

INTRODUCTION

Ectopic Cushing's syndrome (CS) accounts for 10-20% of cases of adrenocorticotropic hormone (ACTH) dependent CS. The diagnosis of ectopic CS and localization of tumors can be challenging. We present a rare case of ectopic CS arising from thymic carcinoma.

CASE PRESENTATION

A 45 year old African American male with one year history of severe peptic ulcer disease requiring a Billroth II procedure and partial gastrectomy presented to University with severe hypokalemia. He had history of hypertension, uncontrolled diabetes, weight loss, and worsening bilateral lower extremity weakness. Initial workup revealed an early morning cortisol of 55.1 (6.0 - 18.4 ug/dL). Given his symptoms and hypercortisolemia additional testing revealed ACTH 193(7.2-63.3 pg/mL) and 24 hour urine free cortisol of 9222(5 -64 ug/24 hr). Low (1mg) and high dose (8mg) dexamethasone testing failed to suppress cortisol levels appropriately: 65 and 59 (6.0 - 18.4 ug/dL) respectively. Serum potassium was 1.9 mEq/L and HCO3 was 48. Computerized tomography (CT) of the chest revealed a heterogeneous soft tissue density in the anterior mediastinum measuring 2.2 x 4.7 cm with calcification. An octreotide scan was done that showed mild increased uptake in the location of the thymic mass consistent with low grade neuroendocrine tumor. Fluorodeoxyglucose-positron emission tomography (FDG PET) whole body scan revealed heterogeneous FDG uptake in the anterior superior mediastinal soft tissue mass. Patient was started on Mifepristone and planned for thymectomy, however complete resection was not achieved due to involvement of the brachiocephalic vein. Immunohistochemical staining of the surgical biopsy material was positive for chromogranin, synaptophysin, and ACTH with a Ki-67 proliferative index of 40-50%. Patient was continued on Mifepristone and scheduled for bilateral adrenalectomy to manage the hypercortisolemia.

CONCLUSION

Hypokalemia and metabolic alkalosis are classic findings of ectopic ACTH production that distinguishes it from other causes of CS. The most common site of ACTH producing tumors is in the thorax, however thymic carcinoma has been described in a limited number of cases. Identifying these tumors can be challenging for the clinician. Once identified, surgical resection of the primary tumor, bilateral adrenalectomy or medical management are the treatment options.

Medical management of Ectopic Cushing's syndrome

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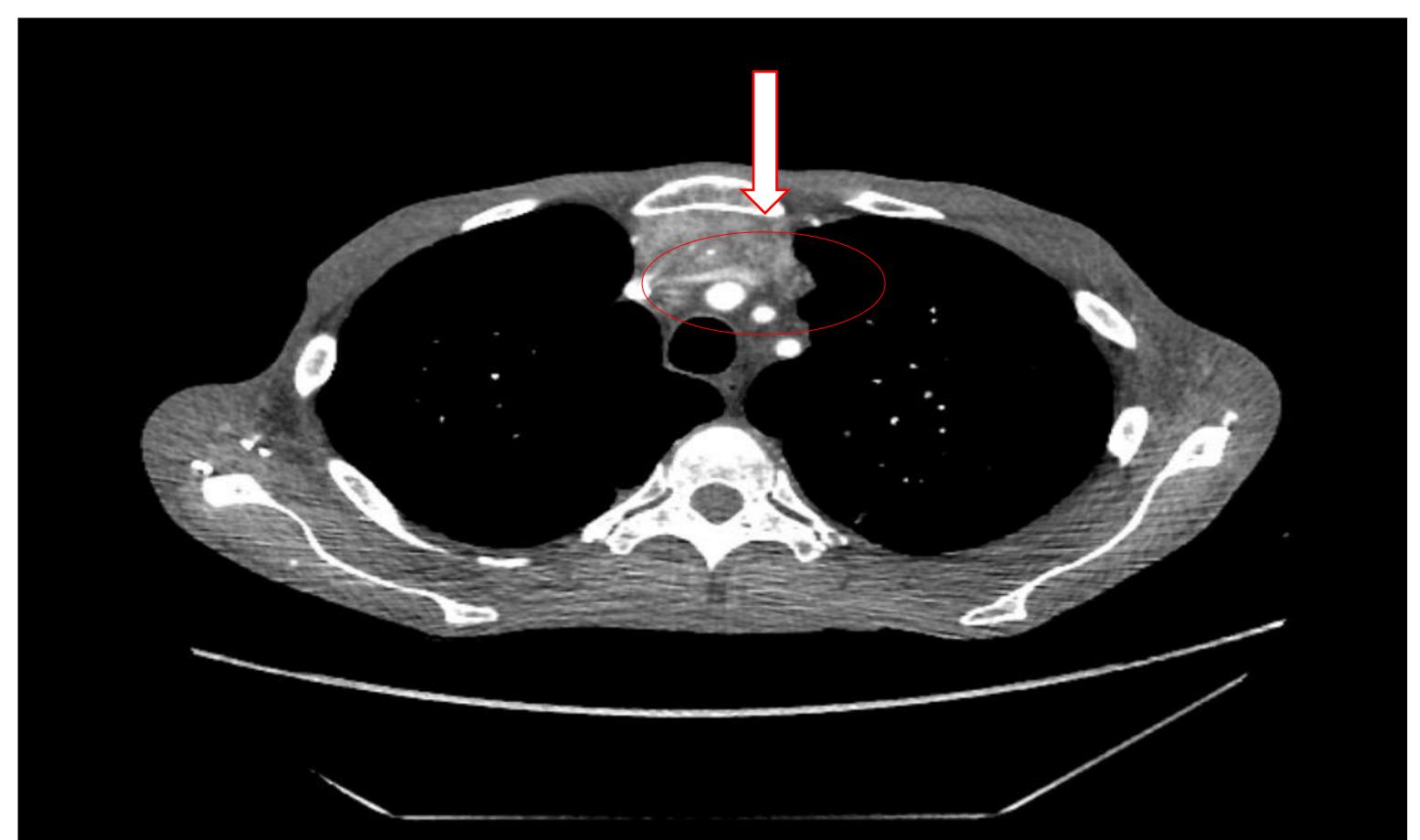


Figure 1: CT chest revealing a heterogeneous soft tissue density measuring 2.2 x 4.7 cm containing calcification in the anterior mediastinum

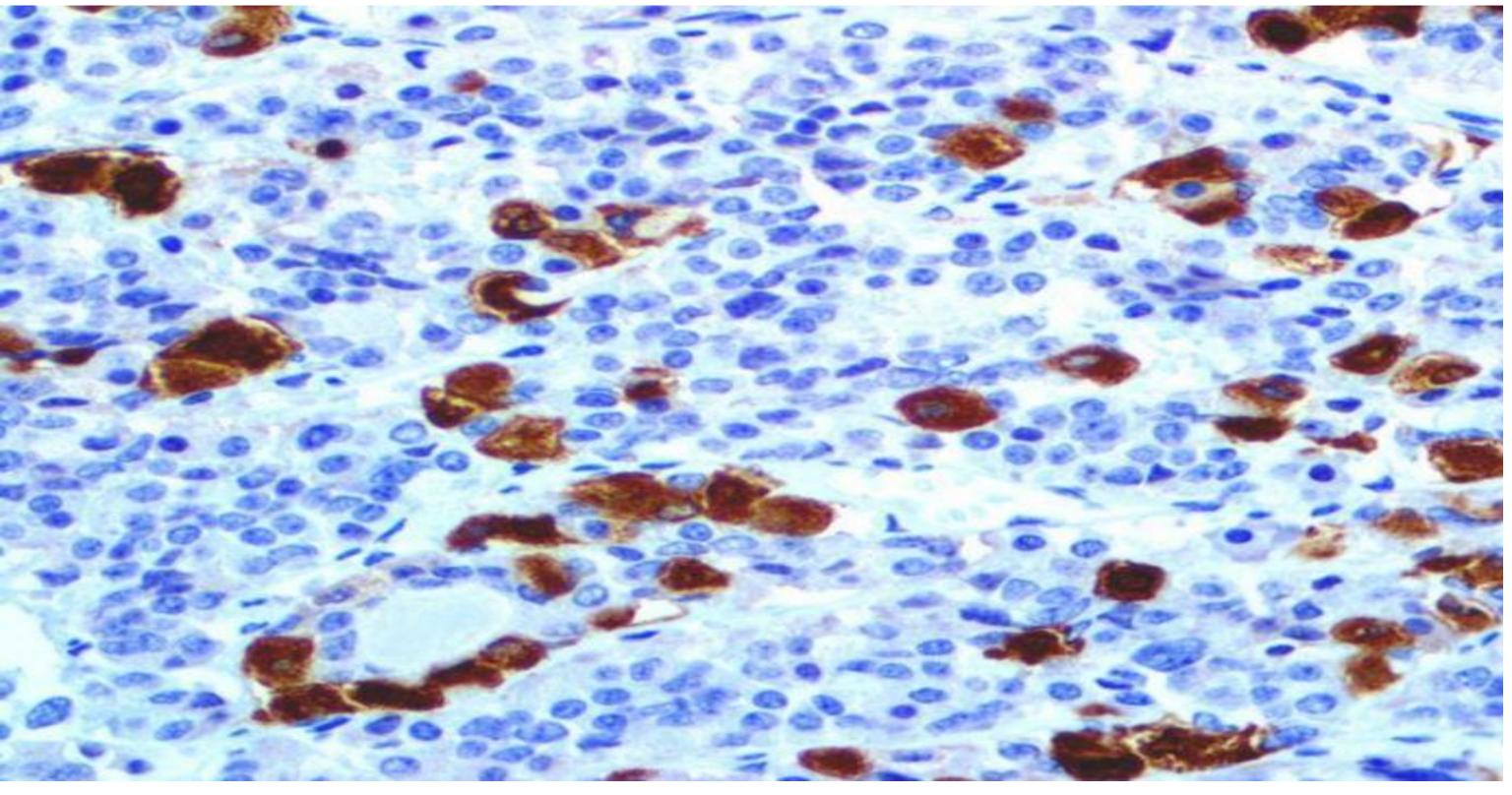


Figure 2: Immunostain shows focal, cytoplasmic ACTH positivity.

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