Roca et al. described a rare case of functional adrenocorticotropic hormone (ACTH) and GH microadenoma and report our findings from a systematic literature review of PHA.

They searched PubMed using the terms “plurihormonal pituitary neuroendocrine tumor,” “ACTH GH pituitary neuroendocrine tumor,” and “acromegaly AND Cushing's disease”. In the 17 articles that were selected for literature review, only 20% (4/20) of patients presented with clinical signs of both diseases. Histologically, 19 were pituitary neuroendocrine tumors composed of two distinct cell populations, while only in 1 case was there evidence of a single cell producing both ACTH and GH. In the case reported here, a 60-year-old woman was incidentally diagnosed with a pituitary microadenoma. Endocrine assessment documented increased levels of insulin-like growth factor 1 and GH; ACTH and cortisol values were within normal ranges. Echocardiography documented ventricular hypertrophy. Because of clinical and biochemical evidence of acromegaly, surgery was recommended. Postoperatively, hormonal replacement therapy was started because of adrenal insufficiency. Her antihypertensive therapy was discontinued due to evidence of normal blood pressure values. Histological examination revealed an ACTH-GH PHA with 2 distinct populations of secreting cells. At 3-year follow-up, the patient showed stable clinical remission and was no longer receiving hormonal replacement therapy.

This is an additional case to the 20 previously reported cases of ACTH-GH PHA. Awareness of this relatively rare entity is clinically relevant. The cytogenesis of ACTH-GH PHA remains a matter of debate, and several hypotheses have been postulated [1)].

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Two cases of a plurihormonal pituitary neuroendocrine tumor expressing the rare combination of ACTH and GH. They both underwent successful transphenoidal hypophysectomy (TSH). Long-term post-operative follow-up revealed no evidence of tumour recurrence. Due to the multiple secretions and plurihormonal characteristics clinical diagnosis of composite pituitary neuroendocrine tumors can be difficult [2)].

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A 40 years old patient presented with sudden weight gain central obesity, hypertension, menstrual disorders, acne, hirsutism suggesting hypercortisolism. Laboratory evaluation confirmed Cushing’s disease (serum cortisol 0 h 234 nmol/l, cortisol after low-dose dexamethasone suppression test 613 nmol/l, ACTH 8 h 56 pg/ml). Furthermore, laboratory data showed elevated IGF1 levels (440 ng/ml, reference range 101–267 ng/ml) with elevated GH after HGPO (3.3 mUI/l). Prolactin, TSH, FSH and LH serum level were in normal range. The sellar MRI demonstrated microadenoma. Transphenoidal removal of the tumor was performed. Immunohistochemistry analyses showed staining for ACTH (40%), GH (80%) and prolactin (50%). Ki67 was less than 0.5%. Laboratory data realized after surgery were normal.

The majority of plurihormonal adenomas produce GH, PRL and TRH because lactotroph, somatotroph and thyreotroph cells have the same progenitor. Association of ACTH and GH secretion with positive immunostaining for GH, PRL and ACTH has not been described previously. These unusual plurihormonal adenomas exhibit aggressive behavior and poor prognosis. Plurihormonality of pituitary neuroendocrine tumors linked with ACTH co-expression seems to predict a higher risk of tumor recurrence. Furthermore, higher morbidity due to mixed ACTH and GH secretion needs a strict follow up [3)]

1)

Roca E, Mattogno PP, Porcelli T, Poliani L, Belotti F, Schreiber A, Maffezzoni F, Fontanella MM, Doglietto

F. Plurihormonal ACTH-GH pituitary neuroendocrine tumor: Case Report and Systematic Literature Review. World Neurosurg. 2018 Jun;114:e158-e164. doi: 10.1016/j.wneu.2018.02.120. Epub 2018 Mar 6. Review. PubMed PMID: 29501516.

2) Rasul FT, Jaunmuktane Z, Khan AA, Phadke R, Powell M. Plurihormonal pituitary adenoma with concomitant adrenocorticotropic hormone (ACTH) and growth hormone (GH) secretion: a report of two cases and review of the literature. Acta Neurochir (Wien). 2014 Jan;156(1):141-6. doi: 10.1007/s00701-013-1890-y. Epub 2013 Oct 1. Review. PubMed PMID: 24081787.

3) https://www.endocrine-abstracts.org/ea/0029/ea0029p1540

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