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Masked pseudomonal skull base osteomyelitis presenting with a bilateral Xth cranial nerve palsy

R. G. Rowlands, M.R.C.S., G. K. Lekakis, F.R.C.S., A. E. Hinton, F.R.C.S.

Abstract
Skull base osteomyelitis classically presents as a complication of severe external otitis, middle ear, mastoid or sinus infection and can lead to multiple lower cranial nerve palsies when the jugular foramen is involved as a consequence of widespread involvement of the skull base. Bilateral skull base osteomyelitis is a recognized phenomenon, but has not previously been reported secondary to pseudomonal infection in the absence of a clinically obvious focus of infection. We report the case of a 77-year-old diabetic patient who presented with dysphonia and dysphagia and had a bilateral Xth cranial nerve palsy. No focus of infection was evident on presentation. Subsequent radiological investigation confirmed the diagnosis of bilateral skull base osteomyelitis.

Key words: Skull Base; Osteomyelitis; Cranial Nerve Diseases

Introduction

Skull base osteomyelitis secondary to pseudomonal infection classically presents in elderly diabetic patients as a severe unrelenting otitis externa leading progressively to the development of a unilateral facial nerve palsy, hearing loss and ultimately lower cranial nerve palsies, the latter condition known as jugular foramen syndrome.[1] Atypical presentations of skull base osteomyelitis[2–4] although rare, do occur and can lead to inevitable delays in treatment if the diagnosis is not suspected.

This case illustrates an atypical presentation of skull base osteomyelitis which only manifested after the development of a bilateral Xth cranial nerve palsy. Clinical examination revealed no evidence of active otitis externa, middle-ear disease or sinus disease. There are no previous documented cases of bilateral skull base osteomyelitis presenting without evidence of an obvious active focus of infection. The case also highlights the limitations of computed tomography (CT) imaging in the early stages of the disease.

Case report

A 77-year-old diabetic man was referred to the ENT clinic at St George's Hospital with a two-month history of dysphonia and progressive dysphagia to solids associated with substantial weight loss. Clinical examination revealed a bilateral vocal fold palsy but the VIIth, IXth, XIth and XIIth cranial nerves were found to be intact. The rest of the ENT examination was also normal with no evidence of active ear disease.

He had previously been reviewed at the time of onset of his dysphonia in his local hospital where a right vocal fold palsy had been observed, but a computed tomography (CT) scan of the area between the skull base and diaphragm had failed to show any abnormality. Eight months previously he had been treated for a severe right otitis externa with an associated right lower motor neurone VIIth cranial nerve palsy. *Pseudomonas aeruginosa* had been isolated from an ear swab and the infection had resolved with ciprofloxacin and topical neomycin ear drops. CT imaging performed at this stage showed no evidence of bony destruction of the temporal bone and at subsequent follow-up the facial nerve palsy and otitis externa had recovered completely. After presentation with the new Xth nerve palsy, urgent panendoscopy was arranged, which merely suggested the presence of oropharyngeal candidiasis, but nothing else of note was demonstrated.

The patient's condition continued to deteriorate and he was admitted to Atkinson Morley hospital for further investigation of his progressive dysphagia that had led to the patient requiring enteral nutrition.

CT brain had shown evidence of a left fronto-parietal infarct, but markers of infection were significantly elevated (CRP 64, ESR 120 mm/hr).

Magnetic resonance image (MRI) scans of the brain demonstrated abnormal signal of the clivus and adjacent petrous bone, initially thought to be suggestive of metastatic disease (Figure 1), but subsequent CT scans showed abnormal bone texture consistent with osteomyelitis (Figure 2).

Two weeks after admission the patient developed a significant chest infection and stridor neccesitating tracheostomy and mechanical ventilation for a period of six days. In order to gain a microbiological diagnosis, a cortical mastoidectomy was performed but no organisms could be cultured from any of the bone samples taken at the time of surgery.

Although no positive microbiological or histological samples could be obtained, serological analysis suggested the presence of an active pseudomonal infection. The patient was commenced on ciprofloxacin and this was combined with ceftazidime on microbiological advice for a further four weeks.

From the Department of Otolaryngology, St George's Hospital, London, UK.
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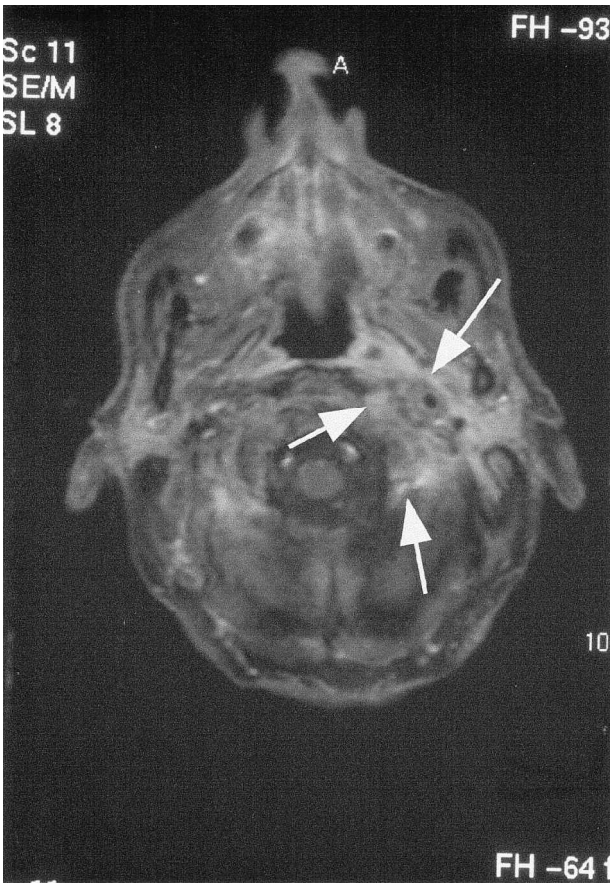

FIG. 1
Fat suppressed post contrast T1-weighted MRI scan (axial view) showing abnormal enhancement on the left side of the skull base (arrowed).

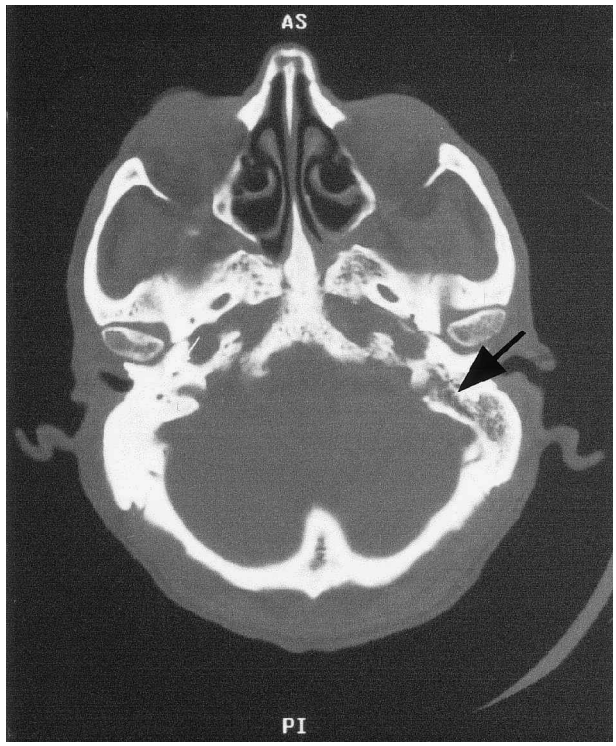

FIG. 2
High resolution CT scan (axial view) indicating destruction within the left petrous temporal bone (arrowed) and abnormal bone texture of the skull base consistent with osteomyelitis.

The patient's clinical condition has substantially improved but there has been no recovery of the bilateral Xth nerve palsy and he has subsequently had a percutaneous gastrostomy tube inserted for the purposes of long-term enteral nutrition.

Discussion

Jugular foramen syndrome is characterized by the development of multiple lower motor neurone palsies and occurs when skull base osteomyelitis involves the jugular foramen, usually indicating a poor prognosis. Atypical presentations of basal skull osteomyelitis producing a unilateral jugular foramen syndrome, in the absence of an obvious focus of infection, in non-diabetic patients have been recognized. Bilateral involvement of the skull base secondary to pseudomonal infection, producing a bilateral tenth cranial nerve palsy, without an obvious active ear or other focus of infection has not previously been described.

Chandler *et al.*[5] described 13 cases of basal skull osteomyelitis, with three having bilateral widespread involvement of the skull base affecting both jugular foramina. However, in all these cases the disease was at a late stage, the patients presenting with significant external otitis together with severe chondritis and perichonditis of the pinna.

Aspergillus infections have been known to lead to skull base osteomyelitis[6] without evidence of otitis externa usually commencing as disease within the middle ear or mastoid, but in this case pseudomonal infection was clearly demonstrated on serology. In addition, of the previous six reported cases of *Aspergillus* infection associated with basal skull osteomyelitis, none were in diabetic patients.

Despite resolution of the active otitis externa several months prior to representing with the new Xth cranial nerve palsy, it is likely that a focus of infection, albeit initially quiescent, existed within the temporal bone and reactivated to produce the slowly progressive, destructive osteomyelitis of the skull base.

Although initial CT scans showed no evidence of bony destruction of the temporal bone, CT may be unreliable in showing bony involvement in the early stages of the disease and is the reason why bone scans should be obtained if any suspicion of osteomyelitis remains. These take the form of technetium and gallium scans, although neither of these tests are specific to the diagnosis.[7,8] Gallium scans can assist in indicating the efficacy of treatment, but markers of infection such as C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) can also enable antibiotic therapy to be monitored.

Conclusion

Progressive bilateral cranial nerve palsies in elderly diabetic patients, even in the absence of an obvious concurrent focus of infection, should still alert clinicians to the possibility of skull base osteomyelitis, particularly if there has been a past history of complicated otitis externa. Where previous CT imaging has proved inconclusive in showing bony involvement of the skull base, appropriate investigations such as bone scans should be instigated.

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Address for correspondence:
Mr R. G. Rowlands,
48 Sandtoft Road,
Charlton,
London SE7 7LR, UK.

E-mail: GRowlands1@aol.com

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