MASSIVE BILATERAL PHEOCROMOCITOMAS: A RARE CASE

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INTRODUCTION: Pheochromocytoma is a rare catecholamine-secreting tumor that arises from the chromaffin tissue of the adrenal medulla. Of the reported cases, only 10% consist in bilateral lesions and the probability of multiple endocrine neoplasia should always be investigated.

CLINICAL REPORT:

Female, 19 years old

History of present illness:

Recurrent episodes, 2 years of evolution:
- Palpitations, headache, nausea, abdominal discomfort

Previous and current medical history: irrelevant

Familiar medical History: Father (56y/o) – Controled hypertension; Mother (45y/o) healty; Sister (7y/o) - healthy

COMPLEMENTARY DIAGNOSTIC EXAMS (AMBULATORY) December/2014

▪ **ABDOMINAL ECOGRAPHY:** "Bilateral peri-renal cystic lesions".

▪ **ABDOMINAL CT:** "Two cystic multiseptated formations, in the dependence of the adrenal glands (AG), right with 11.3cm and left with 7.8cm larger diameter, without infiltrative aspects ".

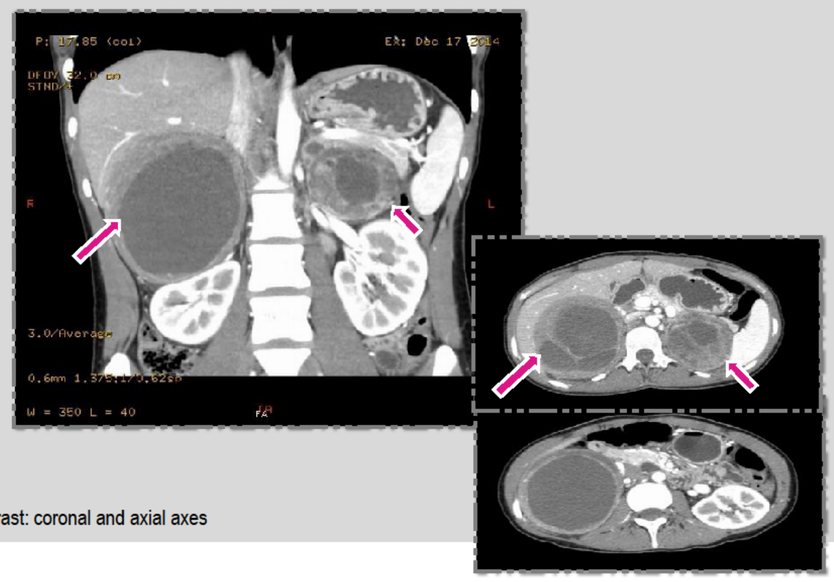


Fig. 1 – Abdominal CT, with contrast: coronal and axial axes

HOSPITAL ADMISSION – ENDOCRINOLOGY DEPARTMENT January/2015

Hormonal Assay

ANALYTE	Result	RV
ACTH pg/mL (8/9h - 23/24h)	14 5.2	-
Cortisol µg/dL (8/9h - 23/24h)	14 2.4	-
Free urinary cortisol µg/24h	31	*10-80*
Cortisol (Low-dose dexamethasone supression test) µg/dL	1.4	-
TSH µUI/mL	**4.8**	*0.4-4.0*
T4L ng/dL	0.9	*0.8-1.9*
PTH pg/mL	41	*9-72*
25-OHD ng/mL	36	*>29*
Calcitonin pg/mL	**55**	*<10*
Plasma free metanephrines pg/mL	**7386.4**	*<60*
Urinary metanephrines µg/24h	**25179.75**	*25-312*
Vanilmandelic Acid mg/24h	**57.35**	*<7.0*
DHEA-SO4 µg/mL	0.7	*0.35-4.3*
Androstenedione ng/mL	2.5	*0.5-3.4*
Compound S ng/mL	3.8	*<8*
Ative Renin µUI/mL	25	*7-76*
Aldosterone pg/mL	62.9	*40-310*

Biochemical Assay

Analyte	Result	RV
Calcium mg/dL	9.5	*8.8-10.6*
Phosphate mg/dL	4.6	*2.5-4.5*
Magnsesium mg/dL	2.1	*1.9-2.5*

▪ **THYROID ECOGRAPHY AND FINE NEEDLE ASPIRATION BYOPSY–** Hypoechoic nodule with 0.8x0.7x1.1cm, in the middle of the LL, with undefined limits and one microcalcification - made cytology. Normal cervical lymph nodes.

Cell-block and immunohistochemistry study: Medular thyroid neoplasia

▪ **CERVICAL AND THORACIC CT:** "Hipodense nodule at the level of the the thyroid LL, 1.1 cm diameter, without evidence of other cervical expansive formations. No changes in the lung parenchyma; without hilar or mediastinal lymphadenopathy."

▪ **ABDOMINAL MRI:** "Bulky masses in both AG, right with 9.4x10.8x10.7cm and left with 6.5x7.8x6.6cm, well defined, with multichystic areas. The right shows extensive internal bleeding. Both kidneys diverted inferiorly and adjacent organs without signs of intrusion."

▪ **MIBG I123 SCINTIGRAPHY:** "Bulky areas of lush and heterogeneous uptake of I123 MIBG in the dependence of the adrenal glands, with smaller areas of central funding in relation to internal bleeding areas (...) these aspects, in this clinical context, are compatible with the hypothesis of bilateral pheochromocytomas. It is also individualized a focus of moderately increased uptake of I123 MIBG in the thyroid LL "

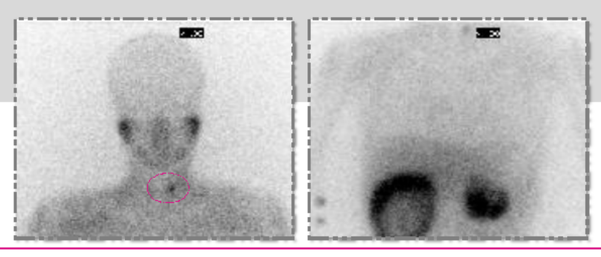


Fig. 2 –MIBG-I123 cervical, thoracic and abdominal scans.

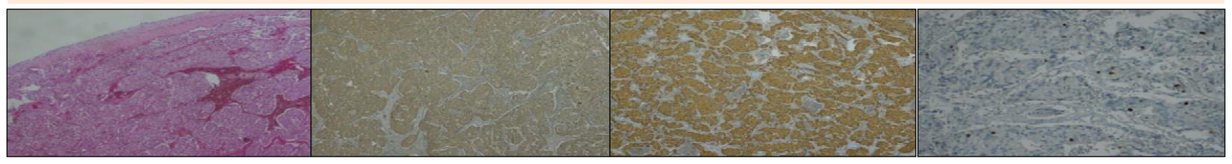
❖ **α-adrenergic blockade (phenoxybenzamine 10mg bid) and β-adrenergic blockade (propranolol 10mg bid): adequate clinical response and tolerability.**

SURGICAL TREATMENT I Urology and Renal Transplant Department February/2015

- Bilateral laparoscopic adrenalectomy
  - Uneventful

Anatomopathological study

BILATERAL PHEOCHROMOCYTOMA WITH BENIGN BEHAVIOUR
Imunohistochemistry: positivity for chromogranin, sinaptofisin and S100; Ki67 3%)



▪ **Post-surgical evaluation (~2Months)**
Patient clinically stable; Medical treatment - Hydrocortisone 15+5+5mg, Fludrocortisone 0.1mg id

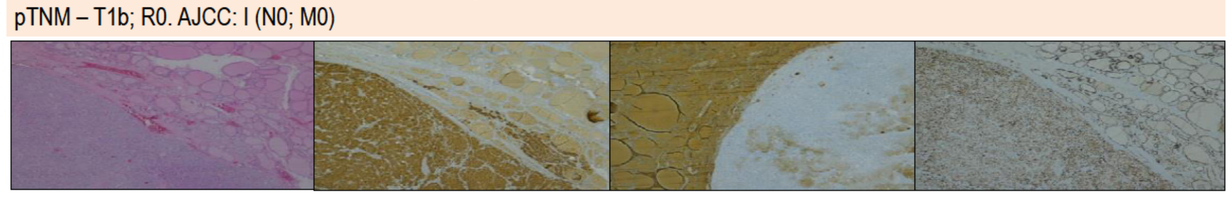
ANALYTE	Result	RV
ACTH pg/mL	12	*9-52*
Cortisol µg/dL	39	*5-25*
TSH µUI/mL	2.4	*0.4-4.0*
T4L ng/dL	1.3	*0.8-1.9*
Calcitonine pg/mL	**60**	*<10*
CEA ng/mL	**8.1**	*<5.4*
Plasma free metanephrines pg/mL	47.2	*<60*
Plasma normetanephrine pg/mL	105.2	*<120*
Urinary metanephrines µg/24h	39.24	*25-312*
Vanilmandelic Acid mg/24h	4.57	*<7.0*
Ative Renin µUI/mL	9.0	*7-76*
Aldosterone pg/mL	26.3	*40-310*

SURGICAL TREATMENT II General Surgery Department April/2015

- Total thyroidectomy with central lymph node dissection (level VI and VII)
  - Uneventful

Anatomopathological study

MEDULLARY CARCINOMA OF THE THYROID; 2 PRETRACHEAL LYMPH NODES WITHOUT NEOPLASIC INFILTRATION
pTNM – T1b; R0. AJCC: I (N0; M0)



▪ **Post-surgical evaluation (~2Months)**
Patient clinically stable; Medical treatment - Hydrocortisone 15+5+5mg, Fludrocortisone 0.1mg id, Levothyroxine sodium 100mcg id

Analyte	Result	RV
ACTH pg/mL	11	*9-52*
Cortisol µg/dL	22	*5-25*
TSH µUI/mL	2.0	*0.4-4.0*
T4L ng/dL	1.5	*0.8-1.9*
Calcitonin pg/mL	<2.0	*<10*
CEA ng/mL	0.8	*<5.4*
Ative Renin µUI/mL	12	*7-76*
Aldosterone pg/mL	24.0	*40-310*

MOLECULAR GENETIC TESTING

▪ **PROBAND:** mutation c.2080 T> C in exon 11 of the RET gene, consistent with the diagnosis of MEN2A (TGC>CGC; p.C634R).

RESULT CONSISTENT WITH THE DIAGNOSIS OF **MEN2A** SYNDROME

▪ **GENETIC SCREENING OF FAMILY MEMBERS (PARENTS AND SISTER):** Negative (not detected mutation in exon 11 of the RET gene).

CONCLUSIONS: This report illustrates an uncommon case of massive bilateral cystic pheocromocitomas in a young patient. In the presence of bilateral adrenal tumors and young age, multiple endocrine neoplasia probability is higher, and should be carried out biochemical, imaging and genetic investigation. If confirmed, genetic evaluation of first degree relatives should be performed. Furthermore, because of the high possibility of recurrence, these patients should maintain close and long-term monitoring.

Bibliography: Lenders JVM et al., *Pheocromocytoma and Paraganglioma: An Endocrine Society Clinical Practice Guideline*. J Clin Endocrinol Metab, June 2014, 99(6): 1915-1942; Brandi ML, *et al. Consensus: Guidelines for diagnosis and therapy of MEN Type 1 and Type 2.* The Journal of Clinical Endocrinology & Metabolism,2001, 86(12):5658–5671; TK Richard *et al. Mmedullary Thyroid Cancer: Management Guidelines of the American Thyroid Association, Thyroid, 2009, Vol.19,Number 6, 565—612*

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Adrenal cortex (to include Cushings)

Diana Martins

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PosterSessionOnline.com