

Inferior petrosal sinus venous sampling in Cushing's Syndrome: Potential Diagnostic Pitfall.

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Abstract

Evaluation of cause of hypercortisolism among patients with Cushing's syndrome may be challenge for the clinicians (1). The identification between pituitary and ectopic ACTH Production is one of the most complex diagnoses in endocrine practice. Here we reported a rare case of Cushing disease which was diagnosed by Inferior petrosal sinus sampling (IPSS) method. We report this case to highlight how IPSS helped to resolve the source of ACTH production.

(Key words: Cushing syndrome, Inferior petrosal sinus sampling, Hypercortisolism.)

Introduction

Cushing's syndrome results from sustained pathologic hypercortisolism caused by excessive corticotropin (ACTH) secretion by tumors in the pituitary gland (Cushing's disease, 70%) or elsewhere (15%), or by ACTH-independent cortisol secretion from adrenal tumors (15%). (1) The diagnosis of Cushing's syndrome should be followed by the measurement of plasma ACTH concentration to determine whether the hypercortisolism is ACTH-independent. (2) In ACTH-dependent patients, bilateral inferior petrosal sinus sampling with measurement of ACTH before and after administration of ACTH-releasing hormone. (3) Evaluation of cause of hypercortisolism among patients with Cushing's syndrome may be challenge for the clinicians. (4)

Case report

A 26-year-old man who remained undiagnosed regarding the source of Cushing syndrome for 10 years was referred for further investigations. He had initially presented with resistant hypertension, obesity and osteoporosis. He had facial puffiness, progressive weight gain, poor sexual characteristics and coarse facial features. He also experienced proximal myopathy, depression, impaired memory

and altered sleep. On examination, he had body mass index of 30.5 kg/m², Cushingoid appearance with moon face, buffalo hump, hyperpigmentation, easy bruising, thin skin, proximal muscle weakness and purplish abdominal striae. His initial biochemical investigation showed elevated 9 a.m cortisol of 1293 pg/ml (normal 123-626) and non suppressible low dose dexamethasone suppression test suggesting Cushing's syndrome (Serum 9am cortisol of 79.1pg/ml). His urinary free cortisol was elevated (1264 pg/ml). His serum ACTH level was 55.7 pg/ml. High dose dexamethasone suppression test did not show suppression (serum ACTH of 34.2pg/ml). His MRI pituitary, CT abdomen, pelvis and chest showed absence of any possible source of ACTH. Venous sampling of Inferior-petrosal-sinus (IPS) was performed to identify pituitary source of ACTH. In the unstimulated IPSS there was a central-to-peripheral ACTH gradient, of 2.6 times (higher in centre) noted suggesting pituitary as source of ACTH. This case illustrates the feasibility of venous sampling in localizing the source of ACTH secretion where imaging studies were inconclusive.

Discussion

The identification between pituitary and ectopic ACTH Production is one of the most complex diagnoses in endocrine practice. It is the integrated evaluation of biochemical tests and imaging techniques. But, none of which has a 100% specificity for accurate diagnosis. (5) However, the clinical, biochemical, and imaging test results are indeterminate resulting in uncertainty regarding the source of ACTH production. In such cases, inferior petrosal sinus sampling (IPSS) can help to resolve this uncertainty by accurately locating the source of ACTH production. Therefore, in Cushing's disease, the concentration of ACTH is expected to be higher in the inferior petrosal sinus draining. We report this case to highlight how IPSS helped to resolve the source of ACTH production.



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