

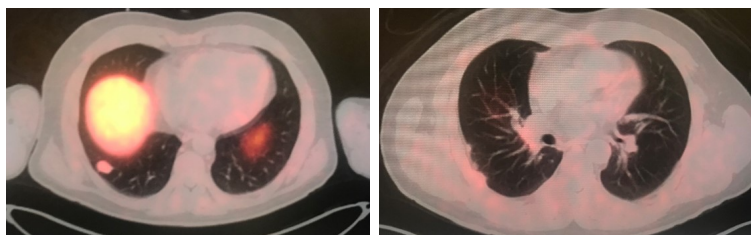
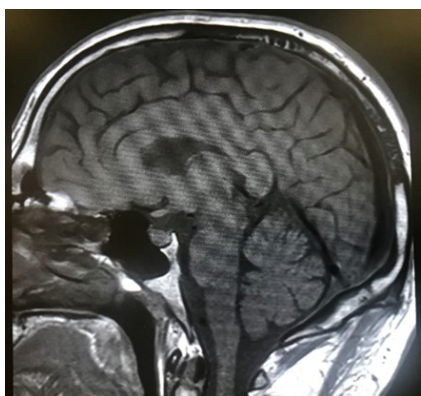


**Introduction**

Ectopic ACTH syndrome (EAS) is a rare condition characterized by tumoral ACTH (adrenocorticotrophic hormone) production and hypercortisolism. Ectopic ACTH secretion has been found to account for approximately 10% of Cushing syndrome (1, 2). Of the causes of ectopic ACTH, pulmonary carcinoids comprise approximately 21% (2).

Pulmonary carcinoid tumours are a rare cause of ectopic ACTH syndrome with the incidence of Cushing syndrome in pulmonary carcinoid tumours being approximately 1% (3). The localization of the source of an ectopic ACTH can be a diagnostic challenge, however is crucial as surgical removal of the offending lesion can be curative for hypercortisolism.

Conventional imaging such as computed tomography (CT) and/or magnetic resonance imaging (MRI) is often used as the initial imaging modality (4, 5), although its sensitivity for the detection of the source of an ectopic ACTH is suboptimal, ranging from 52% to 66%. Gallium-68 DOTATATE PET/CT ([68Ga] -DOTA-(Tyr3) -octreotate) is an FDA-approved high resolution diagnostic tool for imaging neuroendocrine tumours (including carcinoids) (5). Wannachalee et al demonstrated the high sensitivity of [68Ga]-DOTATATE in the localization of ectopic ACTH secreting tumours, for both occult primary tumours and metastatic lesions (5).



Pre-surgery and post-surgery images of Ga-68 DOTATATE Avid Ectopic Adrenocorticotrophic Hormone Secreting Pulmonary Carcinoid tumour

**Case Description**

Here we present a 39 year male who presented with biochemical abnormalities and new onset diabetes and hypertension; a diagnostic work up for Cushing syndrome was undertaken; and it was found that 24-hour urine free cortisol, midnight cortisol and ATCH levels were elevated. The MRI showed no pituitary lesion and CT was unremarkable. This alluded to an ACTH –dependant aetiology and the patient was referred for a [68Ga] -DOTATATE PET/CT scan to assist in the detection and/or localization of a possible ACTH-dependent primary lesion. The scan showed pathological uptake in a parenchymal nodule in the right lower lobe, thought likely to represent the ACTH-secreting tumour. This was confirmed on histopathology post surgical intervention. Patient had resolution of symptoms and was placed on surveillance. A repeat [68Ga] -DOTATATE PET/CT scan was done 4 months later to assess for residual tumour which revealed residual post-surgical inflammatory changes in the region of the right lobectomy

Potassium (mmol/L)	ACTH (mmol/L)	Cortisol (mmol/L)	Urine Cortisol 24hr (mmol/L)	Chromogranin A (mmol/L)
2,7	55,1	1747		
2,5	57,7	1791	3610	90,2
2,5	44,2	1822	4719	
2,6	69,7	1934	524	
4,5	84,6	2589	422	
2,5	79,5	2494		
2.2	71.5	2650		
2.6	63.9	2628		
3.1	62.8	2558		
3.7	58.8	1594		
2.6	30	1712		
3.3	30	1484		
2.7	53.5	1274		
2.5	55.5	1145		
3.6	67.9	1087		
4.2	26	865		
4.0	8.1	572		
4.5	2.0	287		
4.6	6.9	3989		
4.6	2.1	874		
3.7	2.1	1428		
3.9	4.4	658		
4.3	2.9	907		

**Conclusion**

Ectopic ACTH secretion from pulmonary carcinoid tumours (typical or atypical) can present as a challenge in diagnosis and management; however [68Ga]-DOTATATE can be useful as an initial diagnostic modality when this rare condition which has a high mortality and morbidity.

**References:**

Kenchaiah M, Hyer S. Cushing’s Syndrome due to ectopic ACTH from bronchial carcinoid: a case report and review. *Case Rep Endocrinol.* 2012; 2012:215038.

Bukamur H, White C, Khthir R, Browne A, Najar F, Al-Ourani M. Cushing Syndrome by Pulmonary Tumor, *AACE Clinical Case Rep.* 2018;4(No. 6).

Han S, Kim B, Jang H, Kim W, Jeon Y, Kim S, Kim I. Ectopic ACTH syndrome caused by pulmonary carcinoid tumor mimicking long-standing sclerosing hemangioma. *Korean J Intern Med* 2016; 31:794-797.

Varlamov E, Hinojosa Amaya J, Stack M, Fliseriu M. Diagnostic utility of Gallium-68 somatostatin receptor PET/CT in ectopic ACTH-secreting tumors: a systematic literature review and single-center clinical experience. *Pituitary* <https://doi.org/10.1007/s11102-019-00972-w>.

Wannachalee T, Turcu A, Bancosli, Habra M, Avram A, Chuang H, Waguespack S, Auchus R. The Clinical Impact of [68Ga] -DOTATATE PET/CT for the Diagnosis and Management of Ectopic Adrenocorticotropic Hormone – Secreting Tumours. *Clinical Endocrinology.* 2019; 91:288–294.