

Bilateral adrenal hyperplasia complicated with severe ischemic stroke in a young patient

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Abstract

A young patient suffered from acute right hemiparesis, facial weakness and Broca's aphasia with multiple brain lesions due to severe hypertension. His evaluation for secondary causes of hypertension revealed hyperaldosteronism due to bilateral adrenal hyperplasia. Treatment is based primarily on spironolactone and ACE inhibitors. Two years later he was in an outstanding clinical condition with few remained neurological symptoms and his blood pressure well controlled.

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Secondary hypertension representing 5–10% of all cases of hypertension is mainly linked to diseases of the kidneys and the endocrine system. Although correcting the cause of secondary hypertension may lead to cure, diagnosis is probably missed in the majority of patients [1]. Secondary hypertension affects especially young people in whom hypertension is usually undiagnosed, untreated or inadequately controlled. However the consequences of this situation regarding target organ damages could be devastating for a young individual.

A young, right-handed patient, 37 years old, with acute right hemiparesis, facial weakness and Broca's aphasia was admitted to Neurology department during a hypertensive emergency (systolic blood pressure above 250 mm Hg). Computed tomography (CT) scan of the brain within few hours after the onset of symptoms was normal. However, Magnetic Resonance Imaging (MRI) revealed a cerebral infarction at the posterior limb of the left internal capsule as well as multiple small infarcts in the subcortical white matter.

The patient suffered from uncontrolled hypertension at least for nine years prior his admission. Previous incomplete screening for secondary hypertension as an outpatient was negative as far as he knows. Treatment of hypertension with various combinations of antihypertensive drugs, ACE-I included, was not sufficient.

During the following days we revealed the severity of lesions in organ-targets due to hypertension. A moderate to severe concentric hypertrophy of left ventricle and diastolic dysfunction on echocardiogram along with approximately 50% obstructive lesions in both internal carotid arteries as well as grade 2 lesions in both retinas were found. Blood tests revealed just a mild impairment of renal function with serum potassium within normal levels.

A week later the patient still suffered from facial weakness and recessive right pyramidal syndrome. Due to the severity of target organ lesions and the young age of the patient, it was decided for the patient to be transferred to Cardiology department in order to evaluate him for secondary causes of hypertension. There was no sign of coarctation of aorta on echocardiogram. Magnetic Resonance Angiography (MRA) revealed no stenoses in renal arteries. Size and shape of both kidneys were within normal limits; however mild bilateral adrenal hyperplasia was the only finding of an abdomen CT scan. Finally, there was no sign of obstructive sleep apnea.

Regarding patient's endocrinology profile thyroid hormones, catecholamines both in serum and urine and VMA in

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urine were within normal limits. Renin's activity was depressed both in rest and after 2-hour upright position while aldosterone levels were increased above normal limits after 2-hour upright posture (19.5ng/dl → 90.1ng/dl). Aldosterone to renin ratio was 39 in rest and 75 after 2-hour upright posture (normal ratio < 30).

Our findings were indicative for bilateral adrenal hyperplasia. Ischaemic stroke due to multiple brain infarcts was the main consequence of this case of secondary hypertension. Treatment was based primarily on aldosterone antagonist (spironolactone) and ACE inhibitors since no surgical treatment was required. Indeed, patient's blood pressure remained below 120mm Hg when spironolactone 75mg was added.

Four months later the patient was stable with a mild right pyramidal syndrome and he returned to work. A new MRI examination four months later, confirmed the same lesions mentioned above during the acute phase of stroke although more intense (Figs. 1 and 2).

Two years later the patient was in an excellent clinical condition. He managed to lose and remain 20 kg thinner while he was exercising every day. His blood pressure was within normal limits. He performed a symptom-limited cardiopulmonary exercise test for 9.00 min and he achieved a VO_2 max of 28.5 ml/kg/min with a peak blood pressure of 140/80 mm Hg.

Since secondary hypertension may lead to dramatic life threatening complications, the primary determinant for the workup is the index of suspicion. The best screening test for primary hyperaldosteronism is plasma aldosterone to plasma renin activity ratio. A ratio over 30 is suggestive of primary aldosteronism with a sensitivity of 91%. [1]. Bilateral adrenal

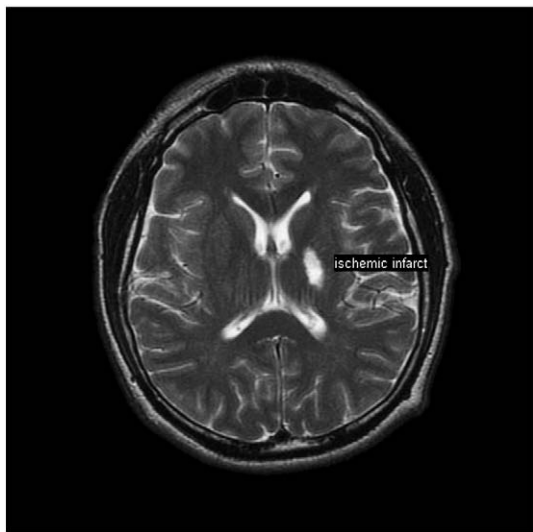


Fig. 1. Axial, T2-weighted MR scan four months after the acute ischaemic stroke. High signal intensity lesion is shown along the posterior limb of the left internal capsule.



Fig. 2. Axial, T2-weighted MR scan four months after the acute ischaemic stroke. Multiple small infarcts in the subcortical white matter (arrow).

hyperplasia represents <30% of the cases of primary hyperaldosteronism [2–4] and treatment is medical. Spironolactone is effective and doses from 25 to 400 mg per day have been used. Other antihypertensive agents may be used concomitantly while diuretics causing hypokalemia should be avoided [1].

Hypertension is a common finding in patients >65 years of age that contributes to cardiovascular morbidity and mortality. However, stroke of undetermined aetiology is the leading cause of neurological complications in a young patient, while hypertension is the third or fourth cause in a descending order [5]. The relatively low frequency of routine stroke aetiologies in patients aged below 45 can be explained by the significantly lower prevalence of traditional risk factors in these patients [6].

According to our knowledge, this is the first reported case of primary hyperaldosteronism due to bilateral adrenal hyperplasia leading to severe neurological complication in a young patient with a favourable long-term outcome.

Acknowledgement

The authors of this manuscript have certified that they comply with the Principles of Ethical Publishing in the International Journal of Cardiology [7].

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A new method to assess ventricular wall stress in patients with heart failure and its relation to heart rate variability

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Abstract

Introduction: Heart failure is characterized by an increase in cardiac load, wall stress and autonomic dysfunction. The neurohumoral imbalance arising from adrenergic activation and parasympathetic withdrawal is associated with worse prognosis. We addressed the hypothesis that an increased left ventricular (LV) wall stress as assessed by cardiac magnetic resonance imaging (CMR) in patients with heart failure is related to a depression of heart rate variability (HRV).

Methods: Cardiac function and mass were measured in 37 individuals with suspected cardiomyopathy using CMR imaging. A thick-walled sphere model was used to calculate ventricular wall stress. Time domain analysis of HRV was obtained by long-term Holter ECG.

Results: Standard deviation of both normal-to-normal (NN) intervals (SDNN) and average NN intervals over 5 minutes (SDANN-i) were negatively correlated with LV enddiastolic wall stress ($r = 0.42$, $P < 0.01$). SDNN and SDANN-i were severely decreased ($P < 0.01$) in patients with increased enddiastolic LV wall stress > 12 kPa (vs. normal range: < 4 kPa).

Conclusion: A relation between increased cardiac wall stress and depressed heart rate variability was observed in patients with heart failure. CMR-based measurement of LV volume and mass is appropriate to calculate LV wall stress which should be considered not only as a potential prognostic determinant but also as therapeutic target.

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Keywords: Heart failure; Ventricular wall stress; Cardiac magnetic resonance; Heart rate variability; Autonomic system; Cardiomyopathy

1. Introduction

Independent of its etiology, heart failure is characterized by an increase in cardiac load and autonomic dysfunction. A neurohumoral imbalance arises from adrenergic activation and parasympathetic withdrawal leading to stimulation of cardiac contractility, fluid retention and vasoconstriction by activation of the renin-angiotensin-aldosterone system. High

ventricular filling pressure and dilatation is associated with increased wall stress that contributes to an adverse cardiac remodeling with various unfavourable consequences for energy metabolism and gene expression [1–3]. Thus, self-perpetuating progression of ventricular dilatation, functional deterioration and autonomic dysfunction is likely to occur, whereby a rise in wall stress appears to have a key role. We addressed, therefore, the hypothesis that ventricular dilatation and a rise in wall stress are associated with depressed heart rate variability (HRV). As established for wall stress calculations in animals, we used a thick-walled sphere model of the left ventricle (LV) based on cardiac magnetic resonance imaging (CMR) to assess wall stress in patients. HRV was measured to assess the autonomic tone.

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