

Title: Aggressive Ectopic ACTH production causing Cushing's
Category: Neuroendocrinology and pituitary

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Ectopic Cushing's syndrome is rare, accounting for 5-10% of all cases of Cushing's. The majority are caused by small cell lung cancer and neuroendocrine tumours.

We report a case of a 59 year old male who presented with osmotic symptoms after newly diagnosed diabetes, hypokalaemia and metabolic alkalosis with clinical features suggestive of Cushing's. Initial random cortisol was > 2000nmol/l and 24hour urine cortisol 14500nmol (99-378nmol). Both low and high dose dexamethasone suppression tests failed to suppress, 2069nmol/L and 2550nmol/l respectively. ACTH was 580ng/L. Pituitary MRI was normal. Adrenal CT showed bilateral adrenal hyperplasia suggestive of ectopic ACTH production. Liver metastases were present. A staging CT revealed a left hilar mass (T2bN3M1b). A liver biopsy confirmed poorly differentiated neuroendocrine tumour. Initial management included insulin and metyrapone. The cortisol levels decreased to 600-800 nmol/l. Diabetes control remained poor. Palliative chemotherapy with Carboplatin and Etoposide resulted in a marginal decrease in the size of the lung mass and liver metastases. Poorly controlled diabetes, recurrent infections, progressive oedema and proximal myopathy increased comorbidity. Follow up scan 2 months later showed increasing size of lung primary with additional metastases in the adrenals and bones with multiple pulmonary emboli. Second line chemotherapy provided some symptom relief however he deteriorated rapidly and died.

Ectopic ACTH production is associated with poor response to chemotherapy, short survival, and a high rate of complications to therapy. Studies have shown variations in clinical course can be ascribed to aggressive transformation resulting in liver and adrenal metastases.

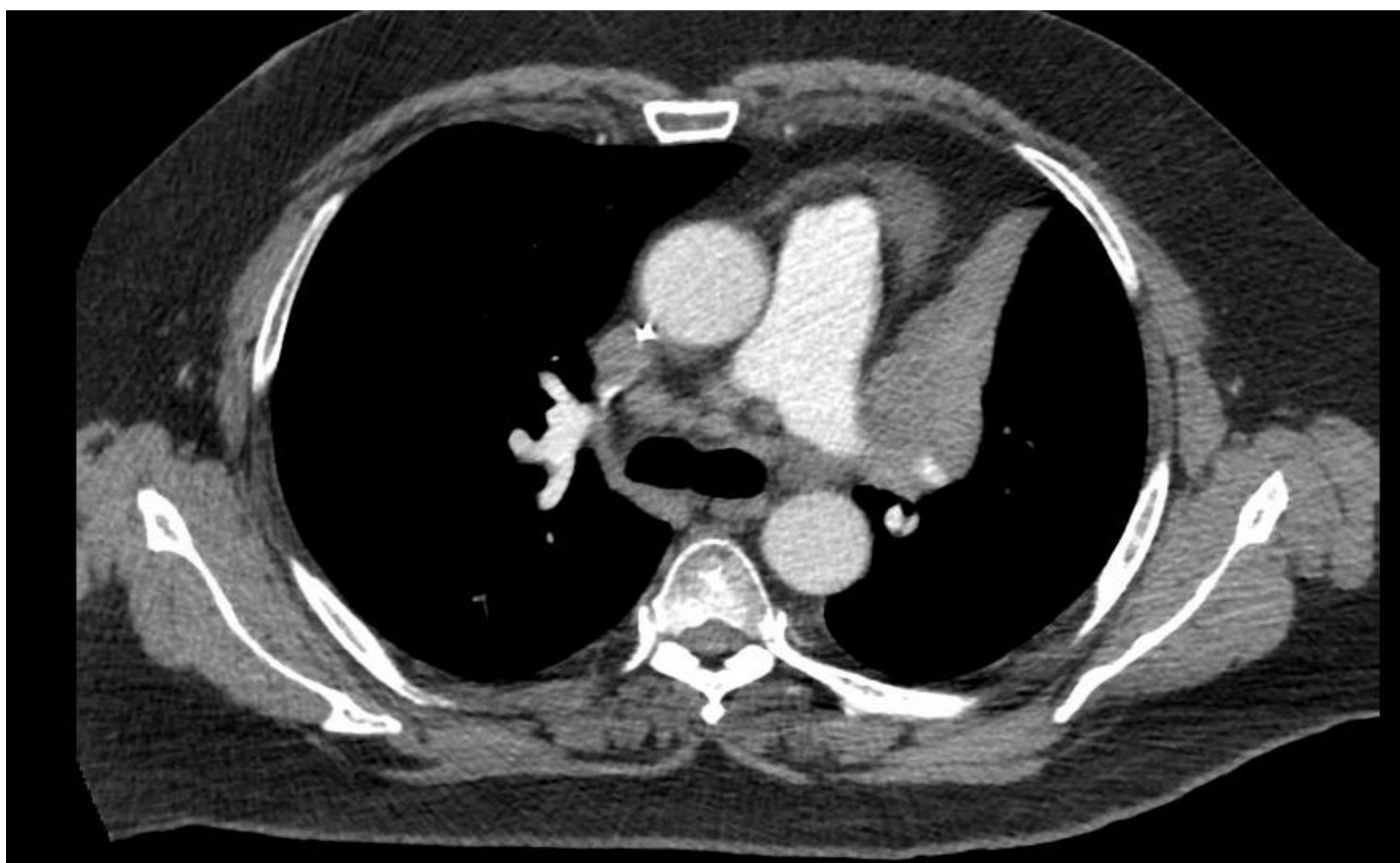


Figure 1. CT Chest showing left hilar mass with left lower lobe collapse.



Figure 2. CT abdomen showing liver metastases.

References:

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