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Case Report

Temporal bone extramedullary hematopoiesis as a cause of pediatric bilateral conductive hearing loss: Case report and review of the literature

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ABSTRACT

Extramedullary hematopoiesis occurs in children with hemoglobinopathy and chronic anemia. The liver and spleen are often affected first, but other foci can develop to support erythrocyte demand. We report a case of a nine-year-old with beta thalassemia and temporal bone extramedullary hematopoiesis causing ossicular fixation and bilateral conductive hearing loss. There is only one case in the literature describing this phenomenon in pediatric patients, and this is the first case report of bilateral hearing loss from this physiologic phenomenon. Otolaryngologists should consider this etiology in patients with chronic anemia and conductive hearing loss in the absence of otitis media.

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1. Introduction

Extramedullary hematopoiesis refers to the formation of blood cells outside of the native physiologic cellular pathways. This finding is not unusual in patients who live in a chronic state of anemia such as those with sickle cell anemia or beta-thalassemia and presents with increased cellularity of marrow space, expansion of blood formation to peripheral marrow cavities, and blood formation outside of the marrow space [1]. We present the first documented case of bilateral conductive hearing loss secondary to extramedullary hematopoiesis in a pediatric patient with beta thalassemia major.

2. Case report

A nine-year-old boy, originally from the Middle East, presented to the pediatric otolaryngology clinic for evaluation with chief complaint of hearing loss over several years. The patient has a known history of beta-thalassemia major, diagnosed at 2 years of age in Jordan, and is undergoing iron chelation therapy with deferasirox and deferoxamine due to iron overload from frequent blood transfusions. He began receiving regular transfusions at 2.5 years of age initially under a hypertransfusion protocol and then continued treatment at our center when he first immigrated to the United States in 2009, adjusting volume for weight. He was maintained at a chelation dosage of 25mg/kg/day, which is within therapeutic goal. He had no associated otologic symptoms and an otherwise unremarkable otologic history.

Clinical examination was notable for frontal bossing, malar prominence, and palpable splenic tip. On otoscopy, a bony lesion was visualized in posterior half of the middle ear through the tympanic membrane on the left (Fig. 1).

An initial audiogram revealed a mild-moderate rising conductive hearing loss on the left and a mild-slight rising conductive hearing loss on the right. On the right ear, otoacoustic emissions (OAEs) were absent, tympanometry revealed type As, speech reception threshold (SRT) was 25dB, and word recognition score (WRS) was 100% at 55dB. On the left, OAEs were absent, tympanometry was type B, SRT was 40dB, and WRS was 100% at 60dB. A repeat audiogram approximately 1 year later revealed similar hearing loss bilaterally with no significant progression of disease (Fig. 2).

Computed tomography (CT) of the temporal bone was obtained to assess for anatomic abnormalities given the visible lesion in the left middle ear. CT revealed diffuse marrow expansion of the entire skull including the bilateral temporal bones. Right predominantly

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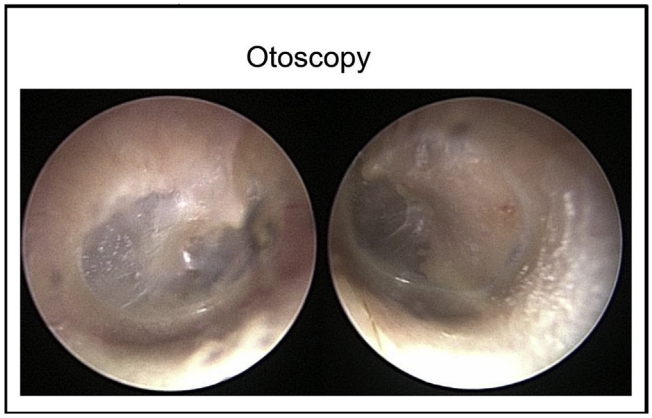


Fig. 1. Otoscopy.

epitympanic extramedullary hematopoiesis involving the heads of the malleus and incus was noted. On the left, mesotympanic and epitympanic extramedullary hematopoiesis was observed involving the ossicles and obliterating the oval window. (Fig. 3).

The patient underwent in-clinic trial with both a bone conduction hearing aid (soft band) and traditional amplification with good results and tolerance. Subjective benefit was reported but no aided testing was performed. Patient and family elected to pursue traditional amplification, however following the study time period and longitudinal testing reported herein, they were lost to follow up.

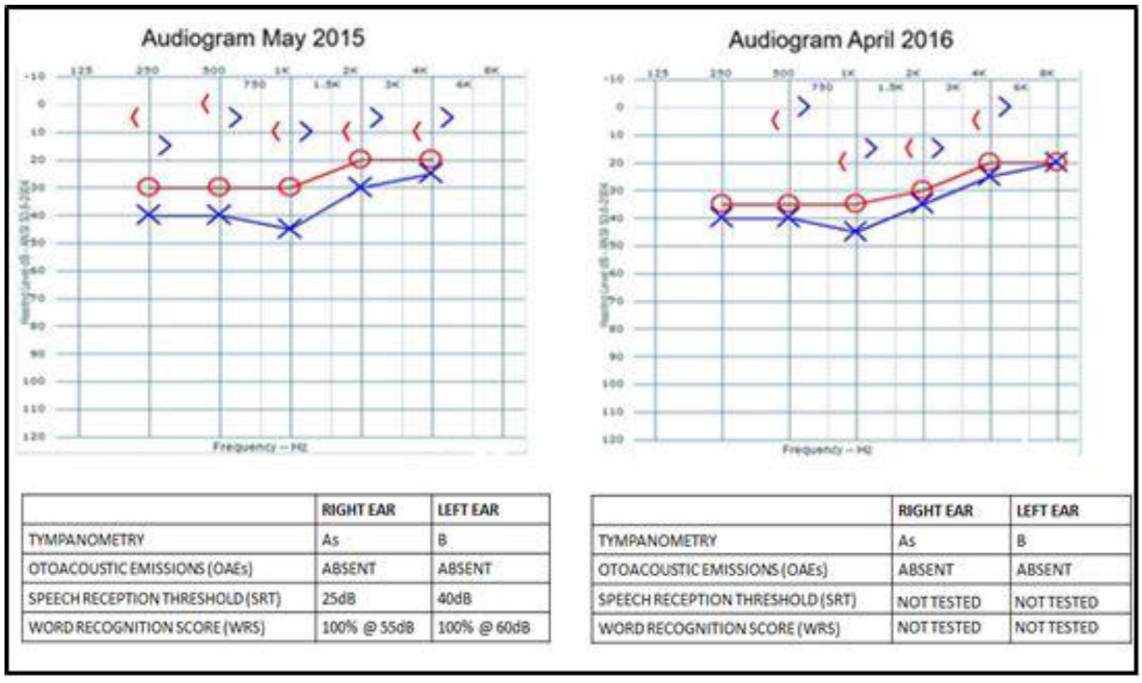
3. Discussion

Alpha and beta thalassemia are inherited disorders in which abnormal protein synthesis of the alpha or beta chains of hemoglobin result in an altered hemoglobin structure and aberrant oxygen carrying ability. Beta thalassemia may present in the forms of thalassemia major, intermedia, or minor with varying degrees of health complications ranging from asymptomatic microcytic anemia to severe anemia requiring frequent blood transfusions and risk of death. Beta thalassemia major manifests due to a relative abundance of alpha chain hemoglobin in the setting of abnormally formed beta chains and results in unstable red blood cell precursors, insoluble unpaired alpha chains, and precipitous destruction within the marrow and spleen resulting in severe hemolytic anemia [2,3]. Extramedullary hematopoiesis ensues with signs of hepatosplenomegaly, bone marrow expansion, and cortical invasion by blood cell precursors [3].

Due to the severe anemias which may occur in thalassemia patients, frequent blood transfusions, particularly in beta thalassemia major, are required. Iron toxicity from serial transfusions is typically treated with iron chelation agents such as deferasirox or deferoxamine. Hearing loss is a known complication of treatment for hemosiderosis and sensorineural loss secondary to side effects from chelating agents may be present in up to 39% of tested ears [4]. Although the mechanism of ototoxicity remains unclear, frequent audiologic evaluation is imperative in patients undergoing chelation therapy.

A less common manifestation of hearing loss in patients with blood dyscrasias such as sickle cell anemia or thalassemia is the presentation of conductive hearing loss. Applebaum and Frankel in 1989 reported on a 22-year old black male with sickle cell anemia who was found to have a bilateral conductive hearing loss. A soft tissue mass in the mesotympanum and epitympanum was identified and histopathology was consistent with extramedullary hematopoiesis, sickling erythrocytes, and blood precursors [1].

In contrast to literature attributing hearing loss to sickle cell anemia, Meara et al. reported the case of a 47-year-old male with thalassemia intermedia, a suspected soft tissue mass in the hypotympanum, and a unilateral conductive loss which was found to be consistent with extramedullary hematopoietic tissue [6]. Unilateral disease was also reported in 2003 in an 18 year old Saudi patient with thalassemia intermedia and middle ear hematopoietic tissue



	RIGHT EAR	LEFT EAR
TYMPANOMETRY	As	B
OTOACOUSTIC EMISSIONS (OAEs)	ABSENT	ABSENT
SPEECH RECEPTION THRESHOLD (SRT)	25dB	40dB
WORD RECOGNITION SCORE (WRS)	100% @ 55dB	100% @ 60dB

	RIGHT EAR	LEFT EAR
TYMPANOMETRY	As	B
OTOACOUSTIC EMISSIONS (OAEs)	ABSENT	ABSENT
SPEECH RECEPTION THRESHOLD (SRT)	NOT TESTED	NOT TESTED
WORD RECOGNITION SCORE (WRS)	NOT TESTED	NOT TESTED

Fig. 2. Audiograms.