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Case Report

Exophthalmos Revealing Cushing's Syndrome: A Case Report

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Abstract

Exophthalmos is typically associated with Graves' ophthalmopathy. Although originally described by Harvey Cushing, exophthalmos is an underappreciated sign of Cushing's syndrome. We present a case of a patient aged 40 years with no pathological history presenting with generalized asthenia associated with weight loss of 15kg for 2 months, epigastralgia and incoercible vomiting. Clinical examination revealed a normotensive, normocardiac patient with pain on epigastric palpation, bilateral exophthalmos, and an unremarkable cervical examination No cushingoid syndrome especially no facial erythrosis, no purple stretch marks, no buffalo hump, no amyotrophy of the lower limbs. An abdominal CT scan performed before the abdominal pain revealed an adrenal incidentaloma, a right adrenal nodule, well limited, of low density, homogeneous, very discreetly and homogeneously enhanced after injection of contrast medium, whose hemodynamic behavior was in favor of a benign adenoma. The patient underwent surgery for right adrenalectomy, with morphological and immunohistochemical findings in favour of adrenocortical carcinoma, with clinical and biological improvement. We discuss the possible mechanisms causing exophthalmos in patients with either endogenous or exogenous hypercortisolemia.

Keywords: Exophthalmos, cushing, hypercortisolemia, incidentaloma.

INTRODUCTION

Cushing's syndrome is a rare endocrine disorder characterized by prolonged, inappropriate excess of cortisol secondary to endogenous or exogenous causes. Exogenous hypercortisolism is usually caused by prolonged administration of corticosteroids for chronic inflammatory diseases. On the other hand, endogenous hypercortisolism can be adrenocorticotropic (ACTH)-dependent or independent. In 1932 by Harvey Cushing 4 of his 12 patients with Cushing's disease, this is an often forgotten clinical sign [1] in patients with CS. Exophthalmos refers to ocular proptosis secondary to endocrinopathies, and is a clinical sign often observed in patients with hyperthyroidism. Cushing's syndrome should be considered in the differential diagnosis of acquired exophthalmos and elevated intraocular pressure and findings of increased orbital fat on orbital imaging. This case highlights the importance of exophthalmos and its association with hypercortisolaemia.

CASE PRESENTATION

A patient aged 40 years with no pathological history presenting with generalized asthenia associated with weight loss of 15kg for 2 months, epigastralgia and incoercible vomiting. Clinical examination revealed a normotensive, normocardiac patient with pain on epigastric palpation, bilateral exophthalmos, and an unremarkable cervical examination No cushingoid syndrome especially no facial erythrosis, no purple stretch marks, no buffalo hump, no amyotrophy of the lower limbs. An abdominal CT scan performed before the abdominal pain revealed an adrenal incidentaloma, a right adrenal nodule, well limited, of low density, homogeneous, very discreetly and homogeneously enhanced after injection of contrast medium, whose hemodynamic behavior was in favor of a benign adenoma. The patient underwent surgery for right adrenalectomy, with morphological and immunohistochemical findings in favour of adrenocortical carcinoma, with clinical and biological improvement.

Figure 1: Exophthalmos bilateral

Figure 2: Surrenal thin-section CT scan

DISCUSSION

The cause of exophthalmos in cushing syndrom is still unknown. Multiple theories have been proposed including fat redistribution and increased retro-orbital fat, associated thyroid disease, and an exophthalmos causative factor. It has been proposed that retro-orbital fat deposition is also part of the fat redistribution seen in cushing syndrom, resulting in increase in volume of the retro-orbital tissues and a consequent rise in intra-orbital pressure [2, 3]. Orbital fat volume was increased in patients with cushing syndrom and orbital muscles are relatively spared [4, 5]. In contrast to patients with Graves' disease the retrorbital fat in cushing syndrom is devoid of inflammatory cell infiltration. Whether differential fat deposition in the orbits is due to increased glucocorticoid receptor density, defective lipolysis or increased lipoprotein lipase activity is not known. When exophthalmos occurs in a patient who 's not thyrotoxic, other diseases need to be considered these include infiltrative disorders affecting the orbit, cavernous sinus thrombosis .it is possible that the retro-orbital dat increase in volume by similar mechanism [6]. Cushing syndrom exophthalmos which's significantly greater than that of patients with treated cushing syndrom's or normal controls. The exophthalmos rarely of clinical signifiance, and appears to improve with treatment of cushing syndrom.

The authors describe a case of iatrogenic Cushing's syndrome in which an emergency orbital decompression was performed. This procedure was necessary because major ocular hypertension and severe bilateral exophthalmos had caused a decrease in visual function and recurrent painful episodes of eyeball luxation [7].

Cushing's disease with elevated endogenous steroid production. Four cases of exophthalmos

associated with prolonged high doses of exogenous steroids are presented. Prior to steroid therapy, these patients exhibited no signs of exophthalmos. They were euthyroid with normal thyroid function studies while on steroid therapy. One case demonstrated a reduction of her exophthalmos after her steroid dosage was markedly decreased. The finding of exophthalmos presents a previously unreported complication of steroid therapy [8].

CONCLUSION

Cushing's syndrome must be considered in the differential diagnosis of acquired exophthalmos and elevated intraocular pressure, as well as orbital imaging findings showing increased orbital fat. Patients with cushing 's syndrom have statistically significat exphotlmos. This rarely causes symptoms and diminishes when cortisol concentrations become normal.

Compliance with Ethical Standards

Acknowledgments

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Disclosure of Conflict of Interest: The authors declare no conflict of interests.

Statement of Ethical Approval: The present research work does not contain any studies performed on animals/humans subject by any of the authors.

Statement of Informed Consent: Informed consent was obtained from all individual participants included in the study.

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