PITUITARY ADENOMA ASSOCIATED WITH PHEOCHROMOCYTOMA/PARAGANGLIOMA

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INTRODUCTION

Pituitary adenomas (PA) and pheochromocytomas/paragangliomas (PHEO/PGL) can occur in the same patient due to either coincidence or as a result of shared pathogenesis. There is evidence that, at least in some cases, classical PHEO/PGL predisposing genes, may also play a role in pituitary tumorigenesis. A new condition called "**the three P Association**" (3PAs) for the combination of PA with PHEO/PGL has been recently described in patients with or without succinate dehydrogenase (*SDHx*) germline mutations.

AIM

To present this new association of multiple endocrine neoplasia, reporting our experience in 3 patients treated in tertiary hospitals.

CASES REPORT

CASE 1

A 54 year old male patient with bilateral pheochromocytoma (figure 1-A) underwent bilateral adrenalectomy.

Three years later he was diagnosed with growth hormone-secreting pituitary microadenoma (figure 1-B) that was completely resected after transsphenoidal surgery.

Genetic screening for PHEO/PGL genes (*MEN-1*, *RET, VHL, SDHB* and *SDHD*) were negative (including sequencing and gross delection analysis).

HORMONAL PROFILE (CASE 1)	VALUE	REFERENCE VALUE
Adrenaline + Norepinephrine (24h urine)	3488 nmol/d	116 - 699
4-hydroxy-3-mandelate (24h urine)	198 µmol/d	15 - 38
IGF-1 (serum)	46,4 nmol/L	8 - 32
GH (serum) - Oral glucose tolerance test	Baseline: 2,7 µg/L 30 min: 2,3 µg/L 60 min: 2,4 µg/L 90 min: 2,5 µg/L 120 min: 2,4 µg/L	< 1

CASE 2

A 38 year old female patient was initially seen for macroprolactinoma (figure 2-A) and chronically treated with dopamine agonist.

Four years later the patient was diagnosed with cervical and unresectable mediastinal PGL (figure 2-B), currently under somatostatin analogue therapy.

Her brother was operated for PGL and gene study revealed a *SDHB* exon 1 deletion (genetic disorder associated with familial PGL type 4). This genetic rearrangement was also detected in her mother and sister.

HORMONAL PROFILE (CASE 2)	VALUE	REFERENCE VALUE
Prolactin (serum)	NA	NA
Adrenaline (24h urine)	3,12 µg/d	4 - 25
Norepinephrine (24h urine)	13,8 µg/d	25 - 125
Dopamine (24h urine)	186 µg/d	190 - 490

NA: No available

CASE 3

A 55 year old female patient was diagnosed with a right pheochromocytoma. She underwent right adrenalectomy.

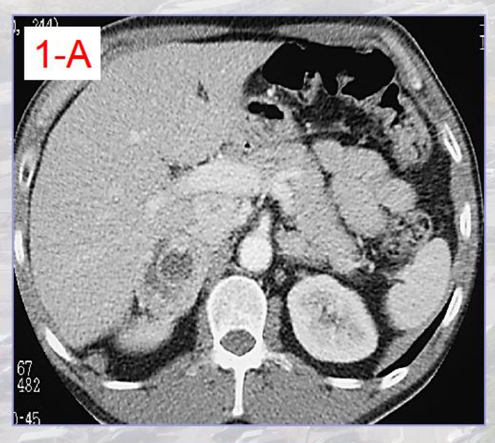
Five years later she was diagnosed with GH-secreting pituitary microadenoma and treated with transsphenoidal surgery.

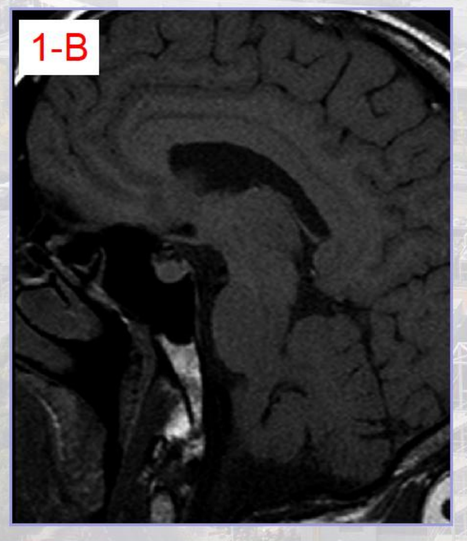
She was also diagnosed with primary hyperparathyroidism without surgical criteria.

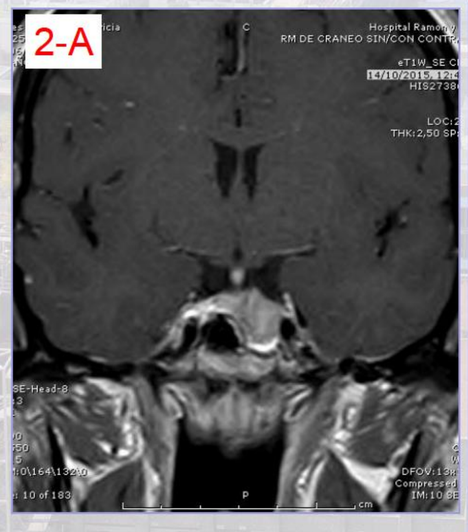
Genetic study for *MEN-1, RET* and *VHL* was negative.

HORMONAL PROFILE (CASE 3)	VALUE	REFERENCE VALUE
Adrenaline (plasma)	162 pg/mL	< 60
Norepinephrine (plasma)	31656 pg/mL	< 300
Dopamine (plasma)	37 pg/mL	< 150
IGF-1 (serum)	839 ng/L	94 - 483
GH (serum) - Oral glucose tolerance test	Baseline: 17,9 µg/L 30 min: 6,3 µg/L 60 min: 4,1 µg/L 90 min: 5,1 µg/L 120 min: 7,2 µg/L	< 1
Calcium (serum)	11,4 mg/dL	8,5 – 10,4
PTH (serum)	87 ng/mL	< 65

IMAGES







FIGURES 1-A Abdominal CT showing bilateral adrenal mass. **1-B** MRI showing a sellar mass. **2-A** MRI displaying a left pituitary mass. **2-B** Octreoscan showing a pathological tracer accumulation in right cervical region.

CONCLUSIONS

The association of PA and PHEO/PGL is an exceptional event, but recent insights provide strong evidence that PA can develop in patients with PHEO/PGL or germline *SDHx* subunit mutations. Genetic testing should be considered in all patients or families with 3PAs.

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