Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 5): May 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.572628 Impact Factor- 3.109

AUTOIMMUNE POLYGLANDULAR SYNDROME (TYPE III C) AND NON-SUSTAINED VENTRICULAR TACHYCARDIA – A RARE CASE REPORT

Dr. Darpanarayan Hazra[1], Dr. Sameer Mehrotra[2], Dr. Shahid Mahdi[3] and Dr. Subhash Chandra[*4]
[1-4] Department of Cardiology, BLK Super specialty Hospital, India

Keywords: *APS TYPE IIIC, APS and NSVT, APS and arrhythmia.*

Abstract

Failure of two or more endocrine glands together requiring hormone replacement is termed as "Autoimmune polyglandular syndrome" (APS). The aim of this case report is to underline the possible etiological link between primary hypothyroidism and non-sustained ventricular tachycardia (NSVT), although supraventricular arrhythmias are common features of hyperthyroidism. Here we present a case of 47 years old patient with repetitive episodes of NSVT followed by ventricular premature complexes (VPC) and ventricular trigeminy. He was diagnosed to have autoimmune thyroiditis 4 years ago and was on thyroxin replacement therapy, which could be an attributed risk for reactivation of arrhythmia.

Introduction

Autoimmune polyglandular syndrome (APS) is associated with two or more endocrine disorders, which are mediated by autoimmune mechanisms and lead to a hypofunctional state. Due to autoimmune pathogenic mechanisms, APS has become an increasingly recognized clinical entity in endocrinology. Patients with APS are currently treated with appropriate replacement therapy for each deficient endocrine organ system.

In the future, early recognition of these disorders and delineation of their cause and pathophysiology may help to improve the clinical scenario and prevent other systemic deterioration. Recent literatures suggest that the initiation of Thyroid supplementation in case of hypothyroidism should be done after ruling out any underlying cardiac conditions, as thyroid hormone replacement therapy can precipitate arrhythmias in an undiagnosed cardiac case.

In this paper, we report an emblematic case of APS type 3c (detailed classifications are discussed below) with NSVT followed by ventricular premature complex and ventricular trigeminy, features not described in previous literatures.

Case Report

A 47 years old gentleman, presented to the emergency room with complaints of acute onset palpitations and chest pain for 1 day, with preceding history of dyspnea and dizziness for 4 days. He was a non-smoker, non-hypertensive, no history of bronchial asthma, or any thromboembolic events (DVT, pulmonary embolism, stroke, MI, amaurosis fugax, limb ischemia) in the past. One year old ECG showed ventricular premature complexes and a 24-hour Holter monitoring had revealed frequent ventricular ectopics in the form of isolated beats, couplets and triplets with occasional supraventricular ectopics, but no episode of supraventricular tachycardia, sustained or non-sustained ventricular tachycardia were recorded, for which he was managed with Tab. Amiodarone 200 mg twice daily for 2 weeks. Patient is a known case of type II diabetes mellitus for the past 8 years and was on oral hypoglycemic agents. Simultaneously, patient is a diagnosed case of primary hypothyroidism for 4 years for which he was on replacement therapy (50mcg once daily). Patient also presented with concerns of alopecia areata, dry scaly skin and vitiligo in the sun-exposed area for which he sought treatment, which however did not subside. Detailed history taking revealed positive family history of chronic thyroiditis with hypothyroidism in the mother and sister, and father had diabetes mellitus type 2.

RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 5): May 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.572628 Impact Factor- 3.109

General examination of the patient showed a conscious, oriented and afebrile patient with heart rate 72/min, B.P-128/98 mmHg, respiratory rate 16/min and SpO2 – 94% on room air. Chest examination and other systemic examinations were unremarkable. Local examination unveiled hypo-pigmented patches in the hands, feet and arms with patchy hair loss and dry scaly skin over the arm and forearm.

Initial blood investigations reported normal haemogram, kidney/liver function tests, cardiac biomarkers and arterial blood gas analysis. Chest X-ray was within normal limits. Old ECG (1 month) showed monomorphic non-sustained ventricular tachycardia and serial ECG at our center revealed frequent ventricular premature complexes (VPC), couplets and occasional run of ventricular trigeminy (Fig 1 -2). 2D echo revealed no regional wall motion abnormality with LVEF – 60%.

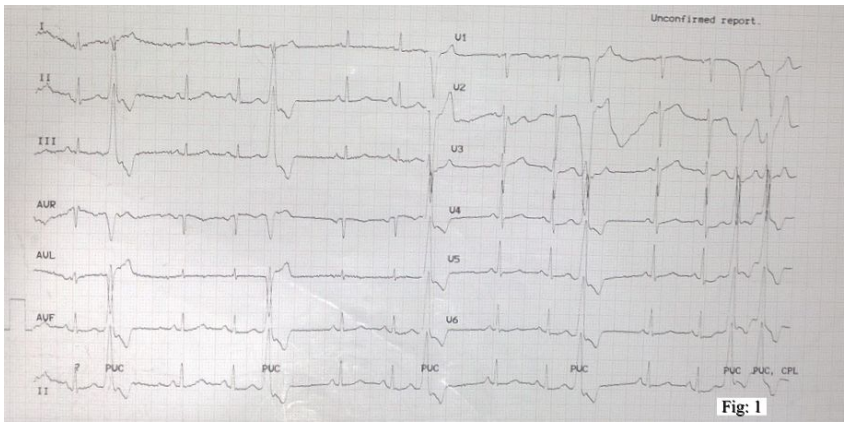


Fig: 1 – Ventricular trigeminy and couplets

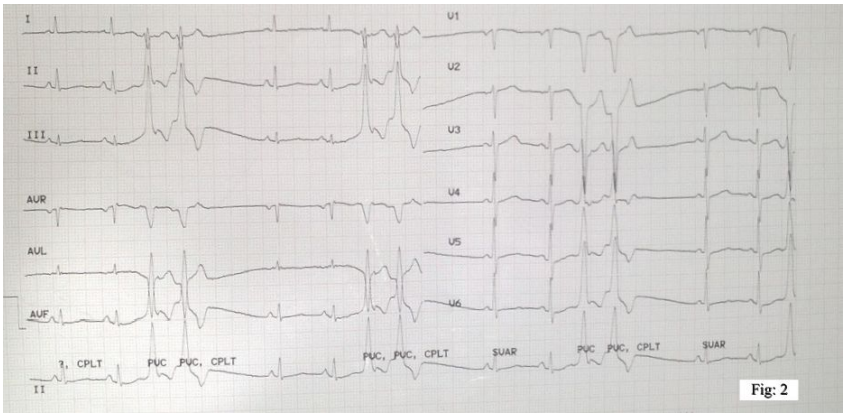


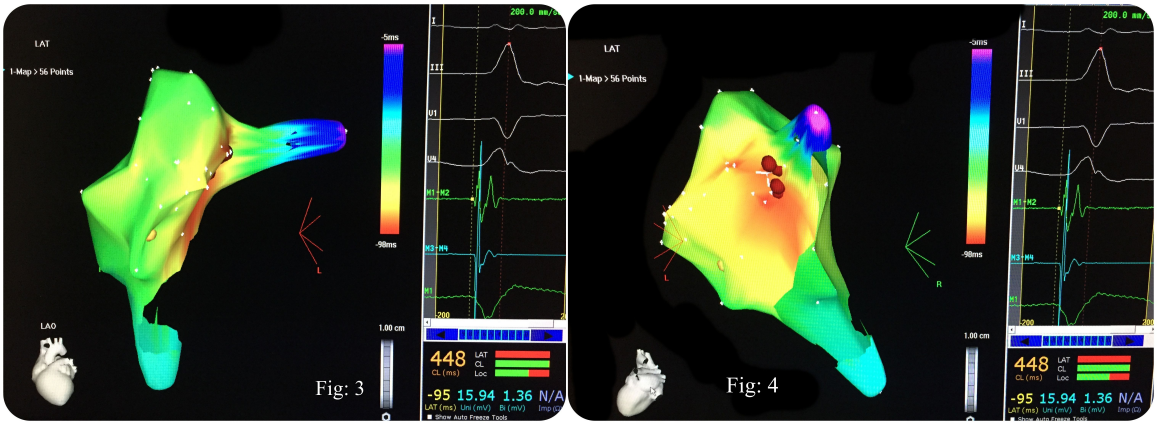
Fig: 2 –Frequent VPCs and couplets

The patient was admitted in ICCU for observation and close vitals monitoring. His initial treatment modality included Amiodarone infusion, dual anti platelets, statins and Inj. NTG 0.3ml/hour infusion.

RESEARCHERID
THOMSON REUTERS
Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 5): May 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.572628 Impact Factor- 3.109

Thyroid function was investigated which revealed T3 – 2.09 pg/mL (2.00-4.40), T4-0.81pg/mL (0.93-1.70), TSH-39.59 uIU/mL (0.27-4.20) and *anti thyroid peroxidase antibody more than1000 IU/ml (0.00-5.61).*

Coronary angiogram was performed to rule out ischemia as the cause of ventricular ectopics and findings were suggestive of non-critical coronary artery disease. Electrophysiological studies showed frequent outflow tract VPCs arising from right ventricle. The VPCs were mapped on 3D mapping (Carto) and were localized to postero septal area of right ventricle outflow tract. Successful radiofrequency ablation at the site resulted in absence of any further VPCs. 2D echo post procedure had no significant changes.



(*Fig: 3-4 VPCs were mapped on 3D mapping (Carto) (LAT view and LAO view) and were localized to postero septal area of right ventricle outflow tract*)

Patient's clinical condition improved progressively, and was discharged on single antiplatelet drug, statin and thyroid replacement therapy.

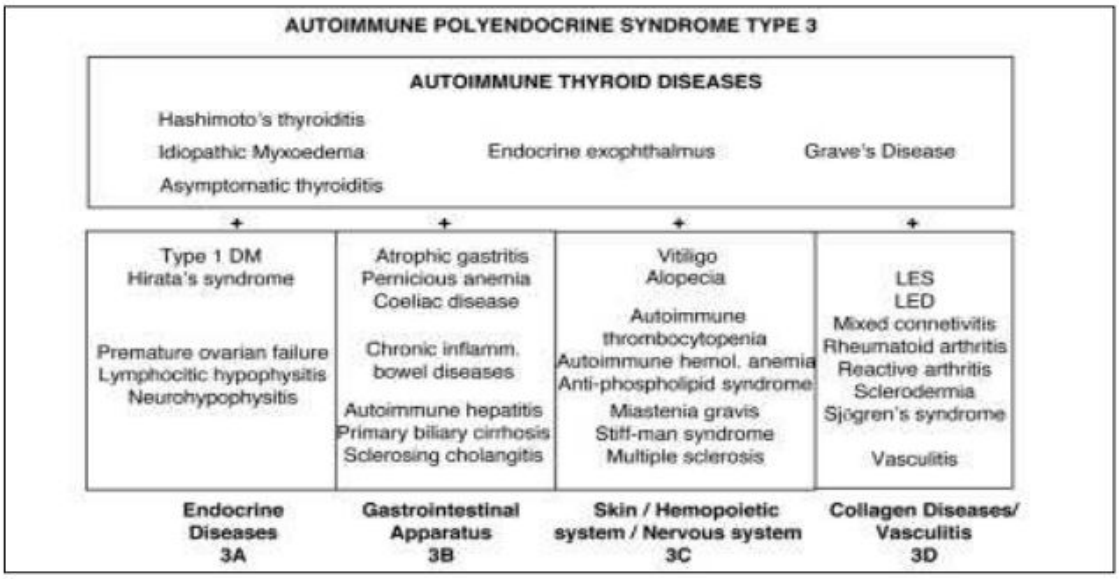
Discussion

Multiple endocrine gland insufficiency that is associated to an autoimmune disease was initially termed as Autoimmune Poly endocrine Syndrome (APS). However in 1980, Neufeld and Blizzard suggested a classification of APS, based on clinical criteria only, and further described them into four main types. (1,2)

- APS-1: chronic candidiasis, chronic hypoparathyroidism, Addison's disease (at least two present).
- APS-2: Addison's disease (always present) + autoimmune thyroid diseases and/or type 1 diabetes mellitus.
- APS-3: Autoimmune thyroid diseases associated with other autoimmune diseases (excluding Addison's disease and/or hypoparathyroidism)
- APS-4: Combinations not included in the previous groups

APS-3 was further sub classified into various entities based on the association of diabetes mellitus, pernicious anemia, vitiligo, alopecia, and hepatic autoimmune disease excluding adrenal involvement, which is depicted in the table below.

Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 5): May 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.572628 Impact Factor- 3.109



Gherbon Adriana et all demonstrated the prevalence of autoimmune chronic thyroiditis (ACT) and DM type 2 in 26.55% (77 patients, 69 F and 8 M). In adults DM type 2 and APS type III was found in 6 (2.06%) cases, of which all had ACT and vitiligo. In these cases the patients with autoimmune disease could have been latent autoimmune diabetes in adults (LADA). Therefore, if we have a patient with two or more autoimmune disease, we should investigate this for another possible autoimmune disease. (3)

Classical ECG manifestations in a patient with primary hypothyroidism are (4):

- Prolonged conduction defects
- Low voltage ECG
- Sinus bradycardia
- Atrio-ventricular or bundle branch blocks.

Tachyarrhythmia in primary hypothyroidism could be due to the following causes (4):

- Alteration of myocyte-specific gene expression
- Interstitial edema
- Myofibril swelling with loss of striation
- Increased arterial stiffness
- Endothelial dysfunction
- Premature atherosclerosis
- Disturbances of the sympathetic-vagal tone with a relative increase in sympathetic tone and autoimmunity.

So far, no case of APS type 3C presenting with non-sustained ventricular tachycardia was found in our search of literature.

This patient presented with non-sustained ventricular tachycardia followed by frequent ventricular premature complexes (VPCs), couplets, occasional run of ventricular trigeminy and co-existing autoimmune thyroiditis along with diabetes mellitus, alopecia areata and vitiligo (APS IIIc). The systemic problems were managed accordingly, with arrhythmia control as top most priority.

Open Access Journal
International Journal of Medical Research and Pharmaceutical Sciences
Volume 4 (Issue 5): May 2017 ISSN: 2394-9414
DOI- 10.5281/zenodo.572628 Impact Factor- 3.109

Conclusion

The presence of an autoimmune endocrinopathy urges the need to investigate other endocrine dysfunction in order to decrease the cardiac morbidity and mortality. Early recognition of such cases and replacement therapy could be lifesaving, bearing in mind the regular monitoring of thyroid function test to avoid over replacement, as it can lead to premature osteoporosis and cardiac arrhythmias (5,6). In cases with life-threatening arrhythmia we advocate appropriate treatment of the arrhythmia in top priority.

Financial support and sponsorship

Nil.

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

References

1. *Update on autoimmune polyendocrine syndromes (APS) Corrado Betterle, Renato Zanchetta Clinical Immunology and Allergology, Department of Medical and Surgical Sciences, Padua, Italy*
2. *Polyglandular autoimmune syndrome: current concepts Jeffrey Meyerson, MB, ChB Emilio E. Lechuga-Gomez, MD Pierluigi E. Bigazzi, MD Paul G. Walfish, MD, FRCPC, FACP*
3. *PREVALENCE OF POLYGLANDULAR AUTOIMMUNE SYNDROME TYPE III IN A GROUP OF ADULTS WITH THYROID DISEASES AND DIABETES MELLITUS Gherbon Adriana MD, PhD Assistant Profesor, Department of Physiology and Immunology University of Medicine and Pharmacy Victor Babes 2A Eftimie Murgu Square, Timisoara, Romania*
4. *Polyglandular Autoimmune Syndrome Type 2 Presenting With Ventricular Tachycardia Olusegun Sheyin*, Taiwo Falade, Olufemi Fasanmade Department of Medicine, Lagos University Teaching Hospital, Lagos Nigeria *Corresponding author: osheyin@yahoo.com Received August 20, 2014; Revised October 03, 2014; Accepted October 10, 2014*
5. *A rare combination of type 3 autoimmune polyendocrine syndrome (APS-3) or multiple autoimmune syndrome (MAS-3)*
6. *Immunologic Endocrine Disorders Aaron W. Michels and George S. Eisenbarth, MD, Ph.D Barbara Davis Center for Childhood Diabetes, University of Colorado Denver, 1775 Aurora Court, MS B140, PO BOX 6511, Aurora, CO 80045*