Unusual presentation of more common disease/injury

Diffuse large B-cell lymphoma: an unusual cause of severe otalgia and facial palsy

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An elderly woman, two months after chemotherapy for diffuse large B-cell lymphoma, presented with left-sided otalgia, discharge and facial nerve palsy. MRI showed an active left mastoid infection with an ear canal lesion, likely to be a cholesteatoma. However, a biopsy of the mass showed recurrent high-grade lymphoma. Following diagnosis, the patient opted for palliative care within the community and consequently passed away a few weeks later.

BACKGROUND

Although rare, the diagnosis of lymphoma should be considered in patients presenting with similar symptoms, who have previously received treatment for high-grade lymphoma.

CASE PRESENTATION

An elderly woman presented with an acute left lower motor neurone facial palsy with a background of a 7-day history of left otalgia with reduced hearing. Five months before presentation, she was diagnosed with stage 2 diffuse large B-cell lymphoma (DLBCL). The DLBCL was found within her neck and left parotid; however, there was no bone marrow involvement. Two months after finishing six cycles of combination chemotherapy containing rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone, a positron emission tomography and CT (PET CT) scan was performed. Low-grade uptake on the left side of the neck was reported and thought to be a residual tumour (for surveillance with repeat PET CT at 2 months). There was also a PET avid area around a right tooth and left mastoid region (see figure 1). This was formally reported as a possible site of inflammation and it was advised that the scan be repeated in 3 months. Two days after the PET CT scan, the patient developed pain in the left ear with no discharge. She was seen by the oncology team who felt this was in keeping with acute otitis media and prescribed a course of co-amoxiclav.

Despite antibiotics, her symptoms deteriorated and urgent referral to the otolaryngology department by her general practitioner was made following the development of a left facial nerve palsy. On initial review, the left external auditory canal (EAC) was inflamed, stenosed and purulent. Facial movement on the left was asymmetrical with a House-Brackmann (H-B) score of 5. An examination of the right ear was normal. An initial diagnosis of left necrotising otitis externa was made and therapy was started with intravenous piperacillin and tazobactam (on

microbiology advice), ciprofloxacin eardrops and eye care. The patient also received four doses of steroids. A CT scan of the temporal bones revealed occlusion of the left external auditory canal with no bony destruction or middle-ear involvement (see figures 2 and 3). A full-body isotope bone scan showed marginally increased uptake in the left mastoid, suggestive of soft-tissue inflammatory changes and specifically no bone involvement. After 8 days of inpatient treatment, all symptoms (including the facial nerve palsy to H-B 3) had improved; therefore, she was discharged with a course of co-amoxiclav, ciprofloxacin eardrops and follow-up.

Figure 1 Axial positron emission tomography CT at the level of the mastoid showing high-uptake area at the region of the left mastoid.

Figure 2 Axial CT scan at the level of the external auditory canal showing a homogeneous opacity within the left canal and some opacification of the mastoid air cells.

The patient returned 6 days later with worsening confusion and neutropaenia. Viral encephalitis was suspected and treatment was started with an antiviral. A magnetic resonance (MR) head scan revealed a T1 low-intensity lesion within the ear canal and in the mastoid region with enhancement after contrast in the adjacent left temporal lobe. There was also leptomeningeal spread (see figure 4). The radiologist felt this was consistent with active mastoid infection and a small intracranial collection. The working diagnosis was subsequently changed to left mastoiditis with intracranial involvement, caused by a cholesteatoma.

A left cortical mastoidectomy was performed. A soft-tissue mass involving the ear canal was noted and a biopsy was done. A histological analysis of the samples was consistent with high-grade B-cell lymphoma. Following a discussion with the oncology team, the patient opted for palliative care support within the community.

INVESTIGATIONS

See above.

Figure 3 Coronal CT scan showing a homogeneous mass filling the left ear canal with no bony destruction.

Figure 4 T1 coronal MRI showing left external auditory canal lesion with leptomeningeal enhancement.

DIFFERENTIAL DIAGNOSIS

The initial working diagnosis was left necrotising otitis externa which was changed to left mastoiditis secondary to a cholesteatoma, in view of the MR head scan findings. However, following histology from the mastoidectomy and EAC mass biopsy, DLBCL was confirmed as the cause of the patient's symptoms.

TREATMENT

See above.

OUTCOME AND FOLLOW-UP

Following a discussion with the oncology team, the patient opted for palliative care support within the community. She passed away a few weeks later.

DISCUSSION

Lymphomas presenting within the ear are rare. In addition, symptoms are often similar to far commoner ear pathologies that can mask the tumour. In this patient, who was immunocompromised, the diagnosis of necrotising otitis externa appeared to be the most likely diagnosis at presentation. In addition to this, findings on imaging did not suggest a malignancy; changes were consistent with an inflammatory process within the ear.

Although rare, several case reports of high-grade lymphoma presenting within the ear canal and temporal bone have been documented. Those arising within the ear canal and external auditory meatus appear to be more common. Fish *et al*¹ reported a case of B-cell lymphoma presenting as a solitary tumour within the external auditory meatus. Merkus *et al*² remarked upon the ability of lymphomas within the ear to present with symptoms far more likely to be attributed to commoner ear pathologies. In a review of cases, Ogawa *et al*³ report otalgia, hearing loss and ear fullness to be the commonest presenting symptoms. Owing to the rarity of this tumour presenting at this primary site, diagnosis is often delayed.

Lymphomas of the inner ear are rarer still and often involve bony destruction and intracranial spread.⁴ As expected they present with symptoms such as hearing loss, tinnitus and vertigo again, implying that diagnosis is somewhat challenging. Our patient was shown to have PET avid response within the left mastoid region shortly after finishing chemotherapy (see figure 1). In the presence of new-onset otalgia and an immunocompromised state after chemotherapy, this was presumed to be otitis media and the patient was treated with oral antibiotics. It is easy, in retrospect, to call on the differential diagnosis of this imaging finding, but recurrence at this stage after treatment is unusual.

Following PET CT, further imaging to investigate possible necrotising otitis externa revealed no changes specific to malignancy. Isotope bone scanning is not specific in differentiating between inflammation and malignancy and temporal bone CT imaging showed opacification consistent with either fluid or soft-tissue mass (and no bony erosion, hence no surgical intervention) (see figures 2 and 3). Later, enhancement on post-contrast MR scanning suggested intracranial collection, which prompted surgical intervention with cortical mastoidectomy. All changes were in keeping with a progressive infective disease of the temporal bone, especially in the presence of facial nerve palsy. Extranodal lymphoma can also share these features, especially those relating to MRI, with the lesion showing intermediate intensity on T1 and T2 with post-contrast enhancement.^{5 6}

The diagnosis of residual extranodal lymphoma was made following biopsy of the mass. Prior to this the patient had failed to respond to the mainstay of treatment for necrotising otitis externa. In this setting, with an opaque mastoid and evidence of possible intracranial extension, mastoidectomy to create a safe ear is the only

option. The ability of this tumour to mimic chronic mastoiditis was ‘uncanny’ and a feature that clinicians should appreciate as the incidence in lymphoma increases.

Learning points

- ▶ Lymphoma within the mastoid can present in a similar manner to other more common ear pathologies.
- ▶ Consider lymphoma as a differential diagnosis in those presenting with otalgia and facial palsy.
- ▶ Be prepared to change working diagnoses when faced with a clinically deteriorating patient.

Competing interests None.

Patient consent Obtained.

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