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# A rare case of an ACTH/CRH co-secreting midgut neuroendocrine tumor mimicking Cushing's disease

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Ectopic ACTH/CRH co-secreting tumors are a very rare cause of Cushing's syndrome and only a few cases have been reported in the literature. Differentiating between Cushing's disease and ectopic Cushing's syndrome may be particularly difficult if predominant ectopic CRH secretion leads to pituitary corticotroph hyperplasia that may mimic Cushing's disease during dynamic testing with both dexamethasone and CRH as well as bilateral inferior petrosal sinus sampling (BIPSS). We present the case of a 24-year-old man diagnosed with ACTH-dependent Cushing's syndrome caused by an ACTH/CRH co-secreting midgut NET. Both high-dose dexamethasone testing and BIPSS suggested Cushing's disease. However, the clinical presentation with a rather rapid onset of cushingoid features, hyperpigmentation and hypokalemia led to the consideration of ectopic ACTH/CRH-secretion and prompted a further workup. Computed tomography (CT) of the abdomen revealed a cecal mass which was identified as a predominantly CRH-secreting neuroendocrine tumor. To the best of our knowledge, this is the first reported case of an ACTH/CRH co-secreting tumor of the cecum presenting with biochemical features suggestive of Cushing's disease.

LEARNING POINTS
The discrimination between a Cushing's disease and ectopic Cushing's syndrome is challenging and has many caveats.Ectopic ACTH/CRH co-secreting tumors are very rare.Dynamic tests as well as BIPSS may be compatible with Cushing's disease in ectopic CRH-secretion.High levels of CRH may induce hyperplasia of the corticotroph cells in the pituitary. This could be the cause of a preserved pituitary response to dexamethasone and CRH.Clinical features of ACTH-dependent hypercortisolism with rapid development of Cushing's syndrome, hyperpigmentation, high circulating levels of cortisol with associated hypokalemia, peripheral edema and proximal myopathy should be a warning flag of ectopic Cushing's syndrome and lead to further investigations.

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