

Rare case of Ectopic ACTH secreting tumour causing cyclical Cushing's syndrome

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Introduction

Cyclical Cushing's syndrome is a rare disorder characterised by the episodic excess cortisol secretion and normal cortisol secretion in between. It is associated with fluctuating symptoms and signs. Pituitary Corticotroph adenoma is the most common cause of Cyclical Cushing's syndrome accounting more than 50% of patients. Around 26% caused by Ectopic ACTH secreting adenomas and 11% from the adrenal tumours and remainders unknown. (1)

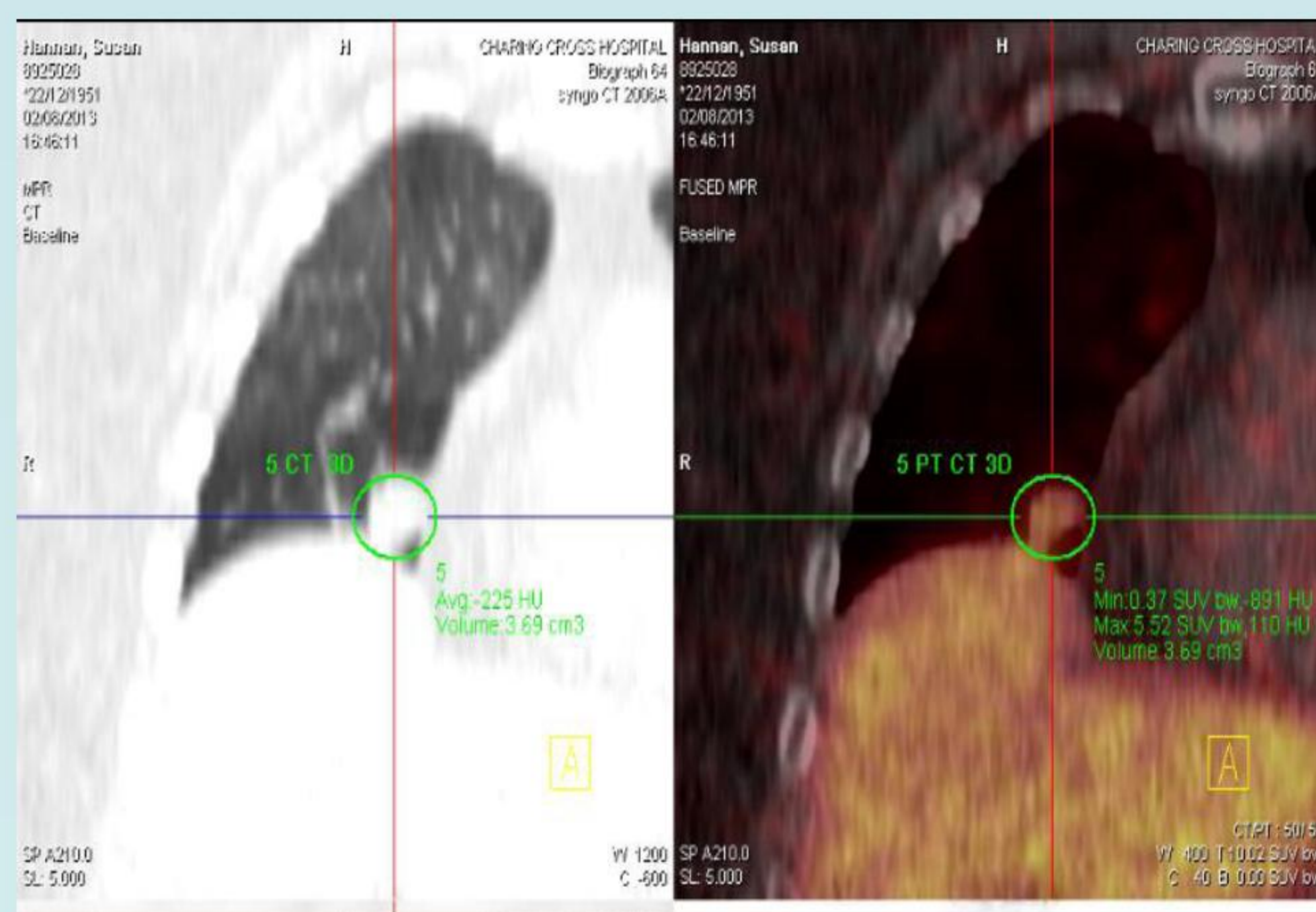
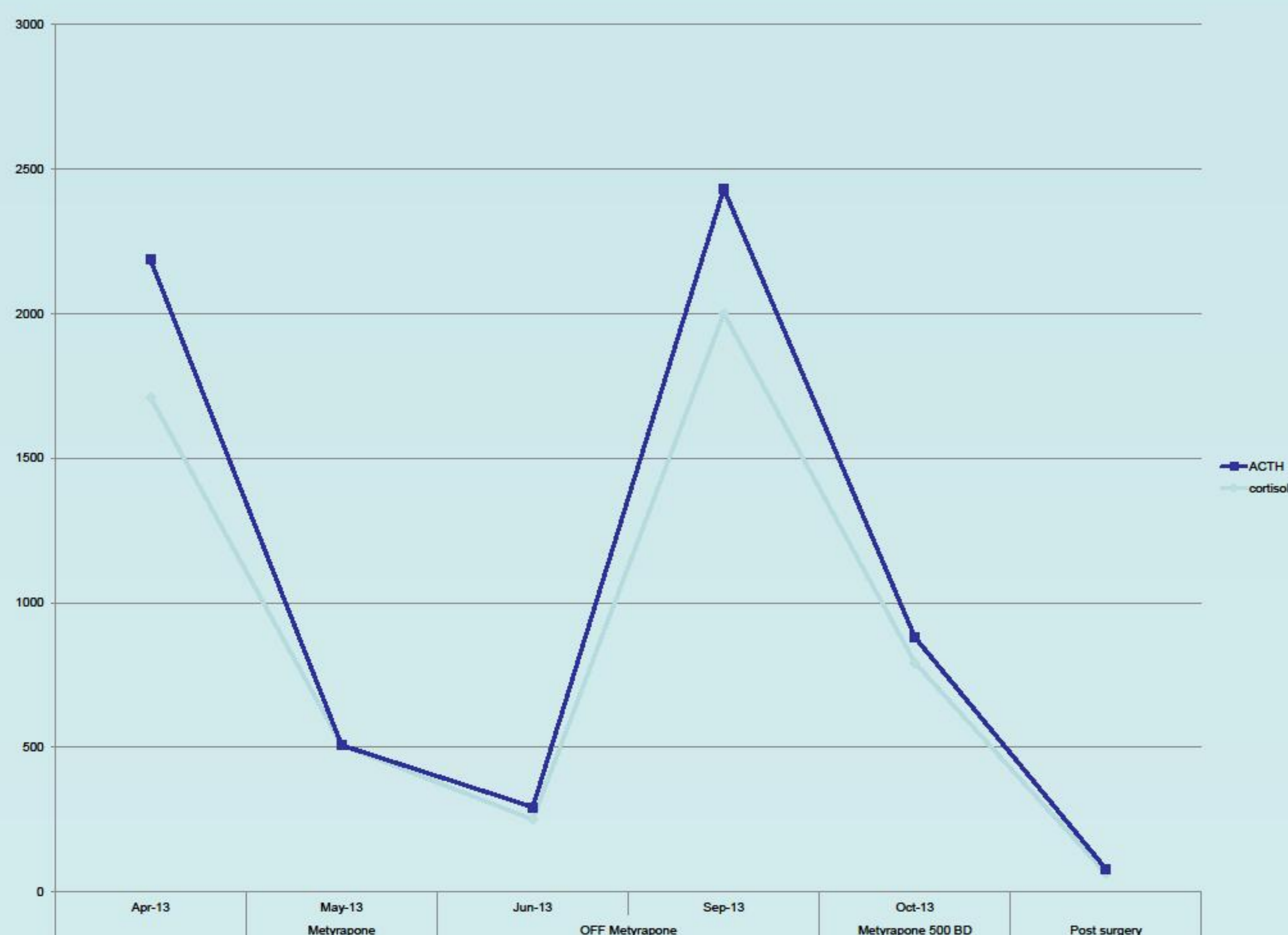
Case report

63 year old lady presented with severe myopathy, easy bruisability and weight gain. She had elevated ACTH of 610 mU/L and midnight cortisol of 1710 nmol/L. Diagnosis of ACTH dependent Cushing's syndrome was made. Her MRI pituitary was normal. Inferior petrosal venous sinus sampling test were not confirmative but suggested possible pituitary source. Her symptoms improved after Metyrapone.

Her cortisol levels started dropping 2 months later and she was weaned off Metyrapone and discharged home once she was symptom free.

She re-presented 4 months later with similar symptoms midnight cortisol of 1400 nmol/L and ACTH 413 mU/L. She underwent Gallium DOTATATE scan showed rt lower lobe gallium avid lesion. Repeat IPSS did not show confirmed ectopic source. She underwent surgical excision 8 months later with histology confirms neuroendocrine tumour Ki67 index less than 3% and she needed hydrocortisone post operatively

Results:



Conclusions:

Cyclical Cushing's syndrome due Ectopic ACTH is extremely difficult to diagnose due to fluctuating clinical picture and biochemistry. In suspected cases specific biochemical and imaging investigation for neuroendocrine tumour is required. Gallium DOTATE scan is more sensitive in localising these tumours

References:

Cyclical Cushing's syndrome ;a clinical challenge Meinardi..et Eur J endocrinology .2007 157(3) 245