Polyglandular autoimmune Syndrome type 2 / Schmidt’s syndrome

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Case history

- 25 years old lady presented to gastroenterologist with recurrent bouts of vomiting associated with abdominal pains, fatigue, muscle aches and dark tan.
- On examination she had dark pigmentation of the palms well hydrated and haemodynamically stable.

- Initial biochemical profile and upper GI endoscopy were normal.
- No family history of autoimmune conditions.
- She had traumatic laceration of the liver as a child, otherwise no significant past medical history.
- She was discharged after rehydration.

- A few days later admitted to A&E with a history of continued vomiting. She was found to be hypotensive with postural drop.
- Biochemical profile revealed hyponatremia and a random cortisol was <20, Addison's disease has been diagnosed. She has been rehydrated and commenced on IV steroids.
- Adrenal antibodies were positive. She was started on oral steroid replacement once haemodynamically stable.
- She had subclinical hypothyroidism with positive thyroid antibodies now has progressed to hypothyroidism on levothyroxine replacement.
- Recently, she has developed alopecia areata been treated with topical steroid cream by dermatologists.

Discussion: PGA type 2 is common of the immune-endocrinopathy syndromes affecting 2 or more endocrine organs and other non-endocrine organ. It is characterised by the occurrence of autoimmune Addison's disease, thyroid autoimmune diseases and/or type 1 diabetes mellitus. Primary hypogonadism, myasthenia gravis, coeliac disease also are commonly observed in this syndrome. Rarely is associated with Alopecia, pernicious anaemia, myasthenia gravis, immune thrombocytopenia purpura, Sjogren's syndrome, rheumatoid arthritis. The prevalence of Schmidt's syndrome is 1.4-2.0 per 100,000 populations, most commonly affects middle aged female.

References: