Cushing's syndrome case series

A mono-centric and longitudinal study was conducted on 19 consecutive patients who underwent synchronous bilateral adrenalectomy (SBA) for ACTH-dependent hypercortisolism between December 2003 and December 2017. This study population was compared to two control groups composed of patients cured after the resection of Pituitary corticotroph adenomas (Group A: 44 patients) and of the ACTH-secreting neuroendocrine tumors (Group B: 8 patients).

Short- or long-term SBA complications or the recurrence of hypercortisolism did not occur. A single patient experienced Nelson syndrome. Clinical features after SBA showed improvement in the glico-metabolic assessment, hypertension, bone metabolism and the occurrence of hypokalemia and infections. The younger the age at the time of Pituitary corticotroph adenoma diagnosis, the longer the duration of active hypercortisolism, higher values of plasmatic ACTH and Cortisol (1 month after pituitary neurosurgery) and higher values of Ki67 in pituitary neuroendocrine tumors were detected in this study population as compared to Group A.

SBA is an effective and safe treatment for patients with unmanageable ACTH-dependent hypercortisolism. A multidisciplinary team in a referral centre with a high volume of patients is strongly recommended for the management of these patients and the identification of patients, for better surgical timing [1)].

In a retrospective study 118 patients with Cushing's syndrome (CS) [102 Cushing's disease (CD), 10 adrenal CS and 6 ectopic adrenocorticotropic syndrome (EAS)]. Thyroid functions [thyroid-stimulation hormone (TSH), T3, free T3 (FT3), T4, and free T4 (FT4)] were measured in all CS at the time of diagnosis and in all CD 3 months after transsphenoidal pituitary tumor resection. Postoperative hormone monitoring within 3 months was conducted in 9 CD patients completing remission. 28 remitted CD patients experienced hormone and antithyroid antibody evaluation preoperatively and on the 3rd, 6th, and 12th month after surgery.

TSH, T3, and FT3 were below the reference range in 31%, 69% and 44% of the 118 CS patients. Remitted CD patients (81/102) had significantly higher TSH (P=0.000), T3 (P=0.000), and FT3 (P=0.000) than those in the non-remission group (21/102). After remission of CD, TSH, T3, and FT3 showed a significant increase, with a few cases above the reference range. By 12 months, most CD patients' thyroid functions returned to normal. Thyroid hormones (including TSH, T3, and FT3) were negatively associated with serum cortisol levels both before and after surgery. No significant changes of antithyroid autoantibodies were observed.

TSH, T3, and FT3 are suppressed in endogenous hypercortisolemia. After remission of CD, TSH, T3, and FT3 increased significantly, even above the reference range, but returned to normal one year after surgery in most cases. Antithyroid antibodies didn't change significantly after remission of CD [2)].

2017

Feng et al. retrospectively analyzed 91 patients with Cushing's syndrome who had either negative findings on Dynamic contrast enhanced MR imaging of the pituitary gland or non-suppressed high dose dexamethasone suppression tests (HDDST). Thin-slice thoracoabdominal computed tomography

(CT) and octreotide scan were also negative to rule out ectopic adrenocorticotropin hormone (ACTH) syndrome. All patients went through inferior petrosal sinus sampling (IPSS) with desmopressin. Afterwards, transsphenoidal pituitary surgery, light microscope pathology and immunohistological staining for ACTH were performed in all patients.

1. Diagnosis of CD. Among the 91 patients included, 90 were confirmed with CD, of whom 89 had positive IPSS findings, therefore the sensitivity was 98.9%. The one patient who was negative for CD also had negative IPSS findings, therefore the specificity was 100%. 2. Tumour lateralization. Among the 51 patients who were ultimately diagnosed with CD and whose lateralization by IPSS and surgery was either left or right, 37 had IPSS lateralization in concordance with surgery, therefore the concordance rate was 72.5%. Patients in the concordant group had a higher frequency of right lateralization by surgery.

IPSS with desmopressin is a sensitive approach in the diagnosis of CD and has moderate accuracy in tumour lateralization, making it an alternative choice to IPSS with CRH [3].

1996

Clinical symptoms of Cushing's syndrome were recognized in approximately 23 of 97 successive hirsute women attending the Endocrinological Department of the Women's Hospital. Endocrine and radiological examinations ultimately confirmed Cushing's syndrome caused by pituitary microadenomas (Cushing's disease) in only 2 women. In 17 of the remaining 21 patients suspected of having Cushing's disease hypercortisolism could not be diagnosed by classical endocrine tests. Thus further endocrinologial tests as corticotrophin releasing hormone (CRH)-tests (100 micrograms i.v.) and insulin tolerance tests were initiated including serum and stress-free salivary cortisol profiles during the day. Diurnal cortisol profiles showed higher values in the morning than during the night, but intermittent elevated cortisol concentrations could be detected. Finally, seven patients could be classified as suffering from a special form of pituitary ACTH-dependent hypercortisolism, named intermittent Cushing's disease (ICD). It is characterized by severe Cushingoid symptoms like marked central adiposity, purple striae, hirsutism and acne, intermittently increased cortisol concentrations, no cortisol response to hypoglycaemia, but preservation of some diurnal variation of serum or saliva cortisol. Three of these seven patients had diagnostic transsphenoidal neurosurgery and pituitary ACTH-producing microadenomas were removed in two women. Repeated determinations of the circadian rhythm of cortisol in saliva samples in combination with an overnight 1 mg dexamethasone suppression and an insulin stimulation test, are recommended to diagnose patients with suspicion on ICD even in an outpatient clinic [4].

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