MESOCORTICOTROPINOMA- ASSOCIATED NELSON' SYNDROME: 28 YEARS FOLLOW-UP

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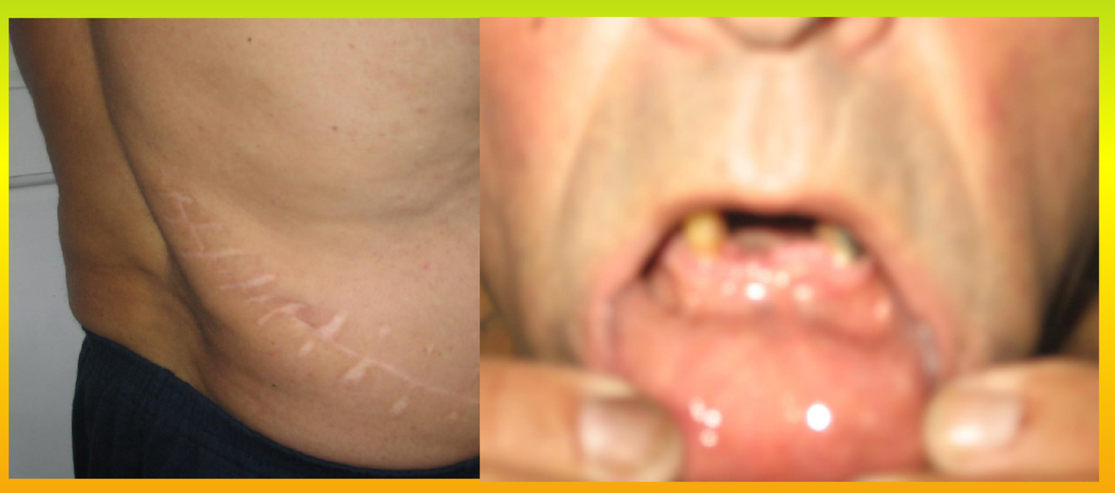
Introduction

Nelson's syndrome (NS) is an exceptionally rare condition diagnosed sometimes after bilateral suprarenalectomy for Cushing's disease (CS) involving rapid enlargement of a pre-existing ACTH-secreting pituitary tumor. The clinical picture varies from hyperpigmentation, headache and visual disturbance to diabetes insipidus and hypopituitarism if the hypothalamic-pituitary portal system is disrupted or normal pituitary tissue is destroyed by the adenoma. The primary treatment for Nelson's syndrome is trans-sphenoid surgery.

Material & Methods

This is a case report revealing the medical history and endocrine profile of a male with NS.



Variable	Result	Normal range
Urinary free cortisol/24h	200.7 µg/24h	50-190 µg/24h
ACTH	24.56 pg/ml	7.2-63.3 pg/ml
TSH	1.24 µUI/ml	0.4-4 µUI/ml
FT4	1.06 ng/dl	0.89-1.76 ng/dl
25-OHD	21.3 ng/ml	30-100 ng/ml
Beta-crosslaps	0.36 ng/ml	≤ 0.584 ng/mL
Osteocalcin	20.9 ng/mL	14-42 ng/mL

Case data

A 65-year old male patient, who had undergone total bilateral suprarenalectomy in 1987, for refractory CS is admitted in 2015 for periodic check-up a NS. In 1988 he received external radiation-therapy for the pituitary ACTH-secreting mesadenoma. In 2003, pituitary MRI performed pointed an 8/10/12 mm-sized mass of oval shape, with intra-sellar expansion. In 2010 a progressive form of NS was diagnosed based on ACTH level of 1250pg/ml (N: 7.2-62.3pg/mL) with increasing levels of 2000pg/ml within 4 years and treated with radiation-theraphy. The eye exam was normal.

On admission, the associated conditions are hypothyroidism after total thyroidectomy for benign nodular goiter, diabetes mellitus and high blood pressure. He has been treated with daily Levothyroxine, Prednison, Fludrocortisone and oral anti-diabetic agents. The ACTH level continues to be high (of 715pg/mL) but decreased compared to previous admissions. Close imagery, endocrine, ophthalmic follow-up is necessary.

Conclusion

The therapeutically resources in mild forms of Nelson's syndrome involves pituitary radiotherapy if surgery is not an option. Despite the current rarity of the condition is has a potential aggressive behavior and close monitoring is required including 28 years after bilateral adrenal remove as seen in this case.