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Case Report

Bilateral objective tinnitus in an infant with tuberous sclerosis

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ABSTRACT

This is the first report of objective tinnitus presenting as audible spontaneous otoacoustic emission in a patient with tuberous sclerosis. The tinnitus was loud, continuous, and high pitched: 7757.8 and 6257.8 Hz. The auditory system may be dysfunctional in patients with tuberous sclerosis. Possible causes of tinnitus in these patients are abnormal myelination and dysfunctional axons and neurons associated with tuberous sclerosis. A disturbance of the outer hair cells or the MOC efferent fibers innervating the outer hair cells is considered to be the source of the loud spontaneous otoacoustic emission.

1. Introduction

Objective tinnitus is caused by various factors of muscular, vascular, tubal, and inner ear origins. In 1962, Loebell first reported a 3½-year-old boy presenting with high-tonal objective tinnitus [1]. In 1979, Kemp proposed that the ear could spontaneously generate otoacoustic emission [2], which is now called spontaneous otoacoustic emission. However, truly objective tinnitus, defined as tinnitus that others can hear, is extremely rare [1,3]. Herein, we present the first case report of bilateral objective tinnitus constituting a loud spontaneous otoacoustic emission in an infant with tuberous sclerosis complex (TSC).

2. Case

The patient was a 1-month-old girl who was born at 40 weeks and 1 day with a birth weight of 3450 g. Family history was noncontributory, and there was no known consanguinity. The child presented with bilateral objective tinnitus noted by her mother. The tinnitus was continuous high-pitched pure tones from both of the child's ears. The tinnitus was louder in the left ear than in the right. The sound could be heard even 15 inches away from the ear. The tinnitus did not change when she bent her neck and was not pulsatile. Her eardrums were intact. Examination with a fiber-optic scope revealed a normal pharyngolarynx, without any abnormal muscular movements such as myoclonus. Auditory brainstem response (ABR) was normal. The ABR threshold was 30 dB (hearing level) on both sides. The latencies of I, III, and V waves evoked by clicks of 90 dB (hearing level) were 1.44 ms, 4.06 ms and 5.98 ms on the right side, and 1.42 ms, 3.90 ms, and 6.18 ms on the left side. The tinnitus was recorded with a microphone (RR-US300, Panasonic Corp) at the distance of about 1 inch from her ear. The recorded microphone signal was analyzed using Fourier-transformation algorithms. The frequency of the tinnitus was 7757.8 Hz (Fig. 1a) and 6257.8 Hz (Fig. 1b) in the right and left ears, respectively. The tinnitus was high-pitched in both ears, but the frequencies were different. The tinnitus was not different when the child was awake or asleep. Brain CT showed calcified subependymal nodules in the walls of both lateral ventricles (Fig. 2a and b) and an area with a high density of cortical tubers, 1 cm in diameter, in the left frontal lobe (Fig. 2c). T2-weighted MRI revealed several low-intensity nodular lesions of subependymal nodules along the walls of both lateral ventricles (Fig. 2d). Echocardiography indicated several masses in the heart. Five hypomelanotic macules were observed on the upper arm, forearm, back, and flank. Shagreen-like patches of elevated lesions were seen on the right upper arm. She experienced seizure without fever at the age of four months. Electroencephalogram indicated sharp waves while she was under sedation induced by midazolam and ketamine hydrochloride. She could turn in bed four months after birth. Growth had not been delayed. Partial seizure with

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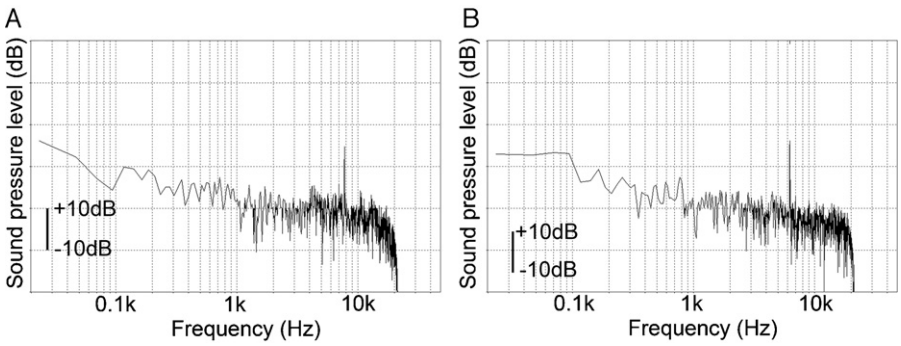

Fig. 1. Strong, spontaneous otoacoustic emissions with a sound pressure level (SPL) of 28 dB at 7757.8 Hz (A) and with a 36-dB SPL at 6257.8 Hz (B) in the right and left ears, respectively.

tuberous sclerosis was diagnosed, because she had at least three major features of the tuberous sclerosis diagnostic criteria, i.e. five hypomelanotic macules, cortical tubers, and subependymal nodules [4]. The tinnitus disappeared in the right ear, and the left-sided tinnitus reduced in intensity six months after birth. This study was approved by the local ethics committee and informed consent was obtained from her patient.

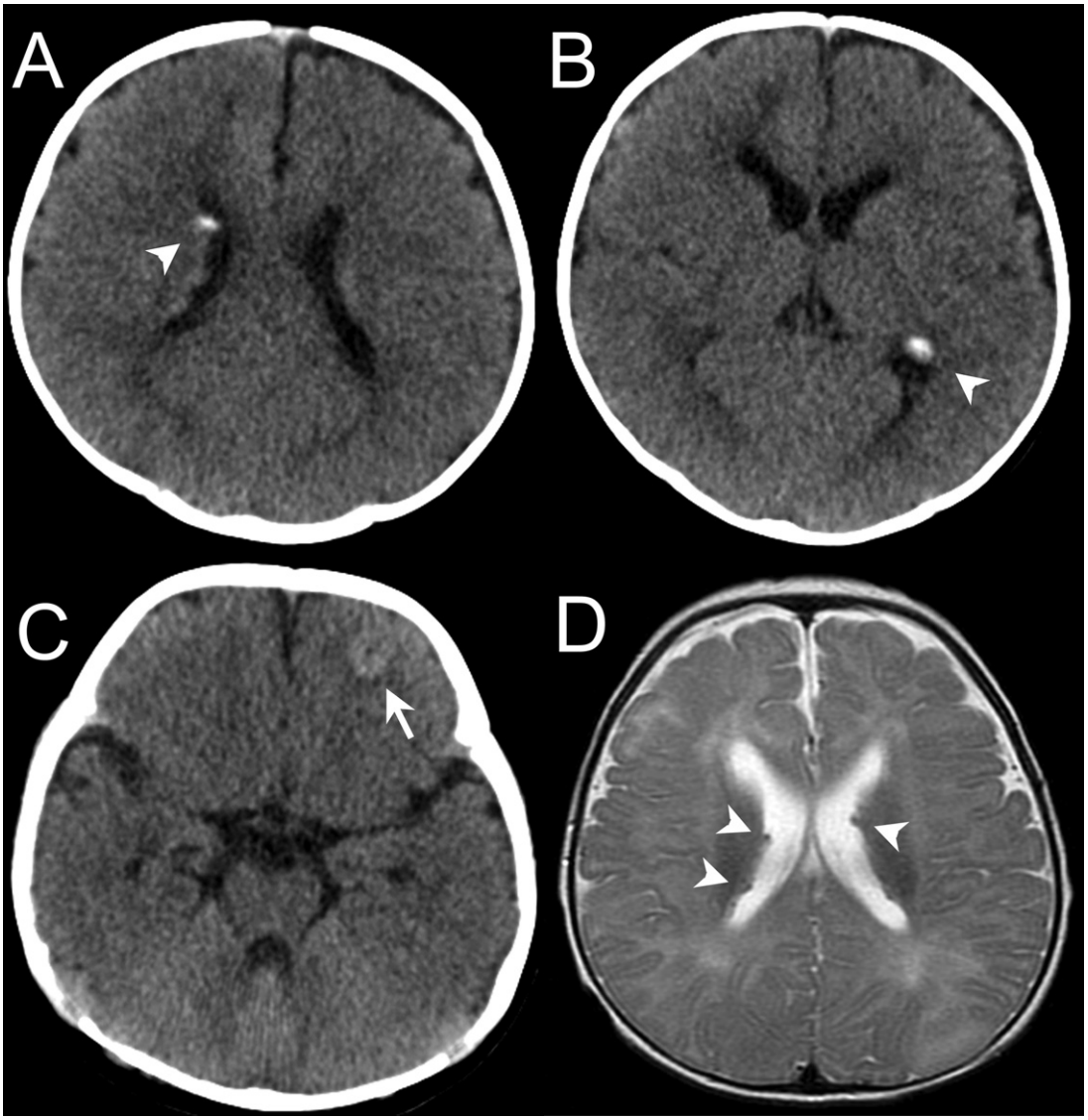

Fig. 2. Brain CT shows calcified subependymal nodules (arrow heads) in the walls of both lateral ventricles (A, B) and an area with a high density of cortical tubers (arrow), 1 cm in diameter, in the left frontal lobe (C). Several low-intensity nodular lesions (arrow heads) suggesting subependymal nodules are seen along the walls of both lateral ventricles on T2-weighted MRI (D).