Autoimmune polyglandular syndrome presenting with pancytopenia in an older male - A case report

Abstract

Background

Autoimmune polyglandular syndromes (APSs) are characterized by multiple endocrine gland dysfunctions with or without non-endocrine organ involvement. These syndromes typically manifest in younger adults, being commoner in females than in males. We report a case of APS-3 B plus C in an older male.

Case presentation

A 64-year-old male presented with malaise, lethargy and exertional dyspnoea for two months. Examination revealed a diffuse goitre with bilateral pitting ankle oedema and multiple depigmented irregular macules over the lower limbs suggestive of vitiligo. Investigations revealed a pancytopenia, vitamin B12 deficiency, histological evidence of atrophic gastritis confirming the diagnosis of pernicious anemia. He also had primary hypothyroidism with positive anti-thyroid peroxidase and anti-thyroglobulin antibodies confirming autoimmune thyroiditis. A diagnosis of autoimmune polyglandular syndrome type 3 B plus C was established. He was treated with blood transfusions, intramuscular vitamin B12 injections and Thyroxine replacement. With this his haemoglobin improved and thyroid profile normalized.

Conclusion

APS can manifest even in older age. Organ involvement in APSs may not conform to a single defined cluster. Patient with an autoimmune organ dysfunction should be screened for other possible autoimmune disorders in order to diagnose complex combinations of APS.

Keywords: autoimmune polyglandular syndrome, autoimmune gastritis, vitilligo, autoimmune thyroiditis.