

Calcification of Auricular Cartilages in Adrenal Insufficiency

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Calcification of the auricular cartilages is a very rare finding observed in metabolic disorders like Addison's disease, adrenal insufficiency or diabetes. Its aetiology and physiopathological mechanisms remain unclear. It may be caused by exposure to cold or trauma in the cartilage but in a significant number of cases, ossification occurs without the identification of the precipitating cause or event.

We present the clinical and pathologic findings of a patient suffering from Addison's disease who developed auricular bilateral calcification. The literature is reviewed and the physiopathological hypotheses discussed.

Key words: Calcification of auricular cartilages. Addison's disease. Adrenal insufficiency.

Calcificación de cartílagos auriculares en la insuficiencia suprarrenal

El hallazgo de pabellones auriculares duros y rígidos a la palpación es un hecho muy infrecuente y poco conocido. Se debe a la calcificación de los cartílagos auriculares que aparece en enfermedades metabólicas como la insuficiencia suprarrenal o la diabetes. Su etiología y su mecanismo fisiopatológico no están del todo aclarados. También se lo ha relacionado con la exposición al frío o traumatismos del cartílago auricular, pero en un significativo número de casos se produce sin conocimiento de su origen o causa precipitante. Presentamos el caso de un paciente con insuficiencia suprarrenal primaria (enfermedad de Addison) en el que se desarrolló una calcificación auricular bilateral, revisamos la literatura y discutimos las hipótesis fisiopatológicas.

Palabras clave: Calcificación de cartílagos auriculares. Enfermedad de Addison. Insuficiencia suprarrenal.

INTRODUCTION

Addison's disease is a primary suprarenal insufficiency most frequently caused by an autoimmune disorder. Other causes are tuberculosis (TBC), infections (meningococcus, histoplasmosis, mycosis, AIDS), bleeding, neoplasias, and accumulative illnesses (haemochromatosis, amyloidosis). It is frequently related to other endocrine pathologies. Speaking physiopathologically, there is a bilateral destruction of the suprarenal cortex due to infiltration of lymphocytes, resulting in a reduction of cortisol in blood (<5 mg/dL) and an increase in corticotropin (>50 pg/mL) due to pituitary feedback.

Typical clinical presentation is hyperpigmentation of the skin and mucosae (due to the excess of corticotropin),

asthenia, fatigue, and weight loss due to hypoglycaemia (due to diminished glyconeogenesis) and low blood pressure due to a deficit of mineralocorticoids which leads to loss of sodium and water and hypovolaemia.

Other less frequent symptoms are depletion of androgens, with hair loss and inhibition of the libido, and more rarely spastic paraplegia, ascending symmetrical paralysis, and calcifications of the auricular cartilages (with barely a score of cases described in the literature), such as the case reported here.

CASE STUDY

Male, 45 years of age, with a history of epilepsy, appendectomy, and smoking, who attends the internal medicine clinic due to progressive asthenia and weight loss (15-20 kg) lasting months. The general examination revealed hyperpigmentation of the skin and mucosae as well as hypotension (80/60 mm Hg). The rest of the cardiopulmonary and abdominal examination was normal. The analytical tests carried out (blood count, clotting times, biochemistry,

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Figure 1. Simple x-ray of the right mastoid region revealing bony/calcified images in the helix and antihelix of the right auricle.



Figure 2. Simple x-ray of the left mastoid revealing the same image of calcification of auricular cartilages.

proteinogram, agglutinations, thyroid hormones, urine, etc.) were normal, except for a positive Mantoux reaction but without subsequent confirmation of active TBC (chest x-ray, lung examination and sputum, all negative). Radiological study of the sella turcica was normal. Negative serology for HIV, meningococcus, etc.

The concentration of corticotropin (229 pg/mL) and cortisol at 08:00 and 20:00 hours (0.24 and 1.1 mg/dL, respectively) were determined. After confirmation of the suspected Addison's disease, treatment was started with hydrocortisone; the patient experienced evident clinical improvement, with reduction of asthenia and normalization of his blood pressure.

After 10 years of endocrinological follow-up, the patient was referred to the otorhinolaryngology clinic as both his auricular cartilages were hard, rigid and non-deformable.

A simple x-ray was taken and revealed calcification of auricular cartilages (Figures 1 and 2) in the context of Addison's disease.

DISCUSSION

At medical clinics, we sometimes encounter physical findings that are very infrequent in the context of a specific illness. The calcification of auricular cartilages found in certain diseases and situations is highly uncommon and its physiopathology has not been clarified precisely. The systemic illness most often related with calcification of auricular cartilages is suprarenal insufficiency.¹⁻⁶ One of the aetiological hypotheses posed is that the acute or chronic shortage of cortisol may produce a long-lasting or transient hypercalcaemia that favours the deposits of calcium in tissues predisposed due to poor peripheral circulation (extremities) or subjected to trauma or cold.¹ Other hypotheses attribute it to an excess of corticotropin^{3,5} produced by positive feedback in view of the deficit of cortisol in the blood. It has also been attributed to lengthy corticosteroid treatments.³

The exact pathogenesis has not been clearly established, as the calcaemia figures are normal in many cases, but when

faced with hypercalcaemia, it is reasonable to consider sarcoidosis, vitamin D intoxication, Burnett's syndrome, and hyperparathyroidism as the possible causes.⁷

Other authors have reported 1 case of diabetes, controlled by dietary measures, which presented bony deposits in the proximity of normal elastic cartilage.² Other cases were linked to gout, acromegaly and periarteritis nodosa.⁷ It has also been related to cold^{2,6} and traumatic aggressions to the auricular cartilage such as acupuncture.⁴ Another related cause is radiotherapy.⁶ In other cases, up to 30%, the condition occurs without any precipitating cause being known.²

A different phenomenon, known as "auricular ossificans," in which the rigidity of the auricular cartilage is attributed to the replacement of the cartilage by bone has been described. Nine cases have been histologically documented.⁶ It has been reported in surfers and has been linked to exostoses of the external auditory canal and exposure to cold.⁶

Hormone replacement therapy to redress the suprarenal insufficiency improves the condition's symptoms but does not alter the auricular calcification, for which there is no treatment.³

In conclusion, the existence of this finding, very infrequent and linked to suprarenal insufficiency, must be taken into account for correct diagnosis.

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