Unusual association of diseases/symptoms

Skull based osteomyelitis due to postsurgery malignant otitis externa presenting as stroke

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Summary

Malignant or necrotising otitis externa is a rare but potentially fatal disease. The classic presentation is one of severe, unremitting, throbbing otalgia, which may progress to osteomyelitis, especially in the elderly diabetic or immunocompromised patient. The case described is of a 72-year-old immunocompetent, non-diabetic man who presented with facial weakness, dysphagia and weight loss. The admitting diagnosis or impression was that of a cerebrovascular event. The eventual diagnosis was that of skull based osteomyelitis secondary to malignant otitis externa complicating mastoid surgery.

BACKGROUND

Necrotising or malignant otitis externa (MOE) is a rare and potentially life threatening disease, which if left untreated can progress to skull base osteomyelitis. The usual presentation is one of unremitting, painful otitis externa in an elderly diabetic or immunocompromised patient. We describe the case of a 72-year-old immunocompetent, non-diabetic patient who presented with multiple cranial nerve palsies. This was initially thought to be due to stroke, which was eventually found to be secondary to skull based osteomyelitis secondary to necrotising otitis externa preceded by mastoid surgery. Stroke is a common condition which is often seen initially by the emergency department. Rapid diagnosis of stroke in the emergency setting is necessary for the timely delivery of thrombolysis. Stroke mimics make up a small but significant proportion of patients presenting to the emergency department with stroke like symptoms. In an American study, 25.3% of patients who triggered the departmental stroke code had a final diagnosis other than stroke. The diagnoses in this group were intracranial haemorrhage, subarachnoid haemorrhage, subdural haematoma, old deficit, hypotension, seizure, intoxication, hypoglycaemia, mass lesion, migraine and others.¹

We describe the case of a patient with an unexpected presentation of skull base osteomyelitis and have undertaken a thorough review of the literature.

CASE PRESENTATION

A 72-year-old man presented to the hospital with a 3–4 week history of dysphagia, choking and weight loss of one stone. On examination, he was found to have a right-sided vocal cord palsy, a House Brackmann Grade III facial weakness and tongue wasting. Otoscopy was not undertaken at this stage. The admission diagnostic impression (medical assessment unit) was that of a cer-

ebrovascular event and he was referred to the medical take team.

INVESTIGATIONS

The initial investigations revealed a granulocytosis – raised white cell count of 14.4, C reactive protein of 75 (0–12), liver function tests, urea and electrolytes and glucose were within the laboratory normal range.

A CT scan (figure 1) demonstrated a soft tissue mass seen within the right posterior nasopharynx medial to the lateral pterygoid; the lesion was obstructing the right internal auditory meatus. There was no fading of bony opacity and hence no visible bony erosion. This indicated either an inflammatory or malignant mass, with a raised likelihood of the latter. An MRI (figure 2) was undertaken which suggested that the same mass was probably inflammatory and did not show any tumour suspicious signal changes.

Perusal of his records revealed a recent ENT consultation 4 weeks prior to his admission. The patient had been under investigation for persistent ear infections with aural polyps. Ten weeks prior to admission, he had undergone a combined approach tympanoplasty with intact canal wall and a temporalis fascia graft. At follow-up, he was noted to have some ongoing problems with discharge which was thought to be due to an infected temporalis graft. The patient was started on topical antibiotic drops for this and was listed for admission for a further exploration and possible removal of infected graft. This previous history and the mass shown on imaging indicated further management by the Head and Neck Team was appropriate. Biopsy of the mass and examination under anaesthesia (EUA) of the ear was undertaken.

The histology of the pharynx biopsy showed inflammatory changes with no evidence of malignancy. The examination of the ear canal showed evidence of discharge and

Figure 1 CT scan showing a soft tissue mass within the right posterior nasopharynx.

Figure 2 MRI scan showing an ill-defined soft tissue mass within the right posterior nasopharynx.

areas of granulation tissue. At the time of the EUA a swab was taken, this grew *Pseudomonas aeruginosa*.

DIFFERENTIAL DIAGNOSIS

The presence of *Pseudomonas* in the ear indicated a possibility that the inflammatory mass might be a 'granuloma' secondary to skull base osteomyelitis secondary to

'malignant' otitis externa. A vasculitic granuloma such as Wegener's, histiocytosis X² or a skull base chemodectoma such as a glomus jugulare were considered less likely while *Pseudomonas* was present. The inflammatory mass at the skull base was considered to have involved and disrupted the function of the facial, glossopharyngeal, vagal and hypoglossal nerves, which elevated this presentation to

stage 3b on the proposed Carney clinicopathological classification of MOE.³

TREATMENT

The patient was treated with intravenous ciprofloxacin as indicated by microbiology, for 6 weeks, and oral ciprofloxacin for a further 6 weeks. His condition began to improve. He did require insertion of a percutaneous endoscopic gastrostomy for his pharyngeal paralysis induced swallowing difficulties, and he had a prolonged hospital stay. On discharge home, the patient had input from the speech and language team as well as a lot of support from his wife.

OUTCOME AND FOLLOW-UP

The patient was discharged to a community hospital 6 weeks after his admission and was treated with a total of 3 months of antibiotics. We can report that all the patient's cranial nerve palsies resolved. Two weeks after discharge, he showed early slight swallowing recovery for sips of water. He benefited from intensive swallowing therapy to restore his deglutition over the next 9 months.

DISCUSSION

The first reported case was published in 1938⁴ and the term 'malignant otitis externa' was coined in 1968, because of its high mortality at that time, which was over 50% of all patients.⁵ This is a rare but potentially fatal disease that occurs principally in elderly diabetics or immunocompromised patients. A review of over 25 papers found that the combination of pain, granulations, otorrhoea and resistance to local therapy for at least 8–10 days is highly sensitive for making a diagnosis of MOE.⁶ Immunocompromised state, including diabetes, *P aeruginosa* grown on culture, a positive bone scan or cranial nerve palsies enhances the specificity of this diagnosis.

Other symptoms include purulent otorrhoea, headache and temporomandibular joint pain and hearing loss. Examination reveals a swollen, tender external auditory canal with granulation tissue.⁷ The main differential diagnosis is that of squamous carcinoma in the temporal bone. MOE can be staged using several different classification systems. Table 1 shows a three stage system which combines clinical and radiological features.

The disease is associated with uncharacteristically invasive behaviour of *Pseudomonas*, which is normally associated with benign otitis externa. This behaves invasively by use of its flagellae and unusual vasculitic properties,

and the failure of defensive immune mechanisms such as the granulocytic response⁸ and/or other immune failures, or the presence of diabetes, to halt its spread. The resultant periostitis can lead to skull based osteomyelitis, first described in 1959,⁷ which may nowadays be classified as Carney clinicopathological classification stage 2, and verified with Tc-99 bone scan. Although the presence of an intact temporal bone indicates the route of invasion would be through the fissures of Santorini and the tympanomastoid suture leading to the stylomastoid and jugular foramina, in this instance the previous surgery provided an entry route as reported by Patel *et al.*⁹

Pseudomonas was reported in 1987 as causing 95% of MOE¹⁰ and fungal invasion is the next likely reported cause of MOE and is associated with immunocompromise. The most common fungal organism is *Aspergillus fumigatus*, although other *Aspergillus* species have been described. There is a suggestion of less diabetic involvement in fungal disease.¹¹

As in this case, the facial nerve is the commonest cranial nerve to be involved¹² followed by glossopharyngeal and vagal; the hypoglossal palsy observed in this case is said to be only rarely present.⁶ Cranial nerve palsies associated with MOE are present in approximately 40% of patients with the disease. There are variable reports on the significance of associated cranial nerve palsies to predicted mortality rates, although some reports⁶ show 80% mortality, in such cases, which are certainly an indication of more extensive disease involvement signalled by multiple cranial nerve involvement. Morbidity, caused by continued palsy is higher in patients with facial nerve palsy.¹³ Conversely, in several more recent studies the authors found that patients with cranial nerve palsies due to MOE had the same survival rate as those presenting with the disease without cranial nerve involvement.¹⁴ This improvement may speculatively be due to recent alleviation of the severity of the disease, supported by this example.

Definitive diagnosis is frequently elusive, requiring a combination of a high index of clinical suspicion, laboratory and imaging techniques and the histological exclusion of malignancy.¹⁶ CT is initially used to evaluate the extent and severity of extra temporal soft tissue involvement and skull based osteomyelitis; however, more than 30% of affected bone will need to be demineralised to appear eroded on CT. Therefore, early findings are limited to soft tissue inflammation.¹⁷ Isotope temporal bone scans (Tc-99) assist the establishment of osteomyelitis¹⁸ Treatment involves prolonged courses of antibiotics as guided by microbiological results. Prior to the widespread availability of quinolones the mainstay of treatment for MOE was surgical debridement, the introduction of monotherapy with ciprofloxacin transformed the outcome in the past being associated with a cure in nearly all patients.¹⁸ There are now concerns about increasing resistance to ciprofloxacin, with case series suggesting that resistance is becoming more common¹⁹ Extensive surgery is limited to patients who fail to improve with antibiotic treatment and is mainly limited to biopsy and debridement of granulation tissue.⁶ Other antibiotics should be guided by local microbiology advice and antifungals are considered in refractory cases. Hyperbaric oxygen therapy has, in some cases, been shown to be of benefit in patients with disease refractory

Table 1 Carney clinicopathological classification of malignant otitis externa³

Stage	Description
1	Clinical evidence of malignant otitis externa with infection of soft tissues beyond the external auditory canal, but negative Tc-99 bone scan
2	Soft tissue infection beyond external auditory canal with positive Tc-99 bone scan
3	As above, but with cranial nerve palsies
3a	Single
3b	Multiple
4	Meningitis, empyema, sinus thrombosis or brain abscess

to standard treatment.^{20 21} However, the need for a course of daily decompressions and limitations on its availability has prevented further research on its effectiveness in this disease.

Learning points

- Investigation of cranial nerve disorders must include otoscopy.
- Otitis externa that is refractory to treatment must be investigated further, and MOE must be considered especially in immunocompromised or diabetic patients and/or with aural *Pseudomonas*.
- Painful and refractory otitis externa may require biopsy to exclude squamous carcinoma.
- Treatment should be in accordance with microbiology results and guidance.

Competing interests None.

Patient consent Obtained.

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