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Case Report

Multiple myeloma with otologic manifestation: case report

Mieloma múltiplo com manifestação otológica: relato de caso

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ABSTRACT: Introduction: Multiple myeloma is a neoplasm that results in the disordered multiplication of plasma cells, production of monoclonal immunoglobulin and normally presents as osteolytic lesions. Otologic changes associated with multiple myeloma are rare and can be confused with several chronic ear manifestations. Objective: To describe the case of a patient with involvement of the temporal bone by multiple myeloma presenting otologic manifestations. Method: Case report collected through clinical interviews and patient's consent after signature of the informed consent form. Results: Male patient, 74 years old, with signs of hypoacusis, physical examination without significant changes, and asymmetric audiometry with mixed hearing loss in the left ear and sensorineural hearing loss in the right ear. The diagnosis was suspected by computed tomography that showed hypoattenuating content filling the cells of the left mastoid, with sclerosis of the internal cortical bone suggesting malignant neoplasm or calcium absorption disorder. He was referred to a hematologist, who confirmed multiple myeloma and started treatment. Considerations for the practice of general practitioners: Awareness that certain otologic manifestations may be indicative of systemic conditions, such as multiple myeloma, highlights the relevance of the general practitioner in early detection and appropriate referral to a multidisciplinary approach.

KEY WORDS: Temporal Bone; Multiple Myeloma; Hypoacusis.

RESUMO: Introdução: Mieloma múltiplo é uma neoplasia que cursa com a multiplicação desordenada de plasmócitos, produção de imunoglobulina monoclonal e normalmente se apresenta como lesões osteolíticas. As alterações otológicas associadas ao mieloma múltiplo são raras e podem ser confundidas com diversas manifestações crônicas do ouvido. Objetivo: Descrever o caso de um paciente com envolvimento do osso temporal pelo mieloma múltiplo apresentando manifestações otológicas. Método: Relato de caso coletado através de entrevista clínica e com consentimento do paciente por meio da assinatura do termo. Resultado: Paciente do sexo masculino, 74 anos, com sinais de hipoacusia, exame físico sem alterações significativas e audiometria assimétrica, com perda auditiva mista no ouvido esquerdo e neurossensorial no direito. O diagnóstico foi suspeitado por tomografia computadorizada, que mostrou conteúdo hipoatenuante preenchendo as células da mastoide esquerda, com esclerose da cortical óssea interna, sugerindo neoplasia maligna ou distúrbio de absorção do cálcio. Foi encaminhado ao hematologista, que confirmou mieloma múltipo e iniciou tratamento. Considerações para prática do médico generalista: A conscientização de que certas manifestações otológicas podem ser indicativas de condições sistêmicas, como o mieloma múltiplo, ressalta a relevância do médico generalista na detecção precoce e encaminhamento adequado para uma abordagem multidisciplinar.

PALAVRAS CHAVE: Osso Temporal; Mieloma Múltiplo; Hipoacusia.

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INTRODUCTION

Multiple myeloma (MM) is a hematological neoplasm that involves proliferation of malignant plasma cells in the bone marrow and a large production of light chain or heavy chain monoclonal immunoglobulins identifiable in serum or urine[1]. It accounts for approximately 1% of malignant neoplasms and is the second most common hematological neoplasm[2].

Multiple myeloma promotes organ infiltration of neoplastic plasma cells, production of excess immunoglobulins and suppression of normal humoral immunity. As a consequence, severe anemia, bone damage, renal failure, recurrent infection, hypercalcemia, coagulation abnormalities, neurological symptoms and vascular manifestations of hyperviscosity are observed[2,3].

The most common manifestations occur in the bones, as osteolytic lesions, but they can also manifest in the form of isolated bone or extramedullary plasmacytomas[4]. Multiple myeloma presents with multiple lytic lesions, while plasmacytoma presents with a mass lesion[5]. The ribs, vertebrae, cranial vault and pelvis are the most affected bones, characterized by regional edema, increased local temperature, poorly localized pain and pathological fractures[3-6].

Although rare, isolated involvement of the temporal bone by MM can cause secretory or purulent otitis media, mastoid effusion, mastoid, middle ear and auditory tube hemorrhage, degeneration of the organ of corti, labyrinthine hydrops and ventricular hemorrhage[6].

The diversity of otologic manifestations similar to other chronic ear diseases poses a challenge to diagnosis. The objective of this study is to report a rare case of MM in the temporal bone, suspected from an audiometry with asymmetric hearing loss.

METHOD

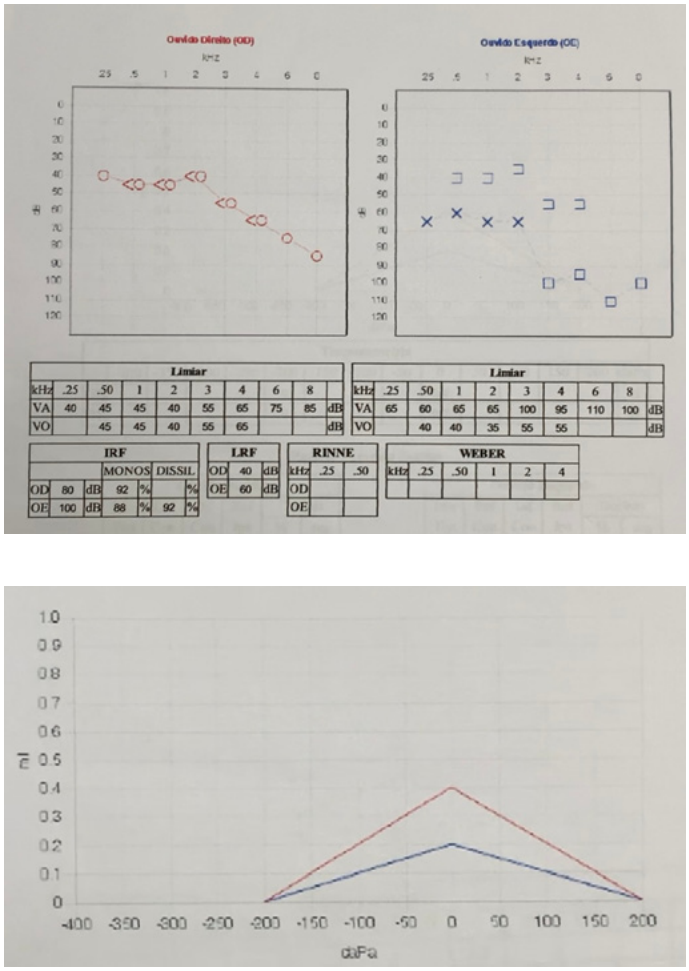
Case report collected through clinical interview with consent of the patient and signature of the informed consent form.

CASE REPORT

Male patient aged 74 years old attended the Otorhinolaryngology office. The companion complained that the patient showed indirect signs of hypoacusis. The patient denied tinnitus, dizziness, previous ear infections or any history of previous work with exposure to noise. History of hypertension, dyslipidemia, diabetes mellitus and chronic pain in the lower back. Using polymedication. He denied smoking, drinking alcohol or using other substances.

No significant changes were evident on physical examination. The otorhinolaryngological examination showed bilateral intact and opaque tympanic membrane, mild septal deviation to the right, area II grade I, grade I tonsils, absence of palpable lymph nodes in the cervical chains. Patient complained of low back pain with previous x-ray.

The audiometric evaluation demonstrated moderate to severe sensorineural loss with a descending curve in the right ear, a Word Recognition Score (WRS) 92% and Speech Recognition Threshold (SRT) 40 dB, Ar tympanometric curve. There was mixed hearing loss in the left ear with WRS 88% and SRT 60 dB, Ar tympanometric curve.



Figures 1-2 - Pure tone audiometry of the right and left ears. Sensorineural hearing loss in the right ear and mixed hearing loss in the left ear. Left and right ear tympanometry with Ar curve.

Due to asymmetric hearing loss and no history of previous otitis, additional imaging examination was requested; mastoid tomography. After a month, he returned with a tomography result showing diffuse bone textural changes in the cranial vault and base bones characterized by areas of sclerosis permeated by lytic lesions, some showed rupture of the cortical bone. Hypoattenuating content filled the cells of the left mastoid, with sclerosis of the internal cortical bone. Such a finding corroborated a wide range of differential diagnoses, including calcium metabolism diseases, myeloma-type neoplastic lesions and metastases from primary tumors. Pneumatized right mastoid with normal transparency and other structures without abnormalities.

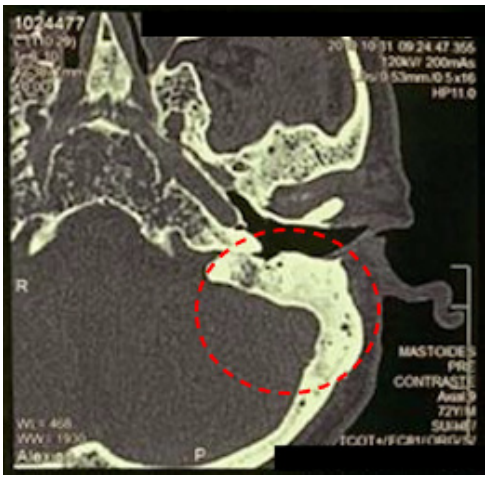


Figure 3 - Axial computed tomography of the left mastoid showing hypoattenuating content and sclerosis of the internal cortical bone.

Due to suspected neoplasia, he was urgently referred to a hematologist as well as to the endocrinology department by suspected calcium metabolism disorders. Additional laboratory tests were requested.

Subsequently, he underwent a bone marrow biopsy with results consistent with MM with monoclonal lambda. He also performed x-rays of the pelvis, arms, forearms, femoris, knees, legs, skull, hemithorax, cervical, dorsal and lumbosacral spine with results of lytic lesions in the pelvic bones, femoris, cranial vault and costal arches. After diagnosis, the patient underwent 23 chemotherapy sessions (bortezomib, cyclophosphamide, dexamethasone) in eight months of treatment. Low back pain and other systemic symptoms improved exponentially with the aforementioned treatment. The disease is currently in remission with hematological follow-up.

At the beginning of chemotherapy sessions, he also developed deep venous thrombosis of the femoral, popliteal, peroneal and gastrocnemius veins of the left lower limb and thrombophlebitis of the great saphenous vein. He was referred to the emergency room, hospitalized and clinical treatment was carried out with improvement in the condition.

During otorhinolaryngological follow-up, the patient was advised to use an individual sound amplification device to gain hearing and had good adaptation.

DISCUSSION

The initial assessment of patients with hearing loss consists of obtaining their otologic history. One should be asked about the onset of the symptom, its progressive nature, unilateral or bilateral involvement, previous trauma, otorrhea, pain, associated vertigo, previous and continuous exposure to noise, use of ototoxic medications, previous surgeries and family history of hearing loss. The physical examination can determine the patency of the external auditory canal, integrity and appearance of the tympanic membrane, presence of otorrhea, among other findings[7]. Audiometric assessment via air and bone is important in continuing the assessment. It identifies the nature, degree of hearing loss and whether one or both ears are involved[8]. Peripheral hearing loss is classified as conductive when caused by impairment of the external or middle ear, which makes it difficult for the sound to get through the inner ear; or sensorineural when it affects the inner ear, caused by dysfunction in the cochlea or spiral ganglion. Hearing loss is categorized as mixed when it has both conductive and sensorineural components[9].

Hearing loss in the external ear can occur due to impacted earwax, inflammation, foreign body, neoplasia, exostoses or osteomas. The middle ear can be affected by trauma, acute or chronic otitis media, cholesteatoma, fenestra and benign otosclerosis, semicircular canal dehiscence or neoplasia. The most relevant imaging modalities for analyzing hearing loss are computed tomography (CT) and magnetic resonance imaging (MRI)[7].

As observed in the case described, the patient's audiometry demonstrated sensorineural hearing loss in the right ear and mixed hearing loss in the left ear, no history of previous otologic diseases or the existence of significant abnormalities in the physical otological examination. The mixed hearing loss in the left ear suggests that in addition to the sensorineural component there is conductive hearing loss, with probable involvement of the middle ear, given that the patency of the external ear was visualized. Since there are no associated signs or symptoms and there is an asymmetric hearing loss, imaging becomes a valuable alternative to elucidate the condition.

The presence of hypoattenuating content in the left mastoid with sclerosis of the internal cortical bone seen on the CT scan of the patient in question is suggestive of bone erosion. This finding corroborates calcium absorption disorders or malignant neoplasms, since benign neoplasms can expand, but not promote erosion[7]. In addition to this finding, the suspicion of MM increases as diffuse bone textural changes in the cranial vault and in base bones were also seen[3-6]. The referral to a continuous diagnostic approach involving collaboration with specialists in hematology and endocrinology is fundamental.

The diagnosis of MM is established when there is the presence of ≥10% of clonal plasma cells in the bone marrow or through a biopsy confirming the existence of plasmacytoma associated with evidence of one or more disease-defining events (hypercalcemia, renal dysfunction, anemia and/or osteolytic lesions), in addition to the presence of three specific biomarkers: clonal plasmacytosis in the bone marrow ≥60%, proportion of involved/uninvolved serum free light chains ≥100 (assuming that serum free light chains are ≥100mg/L), or >1 focal lesion detected on MRI[10]. Osteolytic lesions can be visualized on radiographs, which reveal a possible plasmacytoma, myelomatosis, diffuse osteopenia and sclerosing lesions; by CT scan, which shows perforating osteolytic lesions, expansive lesions with masses in soft tissue, diffuse osteopenia, fractures and rarely, bone sclerosis; and by MRI, which allows the direct visualization of the tumor in the bone marrow, the precise determination of the extent of the disease, and the monitoring of the progress of treatment[11].

The temporal bone harbors a wide variety of injuries. Few reports on isolated otologic manifestations due to temporal bone involvement by MM have been described. Some mention that auditory symptoms occur mainly in the terminal phase of the

disease[12]. However, at this stage, they are overshadowed by diffuse skeletal and systemic manifestations[13]. Hypoacusis was described in some individual case reports and mainly accompanied by dizziness, tinnitus or diffuse signs of MM[6,13].

Authors consider that injuries to the temporal bone and symptoms such as tinnitus and vertigo are considered poor prognostic factors. However, this relationship is debatable, as the worsening of the condition may be related to the patient's clinical conditions, such as renal toxicity caused by chemotherapy or the delay in diagnosis[6].

CONSIDERATIONS FOR THE PRACTICE OF A GENERAL PRACTITIONER

In the face of patients with hearing loss, it is mandatory to collect an otologic history, followed by a thorough physical examination including assessment of the permeability of the external auditory canal and the integrity of the tympanic membrane. An audiometric assessment plays an essential role, enabling the categorization of hearing loss and determining its incidence in one or both ears. Knowledge of the different etiologies is crucial to guide subsequent diagnostic steps, including the use of imaging tests. The awareness that certain otologic manifestations may be indicative of systemic conditions such as MM highlights the relevance of the general practitioner in early detection and appropriate referral to a multidisciplinary approach.

Participation of the authors: Flávia Caroline Klostermann: carried out the clinical interview, collected the data, supervised, reviewed the literature, wrote and made a critical review. Eduardo Vieira Couto: supervised, analyzed and interpreted the data, literature review, wrote and made a critical review. Lucas Zanon de Freitas: analyzed and interpreted data, literature review, wrote and made a critical review.

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