Auricular ossification in a patient with congenital anterior pituitary hypoplasia

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ABSTRACT

Introduction: Ossification of auricular cartilage was first described in 1866 and its incidence is unclear, it is most commonly caused by trauma, frostbite, and inflammation but metabolic disorders have also been associated with it. Case Report: A 38-year-old male with long-term diagnosis of hypopituitarism with poor adherence to treatment presented with bilateral auricular calcification. The patient had improvement of pain in the ears once reported compliance with medication but calcification of ears showed to be not reversible with treatment. Conclusion: The pathophysiology of bilateral auricular calcification in metabolic cases remains unclear but cortisol deficiency seems to have an important role. Treatment with hormonal replacement shows improvement of pain but there was no reversibility of auricular calcification.

Keywords: Auricular calcification, Cortisol deficiency, Petrified ear

INTRODUCTION

The most common etiologies of auricular cartilage hardening are local trauma, frostbite, and inflammation [1–3]. The metabolic disorders that have been reported to be associated with it are ochronosis [4], acromegaly [5], hypercalcemia [6], and adrenal insufficiency [1]. We report a case of progressive stiffening of the ears in a patient with long standing pan hypopituitarism.

CASE REPORT

A 37-year-old white male with pan hypopituitarism is followed in the Endocrine clinic. Diagnosis was made a few weeks after birth, following a complicated early neonatal period which required frequent hospitalizations and meningitis. The patient was born at term following caesarian section with no significant antepartum complications. Anterior pituitary hormone replacement was initiated from early neonatal period. Growth hormone replacement was needed briefly for two years. The patient was able to achieve puberty and midparental height by 15 years of age. The patient never required desmopressin replacement. No developmental delays were noticed however he did not finish high school, and never held any steady job. Currently on levothyroxine 112 mcg, Androgel...
1.62% two pump depressions daily, Hydrocortisone 15 mg in the morning and 5 mg in the evening. The patient reported 50% compliance with the medications. Recent labs showed normal Free T4, and low testosterone level at 145 ng/mL.

Previous magnetic resonance image (MRI) showed hypoplasia of anterior lobe of the pituitary gland along with infundibulum and ectopic posterior pituitary gland along the undersurface of optic chiasm. Cortical dysplasia along the left parietal gyrus was noted as well.

DM-2 was diagnosed at age 33 and well controlled with oral hypoglycemic use. He developed premature cataracts at age 31 and required surgery for right eye. Over the last few years he complained of hardening of his external ears with progressive difficulty and pain on bending them. Pain could wake him up from sleep while turning in bed.

Examination showed normal visual field and bilaterally hard, stiff ears, no redness and warmth on palpation was noted. No tenderness to palpation of nasal and tracheal cartilage was noted. No other significant findings were noted on physical examination. Computed tomography (CT) scan of the face and external ears showed extensive calcifications of the external ears (Figure 1).

Since the patient had been nonadherent to treatment, we educated the patient regarding importance of following treatment, and after that, we noticed improvement in his symptoms of ear pain, the calcification of the ears though remained the same.

**DISCUSSION**

The word petrified means changed to stone or terrified to the point that one is unable to move, that is why the ossification of the auricle is sometimes called, especially in advanced cases, petrified ears, referring to the stiff, inflexible consistency of the external ear that occurs due to the calcification of its cartilaginous parts [7]. Although the most common etiologies of auricular cartilage calcification are local trauma and frostbite and inflammation [1–3], several cases related to systemic processes have been described [1], and sometimes it may be the only cutaneous marker of an underlying endocrinopathy [8].

After reviewing several cases of it, Randall and Spong proposed a classification based on three groups: local tissues injury, metabolic disorders, and familial [1]. From the metabolic disorders, adrenal insufficiency is the most common one [1].

The first reported case of it was in 1866 by Bochdalek [9], and since then, its true incidence is still unclear, “although it seems that it is infrequently seen in clinical practice” [2]. A study of 300 patients examined for inflexibility of the ears without regard to diagnosis found calcification of the ear cartilage in 3% of patients [2, 10].

The exact pathophysiology remains unclear though other factors have been proposed as a possible etiology [hypercalcemia, high adrenocorticotropic hormone (ACTH) level, desoxycorticosterone use, high growth hormone (GH) levels] [1, 11]. Review of literature indicates that cortisol deficiency seems to be the most likely to have an important role [12]. Initially, when patients with adrenal insufficiency were treated with desoxycorticosterone, it was thought that it may have a causative effect, especially since in some experiments with adrenalectomized rats it was noticed more skin calcifications in adrenalectomized rats given desoxycorticosterone along with a glucocorticoid than in animals maintained with glucocorticoids alone [1, 13], but cases of external ear ossification continue to be reported in the absence of desoxycorticosterone use, including ours [2, 12]. Cases of external ear ossification related to hypercalcemia have been reported [6] though adrenal insufficiency can cause hypercalcemia. Several cases of external ears ossification in the setting of adrenal insufficiency have been reported without hypercalcemia [2, 11].

Our review of literature and experience with this patient demonstrates that calcification of external ears may present improvement in pain once treatment is started but the calcification itself has shown to be not reversible.

**CONCLUSION**

Auricular ossification can be a complication of long standing secondary adrenal insufficiency. Although its exact pathophysiology is not known, it is likely related to cortisol deficiency, and it is an important sign to keep in mind.
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Data Availability

All relevant data are within the paper and its Supporting Information files.

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