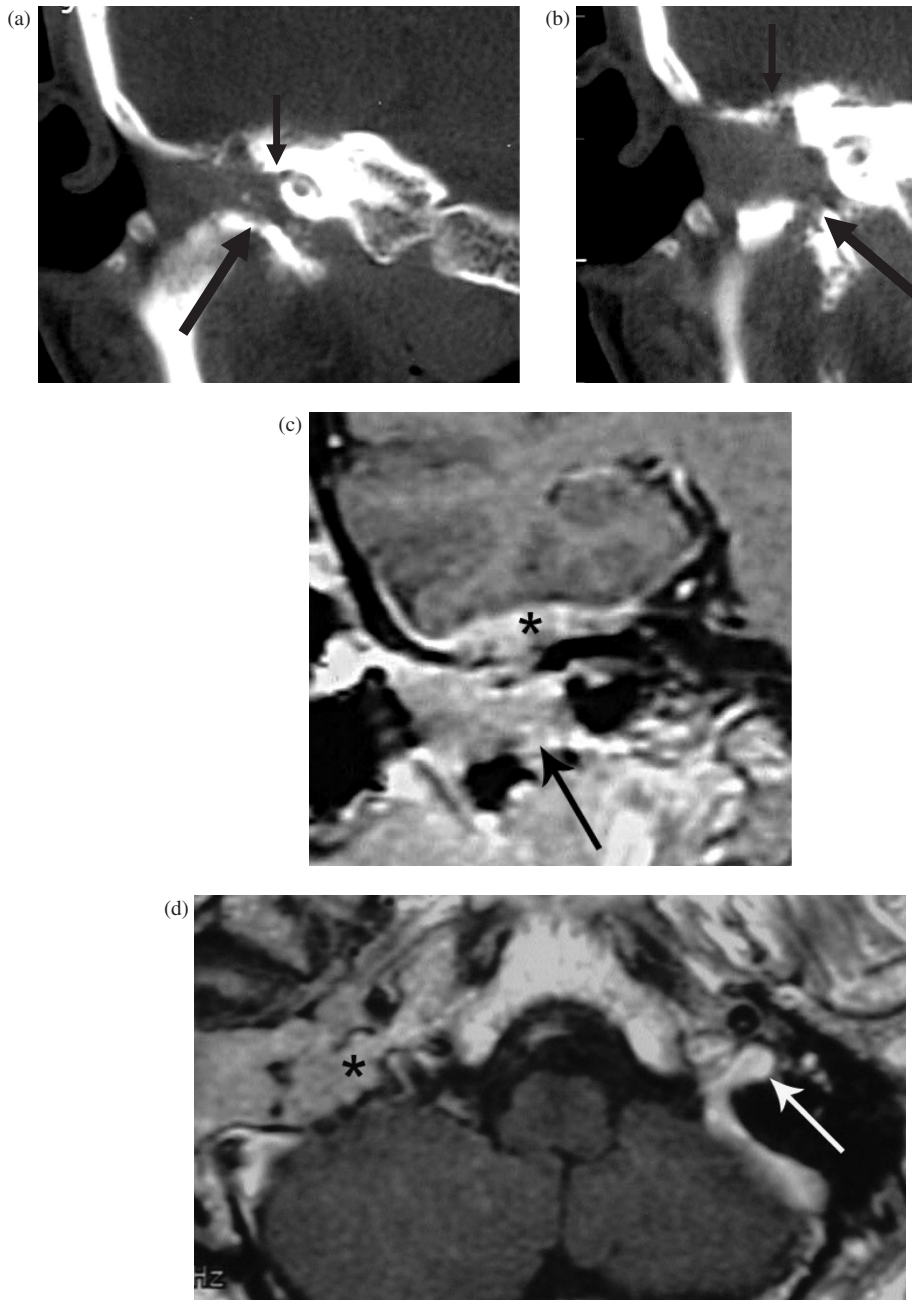

Figure 6 **A 57-year-old male with carcinoma of the right EAC showing intracranial infiltration and spread below the skull base. (a) Coronal CT shows tumour in the right EAC eroding the cochlea (short arrow) and the floor adjacent to the carotid canal (long arrow). (b) Coronal CT (posterior to (a)) shows permeative erosion of the roof of the right middle ear (short arrow), and the floor adjacent to the jugular foramen (long arrow). (c) Coronal contrast-enhanced MR image shows enhancing tumour in the right EAC spreading into the right middle cranial fossa causing gross dural thickening (asterisk). Note tumour extension through the floor of the middle ear cavity (arrow). (d) Axial contrast-enhanced MR image shows moderate tumour enhancement. The tumour has obliterated the right jugular foramen and vein (asterisk). Note the normal enhancing left jugular vein (arrow) for comparison.**

Lymph node disease in carcinoma of the EAC is a significant negative prognostic factor[6]. Any malignant nodal involvement should therefore be classified as advanced disease. The parotid nodes are the first echelon nodes[5,6]. Malignant pre- and post-auricular lymphadenopathy is also commonly seen.

Discussion

The incidence of carcinoma of the EAC is estimated as between one and six cases in every one million population. The majority of tumours occur in the fifth and sixth decades of life. Pooled data shows approximately even

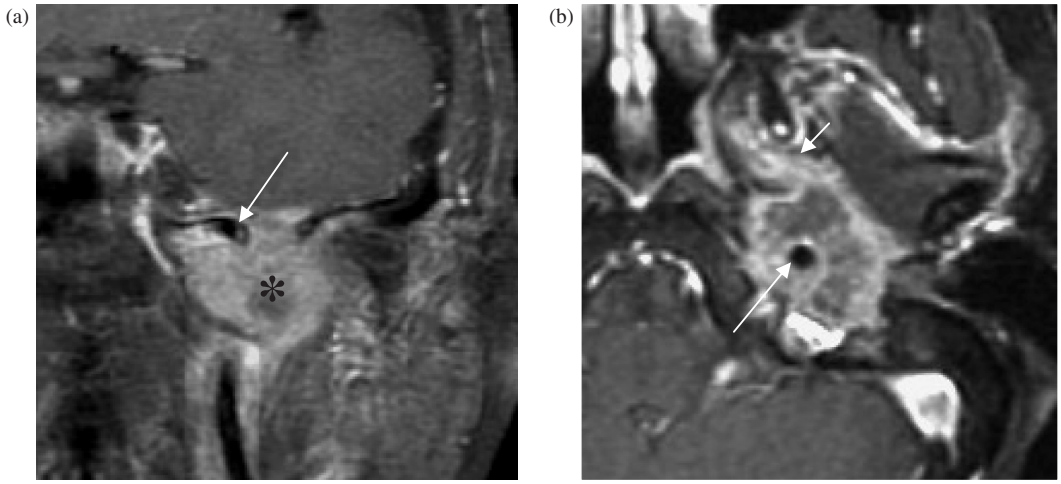

Figure 7 **(a) Coronal contrast-enhanced MR image shows a left EAC tumour (asterisk) spreading and involving the left internal carotid artery (arrow). (b) Axial contrast-enhanced MR image shows the tumour with heterogenous enhancement surrounding the left petrous internal carotid artery (long arrow). Note tumour infiltration along the left Eustachian tube anteromedially (short arrow).**

gender distribution[8]. Several aetiological factors have been implicated, including exposure to ultraviolet radiation, chlorinated disinfectants and human papillovirus infection. Chronic otitis media has been implicated for a long time as an aetiological factor but no definite correlation has been proven. A high association has also been reported in patients previously treated with radiation therapy[11]. Histologically, most of these tumours are squamous cell carcinomas, others being mucoepidermoid and adenoid cystic carcinomas.

Carcinoma of the EAC is treated with surgical resection with or without post-operative radiation therapy. Tumours confined to the cartilaginous portion of the EAC are removed by local wide excision. Tumours arising from or spreading to the bony EAC but lateral to the tympanic membrane are treated by limited partial temporal bone resection or 'sleeve' resection with subsequent reconstruction by a local flap or a microvascular myocutaneous free flap, and refashioning of an auditory canal for preservation of hearing. Tumour involvement of the tympanic membrane, malleus, incus and tympanic bone may necessitate lateral temporal bone resection, which was first reported by Conley and Novack[12].

Subtotal temporal bone resection, pioneered by Parson and Lewis[13], is a combined extracranial and intracranial procedure for treating advanced cancers involving the middle ear, the temporomandibular joint and the middle cranial fossa. The facial nerve is almost invariably sacrificed, as opposed to lateral temporal bone resection. However, the inferior margins of resection are similar in both lateral temporal bone resection and subtotal temporal bone resection, being just lateral to the internal jugular vein and internal carotid artery.

In patients with even more extensive disease, total temporal bone resection may improve patient survival as well as increase the palliative benefits, notably decreased pain and improved hygiene[14]. However, there is increased risk of morbidity, including damage to the cavernous sinus and notably, the internal carotid artery as well as the third, fourth, fifth and sixth nerves. A viable alternative in advanced disease is piecemeal resection of all visible tumour beyond the margins of a subtotal temporal bone resection, followed by post-operative radiation therapy[15].

En bloc surgical resection with complete tumour excision remains the primary aim and the critical determinant of patient survival. In two retrospective studies, the 5-year survival rate was almost 80% for patients with negative resection margins at the time of surgery, significantly higher than those with positive margins (0–35%)[3,16]. In order to achieve clear margins, most cases will invariably require a superficial or total parotidectomy and not uncommonly, a partial mandibulectomy and clearance of the infratemporal fossa.

Surgical planning therefore, involves meticulous preoperative radiological tumour mapping. A high-resolution, non-enhanced bone algorithm temporal bone CT and contrast-enhanced MR imaging are recommended for this purpose.

For early disease confined to the EAC, CT is sufficient. CT can also readily detect erosion of the bony external auditory canal, which is often the first sign of local spread of tumour. Axial and coronal sections are routinely acquired. Sagittal reconstruction may provide additional value for complex tumour infiltration of the temporal bone. However, CT is inadequate to demonstrate the extent of tumour spread beyond the EAC. Contrast-enhanced MR imaging is thus preferred over contrast-enhanced CT.

Post-contrast T1-weighted sequencing with fat saturation in the axial, coronal and sagittal planes allows the

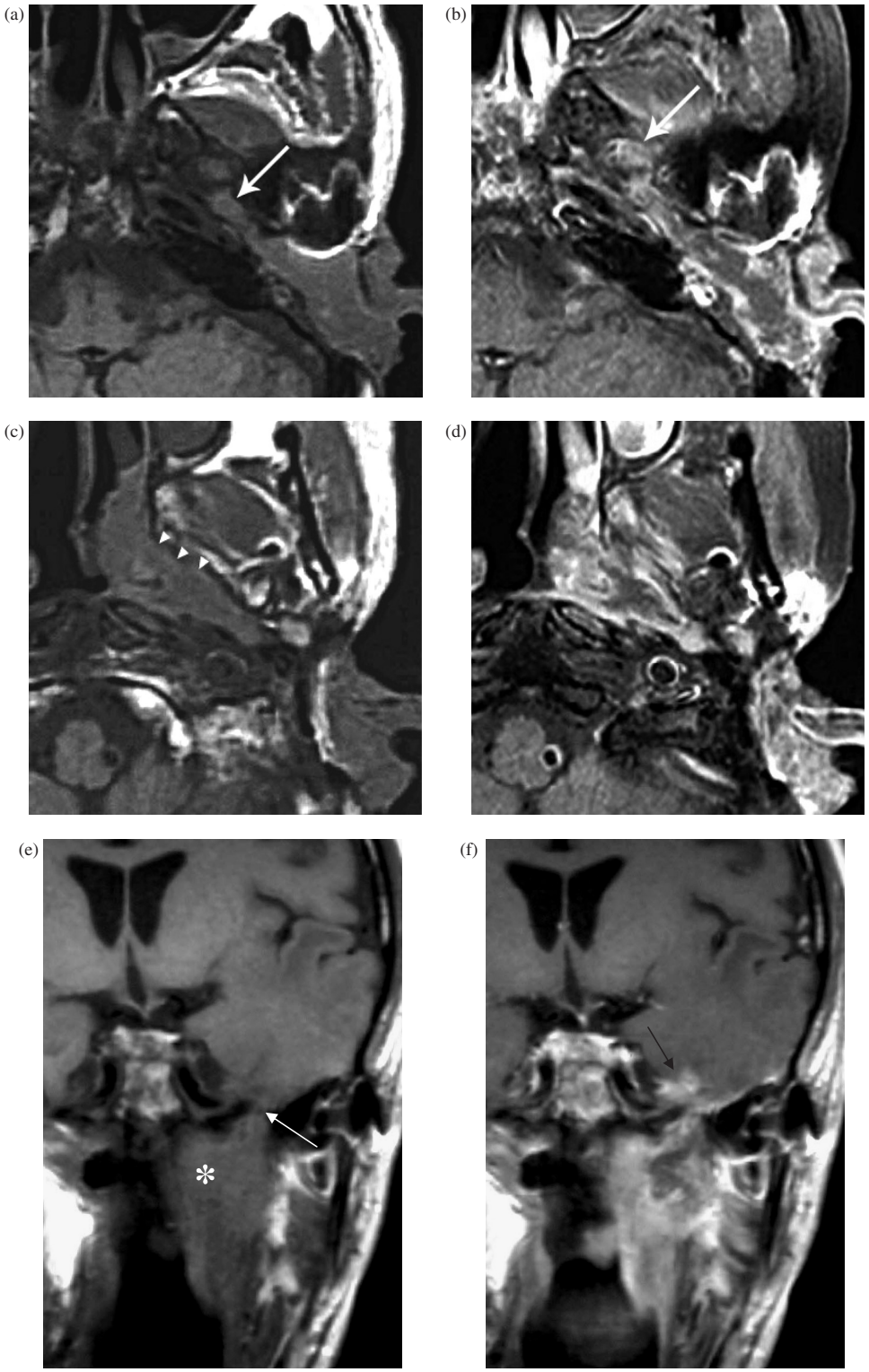

Figure 8 **A 67-year-old male with recurrent carcinoma of left EAC with spread to the nasopharynx. (a) Axial T1-weighted MR image shows an intermediate signal intensity tumour in the left EAC and middle ear extending anteriomedially along the Eustachian tube (arrow). (b) Axial contrast-enhanced image shows heterogeneous enhancement of the tumour with non-enhancing areas, probably due to necrosis. Note enhancement in the left foramen ovale (arrow). (c) Axial T1-weighted MR image shows tumour in the left parapharyngeal space and nasopharynx (arrowheads). (d) Axial contrast-enhanced image shows heterogeneous tumour enhancement. (e) Coronal T1-weighted MR image shows an intermediate signal intensity tumour in the left nasopharynx (asterisk) extending to the left foramen ovale (arrow). (f) Coronal contrast-enhanced MR image shows tumour enhancement and intracranial spread (arrow). Note the oedematous left temporal lobe, which may be due to radiation therapy, intracranial tumour or both.**

best evaluation of the extent of tumour spread into the surrounding extra-temporal anatomic landscape. The entire parotid space should be included for evaluation of echelon lymph nodes.

Intracranial extension is much better seen on coronal sections of contrast-enhanced MR images. The earliest sign seen on MR imaging is mild dural thickening and enhancement along the floor of the middle cranial fossa in the vicinity of the tegmen tympani. Early dural involvement is difficult, if not impossible, to recognize on CT. Although CT demonstrates erosion of the cortical bone of the inner ear very well, spread into the labyrinth is better demonstrated on MR imaging. Perineural involvement of the seventh and eighth cranial nerves cannot be diagnosed with confidence without contrast-enhanced MR imaging. Similarly, MR imaging provides further information on the status of the skull base neurovascular structures when bony erosions are identified on CT.

The differential diagnosis of EAC carcinoma includes necrotizing external otitis and osteoradionecrosis. Necrotizing external otitis is typically described in older patients with diabetes. Diabetes mellitus is found in 95% of adult patients with necrotizing external otitis. Necrotizing external otitis tends to show comparatively more extensive inflammatory changes with cellulitis and abscesses involving the adjacent deep cervical spaces, most commonly the parotid space[17,18]. Osteoradionecrosis of the temporal bone is a delayed complication following radiotherapy for head and neck tumours. The temporal bone is particularly susceptible due to its relatively poor blood supply. While cortical erosion of the bony EAC may simulate carcinoma, the prominent features in osteoradionecrosis involve the mastoid air cells with disruption of the air cell septation, usually on a background of underlying radiation-induced otomastoiditis[19]. Diffuse permeative demineralization of the skull base beyond the temporal bone can be seen in patients with severe osteoradionecrosis.

Nevertheless, the clinical presentation and imaging appearance of EAC carcinoma may exactly mimic necrotizing external otitis or osteoradionecrosis. As early diagnoses significantly affect the eventual outcome, any clinically suspicious EAC lesion with bony erosion should be subjected to early biopsy. As with other squamous cell carcinoma of the head and neck, more than one biopsy should be taken as there may be non-diagnostic changes at the periphery of the tumour and malignant cells may be missed with only a single biopsy[21].

Conclusion

In addition to CT, MR imaging is essential for precise delineation of the extent of tumour spread beyond the EAC, which at presentation, is the single most important prognostic factor. This accurate pre-operative tumour mapping is also critical in deciding surgical options, with a clear resection margin the primary objective.

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