Suspected Conn's Syndrome in A Female Adult Hypertensive: A Case Report.

Type of Article: Case Report

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ABSTRACT

Background: The presentation of adrenal adenoma with ventricular fibrillation and sudden death is rare. Ventricular fibrillation is the most common cause of sudden death. Coronary artery disease, cardiac valvular or myocardial diseases, and non -cardiac abnormalities may also lead to ventricular fibrillation. We present a patient with recurrent ventricular fibrillation and associated adrenal tumour.

Methods: The case records of the index patient and a review of the literature on the subject.

Result: A 39 year old hypertensive woman presented in the accident and emergency department of the University of Port-Harcourt Teaching Hospital with a history of extreme weakness and inability to walk of four day duration. While being evaluated, she had a cardiac arrest and was successfully resuscitated. She had two more episodes of cardiac arrest in the Intensive care unit. Investigations revealed ventricular fibrillation from ventricular tachycardia and severe hypokalaemia with metabolic alkalosis. Abdominal CT scan revealed a right adrenal mass. ACTH level, serum and urinary cortisol levels were normal. A diagnosis of Conn's syndrome was made and patient did well on spironolactone and other anti-hypertensives as she awaits surgery.

Conclusion : There is a need for high index of suspicion for the secondary causes of hypertension while dealing with young patients and whenever there is an unusual presentation. This case highlights the need for proper investigation of patients.

Key Words: Conn's syndrome; Cardiac Arrest; Adrenal tumor; Nigerian Female.

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INTRODUCTION

Primary hyperaldosteronism is one of the endocrine causes of secondary hypertension and accounts for 0.05-2% of patients with secondary hypertension[1]. However, recent studies have shown that using the Plasma Aldosterone/Plasma Renin activity ratio as a screening test followed by aldosterone suppression confirmatory testing has resulted in much higher prevalence estimates (513% of all hypertensives) for primary aldosteronism[2].

Conn's syndrome was first described in 1955 by J.W Conn in a patient with a unilateral aldosterone-producing adenoma[3]. It is characterized by increased aldosterone production, suppressed plasma renin activity (PRA), hypertension, hypokalaemia and metabolic alkalosis. The term primary hyperaldosteronism is used to describe Conn's syndrome and other etiologies of primary hypersecretion of aldosterone. These include; bilateral adrenal hyperplasia, idiopathic hyperaldosteronism in which the adrenals appear normal and glucocorticoid-remediable aldosteronism in which ACTH stimulates aldosterone production. Rarely large adrenocortical carcinomas, adrenal embryologic nest neoplasms within the kidney and ovary secrete aldosterone. Hypokalaemia in a patient with hypertension who is not on diuretics should be investigated for primary aldosteronism. The presentation of adrenal adenoma with ventricular fibrillation and sudden death is rare[4]. Ventricular fibrillation is the most common cause of sudden death. Coronary artery disease, cardiac valvular or myocardial diseases, and non -cardiac abnormalities may also lead to ventricular fibrillation[5].

CASE REPORT

A 39 year old business woman, who had been hypertensive for four years for which she was taking 20mg of Nifedipine retard tablet daily presented with a history of generalized weakness and muscle cramps of about one week duration. Symptoms were not worse with activity. There was no history of exertional dyspnoea, orthopnoea or paroxysmal nocturnal dyspnoea. There were no associated headaches, tinnitus, palpitations, jaundice, anorexia or easy satiety. There was no facial or leg swelling, oliguria or cold intolerance and no past history of chronic use of corticosteroids. There was a family history of hypertension (paternal grandmother) and diabetes mellitus (father).

Physical examination revealed an obese woman with a body mass index (BMI) of 31.41kg/m^2, a buffalo hump and multiple striae. She was conscious and alert, anxious, not pale, afebrile, anicteric, not cyanosed, not dehydrated and had no peripheral oedema. Her pulse was 74beats/min and irregular while her BP was 140/80 mmHg sitting. Jugular venous pressure was not raised. Apex beat was located at the 6th left intercostal space, lateral to the mid-clavicular line, heaving in character. Heart sounds were I and 2 only. There were no cranial nerve deficits, no neck stiffness and Kernig's sign was negative. Muscle power was grade 2 globally. Tone was normal in all the limbs but deep tendon reflexes were depressed globally. While awaiting transfer to the ward from the accident and emergency department (A&E) she had a cardiac arrest and was resuscitated by cardiac massage and defibrillation. She was then transferred to the intensive care unit (ICU) where she had two further episodes of cardiac arrest and was successfully resuscitated. A diagnosis of hypertensive heart

disease with arrhythmias was made to exclude coronary heart disease in a known hypertensive patient.

Electrocardiography (ECG) revealed a heart rate of 80beats per minute, normal QRS axis, multiple ventricular ectopics with bigeminy and R on T phenomenon. There were no u waves. Echocardiography (ECHO) revealed normal looking pericardium and heart valves, asymmetric left ventricular hypertrophy (LVH) with preserved diastolic and systolic function. There was neither abnormal wall motion nor intra-cardiac thrombi.

The ECHO features confimed hypertensive heart disease. Brain CT scan was normal. Abdominal CT scan showed an oval shaped hypodense non-enhancing mass in the right suprarenal area measuring 25 x 29mm with an area of approximately 431mm^2. Its hounsfield unit was 19.04 (post contrast) indicating a soft tissue mass. Other organs were normal. Impression was right-sided suprarenal mass.

Differential diagnoses were adrenal adenoma and adrenal hyperplasia. Kidney function test showed Na+ - 144mmol/L, K+ -1.2mmol/L, HCO3- 30mmol/L, Urea - 2.6mmol/L, Creatinine 70umol/L. Serial electrolyte estimations showed persistent hypokalaemia (Table 1). The 9am serum cortisol was 114ng/ml (60-230), 24hr urine cortisol :560 mmol/24h (152-789) Urine volume :1360mls (600-2400)
Urine creatinine: 13.6mmol/24hr (7.1-15.9) ACTH: 4.1 (1.1-13.2)

A diagnosis of Conn's syndrome was made and treatment was commenced with Tabs Disopyramide 100mg tds, Tabs Amlodipine 10mg dly, Tabs slow K 600mg bd, Tabs Losartan 100mg dly, Tabs Methyldopa 500mg bd, Tabs Nifedipine ® 20mg bd, Tabs Spironolactone 50mg bd. Patient has

TABLE 1:
Serial Serum Potassium Estimates

DATE	6/5	8/5	9/5	10/5	11/5	12/5	15/5	17/5	18/5
K+ VALUE	1.6	2.4	1.3	1.4	1.3	1.3	1.7	1.3	1.4

improved remarkably and is awaiting surgical intervention .

DISCUSSION

This case highlights a patient with what was thought to be essential hypertension but on proper investigation was discovered to have hypokalaemia and a unilateral adrenal mass. She presented with recurrent cardiac arrest and weakness. These were a likely consequence of hypokalaemia. Hypokalaemia can cause atrial or ventricular arrythmias, prolonged QT interval and prominent U waves on ECG[5].

A similar case to ours was reported in Turkey in a 58-year-old man[1] who collapsed while on vacation from ventricular fibrillation. His serum potassium level was 1.8 mEq/L after successful cardioversion. Coronary angiography showed a normal heart with no structural defects, but 12-lead electrocardiography showed indications of left ventricular hypertrophy, which was confirmed by echocardiography. Laboratory examinations showed a suppressed renin level and an elevated serum aldosterone level. Computed tomography then revealed a right adrenal mass . We did not find any similar case in literature from Nigeria as Conn's syndrome is said to be rare in Nigerians[7]. Primary hyperaldosteronism is one of the endocrine causes of secondary hypertension and is said to account for 0.05-2.5% of patients with secondary hypertension. However, using the Plasma Aldosterone/Plasma Renin activity ratio as a screening test followed by aldosterone suppression confirmatory testing has resulted in much higher prevalence estimates (513% of all hypertensives) for primary aldosteronism[8]. Rarely large adrenocortical carcinomas, adrenal embryologic nest neoplasms within the kidney and ovary secrete aldosterone. Hypokalaemia in a patient with hypertension who is not on diuretics should be investigated for primary aldosteronism.

Conn's syndrome is characterized by increased aldosterone production, suppressed plasma renin activity (PRA), hypertension, hypokalaemia and metabolic alkalosis. The term primary hyperaldosteronism is used to describe Conn's syndrome and other etiologies of primary hypersecretion of aldosterone. These include; bilateral adrenal hyperplasia, idiopathic hyperaldosteronism in which the adrenals appear normal and Glucocorticoid-remediable aldosteronism in which ACTH stimulates Aldosterone production.

Patients with Conn's syndrome present with hypokalaemia which may be severe. However, normokalemia does not exclude primary hyperaldosteronism, but in these patients there is usually a history of low salt intake and salt loading usually unmasks the hypokalaemia[9]. A diagnosis of Conn's syndrome is made with biochemical and imaging studies. A plasma Aldosterone/Renin activity ratio of >20 with a plasma Aldosterone level equal to or more than 15ng/dl is highly suggestive and is the screening test of choice[10]. It has been suggested that captopril administration may optimize the PAC/PRA test[11]. The most commonly used confirmatory test is a 24-hour urine aldosterone level obtained after 3 days of salt loading. The patient can be instructed to maintain a sodium intake of at least 200 mEq/d (one teaspoon of salt 3 times daily) for 3 days. A 24-hour aldosterone excretion rate of greater than 14 mcg (with a concomitant 24-h urine sodium >200 mEq) is diagnostic of primary hyperaldosteronism[8]. Postural studies are cumbersome and not routinely done. The saline infusion test in which 2 litres of isotonic normal saline are infused over 2-4 hours and blood samples for aldosterone collected before and after the infusion can also be done. In patients with essential hypertension, plasma aldosterone reduces while in patients with adenoma or idiopathic hyperaldosteronism, the plasma aldosterone fails to suppress. This distinguishes primary aldosteronism from low-renin essential hypertension.

Abdominal CT scan is the imaging procedure of choice. Overall, CT scanning has a sensitivity of 67-85% in patients with primary hyperaldosteronism[12]. Adrenal venous sampling following cosyntropin stimulation is used in patients with equivocal CT scan findings.

The drug of choice is spironolactone in doses of up to 400mg/day. Hypokalaemia tends to correct with adequate doses of spironolactone and potassium supplementation may

not be required. Eplerenone is a newer mineralocorticoid antagonist with less anti-androgen and anti-progesterone effects and may be preferable. Amiloride, a potassium-sparing diuretic may be used in patients who are intolerant of mineralocorticoid antagonists but it is not very effective and it lacks the mineralocorticoid receptor antagonist benefits. Unilateral laparoscopic adrenalectomy is the surgical procedure of choice in patients with Conn's syndrome[13] and its long-term cure rates average 69% (range from 30-60%)[15]. In fact, when a solitary unilateral macroadenoma (<1 cm) and normal contralateral adrenal morphology are found on CT in a young patient (<40 yr old) with primary aldosteronism, unilateral adrenalectomy is a reasonable therapeutic option[15].

In our patient, we were unable to assay the Aldosterone and Renin levels as our laboratories did not have the capacity to do so. However, the presence of severe hypokalaemia and hypertension in the absence of diuretic therapy and with the presence of a unilateral adenoma strongly suggests the diagnosis of Conn's syndrome. In addition, the patient has done well on spironolactone therapy and is being prepared for surgery.

CONCLUSION

This case points out the need for a high index of suspicion and proper investigation of young hypertensive patients for secondary causes of hypertension. This is even more so when they have unusual clinical presentations.

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