ELECTIVE ADRENALECTOMY FOR PHEOCHROMOCYTOMA MULTISYSTEM CRISIS: CAN EARLY BIOCHEMICAL DIAGNOSIS BE THE KEY?

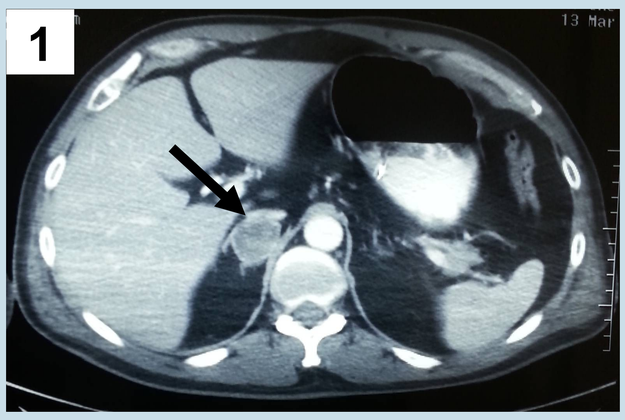
M. Cayón[1], C. García-Figueras[2], A. Gil[3], F. Mateos[4].
Endocrinology and Nutrition Unit [1]. Internal Medicine Unit [2]. Critical Care Medicine Unit [3]. Surgery Unit [4]
Hospital SAS. Jerez de la Frontera, Jerez de la Frontera, Spain.

INTRODUCTION

Pheochromocytoma multisystem crisis (PMC) is the most fulminant clinical expression of pheochromocytoma.
The appropriate timing and judgment for the start of surgery and the adequate preoperative medical treatment are unclear.
We report a case of PMC successfully treated with elective adrenalectomy after doxazosin blockade.
The importance of an early biochemical diagnosis is discussed.

CLINICAL PRESENTATION

A 47-year-old healthy man consulted for sudden onset dyspnea and thoracic discomfort.
In the Emergency Department, his level of consciousness upon arrival was 12 (Glasgow Coma Scale) and he was found to be in acute respiratory distress with severe hypoxemia, requiring intubation in Intensive Care Unit. His blood pressure was 165/110 mmHg and his body temperature, 39,9°C.
Electrocardiography revealed atrial flutter.
The laboratory data showed polycythemia, severe renal and hepatic failure, rhabdomyolysis and hyperglycemia. A blood sample drawn on the patient's arrival at the hospital showed high serum norepinephrine [>758 pg/mL; (normal<370)]. Epinephrine and dopamine concentrations were normal. (table 1).
All infectious screens were negative.
An abdominal CT scan revealed a 3 cm. mass, located in the right adrenal gland and a MIBG scan was strongly positive (figures 1 and 2).
After reaching clinical stability, elective right laparoscopic adrenalectomy was performed under previous α-blockade with doxazosin. Pathology examination revealed a 3 cm pheochromocytoma without evidence of malignant involvement, necrosis or haemorrhage (figures 3 and 4). Six months after, he is asymptomatic and his level of catecholamines is normal.



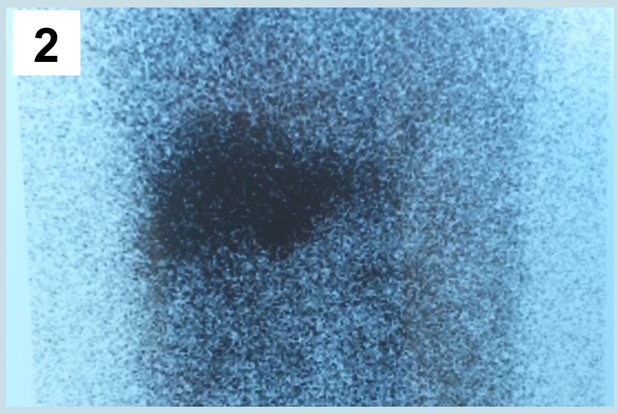
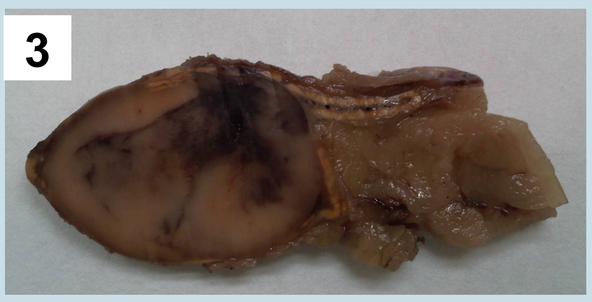


Figure 1.
CT scan image showing a 3 cm mass, located in the right adrenal gland (arrow).
Figure 2.
MIBG scan image of patient's abdomen showing high uptake of the tracer in the right adrenal gland.

Table 1: Clinical and biochemichal parameters

	On admission (ICU)	After doxazosin blockade (2 weeks after)
Blood pressure (mm Hg)	165/110	110/65
Body temperature (ºC)	39,9	36,6
Heart rate (bpm)	150 (atrial flutter)	68 (sinus rhythm)
Leukocytes/mm^3	31900	7000
Haemoglobin (g/dL)	8,5	11,9
Haematocrit (%)	52,8	36,4
Thrombocytes/mm^3	128000	291000
Prothrombin time (seconds)	25	12,5
International Normalized Ratio	3,22	0,95
Thromboplastin partial time (seconds)	49,1	36
Glucose (mg/dL)	585	81
Creatinine (mg/dL)	6,22	1,21
Creatin-kinase (U/L)	3822	34
Myoglobin (ng/mL)	1794	44
AST/ALT/GGT (U/L)	10133/8721/145	22/30/156
Troponin (ng/L)	>10000	0,2
Serum norepinephrine (pg/mL) N<370	>758	<20
Serum epinephrine (pg/mL) N<150	<20	<20
Serum dopamine (pg/mL) N<200	<20	<20



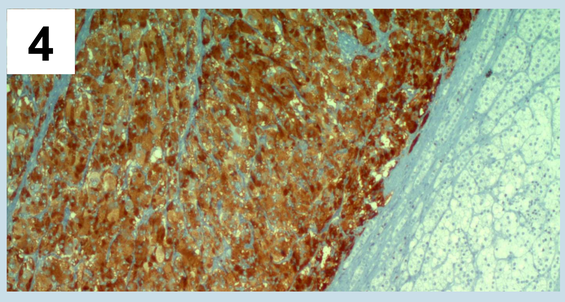


Figure 3.
Right adrenal gland photo after surgical removal showing a 3 cm tumour.
Figure 4.
Pathology examination revealing pheochromocytoma.

CONCLUSIONS

- Most reported PMC cases requiring emergency surgery or died were mixed producers of epinephrine and norepinephrine or epinephrine only.
- The high rate of initial suspicion allowed us an early identification of amine secreted by the tumour and it allowed us to plan a specific therapeutic strategy.
- Further studies with a major number of cases are needed; nevertheless our observation suggests that pure norepinephrine secretor tumours may have a more favourable clinical course and this knowledge could help to identify candidates to elective surgery after doxazosin blockade.

REFERENCES

- Prejbisz A, Lenders JW, Eisenhofer G, Januszewicz A. Cardiovascular manifestations of phaeochromocytoma. J Hypertens. 2011 Nov;29(11):2049-60.
- Davlouros PA, Velissaris D, Tsiola A, Filos KS, Alexopoulos D. Fever with multiple organ failure: not always sepsis. Anaesth Intensive Care. 2010 Nov;38(6):1090-3.
- Uchida N, Ishiguro K, Suda T, Nishimura M. Pheochromocytoma multisystem crisis successfully treated by emergency surgery: report of a case. Surg Today. 2010 Oct;40(10):990-6.
- Moran ME, Rosenberg DJ, Zornow DH. Pheochromocytoma multisystem crisis. Urology. 2006 Apr;67(4):846.