ARCHIVES OF HEAD AND NECK SURGERY
ISSN 2595-2544

CASE REPORT

Extramedullary hematopoiesis of the middle ear

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Abstract

Extramedullary hematopoiesis is characterized by the production of blood at sites outside the bone marrow that occurs due to an increased demand of the hematopoietic system as a consequence of chronic anemia associated with ineffective bone marrow response. Theoretically, any organ may be involved in extramedullary hematopoiesis. In this case report, a patient with sickle cell anemia developed an extremely rare presentation of extramedullary hematopoiesis in the middle ear. Therefore, extramedullary hematopoiesis should be included as a differential diagnosis of middle ear masses in patients with chronic anemia.

Keywords: anemia, sickle cell; ear, middle; hematopoiesis; hematopoiesis, extramedullary; hemoglobin sickle cell disease.

How to cite: Orlando VR, Sousa MCA, Sousa NJA. Extramedullary hematopoiesis of the middle ear. Arch Head Neck Surg. 2020;49:e00202019. https://doi.org/10.4322/ahns.2020.0002

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Financial support: None.
Conflicts of interest: No conflicts of interest declared concerning the publication of this article.
Submitted: May 20, 2019.
Accepted: December 22, 2019.

The study was carried out at Faculdade de Ciências Médicas de Minas Gerais Belo Horizonte MG ,Brasil

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Introduction

Extramedullary hematopoiesis (EMH) is characterized by the production of blood cells outside the bone marrow - a common phenomenon in patients with chronic hematological pathologies such as hemolytic anemia, thalassemia, myelofibrosis, lymphoma, leukemia, and sickle cell anemia. Such pathologies generate a condition of ineffective hematopoiesis[1] that needs to be compensated[2].

The extramedullary tissue that produces blood cells is called extramedullary hematopoietic tissue, and there are two possible situations of EMH: normal hematopoietic tissue may migrate to sites outside the bone marrow or a previously latent hematopoietic tissue can be reactivated, generating nonspecific presentations that can mimic diseases, including malignancies[1,2].

The most commonly involved sites are the liver, spleen, lymph nodes, paravertebral space, thymus, heart, prostate, ligaments, adrenals, pleura, retroperitoneal space, skin, and nerves. Rarely, EMH extends to the head and neck, but it may appear in the nasopharynx, paranasal sinuses, thyroid, and close to the trachea and middle ear[1,2].

Sickle cell (SC) anemia, or SC disease, is the most common condition among chronic hereditary hemolytic diseases, affecting mainly African descent populations[3]. It is a disease characterized by structural hemoglobinopathy

and chronic hemolytic anemia, with systemic repercussions due to hypoxia and vaso-occlusive crisis, which can occur in several organs such as the heart, bones, kidneys, and brain[4].

Case report

A 28-year-old, African descent, female patient with SC anemia was assisted with nasal complaints. She reported no hearing loss, pulsatile tinnitus, vertigo, or otalgia. During the otorhinolaryngological examination, she had a retrotympanic wine-red mass on her left ear's posteroinferior quadrant. The right tympanic membrane was intact, unalteredintact. She had a normal audiomatry test. (Figure1). Computed tomography (CT) of the temporal bones showed a soft tissue density mass in the left mesotympanum, with no bone erosions including in the ossicular chain (Figure 2). As the patient had been previously diagnosed with SC anemia, the use of contrasts during CT was contraindicated due to the increased risk of red cell sickling, and this made it impossible to differentiate an inflammatory lesion from a possible glomus tympanicum. As there was suspicion of glomus tympanicum, the patient underwent surgery to excise the left ear lesion retroauricular e transcanal

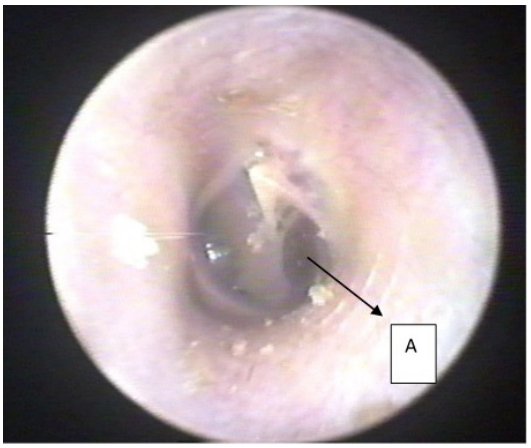

Figure 1. Right Tympanic with retrotympanic wine-red mass on her left ear's posteroinferior quadrant.

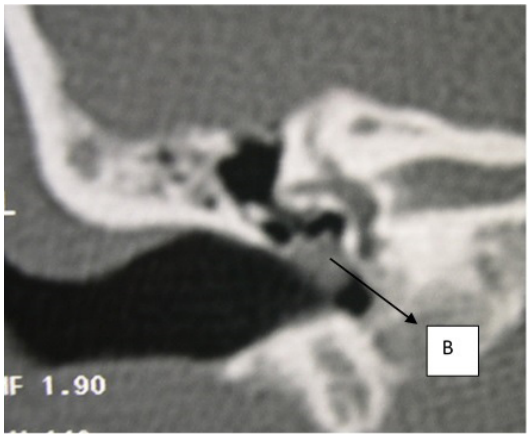

Figure 2. Computed tomography (CT) of the temporal bones : tissue density mass in left mesotympanum.

approach, with a tympanomeatal flap that provided a good exposure of the lesion. After curettage of the posterosuperior bone ridge with good visualization of the ossicular chain, the soft tissue mass was detached from the cochlear promontory without major difficulties with minimal perioperative bleeding (Figure 3). With no evidence of glomus tympanicum at anatomopathological examination, the slide was sent for immunohistochemistry (IHC), which revealed the presence of EMH.

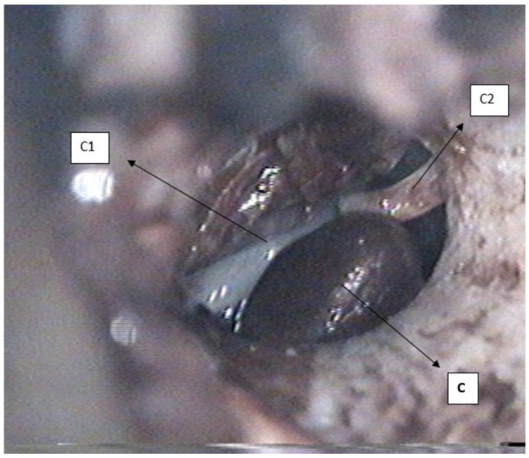

Figure 3. C: Retrotympanic wine-red mass; C1: Tympanomeatal flap; C2: Malleus.

Discussion

The most common vascular neoplasia of the middle ear is the glomus tympanicum, which was our diagnostic hypothesis in the preoperative and postoperative periods, despite the fact that the patient did not present hypoacusis or pulsatile tinnitus. The correct diagnosis was only confirmed after IHC study of the lesion.

Unlike other cases described in the literature, only four this patient did not have any otological complaints, and the red-purple mass was a casual finding. As in other cases, CT was performed aiming to perform a more comprehensive analysis of the lesion and a differential diagnosis of other middle ear tumors. The examination showed a well-circumscribed soft tissue density mass located in the mesotympanum with no involvement of the ossicular chain, very similar to a glomus tympanicum.

Extramedullary hematopoiesis is a relatively common phenomenon in patients with chronic anemia, as in SC disease. This occurs because of an increased demand of the hematopoietic system as a result of a condition of chronic anemia associated with ineffective bone marrow response[1,2]. The body responds to this increase in demand in three stages: initially, there is an increase in bone marrow cellularity; subsequently, an expansion of blood production to the bone cortex develops; finally, EMH occurs[1,4].

Seldom, EMH extends to the head and neck, but it may appear in the nasopharynx, paranasal sinuses, thyroid, and close to the trachea and middle ear[1,2].

Although SC disease is associated with sensorineural and central hearing loss, conductive hypoacusis rarely occurs. According to some reports, the incidence

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of sensorineural hearing loss varies between 5 and 7%, and is related to ischemic injuries in the organ of Corti. When conductive hypoacusis occurs, it may be a consequence of bone resorption secondary to vaso-occlusive crisis (previously demonstrated in histopathological studies of the temporal bones), or of the mass effect exerted by EMH itself[3,5].

Despite its rare occurrence, EMH should be included as a differential diagnosis of retrotympanic masses in patients with chronic anemia.

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