

Massive adrenal hyperplasia from paraneoplastic adrenocorticotrophic hormone

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DESCRIPTION

A 63-year-old woman with a smoking history of 20 pack-years presented with 3 months of weight loss. A right lung mass and uniformly enlarged adrenal glands were noted on computed tomography (figure 1A). A week later she developed severe hypokalemia (K 2.3 mEq/L, 3.5–5.5), hypertension and hyperglycemia. Endocrine testing demonstrated adrenocorticotrophic hormone (ACTH) dependent hypercortisolemia: serum cortisol did not suppress with high-dose dexamethasone (65.4 µg/dL, <1.8), ACTH level was 56.6 pmol/L (2.2–13.3). Lung biopsy revealed small cell lung cancer (SCLC) and paraneoplastic Cushing's syndrome was diagnosed. She needed aggressive potassium replacement and was treated with chemotherapy plus adrenal enzyme inhibition with metyrapone and fluconazole. Despite this, her condition progressed. The adrenal glands quadrupled in size over 8 weeks, measuring 6 cm in the largest dimension (figure 1B). Paraneoplastic ACTH can cause extreme cortisol elevations and massive adrenal enlargement, and has a poor prognosis.¹ The median survival for patients

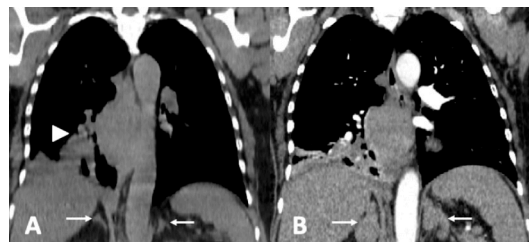


Figure 1 CT imaging demonstrated (A) bilateral enlarged adrenal glands (arrows) and right lung mass (arrow head) at initial presentation and (B) fourfold enlargement of the adrenal glands after 2 months.

Learning points

- ▶ Small cell lung carcinoma (SCLC) is an aggressive neuroendocrine tumour with the capacity to secrete hormones, including adrenocorticotrophic hormone (ACTH), resulting in Cushing's syndrome.
- ▶ SCLC with ACTH secretion carries a poorer prognosis than SCLC without paraneoplastic phenomena.
- ▶ Extreme ACTH secretion causes massive adrenal stimulation leading to adrenal gland hypertrophy and excessive cortisol secretion, resulting in hyperglycemia, hypertension and hypokalemia.

with SCLC is 6.6 months with ACTH secretion compared with 13.1 months without ACTH secretion.¹ The patient died 4 months after diagnosis.

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