

A Case of Schmidt Syndrome Masqueraded by End Stage Renal Disease

Sarahi Herrera- Gonzalez and Martha Bilbatua*

Department of Internal Medicine, Jersey City Medical Center

Corresponding author

Martha Bilbatua, Department of Internal Medicine, Jersey City Medical Center, 355 Grand Street, Jersey City NJ, 07302, USA

Submitted: 17 Jun 2020; Accepted: 25 Jun 2020; Published: 06 July 2020

Background

Autoimmune Polyendocrine Syndromes (APS) are a rare manifestation of autoimmune diseases, this syndrome is composed by 5 types, nevertheless Type I (or juvenile) and Type II (or Adult) syndromes are the most common. APS II englobes Adrenal Insufficiency associated with Autoimmune thyroid disease or/and Diabetes Mellitus Type 1 [1]. We present an interesting case of a patient with Autoimmune Hypothyroidism and Adrenal insufficiency whose diagnosis was delayed by masquerade of diagnosis by concomitant End Stage Renal disease.

Case Report

Our patient is an 80-Year-Old Female with extensive past medical history including Hypertension, coronary artery disease, Diastolic Heart Failure, Atrial Fibrillation, Hashimoto's disease, Ureteral Cancer status post Urostomy and artificial bladder, End Stage Renal Disease (ESRD) on Hemodialysis, amongst others who presented to our facility for evaluation of Abdominal Pain of 2-month duration. She also endorsed decrease appetite and weight loss, as well as fatigue. However, an 8-day history of nausea and vomiting is which finally prompted her to seek medical attention.

Further evaluation was done and Computed Tomography (CT) of the abdomen revealed high grade small bowel obstruction and severe hydro-nephrosis, for which a Nasogastric tube was placed for decompression. Since admission, she was found to have repeated episodes of hypoglycemia with glucose levels of 30-60 mg/dL that were persistent despite dextrose infusion and high starch diet through the NGT tube. Other laboratory abnormalities on admission were notable for Na 122 mmol/L and K 5.8 mmol/L levels that were initially attributed to her known history of ESRD. By Nephrologist recommendation, the patient was started on Prednisone and Fludrocortisone given high suspicion of Adrenal insufficiency.

The patient was then evaluated by Endocrinology 8 days after initiation of treatment. Prior to morning dose of Prednisone, cortisol and ACTH levels were obtained and found to have morning cortisol level of 2 mcg/dL (3-22 mcg/dL), ACTH level of 811 pg/ml (6-50 pg/ml), Thyroid Peroxidase antibody of 50 IU/mL (< 9 IU/mL), Thyroglobulin antibody of 6 IU/mL (< 1 IU/mL) and a negative Adrenal Cortex Antibody, 21 Hydroxylase Antibody was not available at our facility. No further episodes of hypoglycemia

were seen.

Discussion

APS are divided in 5 groups, being I and II the most common syndromes [1]. APS I presents in early years of life and composes a combination of endocrinopathies with susceptibility to fungal infections, mostly oral candidiasis for which is also known as autoimmune polyendocrinopathy-candidiasis ectodermal dystrophy/dysplasia (APECED) [2]. It has been associated to a gene mutation in the 22q22.3 Chromosome called Autoimmune Regulator (AIRE) (1, 2), same gene that also contributes to the T helper cell response to mycosis by cell interaction between them and monocytes [2].

Schmidt Syndrome, or APS II, comprises Primary Adrenal Insufficiency (or Addison's disease) and Autoimmune Hypothyroidism, whereas the addition of Type 1 Diabetes Mellitus, or full triad is known as Carpenter syndrome Hashimoto's disease is known as the most common autoimmune disease, and the likelihood of having multiple associated autoimmune disorders subsequently is higher once the first develops [3-6]. Thus, a high index of suspicion and a low threshold for screening of other endocrinopathies has to be present in a clinician when other subtle signs and symptoms that may not be explained by the primary disease are noted.

Adrenal Insufficiency may present as chronic condition with insidious presentation or as an acute and life threatening emergency in the occurrence of adrenal crisis, most of the times with infection as a precipitating event [7]. Evaluation and diagnosis is based on screening of other endocrinopathies in subject with a primary autoimmune disorder given that in many cases sero-positivity presents before clinical disease [6, 7]. In Schmidt syndrome there is a lymphocytic infiltration and destruction of both glands [8].

Antiadrenal Antibodies and 21 hydroxylase antibodies are generally required for the diagnosis of Addison's disease [9]. Nevertheless, spontaneous disappearance of such antibodies has been reported previously, especially after steroid use, which has been linked to better outcome [10, 11]. Given that Primary Adrenal Insufficiency's most common cause is autoimmune, the lack of presence of the mentioned antibodies in our patient does not exclude autoimmunity as the etiology when there is possibility of spontaneous disappearance as reported.

Treatment of Schmidt Syndrome is basic and consists of deficiencies replacement. It is important to note that glucocorticoids and mineralocorticoids should be provided before thyroid replacement given that a sudden increase of metabolism is expected by thyroid hormone supplementation and can precipitate an adrenal crisis [4].

Conclusion

APS II, also known as Schmidt Syndrome is a rare disorder that generally presented in adults, mostly females, which encompass Adrenal Insufficiency and Autoimmune Hypothyroidism. Patients with one Autoimmune Endocrinopathy must be screen for secondary disorders given its likelihood of coexistence increases once one disorder is identified and given underdiagnosis may become life-threatening whereas its treatment is as simple as hormone and enzyme replacement.

References

1. Frommer L, Kahaly G (2019) Autoimmune Polyendocrinopathy. *The Journal of Clinical Endocrinology & Metabolism* 104: 4769-4782.
2. Sajjadi-Jazi S, Soltani A, Enayati S, Kakavand Hamidi A, Amoli M (2019) Autoimmune Polyglandular Syndrome Type 1: a case report. *BMC Medical Genetics* 20: 143.
3. Siniscalchi C, Moretti V, Cataldo S, Rocci A, Basaglia M, et al. (2018) The Schmidt Syndrome. *Acta Biomed* 88: 499-501.
4. Arya PVA, Kumar J, Unnikrishnan D, Raj R (2019) Case of autoimmune polyglandular syndrome type 2: how we uncovered the diagnosis. *BMJ Case Rep* 12: e227187.
5. Caturegli P, De Remigis A, Rose NR (2014) Hashimoto thyroiditis: clinical and diagnostic criteria. *Autoimmun Rev* 13: 391-397.
6. Martins SC, Venade G, Teixeira M, Olivério J, Machado J, et al. (2019) Autoimmune Polyglandular Syndrome type 2. *Rev Assoc Med Bras* 65: 1434-1437.
7. Bain A, Stewart M, Mwamure P, Nirmalaraj K (2015) Addison's disease in a patient with hypothyroidism: autoimmune polyglandular syndrome type 2. *BMJ Case Rep* 2015: bcr2015210506.
8. Van den Driessche A, Eenkhoorn V, Van Gaal L, De Block C (2009) Type 1 diabetes and autoimmune polyglandular syndrome: a clinical review. *Neth J Med* 67: 376-387.
9. Baxter M, Gorick S, Swords FM (2013) Recovery of adrenal function in a patient with confirmed Addison's disease. *Endocrinol Diabetes Metab Case Rep* 2013: 130070.
10. De Bellis A, Falorni A, Laureti S, Perrino S, Coronella C, et al. (2001) Time Course of 21-Hydroxylase Antibodies and Long-Term Remission of Subclinical Autoimmune Adrenalitis after Corticosteroid Therapy: Case Report. *The Journal of Clinical Endocrinology & Metabolism* 86: 675-678.
11. Zelissen PM, Bast EJ, Croughs RJ (1995) Associated Autoimmunity in Addison's Disease. *Journal of Autoimmunity* 8: 121-130.

Copyright: ©2020 Sarahi Herrera-Gonzalez. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.