Malignant External Otitis With Multiple Cranial Nerve Palsies: The Use Of Hyperbaric Oxygen

S Duvvi, S Lo, R Kumar, J Blanshard

Citation

Abstract
Malignant (invasive) otitis externa is an infection involving the external ear canal primarily caused by Pseudomonas aeruginosa. It may involve widespread areas of soft tissue around the skull base and in more advanced cases may give rise to osteomyelitis and cranial neuropathy. Elderly diabetics are at highest risk, but patients with malnutrition and immunosuppression are also susceptible. We report a case of malignant otitis externa complicated by cranial neuropathy and of temporomandibular joint (TMJ) involvement, which was successfully treated by aggressive intravenous antibiotics and hyperbaric oxygen therapy.

REPORT OF A CASE
A 72-year-old lady was admitted into hospital with symptoms of left sided otalgia, otorrhoea, hoarseness, and puffiness of the left half of her face with trismus. She also had concurrent poorly controlled Type II Diabetes recently commenced on insulin, peripheral neuropathy, and hypothyroidism. She had been seen in the ENT outpatients department three months earlier with left otitis externa, when treatment by routine aural suction and topical Gentisone HC ear drops failed to successfully clear the infection.

On examination she was apyrexial. The left ear canal contained purulent debris underlying which were granulations deep in the canal floor and an intact tympanic membrane. Examination of the cranial nerves revealed palatal asymmetry and a paralysed left vocal fold consistent with 9th and 10th cranial nerve palsies. The remainder of the cranial nerve examination was normal. Ear swabs were taken for culture but no organisms were subsequently grown. Haematological investigations showed leucocytosis and thrombocytosis consistent with inflammation. The erythrocyte sedimentation rate (ESR) was 115 mm/hr, C-reactive protein level (CRP) was 80mg/l and blood glucose 21.3 mmol/L. An audiogram showed a mixed hearing loss of 50-60db in the left ear. Computed tomography (CT) scan was performed which showed soft tissue in the left mastoid air cell system extending to its tip with bony destruction extending into the region of the pterygoids. There was loss of sharpness around the carotid foramen and the jugular bulb could not be identified (Figure 1).

Figure 1
Figure 1: CT scan showing the soft tissue obliterating left external auditory canal left mastoid, infra-temporal fossa, skull base and involving the left TMJ.

A diagnosis of malignant otitis externa was made. The patient was admitted and treated with regular aural toilet, and high dose intravenous antibiotic therapy consisting of ciprofloxacin, metronidazole, polymyxin and topical
Gentisone HC drops for a presumed pseudomonas infection. A myringotomy was performed and some middle ear fluid was sent for microbiology culture which did not isolate any pathogens. After six weeks of intravenous antibiotic therapy her left sided otalgia, otorrhoea, puffiness of left half of face and trismus were improving with her CRP returning to normal but her ESR remained elevated at 110 mm/hr.

Therapy was changed to high dose oral ciprofloxacin and topical Gentisone HC ear drops. 4 weeks later her ESR continued to be elevated at 108 mm/hr. At this stage she continued to have hoarseness, scanty otorrhoea, trismus, and mild otalgia. A CT scan was repeated which showed similar features to the earlier scans. In view of the recognised significant mortality and morbidity of unresolving malignant otitis externa, the patient was commenced on hyperbaric oxygen therapy. The patient received a total of 40 treatments of one hour duration at 22 psi (2-2.5 atm) over two months. Eight weeks after completion of treatment her symptoms of otalgia, otorrhoea, puffiness of the face and trismus resolved and the ESR returned to a steady state of 60 mm/hr. A further CT scan at this stage showed marked improvement in the appearance of the skull base with resolution of the soft tissue swelling and reappearance of fat planes, with only some effusion in the left TMJ (Figure 2).

One and a half years later she remained well with mild residual left vocal fold palsy and left palatal weakness.

**Figure 2**

Figure 2: CT scan showing the reappearance of soft tissue planes with the decrease of the soft tissue swelling after the treatment.

**DISCUSSION**

Malignant (or necrotizing) otitis externa is a potentially fatal aggressive infection of the external auditory canal which is usually caused by Pseudomonas aeruginosa. It was first described in 1959 by Meltzer and Kelemen and the syndrome later termed by Chandler in 1968. To date, the pathogenesis of malignant otitis externa continues to remain unclear. However, diabetic microangiopathy with subsequent hypoperfusion and diminished local host resistance is thought to account for the increased susceptibility to this infection. Patients usually present with severe otalgia, otorrhoea and hearing loss. There are three stages in malignant otitis externa. In stage I disease, the cardinal clinical features include persistent purulent otorrhoea, otalgia and infected granulation tissue in the floor of the external auditory canal with no facial nerve palsy. In stage II disease the process extends into the soft tissues leading to skull base osteomyelitis and involves the posterior cranial nerves XI and XII as they exit their respective foramina. In stage III disease, intracranial extension occurs along with the stage II signs.

Investigation with CT scanning 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 as well as revealing soft tissue swelling in the nasopharynx or parapharyngeal space. Serial CT scans are helpful to evaluate the effectiveness of treatment by demonstrating resolution of central soft tissue swelling, but have not been found as useful in monitoring skull base osteomyelitis. Radioisotope scans using Technetium 99 and Gallium 67 are highly sensitive and have previously been used to monitor response to treatment. They help in revealing areas of high tissue activity but suffer from the draw-back of offering poor anatomical localization. Gallium 67 which is taken up by macrophages and reticular endothelial cells, localizes to areas of intense inflammation and has been used as an alternative to Technetium 99. Positive results with serial Gallium 67 scans imply an area of persistent inflammation. This investigation has been advocated for monitoring disease progression in patients with malignant otitis externa, but on some occasions Gallium scans can be normal despite relapse. MRI plays a complimentary role to CT only in determining the initial severity of the disease as it offers...
superior clarity in delineating soft tissue. There have been a few reports of MR imaging being employed in the diagnosis and follow-up monitoring. Serial measurements of the ESR and CRP are useful investigations to monitor patient's progress, as we have demonstrated in the current case (Figure 3).

Figure 3
Figure 3: Graph showing the CRP and ESR levels during the treatment of the patient

The treatment of this condition has improved in recent years due in part to a better assessment of the extent of the infection by CT, radioisotope scans and MRI, and its favourable response to treatment with quinolone antibiotics which have remarkable anti-pseudomonal activities. Only a limited number of cases of successfully treated malignant otitis externa complicated with cranial nerve involvement have been reported in the world literature. They were all treated by combined topical and oral antibiotic therapy with adjuvant hyperbaric oxygen, when there was evidence of intracranial spread or when the disease remained refractory to routine treatment, hyperbaric oxygen as an adjunct to antibiotic therapy has been shown to be useful.

The main effect of hyperbaric oxygen is the elevation of the oxygen partial pressure in the tissues from hypoxic levels to normal or supranormal levels. This amplifies the oxygen diffusion gradient into avascular areas and allows increased phagocytic oxidative killing of bacteria. In our case the patient received HBO treatment at a pressure of 2.0 to 2.5 atm lasting for 90 to 120 minutes for 40 treatments along with appropriate antibiotic therapy.

CONCLUSION
Malignant otitis externa remains a very serious and potentially fatal infection. A high index of suspicion is essential for the early diagnosis of this condition. It almost exclusively affects elderly diabetics and can result in cranial polyneuropathies following spread of the infection beyond the bone of the external auditory canal via vascular and fascial planes. Radiological modalities, in particular CT, MRI and radioisotope scans, are useful in detecting both soft tissue and bony involvement, and can therefore assess the extent of the infection and monitor change. ESR is the most useful blood investigation in monitoring disease progression. Hyperbaric oxygen therapy appears to be an invaluable adjunct to antibiotic therapy in treating advanced and refractory infections involving the base of the skull with cranial neuropathies and refractory cases.

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CORRESPONDENCE TO
Stephen Lo Department of Otolaryngology-Head and Neck Surgery St. George's Hospital Blackshaw Road London, SW17 0QT UK E-mail: stephenlo@lycos.co.uk Tel: +44-7786-927613 Fax: +44-208-7253306

References
Author Information

S. K. Duvvi, MS MRCS DOHNS
Department of Otolaryngology-Head and Neck Surgery, North Hampshire Hospital

S. Lo, BMedSc MBA MRCS DLO
Department of Otolaryngology-Head and Neck Surgery, St. George's Hospital Medical School

R. Kumar, MS
Department of Otolaryngology-Head and Neck Surgery, North Hampshire Hospital

J. Blanshard, FRCS (ORL)
Department of Otolaryngology-Head and Neck Surgery, North Hampshire Hospital