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Clinical Records

Cerebellopontine angle lymphoma presenting as chronic mastoiditis

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Abstract
Middle-ear tumours are not uncommonly mistaken for mastoiditis on presentation. We report a case of cerebellopontine angle non-Hodgkin's B-cell lymphoma originally presenting as a middle-ear inflammatory process. In mastoiditis not resolving with conventional treatment it is important to look for an underlying cause.

Key words: Cerebellopontine angle; Lymphoma, B-cell; Mastoiditis

Introduction

A 76-year-old European woman presented to the otolaryngology department at Waikato Hospital on the 5th of November 1998. She complained of right-sided facial weakness and otalgia, dysequilibrium and occasional sweating. She had experienced a complete right facial palsy 11 weeks prior, that improved with prednisone. She then developed right-sided otalgia and a middle-ear effusion. A grommet was inserted one week prior to this. Audiology at this time showed a right moderate to severe mixed hearing loss and a left mild to moderate sensorineural loss.

The right tympanic membrane was dull and the external canal roof bulging. There were no symptoms of acute mastoiditis. Physical examination was otherwise normal. Computed tomography (CT) of the mastoid bones showed partial opacification of the mastoid air cells and middle ear. The patient was diagnosed with chronic mastoiditis with facial nerve involvement. Symptoms improved with intravenous antibiotics and she was discharged. She was readmitted on the 18th of November with ongoing otalgia and otorrhoea. She proceeded to right cortical mastoidectomy. Intra-operatively, thickening of the external auditory canal lining and blockage of the attic and aditus with inflamed middle-ear mucosa was seen. The mastoid bone behind the attic was healthy. Biopsies showed a dense lymphocytic infiltrate with T-cell dominance but no clonality. No organisms were cultured. Post-operatively the facial palsy improved.

The patient represented on the 5th of January with persisting hearing loss and worsening right facial palsy (Grade III–IV/VI (House-Brackmann grading system)). The right middle ear was discharging and a luminal polyp blocked the grommet. Prednisone and antibiotics were started. Repeat CT showed the opacification previously seen and erosion of the sinus tympani and tegmen tympani. The facial nerve was almost certainly involved. Her condition was now thought to be due to a granulomatous disorder or a malignant otitis externa. She underwent a repeat wide cortical mastoidectomy. There was thick granulation tissue in the middle ear, mastoid, aditus, attic and adjacent meatal skin, and oval window. The facial nerve was found to be dehiscent and oedematous. Histology was again nonspecific.

The patient re-presented on the 22nd of January with deterioration of her symptoms. The right external meatus was nearly occluded and bare bone was visible in the

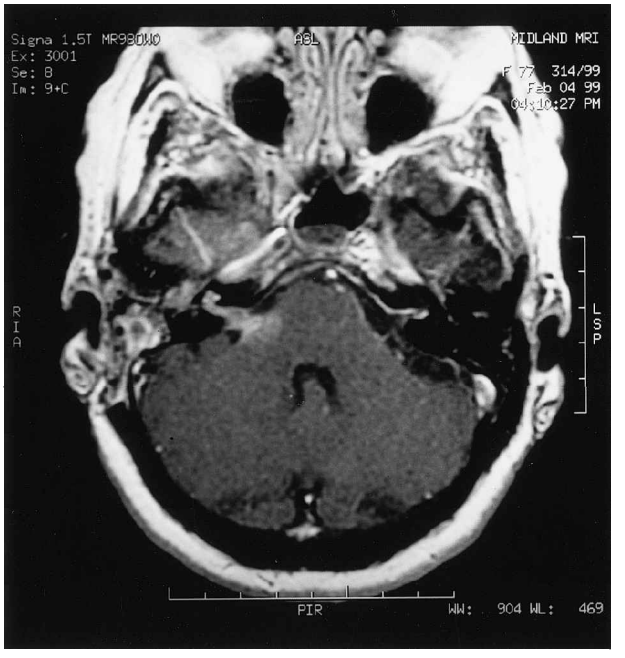

Fig. 1
T1-weighted axial MRI demonstrating the lymphoma within the right CPA.

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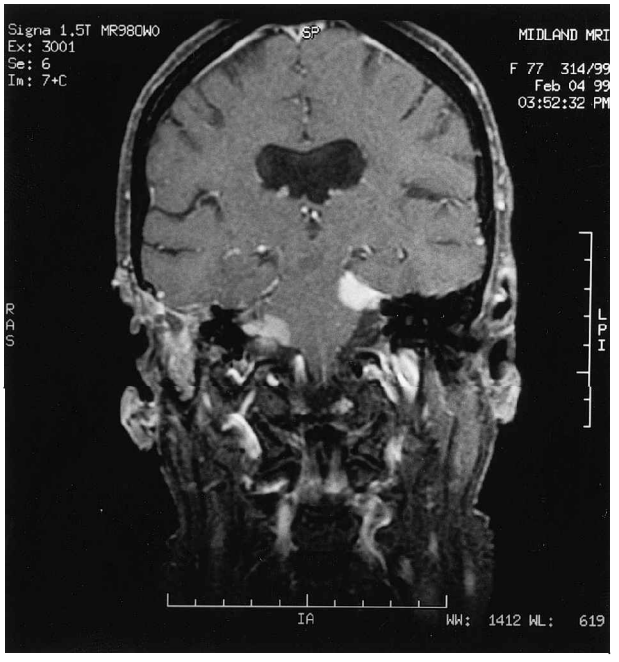

FIG. 2
T1-weighted coronal MRI demonstrating both right and left CPA lesions.

external canal wall, with pulsatile otorrhoea. There was a profound hearing loss on the right. Oral steroids and intravenous antibiotics were started. Pus cultures, full blood count and film, erythrocyte sedimentation rate, autoantibodies and ANCA were unhelpful.

A third CT showed mastoid and middle-ear opacification and erosion of the lateral wall of the facial canal. MRI showed enhancement and fluid in the right mastoid and abnormal enhancement of the right facial nerve and cochlear. There was generalized meningeal thickening. Three distinct homogenously enhancing intracranial lesions were identified. The first extended from the right cerebellopontine angle (CPA) to fill the internal auditory meatus to a depth of 12 mm (Figure 1). The second was dural based on the medial aspect of the right middle cranial fossa. The third was a 12 by 24 mm lesion at the left CPA (Figure 2). Lumbar puncture and bone marrow aspirate were performed with no signs of malignancy on cytology. CSF protein was raised (0.99) but no specific pathology or oligoclonal bands were identified. The patient then developed a right VIth cranial nerve palsy. She underwent a right modified radical mastoidectomy on 22nd of February 1999, the final histology was of a B-cell non-Hodgkin's lymphoma (NHL).

Staging CT of the chest and abdomen identified no abnormalities. She had one cycle of CHOP chemotherapy with intrathecal prophylactic methotrexate but did not tolerate this well, and was admitted on the 14th of May with acute deterioration. No further active management was undertaken and she died on 21st of May 1999.

Discussion

Primary presentation of lymphoma in the middle-ear cleft is uncommon. Tumour cell involvement is not unusual in generalized lymphoma but is rarely symptomatic. An extensive review of the literature identified 36 reported middle-ear cleft lymphomas presenting clinically. Seventeen were thought to represent primary tumour. Seventy-five to 90 per cent of middle-ear cleft tumours are epithelial in origin.[1]

We suspect the aetiology in this case may be tumour migration along the internal auditory meatus of a CPA lymphoma. We are not aware that this has been previously reported. Tumour migration in this case is suggested by the MRI appearances of tumour within the internal auditory meatus. In hindsight, the bilateral sensorineural hearing loss at primary presentation could have been due to masses at both CPAs. The lymphomas could have been disseminated masses from another source. Normal abdominal and chest CT scans, bone marrow biopsy and blood tests make this a low probability. The primary was most likely cerebral.

Approximately 30 per cent of extranodal NHL involve the head and neck structures.[2] NHL accounts for 0.3 to three per cent of all intracranial tumours and 33 per cent present in multiple areas.[3] CPA tumours account for eight to 10 per cent of all intracranial tumours and 80–90 per cent of these are acoustic neuromas.[4–6] A wide literature review of series and case reports found 15 reported CPA lymphomas. Twelve represented primary disease. The incidence of NHL has been increasing over the past 20 years. It is more common in immunocompromised patients and the rising number of human immunodeficiency virus (HIV) positive patients is no doubt contributing to the increasing incidence. It is estimated that six to 30 per cent of HIV positive patients will develop lymphoma, most of these extranodally.[7,8]

Diagnosis of middle-ear cleft tumour is often difficult. A lack of histological diagnosis at the first two mastoidectomies delayed the diagnosis in this case. It is unusual for benign disease to cause facial paralysis. Facial nerve paralysis has been reported in 30–60 per cent of temporal bone malignancies and tends to follow hearing loss and symptoms of middle ear infection.[9] It is important to consider further investigation in apparent middle-ear infection that is not resolving with conventional treatment. Steroid treatment may mask early symptoms of lymphoma. Earlier MRI may have helped diagnosis, but has been reported to be no more sensitive than CT in diagnosing cerebral lymphoma.[10] It can be helpful in distinguishing lymphoma from other tumours.

This case typifies the often confusing presentation of this disorder. This patient's symptoms were attributed to more common aetiologies. Blood workup, initial histology and CT scans contributed little to making the final diagnosis. Only with MRI and tissue biopsies at the third operation was the diagnosis of lymphoma made. We believe that in a patient with persistent or recurrent VIIth nerve palsy and unhelpful CT scan, MRI would be wise to exclude pathology involving the brainstem, CPA and facial nerve itself.

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