02/27/21 Morning Report with @CPSolvers

***Case Presenter**: Thiago Mendes (@mendes.thiagob) **Case Discussants**: Sandra Fonseca (@SandraF73382667) and Maria Luiza (@mluizarod)*

CC: darkening of the skin.

HPI: 17 yo female presenting with darkening of the skin for a long time.

Fatigue, poor school performance (unable to inject insulin in herself - needed help for that)

Vitals: **T**: normal | **HR**: normal | **BP**: 118x72 mmHg | **RR**: normal | **SpO_2**:normal | BMI 20.4 | No postural hypotension

Exam:
Gen: normal
HEENT: normal
CV: normal
Pulm: normal
Abd: normal
Neuro: normal
Extremities/Skin: lesions with increased pigmentation around the mouth and elbow (internal region) associated with some descamation around the mouth

Past Medical History:
Type 1 Diabetes since 9 yo

Meds:
Insulin Glargine 36 UI/day
Insulin Lispro 6-6-6 (before eating)

Family History:
None

Social History:
None

Health Related Behaviours:
None

Allergies:
None

Notable Labs & Imaging:
Hematology:
WBC: 7010 | Hgb: 11,9 | Hct 35,9

Chemistry:
Na: 134 | K: 3,5 | A1c 9.6% | TSH 5,06 | LDL 101 | HDL 36 |Fasting glucose 181 | Total cholesterol 151
Morning cortisol 2,6 (low) | ACTH > 1250 (high)| Aldosterone 6 (low) | Renin > 500 (high)

Final Diagnosis: Adrenal Primary Insufficiency (Autoimmune) or Addison's Disease -> Type-2 Polyglandular syndrome (associated with type 1 diabetes)

Treated with Hydrocortisone, and the skin lesions and the school performance got better

Problem Representation:
ENG: 17yoF w/ PMH of type 1 DM p/w chronic darkening of the skin. Labs showed low cortisol and high ACTH.
ESP: mujer de 17 años con antecedente de diabetes tipo 1 se apresenta con fatiga y oscurecimiento de la piel
POR: Paciente 17f com DM mal-controlado com lesões escurecidas por longo tempo. Exames mostraram baixo cortisol e elevado ACTH.

Teaching Points (Rafa):

- **APPROACHING YOUNG F W/ DARKENING SKIN LESIONS**
  Nutritional deficiencies, drug abuse, primary adrenal insufficiency (high ACTH - increased melanin production - hyperpigmentation of buccal mucosa, palms), Peutz-Jeghers syndrome (hyperpigmented macules on mouth, lips, hands, genital a/w hamartomas throughout GI tract), heavy metals, genetic syndromes, PCOS (insulin resistance - hyperpigmentation).
- **TYPE 1 DM**
  Most common in young patients - autoimmune destruction of B-cells - clinical manifestations include polydipsia, polyuria, polyphagia, weight loss, DKA)
  Pearl: Not seen w/ acanthosis nigricans - insulin resistance is seen in type 2 DM!
- **ADRENAL INSUFFICIENCY**
  Addison disease - autoimmune destruction
  Infectious causes: TB, Paracoccidioides
  Waterhouse-Friderichsen syndrome: acute and d/t aladrenal hemorrhage (DIC, N. meningitidis)
  Clinical syndrome: look for N/V, abdominal pain, fatigue, hyperkalemia (not always present), hyponatremia, NAGMA, hypotension
  Cortisol and ACTH in the morning are important for the dx
- **AUTOIMMUNE POLYGLANDULAR SYNDROME**
  Type 1: chronic mucocutaneous candidiasis, hypoparathyroidism, and AI
  AIRE dysfunction
  Type 2: Addison's disease (AD) associated with autoimmune thyroid disease and/or T1DM