A CASE REPORT OF AUTOIMMUNE POLYGLANDULAR SYNDROME II WITH MYASTHENIA GRAVIS
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Abstract : We describe the case of a 30 yr old man with known type 1 Diabetes for past 10 years who initially was admitted with Left lower lobe consolidation and Diabetic Ketoacidosis, after initial intensive management, further evaluation showed him to have hypothyroidism (hashimotos thyroiditis), hypogonadism, addisons disease and myasthenia gravis. Hence we present a rare case of autoimmune polyglandular syndrome type 2 (Schmidt syndrome) with myasthenia gravis.

Keyword : Type 1 Diabetes Mellitus, Hypothyroidism, Hashimotos Thyroiditis, Addisons disease, Hypogonadism, Myasthenia Gravis, Schmidt Syndrome

INTRODUCTION: Autoimmune Polyglandular Syndrome type II is the most common of the immunoendocrinopathy syndromes. It is characterized by the obligatory occurrence of Addisons disease in combination with thyroid autoimmune disease and type 1 DM. Primary hypogonadism, myasthenia gravis and celiac disease are also commonly observed in this syndrome.

CASE REPORT: The patient is a 30 year old male with known Type 1 diabetes mellitus for past 10 years who was initially admitted to the IMCU in a moribund condition, diagnosed with left lower lobe consolidation – Klebsiella pneumonia and Diabetic Ketoacidosis. After intensive management in the IMCU with IV Fluids, Insulin infusion, Antibiotics and close monitoring of electrolytes he made a significant recovery. A week later he was shifted to the general ward with resolution of lung signs both clinically and radiologically with well controlled blood sugar levels. In the ward a detailed clinical examination showed him to be ill built, poorly nourished with mild pallor, glossitis, dark pigmentation of buccal mucosa, the knuckles, elbows and palmar creases. His vitals were stable except for prominent postural hypotension; Supine BP -110/70 mmHg; standing BP – 86 / 60 mmHg. Neurologically he had bilateral partial ptosis, which was more obvious towards the end of the day, weakness of neck flexion and weakness of proximal muscles of upper and lower limbs. Reflexes were present with prominent pseudomyotonic reflex. Sensory system was normal. His heart sounds were regular, abdomen was soft with no organomegaly, respiratory system showed few resolving crepitations in the left lung base.

His lab investigations were as follows – CBC: Hb – 9g/dL, TC – 10000, Plt – 1.7 L, MCV – 84.5, MCH – 25.8, MCHC – 32. Peripheral Smear showed normochromic normcytic blood picture and iron studies were suggestive of anemia of chronic disease. Ferritin – 398.2 ng/ml (22 – 322 ) Fe – 12 ug/dL (65 to 175 ) TIBC – 233 ug/dL (250 to 450 ) Transferrin – 183 (176 – 280)
The most frequent clinical combination is Addison disease, Type 1 DM, Hashimotos thyroiditis (Schmidt syndrome); while the least frequent combination is Addison disease, Graves disease and type 1 DM (Carpenters syndrome).

Other disorders associated with PGA–II include – Hypogonadism, Idiopathic thrombocytopenic purpura, Myasthenia gravis, Parkinson disease, vitiligo, alopecia, pnicious anemia, Celiac disease, Graves disease and seronegative arthritis.

The pathophysiology of polyglandular autoimmune syndrome type II is poorly understood; some degree of genetic susceptibility must exist in the individual; it is associated with HLA-DR3 &/or HLA DR4. The individual is then exposed to the autoimmune trigger, which could be an environmental or intrinsic factor. The trigger mimics the molecular structure of a self-antigen. An alternative explanation is that a breakdown in immunologic tolerance occurs. Next, a subclinical phase of active production of organ-specific autoantibodies occurs. This phase is followed by autoimmune activity in the respective organ, in which there is progressive glandular destruction. The individual is still asymptomatic. Overt clinical disease subsequently develops when extensive organ damage has occurred.

The differential diagnosis of PGA syndrome should include DiGeorge Syndrome (hypoparathyroidism due to glandular agenesis and thymic agenesis), Neuberger syndrome (hypoparathyroidism, primary hypogonadism, type 1 DM & panhypopituitarism), Wolfraams Syndrome (DIMOAD syndrome) and IPEX syndrome (immune dysregulation, polyendocrinopathy, enteritis and X linked inheritance).

Currently the treatment of the polyendocrine syndromes is dictated by the individual disorders. Our particular patient was put Insulin (Basal bolus regimen), Steroids – Hydrocortisone 10 mg in the morning and 5 mg in the evening. Thyroid replacement with Liotrix 100 ug per day and pyridostigmine 60 mg TID. The patient showed improvement in ptosis and muscle power after starting pyridostigmine. We plan to commence him on testosterone supplements on subsequent follow up.

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