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Case Report | Olgu Sunumu

A CASE OF PURE AUTONOMIC FAILURE PRESENTING WITH SYNCOPE; HYPOTENSION AND HYPERTENSION ATTACKS

SENKOP İLE BELİRTİLEN BİR SAF OTONOM BAŞARISIZLIK OLGUSU; HİPOTANSİYON VE HİPERTANSİYON ATAKLARI

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ABSTRACT

A 67-years old female patient was admitted to a cardiology specialist with the complaints of hypertension, syncope, and presyncope attacks for the last one year. While blood and urine tests were normal, follow-up of the patient's vital signs showed hypotensive attacks with presyncope and dizziness. Cranial Magnetic Resonance Imaging (MRI), carotid and vertebral artery color Doppler Ultrasonography (Doppler USG), Electroencephalography (EEG) ve Electromyography (EMG) were performed. Despite other tests being normal, cranial MRI showed white matter damage. The patient is still in routine follow-up.

It may be effective to apply different treatments for patients' syncope and other demonstrative and disability symptoms in pure autonomic failure (PAF).

Keywords: Pure autonomic failure, Syncope, Hypotension, Hypertension attacks

ÖZ

Altmış yedi yaşında kadın hasta son bir yıldır hipertansiyon, senkop ve presenkop atakları nedeniyle kardiyoloji uzmanına başvurdu. Kan ve idrar tetkikleri normal iken hastanın vital bulgu takibinde, presenkop ve baş dönmesi ile birlikte hipotansif atakları görüldü. Kranial MRI, karotis ve vertebral arter renkli Doppler Ultrasonografi (Doppler USG, Elektroensefalografi (EEG) ve Elektromiyografi (EMG) yapıldı. Diğer testlerin normal olmasına rağmen, kranyal Manyetik Rezonans Görüntüleme (MRG)'de beyinde beyaz cevher hasarı saptandı. Hastanın rutin takibi devam etmektedir.

Saf otonomik yetmezlikte, hastaların senkop ve diğer belirtileri için farklı tedavi seçenekleri uygulamak etkili olabilir.

Anahtar Kelimeler: Saf otonomik bozukluk, Senkop, Hipotansiyon, Hipertansiyon atakları

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Introduction

Pure autonomic failure (PAF) is not a well-known cause of orthostatic hypotension. A neurodegenerative disorder of autonomic nervous system clinically characterized by orthostatic hypotension was first described in 1925.[1] Bannister and Oppenheimer described a new entity in 1982 showing autonomic failure without other neurological symptoms of the Shy-Dragger Syndrome (also cerebellar ataxia, parkinsonism, and upper and lower motor neuron symptoms with orthostatic hypotension).[2] The hallmark of orthostatic hypotension is failure of releasing norepinephrine upon standing up. Dysfunction or loss of peripheral sympathetic nerves leads to impaired secretion of norepinephrine and other catecholamines. Conversely, patients usually have normal blood pressure while seating and sometimes have high blood pressure while lying down. Approximately, half of the patients have concomitant supine hypertension.[3] The cerebral white matter damage and left ventricular hypertrophy can accompany hypertension and may lead to end organ damage.[4,5] When the diagnosis is established as PAF for the patients presenting with orthostatic hypotension, several different treatments may be beneficial for syncope and other demonstrative and incapacitating symptoms.

Case Report

A 67-years old female patient was admitted to a cardiology specialist for hypertension, syncope, and presyncope attacks for the last 1 year. The patient did not have any known disease except hypertension with no medical treatment. Investigations for the etiology of her hypertension attacks, syncope, and presyncope were carried out. Ambulatory 24 hours cardiac rhythm and blood pressure monitoring showed slight sinus bradycardia at night (lowest 47 bpm) and hypertensive attacks. Transthoracic ECG revealed normal left ventricle ejection fraction with left ventricle hypertrophy. Because of the hypertensive attacks, hormone tests (Thyroid Stimulating Hormone; TSH, Adrenocorticotropic hormone; ACTH, cortisol, renin) and norepinephrine and metanephrine levels in 24 hours urine test were also performed. While all blood and urine tests were normal, follow-up of the patient's vital signs showed hypotensive attacks with presyncope and dizziness. Patient faints upon standing up. Moreover, when asked in details patients admitted having symptoms like frequent constipation and urinary incontinence. Neurology specialist also consulted the patient for the neurological causes of syncope. Cranial MRI, carotid and vertebral artery color Doppler USG, EEG, and EMG were performed. Despite other tests being normal, cranial MRI showed white matter damage (Figure 1). Sympathetic nervous system nuclear imaging as 123I-metaiodobenzylguanidine single-photon emission computer tomography (123I-MIBG-SPECT) was planned with the suspicious diagnosis of pure autonomic neuropathy and multisystem atrophy.

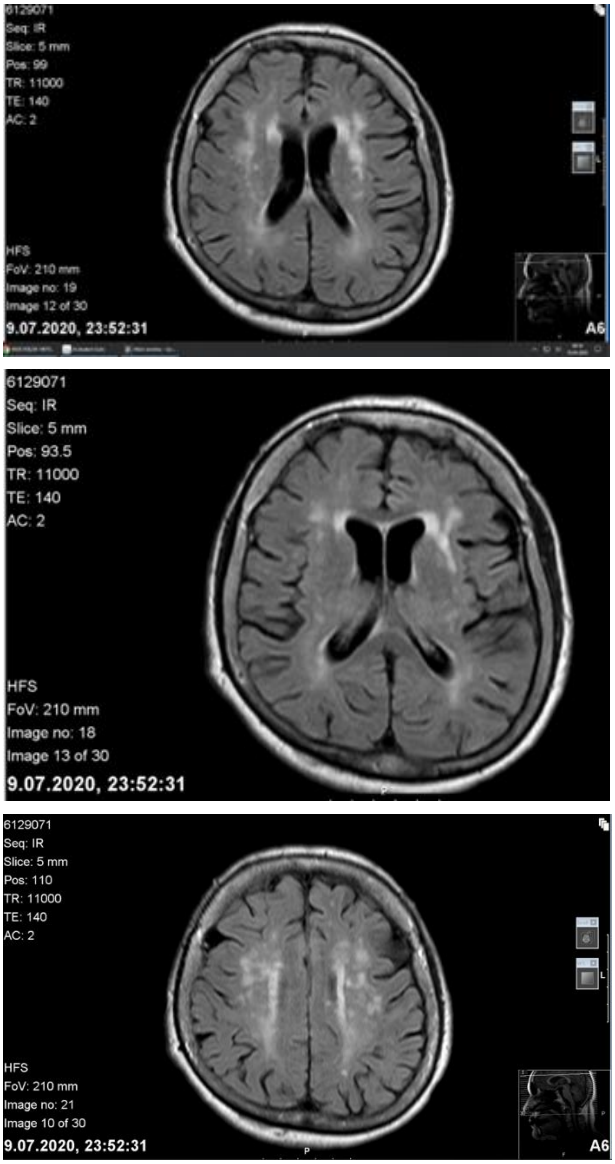

Figure 1. Flair images showing ischemic white matter lesions

The patient was planned to be referred to another hospital after discharging due to the unavailability of access to contrast media for scintigraphy in our hospital. Before the discharge of the patient, she was put on fludrocortisone as a treatment after which no more presyncope and syncope with hypotension attacks were seen. Cardiac sympathetic denervation at 123I-MIBG-SPECT was in line with the diagnosis of pure autonomic neuropathy (Figure 2). The patient did not have any neurological signs of parkinsonism, ataxia, or motor neuron symptoms. The patient is still in routine follow-up without any new complaints.

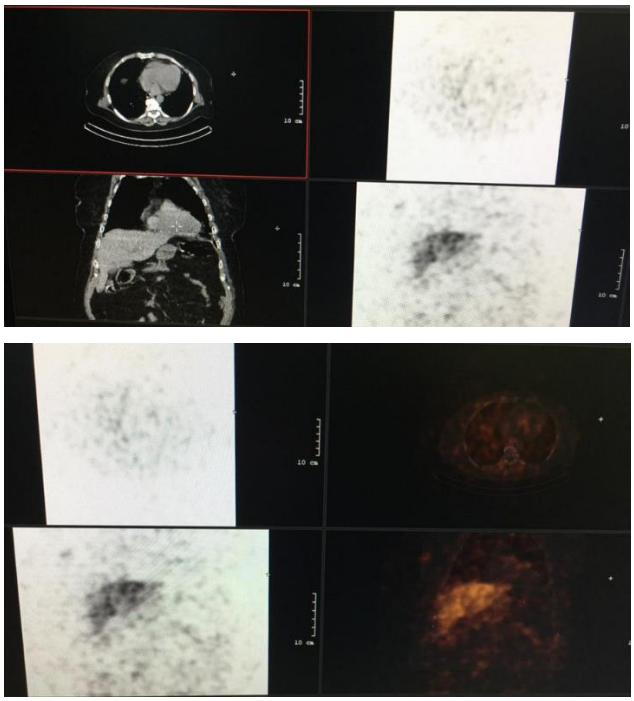

Figure 2. Pure autonomic neuropathy imaging with Spect 123I MIBG spect

Discussion

Autonomy PAF is a rare, sporadic, neurodegenerative disorder of the autonomic nervous system. Its main clinical symptoms are neurogenic orthostatic hypotension and urinary and gastrointestinal autonomic dysfunctions.

Orthostatic hypotension is defined as a reduction in systolic blood pressure by at least 20 mmHg or diastolic blood pressure of 10 mmHg within 3 minutes of standing or 60-degree head-up tilt.[6] When supine hypertension present greater than 140 mmHg or diastolic blood pressure than 90mmhg, then 30-mmHg systolic blood pressure reduction than 30mmhg is presumed more appropriate to meet the diagnosis of orthostatic hypotension.

Mechanically, orthostatic hypotension is related to venous pooling on lower extremities and, splanchnic vascular beds (approximately 300-1000 ml) on standing, which then leads to a reduction in venous return to the heart. Cardiac and stroke volumes are also reduced. In healthy individuals increase in cardiac contractility and heart rate leads to an increase in sympathetic outflow through baroreflex. In PAF, inadequate sympathetic response to standing is the leading cause of orthostatic hypotension. However, differential diagnosis of PAF is tough. It includes non-neurogenic orthostatic hypotension, syncope, neurogenic orthostatic hypotension, autonomic neuropathies, and inherited disorders. If the patient has syncope with hypotensive and hypertensive attacks and systolic blood pressure decrease on standing, interrogation of other symptoms like urgency, anhidrosis, dizziness, constipation, and incontinence may be helpful for the suspicion of PAF.[7] Tilt table test, cardiac sympathetic innervations imaging like ^{123}I-MIBG-SPECT and standing up blood pressure testing will set the diagnosis of PAF. Once the diagnosis is established, the treatment options may improve the quality of life of the patients.

Several strategies ranging from non-pharmacological measurements to medical treatment are available in PAF. Non-pharmacological measurements include an increase in fluid and salt intake, avoiding maneuvers increasing intrathoracic pressure (coughing, straining, etc.), encouraging isotonic exercise, and gradual movement change with postural change. On the other hand, medical treatment include fludrocortisone, midodrine, and another sympathomimetic agent, erythropoietin, caffeine, clonidine, droxidopa, and beta-blockers (with intrinsic sympathomimetic activity).

Conclusion

To increase the awareness of PAF, we here presented a PAF patient who responded to the first line therapy with 0,1 mg fludrocortisone which is available in our country. We would like to point out the use of ^{123}I-MIBG-SPECT imaging to test sympathetic innervations. Consideration of medical and non-pharmacological treatment options during careful follow-up of PAF patients may increase their quality of life.

Compliance with Ethical Standards

Written informed consent was obtained from the patient for the publication of this case report.

Conflict of Interest

All author declared no potential conflict of interest with respect to research of this article.

Author Contribution

All authors contributed equally to this article.

Financial Disclosure

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