Carcinoma of temporal bone presenting as malignant otitis externa

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Abstract
A 69-year-old man presented with a three-month history of otalgia and tenderness of the right ear and a one-week history of a painful right parotid swelling. Examination revealed granulation tissue in the right ear canal with normal looking tympanic membranes and a parotid abscess. Repeated biopsies from the ear canal and parotid showed non-specific inflammation. Repeated cultures from both areas grew *Ps. aeruginosa*. The patient’s condition improved following three weeks of intensive treatment for malignant otitis externa only to relapse five weeks after the end of treatment. He received a second course, only to improve temporarily. He developed a right facial nerve palsy five weeks after he was first seen, followed four months later by palsies of all cranial nerves except the olfactory, before dying, seven months after his first appointment.

The radiological, histological and post-mortem findings are discussed.

Introduction
Malignant otitis externa is a severe progressive form of otitis externa usually occurring in elderly diabetics (Chandler, 1968). Over 91 per cent are over the age of 55 and 93 per cent are diabetics (Zaky *et al.*, 1976). Two cases were reported to have occurred in children who were not diabetic but both suffered from anaemia and malnutrition (Joachims, 1976). To date there are 11 case reports of malignant otitis externa in children in the English literature (Nir *et al.*, 1990). Diagnosis is mostly a clinical one, and requires a high index of suspicion. The characteristic clinical manifestations are otalgia, otorrhoea, the presence of granulation tissue at the isthmus of the ear canal and tenderness of the tissues around the ear and mastoid.

Malignant otitis externa, which is predominantly caused by *Pseudomonas aeruginosa*, begins in the ear canal skin and may extend inferiorly to the soft tissues at the base of skull, through the fissures of Santorini or at the isthmus, to involve the parotid gland, cartilage, bone, nerves and blood vessels (Chandler, 1987). The infection is resistant to ordinary methods of treatment and if not arrested may progress to osteomyelitis of the temporal bone, skull base, facial palsy and other multiple cranial nerve palsies, intracranial complications and even death.

Facial nerve palsy is a bad prognostic sign. The disease carries a high mortality rate of 30 to 80 per cent of patients (Chandler, 1977; Damiani *et al.*, 1979; Mills, 1986).

This case is believed to be the first case of carcinoma of temporal bone presenting as a classic malignant otitis externa with neoplastic changes seen at post-mortem and provides a cautionary note in the diagnosis of malignant otitis externa.

Case report
A 69-year-old male patient presented on 8 September 1981 with a history of right otalgia and tenderness of the ear for the previous three months. A week before he had developed a painful swelling in the right parotid area. His past history was unremarkable.

On examination there was some granulation tissue in the right ear canal but the ear drum appeared normal. There was a diffuse, tender swelling over the right parotid area. Other ear, nose and throat examinations were negative. All blood tests including fasting blood sugar, urinalysis and X-rays of the chest, skull base and temporal bones showed no abnormalities. An audiogram showed a bilateral symmetrical high frequency hearing loss, other audiometric tests were normal.

He was admitted to hospital and the right ear was examined under general anaesthesia on the following day. It was found that the ear canal was narrowed by granulation tissue but the ear drum was normal. Granulation tissue was sent for histological examination and a gentamicin wick was inserted.

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FIG. 1
Showing the wall of the external auditory meatus lined by a thick layer of well differentiated invasive squamous cell carcinoma arising from the epidermis of the meatus. H&E x65.
To show the squamous cell carcinoma deep in the external auditory meatus where it seems to be spreading into the matrix of an epidermoid cholesteatoma probably in the middle ear. H&E x65.

was drained. A narrow track between the parotid abscess and the right ear canal was located. Swabs from the parotid abscess and the ear canal were sent for culture and sensitivity and grew *Pseudomonas aeruginosa*. Biopsies from the parotid gland and ear canal showed non-specific chronic inflammation.

A sialogram of the right parotid showed free communication of the remaining normal parotid tissue with the abscess cavity. A sinogram of the abscess track showed that the track extended to the floor of the right ear canal and upwards towards the base of the skull.

A three week course of intravenous carbenicillin 5 gm six-hourly together with gentamicin 80 mg eight-hourly and gentamicin wick in the ear canal was initiated. The patient’s blood gentamicin level and kidney function were monitored.

Five weeks after his admission the patient developed a right facial weakness which progressed to a complete lower motor neurone paralysis. This was confirmed by electromyography.

Repeated biopsies from the ear canal and parotid gland showed only chronic non-specific granulation tissue. Repeated culture from the ear canal and parotid grew *Ps. aeruginosa*.

The patient’s condition had improved towards the end of October 1981 and he was discharged home on 3 November. However, he was re-admitted on 4 December following a relapse. He was given intravenous azlocillin 2 gm eight hourly together with gentamicin 80 mg eight hourly for three weeks with full monitoring of his renal function, and blood gentamicin levels.

The facial palsy was total and an electromyogram showed no sign of recovery. A right tarsorraphy was performed to protect the patient’s eye. His condition had improved while on treatment but his hearing had not altered, as confirmed by a series of pure tone audiograms.

On 8 December 1981, the right ear was examined under general anaesthesia and apart from granulation tissue around the isthmus of the ear canal, the rest of the ear canal, ear drum, middle ear and mastoid area were normal. The granulation tissue was removed and gentamicin wick was inserted. The histology results on the granulation tissue again showed chronic non-specific inflammation.

On 2 February 1982, the patient developed paralysis of the lower four cranial nerves in rapid succession. There was also an increase in the discharge from the parotid area.

A skull CT scan showed no abnormality apart from soft-tissue swelling around the right parotid gland and base of skull. Nasogastric feeds were established owing to the patient’s dysphagia and ampicillin was prescribed for a chest infection due to aspiration.

On 16 February 1982, the right abducens nerve was paralyzed and a CT scan showed signs of skull base erosion.

On 27 March, he developed right proptosis with loss of vision and chemosis. An orbital venogram showed slow filling of the right cavernous sinus. A right common carotid angiogram showed appearances consistent with cavernous sinus thrombosis.

Biopsies from the right ear canal and parotid gland showed only chronic non-specific inflammation. Panendoscopy showed no abnormality apart from right vocal fold and right pharyngeal and soft palate paralysis. Blood cultures were sterile throughout the disease.

On 4 April, there was paralysis of all the cranial nerves on the right side except for the olfactory nerve. The patient began to lose weight, became very irritated and depressed and finally very confused before dying on 17 April 1982, seven months after his admission.

Post-mortem examination was carried out and the following positive findings were recorded:

1. There was a swelling anterior to the right ear with a sinus at the angle of the jaw. This communicated with an extensive...
Abscess in the region of parotid gland. A parotid frozen section showed no signs of malignancy. The nose, pharynx, pituitary, mouth, larynx, thyroid gland, trachea, thorax and abdomen were all normal apart from signs of confluent bronchopneumonia probably due to aspiration.

2. There was a mass of friable tissue arising from the apex of petrous bone and involving the anterior part of the right temporal bone. The right petrous bone was almost completely destroyed by friable tissue which on frozen section and smearing was consistent with keratinizing squamous cell carcinoma (Figs. 1–3). The right cavernous sinus was completely occluded and invaded by keratinizing squamous epithelium. Apart from the involvement of the anterior part of the right temporal lobe, the rest of the brain was normal.

3. Examination of the rest of the body showed no gross pathology.

Discussion

Most cases of malignant otitis externa described in the literature have common features such as otalgia, ototympanitis, tenderness around the ear, granulation tissue in the isthmus of the ear canal, normal ear drum, normal middle ear cleft, a tendency of the disease to spread to the deep tissues of the base of skull involving the cranial nerves and intracranial cavity and ultimately leading to death (Schwart et al., 1971; Cohen et al., 1987). It has generally been accepted that the disease is due to inflammation of the ear canal by Ps. aeruginosa in the elderly diabetic patient. However if the literature is closely examined, not all cases conform with these features. In the case described above, though the patient was old and Ps. aeruginosa was isolated, he was not a diabetic. Nevertheless, the disease progressed in spite of the most energetic treatment with accepted remedies. Furthermore, although the disease process was relentless and ended in the death of the patient, at no time was there any clinical feature or surgical or histological evidence to indicate the presence of cancer. We were therefore surprised at the post-mortem histological findings which are uncontroversial. This only became apparent after decalcification of the specimen and examination by a neuropathologist skilled in this procedure. This therefore raises the question that some cases presenting as malignant otitis externa and ascribed to Ps. aeruginosa may be due not to inflammation only but to a neoplastic changes. This would certainly explain the relentless course of the disease, particularly in those cases where death occurs eventually.

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References


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