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Clinical Records

Malignant otitis externa with optic neuritis

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Abstract
Malignant otitis externa is a serious condition that presents difficulties in treatment, and also in monitoring its progress. A case of malignant otitis externa with optic neuritis is presented that remained refractory to standard treatment but was cured by adjuvant hyperbaric oxygen therapy. This is the only reported case that has survived this disease with optic neuritis. The usefulness of imaging techniques in this condition is discussed, as well as the ESR, in evaluating the effectiveness of treatment.

Key words: Otitis externa; Hyperbaric oxygenation; Optic neuritis

Introduction

Malignant otitis externa is an uncommon, life-threatening disease that occurs primarily in elderly diabetics. It is a severe, progressive skull base infection that begins in the external ear canal or middle ear, usually caused by *Pseudomonas aeruginosa.* It was first described in 1959 (Meltzer and Kelemen, 1959) and the syndrome later termed by Chandler in 1968 (Chandler, 1968).

Its presentation is similar to otitis externa. Recognized features include purulent otorrhoea with granulation tissue in the external ear canal. However, an important hallmark of this condition is of a severe unremitting headache/otalgia that appears disproportionate to the clinical signs even after adequate local treatment. Progression of the disease leads to multiple cranial neuropathies. Intracranial complications can ensue, with meningitis and septic venous sinus thrombosis, which may be fatal (Rubin and Yu, 1988). In the only two cases reported of malignant otitis externa complicated with optic nerve involvement the mortality is 100 per cent (Dinapoli and Thomas, 1971; Holder *et al.*, 1986).

The treatment of this condition necessitates early diagnosis and long-term high dose anti-pseudomonal antibiotics (Strauss *et al.*, 1982; Leggett and Prendergast, 1988; Lang *et al.*, 1990; El-Silimy and Sharnuby, 1992). This may be combined with surgical debridement of areas of necrotic tissue (Doroghazi *et al.*, 1981), and also more recently hyperbaric oxygen therapy (Mader and Love, 1982; Shupak *et al.*, 1989; Davis *et al.*, 1992). However, difficulty remains in monitoring the course of the disease due to its 'deep-seated' nature. The decision as to when treatment can safely be stopped remains arbitrary and a number of cases are refractory to treatment.

Case report

A 46-year-old Caucasian insulin-dependent diabetic man presented with otalgia and otorrhoea from his left ear. Examination revealed an otitis externa with an inflamed ear canal containing pus and granulation tissue and a peri-auricular cellulitis. Swabs were taken for culture, but no organism was subsequently grown. The condition responded rapidly to intravenous antibiotics and antibiotic ear-drops, and the patient was discharged home within a few days.

Two weeks later the patient was re-admitted with increasing otalgia and otorrhoea. He had also noticed some weakness of the left side of his face. Examination revealed an infection of the left ear canal and a left-sided facial nerve palsy but no other neurological abnormalities. A computed tomography (CT) scan was performed which showed soft tissue in the left mastoid air cell system

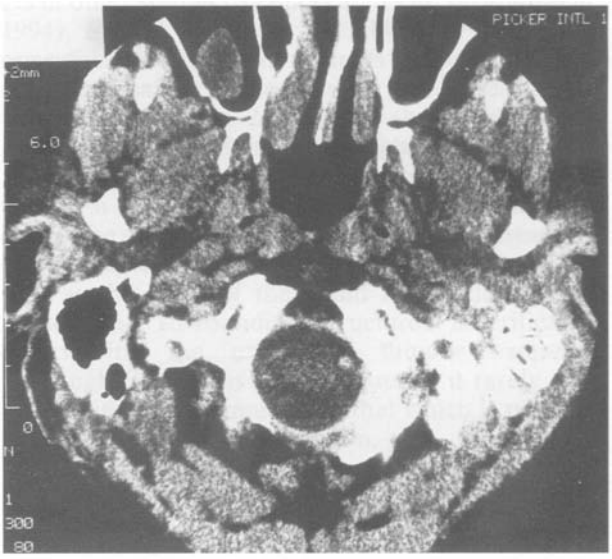

Fig. 1
CT scan showing soft tissue obliterating the left mastoid air cell system.

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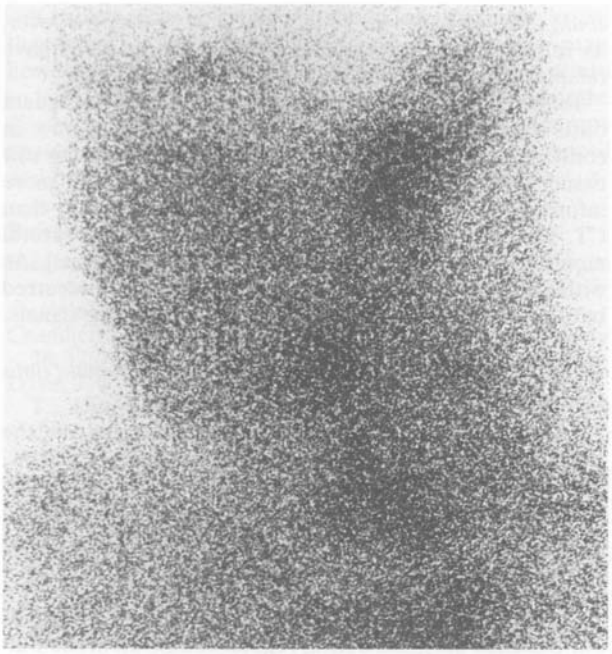

FIG. 2
Gallium scan (left lateral oblique view) showing increased uptake in the left temporal bone.

extending to its tip with bony destruction (Figure 1). A diagnosis of malignant otitis externa was made and treatment initially commenced with an intravenous cephalosporin. It was decided, on this evidence, to decompress the facial nerve. An extended cortical mastoidectomy was performed, the inflammatory tissue was excised and, the horizontal and vertical segments of the facial nerve decompressed. Histological examination of the tissue in the mastoid cavity confirmed this to be inflammatory granulation tissue and culture of this tissue did not isolate a pathogen.

Given the likelihood of a pseudomonas infection, the antibiotic therapy was changed to ciprofloxacin and the patient was discharged home 10 days later on oral

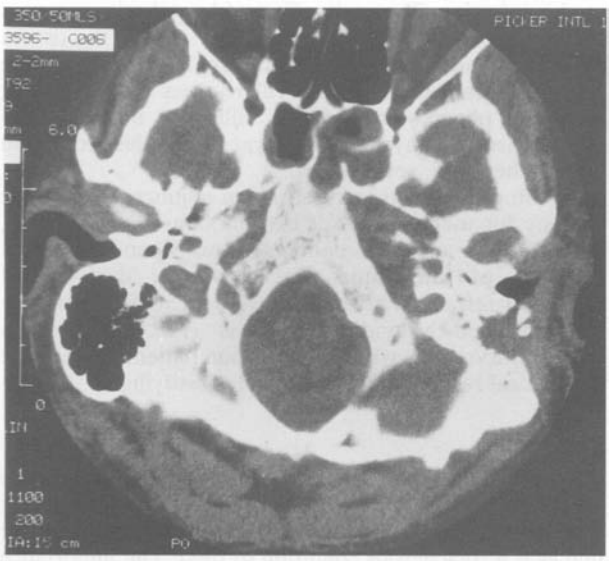

FIG. 3
CT scan showing erosion of the left temporal bone with soft tissue swelling of the post-nasal space (after the initial extended cortical mastoidectomy).

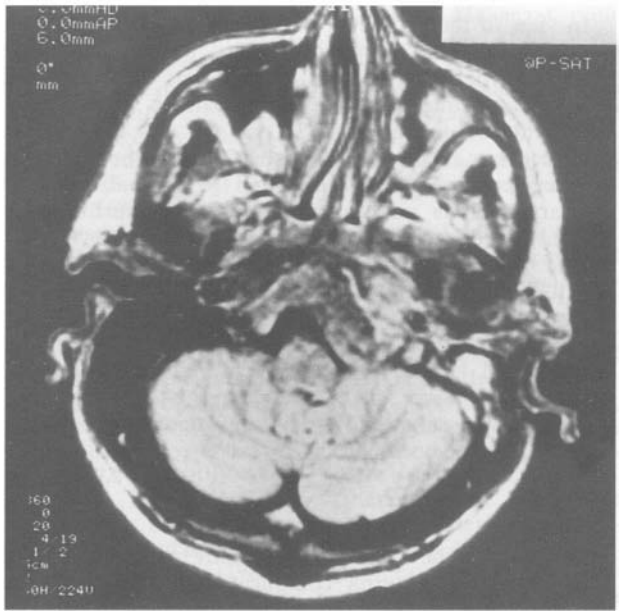

FIG. 4
MRI scan showing abnormal signal of the petrous part of the left temporal bone with compression of the brain stem.

antibiotics. A base line gallium scan predictably showed increased uptake in the left temporal bone (Figure 2). Two subsequent gallium scans were performed at two monthly intervals and these were normal. The antibiotics were eventually discontinued some five months after the initial presentation.

Whilst off treatment the patient began to get headaches that became increasingly severe over the following month and led to his re-admission. He had lost weight and felt generally unwell. On examination, he was apyrexial and his left ear canal contained purulent debris with bare bone exposed in its floor. Examination of his cranial nerves revealed a left-sided facial nerve palsy, which had not recovered its function since his previous admission, palatal asymmetry and a paralysed left vocal fold. Haematological investigations showed a moderate leucocytosis and thrombocytosis consistent with a chronic inflammatory condition. The erythrocyte sedimentation rate (ESR) was 84 mm/hr and the C-reactive protein (CRP) was 165 mg/l. A CT scan was performed and showed extensive erosion of the temporal bone (Figure 3) along with soft tissue swelling of the post-nasal space. Treatment re-commenced for his malignant otitis externa after discussion with the microbiologists. High dose intravenous triple antibiotic therapy – ceftazidime, metronidazole and amoxycillin – was chosen. A further examination under anaesthesia was performed and the mastoid re-explored. This revealed some fluid and fibrous tissue in the mastoid cavity and a large meatoplasty was formed. Histologically biopsy specimens confirmed a chronic osteomyelitis.

After three weeks of intravenous therapy, the CRP had returned to normal but the ESR was still elevated at 77 mm/hr. At this stage the patient had begun to notice some visual disturbance. Examination by an ophthalmologist revealed bilateral papilloedema that was more marked on the left. An magnetic resonance image (MRI) scan was arranged and showed the whole of the left petrous bone to be abnormal along with the sphenoid and clivus. There was evidence of dural involvement with compression of the basal cisterns and brain stem, and also decreased flow was noted in the internal carotid artery (Figure 4).

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In view of the progression of the disease process, with the development of papilloedema, despite antibiotic therapy, hyperbaric oxygen therapy was added to the treatment regime. In total, the patient received 27 treatments of one hour at 22 psi. On this therapy his papilloedema resolved and his vision returned to pre-disease acuity. His ESR returned to normal. He was discharged home on oral antibiotics for a further four months. A repeat MRI scan showed improvement in the appearance of the base of the skull with resolution of the soft tissue swelling, although the petrous bone remained extensively abnormal.

Five years after the course of hyperbaric oxygen he remains well. He has a residual vocal fold palsy and facial weakness. However, the papilloedema has not recurred and his ESR remains low. His malignant otitis externa appears to have been cured.

Discussion

In reviewing the world literature, only two other cases of malignant otitis externa complicated with optic nerve involvement have been reported (Dinapoli and Thomas, 1971; Holder *et al.*, 1986). In both cases, the patients were blinded by progression of the disease and eventually succumbed despite aggressive antibiotic therapy. Assessment of the extent of the disease radiologically, the usefulness of the ESR in monitoring the course of this condition and the beneficial effect of hyperbaric oxygen therapy for refractory cases of malignant otitis externa are discussed.

Radiological assessment of malignant otitis externa

The importance of assessing both the extent of the infection, and its response to treatment, has lead to an interest in its radiological investigation. Three methods in particular may be of use – CT, radioisotope scans and MRI.

CT scan is the current modality of choice for defining the anatomical extent of the disease in malignant otitis externa. It is particularly useful in showing subtle differences in bone density in the skull base and also revealing soft tissue swelling in the nasopharynx or parapharyngeal space. Previous studies have considered the use of serial CT scans to evaluate the effectiveness of treatment. Whilst repeat scanning has demonstrated resolution of central soft tissue swelling, it has not been found as useful in monitoring skull base osteomyelitis (Mendelson *et al.*, 1983; Gold *et al.*, 1984). This is because of the long time interval required for re-mineralization of the affected bone, and thus of the CT scans to return to normal.

Radioisotope scans have also been used previously to monitor the response to treatment. They help on a pathophysiological basis in revealing areas of high tissue activity but suffer from offering poor anatomical localization. Technetium 99 and gallium 67 have been used. Technetium 99 is taken up preferentially by osteoblasts and shows areas of increased osteoblastic activity enabling detection of osteomyelitis. This investigation is highly sensitive (Strashun *et al.*, 1984), and, as bone remodelling persists for some time after a cure has been achieved, it can not provide the necessary information required to know when it is safe to stop treament.

Gallium 67 has been used as an alternative to Technetium 99. This is taken up by granulocytes and so localizes to areas of intense inflammation. Serial scans with positive results imply an area of persistent inflammation. For this reason this investigation has been advocated for monitoring patients with malignant otitis externa (Parisier *et al.*, 1982; Reiter *et al.*, 1982; Gold *et al.*, 1984). However, its reliability has been questioned because patients have relapsed despite normal scans (Gherini *et al.*, 1986).

Relatively little experience of MRI exists in malignant otitis externa (Gherini *et al.*, 1986). This technique, in contrast to CT, offers superior clarity in delineating soft tissue structures. In our experience, this gave more information of the soft tissue changes intracranially than CT. However, even after clinical improvement of several months duration the images still remained abnormal. As with CT, resolution of the soft tissue changes had occurred but there was a persistence of the abnormal bone signals.

Does the ESR offer effective monitoring of malignant otitis externa?

Interestingly, we found that serial measurements of the ESR correlated well with the patient's condition. Following the hyperbaric oxygen therapy, the patient improved clinically with a reduction in the ESR. This simple blood test is often employed in orthopaedic surgery to monitor chronic osteomyelitis. A similar finding has been reported by Rubin and Yu (1988), in patients with malignant otitis externa. This provided a useful investigation to monitor our patient's progress during regular follow-up in the out-patient department.

Hyperbaric oxygen therapy for refractory cases of malignant otitis externa

The treatment for this condition has improved greatly in recent years with the introduction of the quinolone antibiotics with their remarkable anti-pseudomonal activity (Leggett and Prendergast, 1988; Lang *et al.*, 1990). There have also been anecdotal reports of curing refractory malignant otitis externa with hyperbaric oxygen therapy where antibiotic therapy has failed or not been tolerated by patients (Mader and Love, 1982; Shupak *et al.*, 1989). The pathogenesis of malignant otitis externa still remains unclear. However, the most attractive hypothesis is of a diabetic microangiopathy (Chandler, 1968). Indeed, histopathological studies of the capillaries in the tissues overlying the temporal bone have confirmed a thickening of the subendothelial basement membrane (Nadol, 1980). A resultant hypoperfusion and diminished local host resistance would account for the increased susceptibility to this infection. The main effect of hyperbaric oxygen is the elevation of the oxygen partial pressure in the tissues (Bingham and Hart, 1977), thus, amplifying the oxygen diffusion gradient into avascular areas and allowing increased phagocytic oxidative killing of bacteria. The employment of hyperbaric oxygen for this condition would, therefore, seem logical.

In using hyperbaric oxygen as an adjunct to antibiotic therapy for malignant otitis externa, Davis *et al.* (1992), cured all 16 of their patients, even when there was evidence of intracranial spread or the disease had remained refractory to other treatment. Our experience has confirmed the findings of Davis *et al.* (1992), when hyperbaric oxygen was added to our patient's treatment the clinical response was immensely gratifying.

Conclusion

Whilst great advances have been made in reducing the mortality of malignant otitis externa, it remains a very serious and potentially fatal infection. Its tendency to recur makes it a treacherous condition to treat. The importance of monitoring the activity of the disease to judge both the efficacy of treatment, and when to stop, is imperative. The ESR was the most useful investigation in this respect. Radiological modalities are useful in detecting both the

soft tissue and bony involvement and can, therefore, assess the extent of the infection. Changes in the scans may, however, take considerable time, or indeed, never return to normal, which possibly limits their use in following the disease activity. Finally, hyperbaric oxygen therapy appears to be an invaluable adjunct in treating advanced and refractory cases of malignant otitis externa.

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