ADDISON DISEASE IN ANTIPHOSPHOLIPID SYNDROME – CASE REPORT
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INTRODUCTION
- Primary adrenal insufficiency or Addison Disease (AD) is a potentially fatal condition if not diagnosed in time.
- Antiphospholipid syndrome (APS) is characterized by recurrent arterial/venous thrombotic events due to complex interactions between antiphospholipid antibodies, endothelium, and platelets.
- AD is a rare APS complication (< 0.5% of APS cases), although it is the most common endocrine manifestation of the syndrome, and it is caused by adrenal venous thrombosis and consequent hemorrhagic infarction.

CASE REPORT
- Caucasian woman, 36 years old
- APS diagnosis since the age of 24; arterial hypertension
- Medical history of multiple thrombotic events
  - deep vein thrombosis
  - spontaneous abortions (G3 P0)
  - splenic vein thrombosis; hypersplenism; thrombocytopenia
  - iatrogenic hemorrhagic events
- 9 – 22/11/2010: hospitalization for acute renal failure (creatinine 3.63mg/dL) after spontaneous abortion. Discharged under corticosteroid therapy (prednisolone 1mg/Kg/day, progressive dose reduction)
- Abdominal CT 10/11/2010: “heterogeneous nodular lesion with high spontaneous density, suggestive of hematoma, in the right adrenal gland (72 x 50mm); left gland poorly visualised; voluminous splenomegaly”
- 26/12/2010 (1 week after suspending prednisolone): admitted to the emergency department:
  - NAUSEA, PERSISTING VOMITING, ASTHENIA (4 days)
  - Medicated then with warfarin, pantoprazole, nifedipine and furosemide
  - Normotensive (patient prone to hypertension)
  - Initial analytical study: HYponATREMIA (114 mmol/L) and HYPERKALIEMIA (7.9 mmol/L), significative worsening of renal function (creatinine 4.90 mg/dL)

- Readmitted and restarted on therapy with prednisolone. 29/12/2010 (9 a.m.), under prednisolone 40mg id:
  - cortisol 1,3 ug/dL (5-25)
  - ACTH 15 pg/mL (9-52)
- Probable diagnosis: PRIMARY ADRENAL FAILURE
  - She initiated glucocorticoid and mineralocorticoid replacement with good clinical and analytical response (discharged with hydrocortisone 10mg +5mg +5mg and fludrocortisone 0.05mg id)

Further study after stabilization (Endocrinology Department) 1/03/2011 (8 a.m.), after suspending hydrocortisone for 48h: cortisol <1 ug/dL (5-25)
- ACTH 138 pg/mL (9.52)
- Diagnostic confirmation
  - aldosterone < 7.0 pg/mL (40-310)
  - active renin 4.9 uU/mL (7.76)
  - DHEA-SO4 < 0.2 ug/mL (0.35-4.3)
  - androstenedione < 0.3 ng/mL (0.5-3.4)
  - 17-OH-progesterone 1.70 ng/mL (0.2-1.8)
  - 21-hydroxylase antibodies negative
  - no anterior pituitary deficits
  - normal thyroid function
  - creatinine 1.68 mg/dL, normal ionogram

Abdominal MRI 14/05/2015 (Fig 1): small nodule with T2 hypointensity and T1 hyperintensity – reabsorption of adrenal hematoma

Currently treated with hydrocortisone 15mg + 10mg + 5mg, fludrocortisone 0.05 mg id, warfarin and antihypertensives. Continues to be followed in regular Endocrinology consultation.

CONCLUSION
- Based on the clinical presentation, analytical study and adrenal imaging, the diagnosis of AD was established in this case.
- High index of clinical suspicion for AD in APS is needed – in suspected cases, the diagnosis should be investigated and treatment promptly initiated.
- Conversely, the possibility of APS should be considered in patients with AD of unknown etiology, after exclusion of more common causes.