Subclinical Cushing Syndrome Associated with an Empty Sella Turcica

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Picture.

The coexistence of adrenocorticotrophic hormone (ACTH)-independent Cushing syndrome (CS) and an empty sella (ES) is rare (1, 2). A 53-year-old woman presented with an ES (Picture A, B) and a 40×25-mm left adrenal tumor (Picture C). She had no Cushingoid features, hypertension or diabetes. The levels of morning plasma ACTH, cortisol, dehydroepiandrosterone sulfate and urinary free cortisol were

1.8 (reference range, 7.2-63.3) pg/mL, 5.7 (4.0-19.3) µg/dL, <50 (180-2,100) ng/mL and 9.6 (11.2-80.3) µg/day, respectively. Overnight 1- and 8-mg dexamethasone suppression tests indicated unsuppressed morning cortisol levels (3.2 and 2.8 µg/dL, respectively). ¹³¹I-adosterol scintigraphy revealed a strong left adrenal uptake (Picture D). These findings suggested subclinical CS (SCS).

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A diagnosis of secondary or tertiary adrenal insufficiency was also considered because repeated examinations showed low levels of morning plasma ACTH (1.5-2.1 µg/dL) and relatively low morning cortisol levels (3.9-6.6 µg/dL). The midnight (23-h) ACTH and cortisol levels were 1.2 pg/mL and 3.4 µg/dL, respectively. The administration of corticotropin-releasing hormone increased the plasma ACTH (4.4-25.5 pg/mL) and cortisol levels (6.9-18.4 µg/dL), indicating that the low plasma ACTH level was caused by hypothalamic dysfunction rather than SCS. The levels of other pituitary hormones were normal. The patient may have had SCS and isolated hypothalamic ACTH deficiency resulting from ES.

The authors state that they have no Conflict of Interest (COI).

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