A CASE OF POLYGLANDULAR AUTOIMMUNE SYNDROME TYPE 1 WITH HYPERCALCEMIA AND HYPOTENSION

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Objectives:

Autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy (APECED) is also known as autoimmune polyendocrine syndrome type 1 (APS-1). We present a case of autoimmune polyendocrine syndrome type 1 with hypercalcemia of adrenal insufficiency during the calcium treatment because of hypoparathyroidism.

Methods:

A 20-year-old female patient was diagnosed with APS-1 in 2004. She applied to ER with the complaints of nausea and vomiting. Her laboratory findings proved hypercalcemia, hyponatremia and hypotension. She'd had an upper respiratory infection a week prior to her application to our hospital. In her ECG, t-waves were found to be commonly inverted. Her echocardiography revealed increased pericardial brightness. Thus, indomethacin 2x1 was started as pericardite was suspected. BP of 90/40 mmHg and dry skin were found in the physical examination. Horizontal ridges and trophic deformities on both finger nails and toes were observed **(Picture 1).** The laboratory findings were as follows: BUN:58 mg/dl, creatinine:0.82 mg/dl, Na:127 meq/l, K:4.6 meq/l, Ca:12.6 mg/dl, FSH:4.42 mIU/mL, LH:5.63 mIU/mL, ACTH:124 pg/mL, cortizol:0.40 µg/dL, DHEAS:4.6 µg/dL, free T3:1.94 pg/mL, free T4:1.07 ng/dL, TSH: 3.26 µIU/mL, AntiTPO:14.7, PTH:0.01 pg/mL. Hypercalcemia was associated to adrenal insufficiency. IV hydration and steroid of stress dose was started because of hypercalcemia and hypotension. We applied maintenance doses of hydrocortisone (30 mg a day) and fludrocortisone (0.1 mg x 1/2 a day) to the improved patient. Calcitriol and calcium were restarted to the patient as her Ca level was 6.5 mg/dl in the follow-up period. The patient had a leukoplakic lesion characterized with candida plaque on the right buccal mucosa. Thus, sodium bicarbonate and mycostatin suspensions were added to the treatment.

Conclusions:

The most important components of OPS Type 1 are hypoparathyroidism, Addison's disease and candidiasis with other disease associations of hypothyroidism, hypogonadism and infertility, alopecia (baldness), malabsorption, chronic active (autoimmune) hepatitis. It is a genetic disorder inherited in autosomal recessive tendency due to a defect in the AIRE (Auto immune regulator) gene located on chromosome 21.q223.

Picture 1
Dystrophic view of finger nails of our case with APECED.

In our case, hypercalcemia was thought to develop as a result of calcitriol and calcium treatment and addition of adrenal insufficiency. One should always remember that calcitriol and calcium treatment simultaneously with adrenal crisis triggered by stress assist the development of hypercalcemia in the cases with APECED.

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