Case Report on Autoimmune Polyendocrine Syndrome

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Abstract: Polyglandular autoimmune syndrome is a very rare disorder, it occurs in certain homogenous ethnic populations due to consanguineous marriages. This is a case of 24-year-old male came with the complaints of swelling in front of neck and slowness of speech for 1 year and feeling difficulty while swallowing since 1 year, he was also suffering with hair loss, premature graying of hair, and weight loss. Patient is a known case type I DM in the past 10 years and is on ayurvedic medicines. As the patient was diagnosed with autoimmune polyendocrine syndrome, the treatment is more complex for multiple glandular deficiency than the treatment of an isolated endocrine deficiency. So, symptomatic treatment is preferred for this patient. For type 1 DM Glicazide 80 mg OD is recommended and Candid mouth paint is given for oral candidiasis, along with Tab Fluconazole. Also, vitamin supplement, Inj rabeprazole and soliwax ear drop were recommended.

Keywords: polyglandular, autoimmune, deficiency, endocrine, candidiasis

1. Introduction

Autoimmune polyendocrine syndromes (APS), also known as polyglandular autoimmune syndromes (PGAS). APS is a heterogeneous group of rare diseases characterized by autoimmune activity against more than one endocrine organs. Sometimes, non-endocrine organs also get affected. The two major autoimmune polyendocrine syndromes, (type1-type2/APS-1 and APS-2), both have Addison's disease as a prominent component. Further autoimmune polyendocrine syndromes include APS3 and APS4. The major autoimmune polyendocrine syndromes have a strong genetic component with the type 2 syndrome occurring in multiple generations and the type 1 syndrome in siblings.

2. Case report

A 24-year-old male came with the complaints of swelling in front of neck and slowness of speech for 1 year and feeling difficulty while swallowing since 1 year, he was also suffering with hair loss, premature graying of hair, and weight loss. He was complained breathlessness and chest pain for the past 3 months. Patient is a known case type I DM in the past 10 years and is on ayurvedic medicines. He also complains weight loss from 45 kg to 37 kg within 5 months. He complained about progressive loss of hearing of left ear and pain in the same. He also complained about skin tags in his body. Family history tells about 3rd degree consanguineous marriage. The patient presented with male pattern baldness, oral candidiasis, hypoplastic crowded teeth, Madorosis, thyroid swelling, skin tags on right shoulder, onychomycosis. Laboratory investigations reveals that PPBS 243 mg/dl and TSH level were seems to be elevated 3.45 μIU/ml and USG of neck shows goiter with multiple heterogeneous nodules.

3. Intervention

Patient was diagnosed with Polyglandular autoimmune syndrome

4. Discussion

Polyglandular autoimmune syndrome is a very rare disorder, it occurs in certain homogenous ethnic populations due to consanguineous marriages. Its hallmarks are chronic mucocutaneous candidiasis, hypoparathyroidism and adrenal insufficiency. It is more prevalent among some populations such as Finns that is 1 out of 25000 then Sardinians that is 1 out 14,400 and Iranian Jews.

Symptomatic treatment is preferred for Polyglandular autoimmune syndrome. For type 1 DM Glicazide 80 mg OD is recommended and Candid mouth paint is given for oral candidiasis, along with Tab Fluconazole. Also, vitamin supplement, Inj rabeprazole and soliwax ear drop were recommended.

5. Conclusion

As the patient was diagnosed with autoimmune polyendocrine syndrome, the treatment is more complex for multiple glandular deficiency than the treatment of an isolated endocrine deficiency. So, symptomatic treatment is preferred for this patient.

References


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