Case Report

Middle Ear Extramedullary Hematopoiesis: A Case Report

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Background: Extramedullary hematopoiesis (EMH) in the middle ear is exceedingly rare, with fewer than five cases reported. The authors report the first Thai case of middle ear EMH.

Case Report: A 32-year-old Thai thalassemic man presented with complaint of right-sided hearing loss from a middle ear mass. The CT/MRI was done and the diagnosis of EMH was confirmed by a pathological examination after a surgical removal.

Conclusion: A differential diagnosis of EMH should also be done in thalassemic patients presented with a middle ear mass.

Keywords: Extramedullary hematopoiesis, Middle ear, Thalassemia

During the fetal period, the liver, the spleen, and yolk sac are blood-producing organs. After birth, hematopoiesis occurs in bone marrow of long bones, sternum, vertebral bodies, and ribs. If there is an increased demand for blood due to chronic anemia or bone marrow infiltrating diseases that cause ineffective blood production from bone marrow, such as myelofibrosis or leukemia, extramedullary hematopoiesis (EMH) can occur in various parts within the body. The more common presentation is hepatosplenomegaly. The less common locations of EMH are renal, intracranial and in the lungs. In the head and neck areas, there are rare cases of EMH involving the lacrimal gland(1), the middle ear (2-5), cervical lymph nodes, the thyroid gland, subglottic(6), and paranasal sinuses(7). EMH in the middle ear is exceedingly rare with only five cases reported. Although one percent of the Thai population is affected with thalassemic diseases and about 30 to 40% of the population has abnormal gene carriers, the present paper reported the first Thai case of middle ear EMH.

Case Report

A 32-year-old Thai man known thalassemia hemoglobin AE Bart’s disease, presented with complaint of decrease in hearing in his right ear over two-year period, without tinnitus or vertigo. He denied ear pain or any ear discharge. On physical examination, the right tympanic membrane appeared bulging due to a large multilobulated blue mass in the middle ear. The Weber test was lateralized to the right and the Rinne test was negative in the right ear (Fig. 1). An audiogram showed right mixed hearing loss with a 25 dB air bone gap and speech reception threshold (SRT) of 50 dB (Fig. 2).

Computed tomography (CT) scan showed a generalized widening of the medullary cavity of the bone marrow and coarse trabeculations of the skull base, ethmoid sinuses and temporal bones. There was a soft tissue mass filling in the right epitympanum, mesotympanum, and hypotympanum with extension into the Eustachian tube. The ossicular chain was preserved. There were also soft tissue masses

Fig. 1 Otoscopic examination showing blue mass in the right middle ear.
expanding into the posterior ethmoid sinuses and sphenoid sinus (Fig. 3). The magnetic resonance imaging (MRI) demonstrated low signal T1-W and intermediate to slightly high signal T2-W of the right middle ear mass with faint enhancement. However, this lesion had iso-signal intensity with the bone marrow of the skull base of the sphenoid bone, petroclival region, occipital bone as well as upper cervical spine (Fig 4, 5). Blood test revealed a hematocrit of a 23% and white blood cell count of 7,296 cell/μL.

The postauricular approach with exploratory tympanotomy was done to expose a red-colored, multi-lobulated mass loosely attached to middle ear mucosa wall of the mesotympanum, epithympanum extending into the Eustachian tube. The middle ear mass was removed easily with minimal bleeding. The ossicular chain was intact. A postoperative audiogram showed an improved conductive component of hearing with SRT at 25 dB (Fig. 6). A histologic examination revealed

Fig. 2 Preoperative audiogram showing right 25 dB conductive loss and SRT of 50 dB.

Fig. 3 Axial (A) and coronal (B) CT scan showing mass in middle ear (arrows) and sphenoid (arrow head).

Fig. 4 Axial T1 (A) and T2 (B) fat suppression MRI showing middle ear mass.

Fig. 5 Post-contrast T1 axial (A) and coronal (B) MRI showing faint enhancement of middle ear mass with intact ossicles.
hematopoietic precursor cells with mature erythroid and granulocytic elements with multi-nucleated giant cells that were morphologically consistent with megakaryocytes. Scattered lymphocytes were also seen. The diagnosis of EMH was made (Fig. 7).

Discussion

EMH in the middle ear is exceedingly rare. From three previous middle ear EMH case reports, two cases are sickle cell anemia and thalassemia with Von Willebrand’s disease, all of them presented with hearing loss from middle ear masses. There are two theories on the origin of EMH. Firstly, the paraosseous foci is proposed in patients with hyperactive bone marrow that causes bone trabeculae to become very thin resulting in the medullary tissue herniating out from the underlying bone. The second is the extraosseous or intraparenchymal foci, which may arise from hematopoietic stem cells that came via blood circulation or fetal cell rest. The second theory usually is proposed when the bone marrow has failed to function. The presented case is a known case of thalassemia and the CT scan showed a hyperactive bone marrow that has caused the widening and thinning of the adjacent temporal bone and the skull base. Therefore, in the present case, the first theory is likely to be the cause of EMH in the middle ear, posterior ethmoid sinuses and sphenoid sinuses.

Rather than the chronic hypoxia that usually causes sensorineural hearing loss or vestibular symptoms in anemic patients, this slow-growing mass in the tympanic cavity can result in a decrease in the conductive component of hearing. The pulsatile tinnitus, which is common in glomus tumor, is less seen in EMH due to its low vascularity.

Characteristic findings in CT scans are soft tissue masses with minimal aggressive nature that are unlikely to erode ossicular chains or I-S joints. Widening of trabecular bones is also commonly seen in typical cases. MRI T1- and T2-W images reveal either low or high signal intensity of the soft tissue mass which usually has the same signal intensity with the expanding bone marrow cavities. Bone marrow cavities in thalassemic patients usually have exceedingly high red marrow activities, which are caused by conversions of fatty marrow to red marrow. This results in diffused low signal intensity in T1- and slightly high signal intensity or low signal intensity in T2-W images. Some patients with high fatty marrow may have high signal intensity in both T1- and T2-W images.

Surgical excision is recommended in symptomatic EMH of the middle ear. Surgery can be done easily with satisfactory outcomes due to low vascularized mass. Because of its benign nature, extensive surgery should be avoided. EMH is usually asymptomatic, however, in some locations, acute compressive symptoms, such as airway compromise or visual loss, can occur. In such cases, emergency treatments are required. As for other treatment modalities, EMH is sensitive to low dose radiation therapies that are used in patients who need rapid mass decompression. Blood transfusions can reduce the hematopoiesis process and hydroxyurea treatment has been used to improve the efficacy of erythropoiesis.

Conclusion

Although EMH in the middle ear is extremely rare, the differential diagnosis should be done in patient with underlying thalassemia or chronic hematologic
diseases. Radiological investigations may show benign characteristics of the mass. Middle ear conservative excisional surgery can be easily performed with good outcome.

**What is already known on this topic?**

When a patient presented with a middle ear mass, the common differential diagnosis are glomus tumor, cholesterol granuloma, cholesteatoma, or middle ear vascular anomalies. Due to very few case reports of the middle ear EMH, an otolaryngologist may not have knowledge of this entity and did not included in the differential diagnosis.

**What this study adds?**

This study reports a very rare case of EMH in middle ear that usually occurs in patients with chronic hematological diseases such as thalassemia, which is a common genetic disease in Thailand. If an otolaryngologist is aware of this entity and characteristics findings of the CT/MRI, the diagnosis can be made easily. A non-radical surgery is the appropriate treatment.

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**Potential conflicts of interest**

None.

**References**