Left adrenal Ectopic Cushing Syndrome even more challenging

B. Biagetti[1], O. Simó-Servat[1], A. Ortiz-Z[1], C.Iglesias[2], E.Caubet[3], J.Castell-Conesa[4], A. García-Burillo[4], R.Ferrer[5], B.Dalama[1], G. Obiols[1], J Mesa[1]

1 Department of Endocrinology 2 Department of Pathology 3 Department of Surgery,4 Departament of Nuclear Medicine 5Biochemistry Laboratory : Hospital Universitari Vall d'Hebron, Universitat Autonoma de Barcelona, Pg. Vall d'Hebron 119-129, Barcelona 08035, Spain

Background: The Ectopic ACTH Syndromes (EAS) often associated with severe hypercortisolism (HC) are really a challenge.

Case presentation: We report the case of a 48-year-old woman with ectopic adrenocorticotropic hormone secretion (EAS) confirmed biochemically and by bilateral inferior petrosal sinus sampling. Not conclusive left-adrenal topography in all imaging studies was found, even somatostatin receptor scintigraphy (SSRS) and 123I-metaiodobenzylguanidine. Due to positive SSRS, medical treatment with high-dose extended-release somatostatin analogs (SSAs) was initiated but patient's condition worsened dramatically one week later. Immediately after cortisol normalization with ketoconazole the patient underwent surgery. Her left adrenal gland and a contiguous lesion, that resulted to be, an associated extra-adrenal ganglioneuroma, were removed but surprisingly only the adrenal medulla had significant staining of adrenocorticotropic hormone (ACTH). After surgery, the patient improved, blood pressure normalized without medication, and her hypercortisolism and clinical symptoms were resolved

Figure 1: Nuclear Medicine Imaging showing left adrenal topography

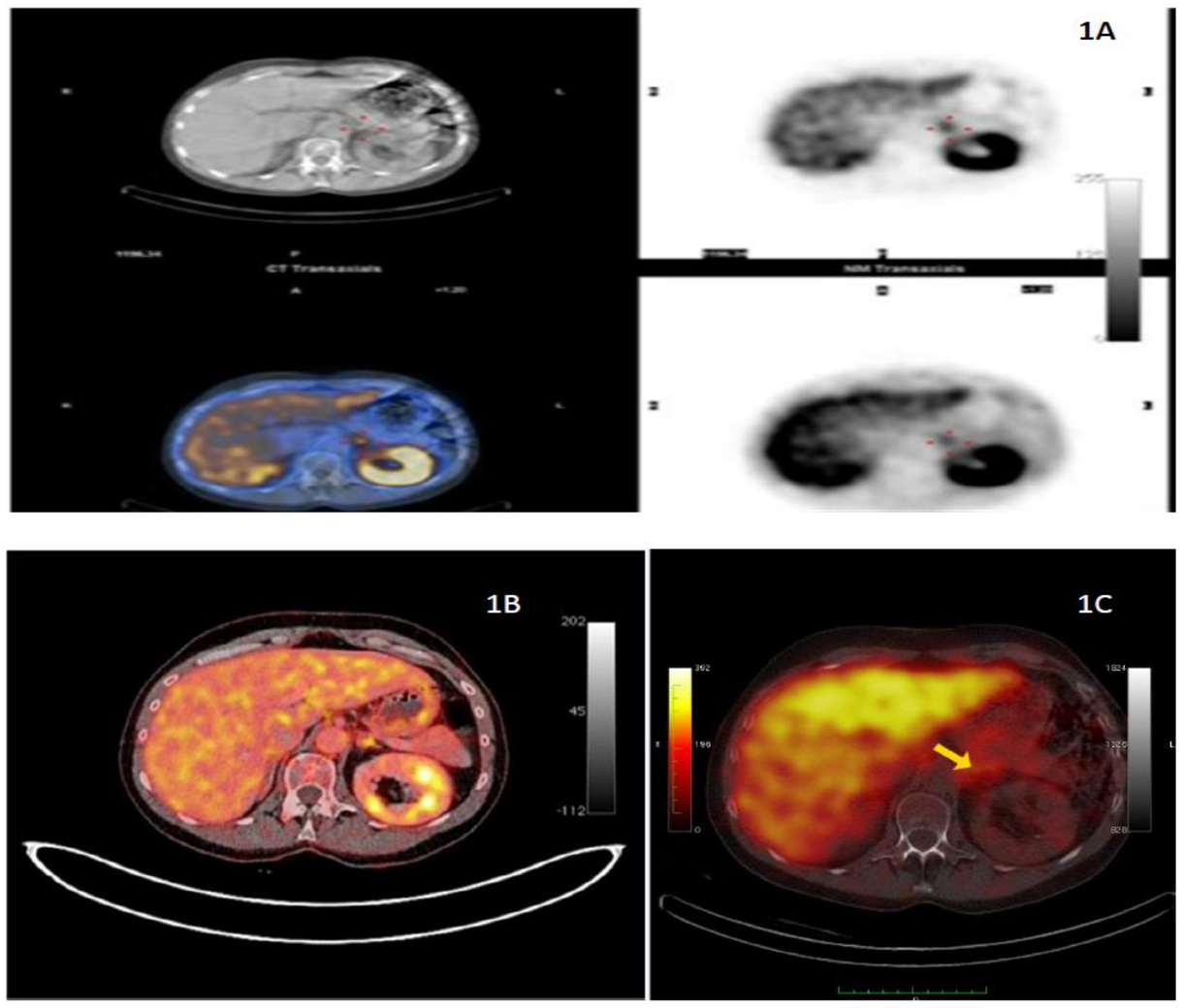

Figure 2: Left 2A: Adrenal medulla. Right 2B: Extra-adrenal ganglioneuroma

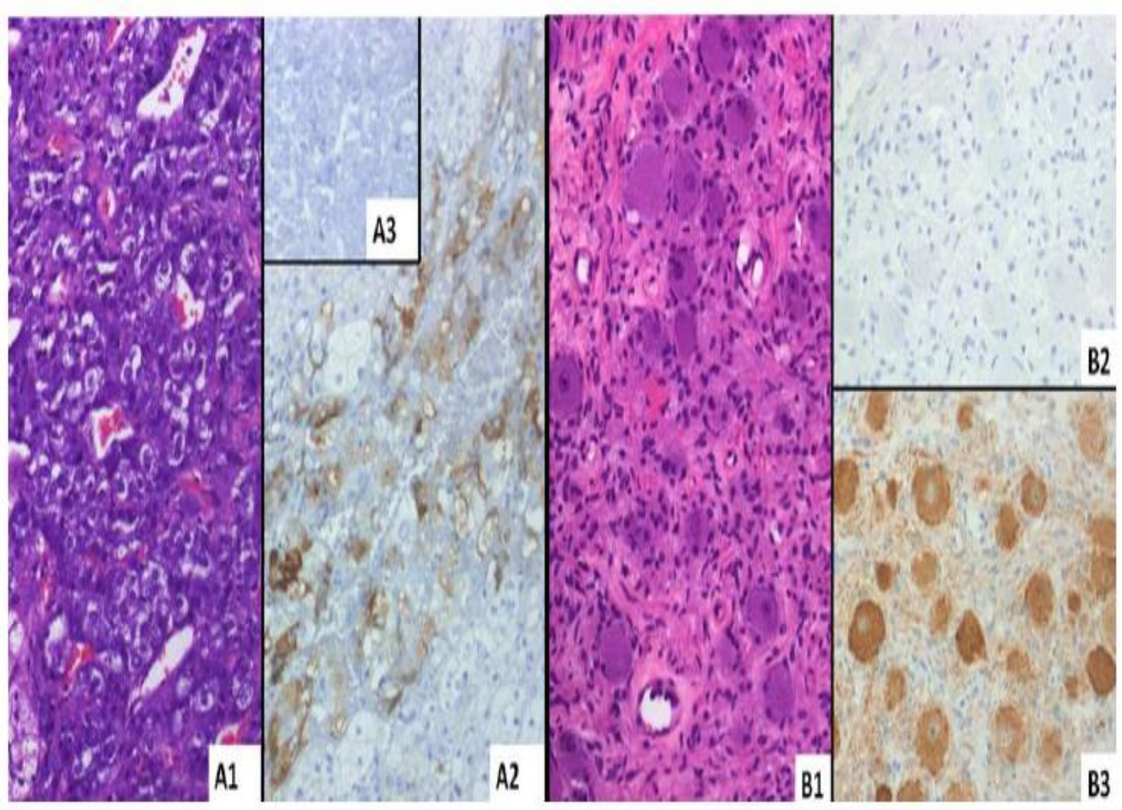

Figure 2A. A1: Adrenal medulla of the present case, without morfological abnormalities (H&E, x20); Figure A2: positive ACTH immunostain of the same histological area as A1; A3: absolute negative ACTH stain in an adrenal control case

Figure 2B. B1: extra-adrenal ganglioneuroma (H&E, x20); B2: ACTH immunostain, absolutely negative; B3: NSE positive expression in neuronal component, according to the diagnosis

Fig. 1A - Somatostatin Receptor Scintigraphy (SSRS)
The Technetium-99m-labeled octreotide acetate scintigraphy (SPECT-CT) showed an assimetric adrenal gland uptake, with slightly higher concentration of somatostatin receptors in the left one.

Fig. 1B - 18F-FDG PET-CT
The 18F-FDG PET-CT also showed an assimetric adrenal gland uptake, with slightly higher glucose hypermetabolism in the left adrenal gland than in the right one.

Fig. 1C - 123I-MIBG SCINTIGRAPHY (Analog of norepinephrine)
The Iodine-123-metaiodobenzylguanidine SPECT-CT showed a high uptake on the left adrenal gland, expressing an energy-dependent type I amine uptake along with intravesicular hormone storage in the cells of the adrenal medulla.

Learning points:

- EAS with high cortisol levels can be life-threatening
- EAS can occur in the adrenal glands, even in the absence of pheochromocytoma.
- Surgery represents the best treatment option for adrenal EAS, with potentially curative effect.
- Adrenal-blocking agents drugs: are fast, safe, cheap and well known remain being first line drugs.
- HC can affect SSTR2 expression in tumor, with diagnostic and treatment implications
- SSAs may have unpredictable responses

ECE 2016 262-EP

Clinical case reports - Pituitary/Adrenal

Betina Biagetti

Poster presented at: ECE2016 28 – 31 May 2016 MUNICH, GERMANY 18th EUROPEAN CONGRESS OF ENDOCRINOLOGY

PosterSessionOnline.com