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CASE REPORT

Multiple myeloma of cranium with external ear canal swelling and occipital mass: A rare case report

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ABSTRACT

Multiple myeloma (MM), the most common plasmacell neoplasm, manifests relatively often in the head and neck region, but rarely in the ear. We present a patient with fluctuating hearing loss, otalgia, persistent otitis externa and occipital swelling. CT imaging showed a large mass of cranium and the initial radiological diagnosis was plasmacytoma. Multiple osteolytic bone lesions, monoclonal serum protein with serum M component concentration of 35.2 g/L and bone marrow biopsy with abnormal, clonal plasma cells population confirmed the diagnosis of multiple myeloma. Although a rare manifestation in the external ear canal and its surroundings, plasmacytoma or multiple myeloma should be considered as a possible diagnosis in the presence of a scalp mass or a treatment resistant swelling of the external ear canal.

Abbreviations: MM: multiple myeloma; SPB: solitary plasmacytoma; EMP: extramedullary plasmacytoma; CT: computed tomography; PET: positron emission tomography; MRI: magnetic resonance imaging

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KEYWORDS
Multiple myeloma; plasmacytoma; occipital mass; external ear canal

Introduction

Multiple myeloma (MM) is the most common plasmacell neoplasm, and myeloma-related lytic lesions are seen relatively often in the head and neck region. The other forms of plasmacell neoplasms are solitary plasmacytoma (SPB) and extramedullary plasmacytoma (EMP) [1–4]. MM commonly presents with disseminated bone marrow involvement, lytic bone lesions, anemia, renal insufficiency and hypercalcemia. The mean age at the diagnosis of MM is relatively old, that is, 65–70 years, and only 10% of the patients are under 55 years during the diagnosis [1–2]. In this case report, we present a 41-year-old MM patient with persistent ear symptoms.

Case report

Forty-one-year-old man presented to the university hospital otorhinolaryngologic outpatient clinic with persistent otitis externa. Fluctuating hearing loss on his right ear had started 6 months before, and he had noticed soft swelling in the right occipital region four months before (Figure 1(A)). Recently, he had experienced some vertigo but only occasional ear pain. He had no history of nasal obstruction, facial pain or orbital symptoms. He did not have other constitutional symptoms such as weight loss, fever, loss of appetite, fatigue or night sweats.

In the physical examination, the external ear canal was blocked by soft tissue swelling (Figure 1(B)) most notably in the posterior wall, where needle aspiration resulted with low pressure blood. Computed tomography (CT) (Figure 1(C)) revealed a 13.0 cm by 9.3 cm by 5.0 cm homogenously enhanced tumor destructing right temporal bone, occipital bone, and parts of parietal bones. Caudally the tumor surrounded foramen magnum from lateral and posterior aspects. On the right side, tumor infiltrated foramen jugularis and blocked sinus sigmoideus and internal jugular vein. The appearance was consistent with plasmacytoma and the patient was admitted for hematologic evaluation.

At the hematology department, a whole-body positron emission tomography (PET) scan revealed several

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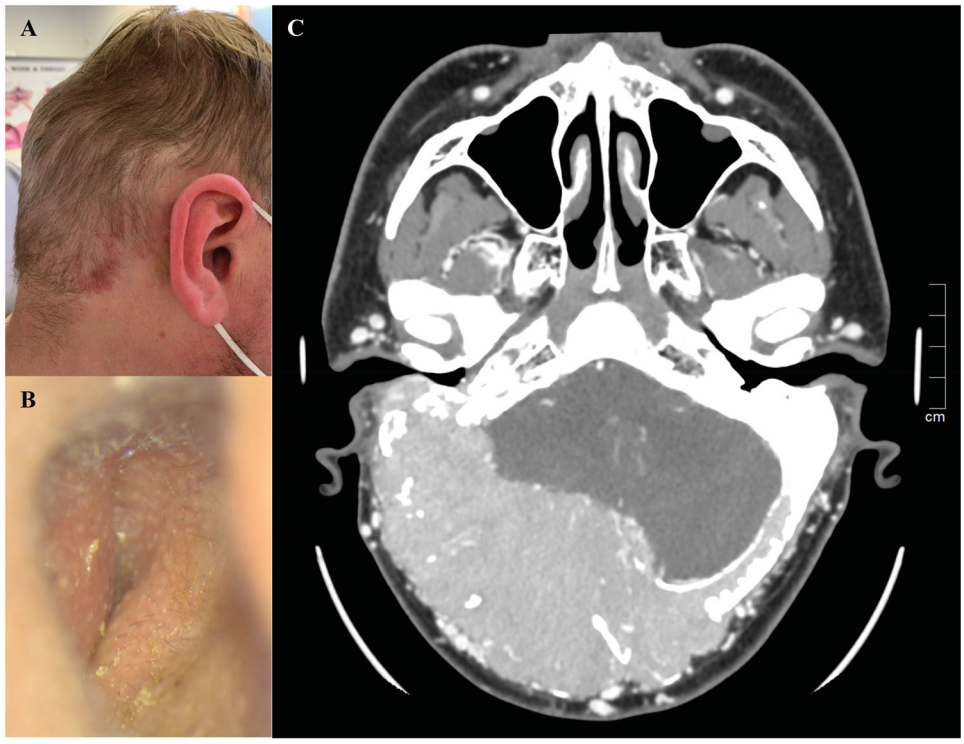

Figure 1. The clinical and radiological findings of the patient. The patient had swelling in the right occipital region (Panel A) and external ear canal (Panel B). Panel C shows the CT scan with large mass destructing right temporal bone, occipital bone, and parts of parietal bones.

active bone lesions. Cranial magnetic resonance imaging (MRI) had similar findings compared to CT scan with $14 \times 10.5 \times 4.8$ cm sized tumor destructing right temporal and occipital bone and processus mastoideus. MRI also showed a minor cerebrospinal fluid flow disturbance. A biopsy from the tumor was taken by neurosurgeon, but the sample was nonpresentative showing only connective tissue. The patient had no anemia (hemoglobin 1.44 g/dL), and his serum calcium and renal function tests were normal (ionized calcium 1.22 mmol/L and plasma creatinine 75 µmol/L). Serum protein electrophoresis showed an elevated total protein levels of 103 g/L with increased gamma globulin level of 41 g/L and M component concentration of 35.2 g/L. Serum immunoglobulin levels were as follows: IgG was elevated 61.2 g/L, whereas IgA and IgM levels were decreased (0.7 g/L and 0.3 g/L, respectively). Serum-free light chains test showed elevated levels of kappa (99.4 mg/L) and normal levels of lambda-free chains (9.5 mg/L) with a ratio of 10.4. Serum albumin was 43.8 g/L and beta 2 microglobulin was 2.6 g/L and his bone marrow biopsy showed a normal total bone marrow cell count with 10–30% plasma cells exhibiting kappa clonality, thus verifying the diagnosis of MM. IgG kappa ISS stage I MM with several osteolytic bone lesions was diagnosed. The treatment was started with the combination of bortezomib, cyclophosphamide and dexamethasone (VCD) in patient's regional hospital, where follow-up was also arranged.

Discussion

Worldwide, the median age at the time of MM diagnosis is 65–70 years, and only 2% of cases occur in patients under 40 [1]. Therefore, the MM patient in this case report was relatively young.

There are several reported cases in literature with MM in the head and neck region and also some cases with temporal bone lesions. According to the most recent and the most comprehensive review article by Sweeney et al., there has been 25 cases of plasmacytoma of the temporal bone between 1950 to 2015, from which 44% were diagnosed with MM [5]. They showed that plasmacytoma in temporal bone is a generally rare and can mimic other tumors, such as jugular paragangliomas, endolymphatic sac tumors, or chondrosarcomas. Some other MM cases with skull involvement has been reported in the literature [6–10].

Although comparably often manifesting itself in the head and neck region, typical MM lesions are found in the spine causing usually prolonged back pain [1]. Compared to this, 80% of EMPs appear in

the head and neck region. Still, EMPs constitute less than 1% of the head and neck neoplasms. EMPs commonly locates in nasal cavity, paranasal sinuses and nasopharynx [3–4]. In this case, the patient had unusually large tumor affecting right temporal and occipital bone and processus mastoideus with uncommon symptoms.

Despite having a large mass of scalp, our patient had rather minor symptoms. Also, initially, the MM's diffuse occipital swelling did not draw the attention of the clinicians, and the symptoms were treated as otitis externa. Finally, as the standard treatments for otitis externa did not resolve the symptoms, the patient was referred to the university hospital otorihinolaryngologic clinic. There clinical examination and needle aspiration of the ear canal mass lead to imaging, which lead to correct diagnosis.

In summary, persistent otitis externa not responding to the standard treatments should raise suspicions of malignancy. Prompt otologic examination with biopsies and appropriate imaging are warranted.

Informed consent

We have obtained informed consent from the patient presented in the case report. Noted that there is no identification of the patient, nor any images that can directly identify the patient.

Disclosure statement

The authors report no conflicts of interest.

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