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Hypokalemic Periodic Paralysis: A Rare Presenting Manifestation of Conn's Syndrome

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Abstract

Primary aldosteronism (Conn's syndrome) is the most common secondary form of hypertension. Here we report a case of rare presenting manifestation of primary aldosteronism where disease came to clinical picture when patient presented to endocrine department with hypokalemic periodic paralysis.

Keywords: Hyperaldosteronism; Adenoma; Hypokalemic paralysis; Hypertension

Introduction

Hypokalemic periodic paralysis is a rare disorder characterized by transient attacks of flaccid paralysis of varying intensity and duration [1]. The condition has the potential to be life-threatening. Early detection and rapid diagnosis are crucial, as some of the underlying causes are correctable. Although mostly familial in etiology, several sporadic cases of different etiologies have been reported, including rare cause like primary hyperaldosteronism (PA). This article reports the case of a middle aged man presented as quadriparesis due to hypokalemia as a result of PA.

Case Report

A 45-year-old male presented in emergency department with chief complaint of weakness of all four limbs from last 3 days. He had hypertension since last 5 years and irregularly on treatment. Weakness first appeared in bilateral lower limbs, symmetrical in onset and in next 6 - 8 h progressively involved upper limbs. He had no respiratory or swallowing difficulty and was able to move his neck and facial muscles. He had no history of similar episode in past. He denied any history of upper respiratory tract infection and diarrhea.

On physical exam, the patient's pulse rate was 104/min, regular and blood pressure was 200/110. No jugular venous distension, goitre or lymphadenopathy were appreciated. Cardiac examination revealed tachycardia with a regular rhythm and no murmurs. Examinations of the chest and abdomen were unremarkable. There were no deformities or edema of the extremities and distal pulses were present and equal bilaterally. Neurologic exam revealed flaccid paralysis of all extremities which involved the proximal and distal muscles and included the hips and shoulders. Powers in lower limbs and upper limbs were 0/5 and 1/5 respectively. Sensation was intact but deep tendon reflexes were absent. Cranial nerve function was grossly intact. He denied any bowel, bladder involvement.

Investigations revealed low potassium levels (1.3 mEq/L) and normal renal function, liver function and thyroid function test. ECG finding revealed flattening of T wave with prolongation of PR interval and QRS duration suggestive of hypokalemia. Arterial blood gas analysis (ABG) showed metabolic alkalosis (pH: 7.55, HCO_3: 32, CO_2: 37.5) and urinary chloride level was more than 20 mEq/L. Patient was managed on the line of hypokalemic periodic paralysis with potassium supplementation. USG for abdomen revealed no adrenal mass. Overnight dexamethasone suppression test (ONDST) was performed to rule out any possibility of Cushing's syndrome and serum cortisol level was found to be 1.43 µg/dL (normal < 1.8 µg/dL).

In view of hypertension, hypokalemia, metabolic alkalosis and normal ONDST, serum aldosterone and plasma renin activity were measured after normalizing plasma potassium level. Plasma aldosterone concentration (PAC) and plasma renin activity (PRA) were 55 ng/dL (N < 16 ng/dL) and 0.68 ng/mL/h (0.7 - 3.3 ng/mL/h), respectively and PAC/PRA ratio of 80.88 was compatible with the diagnosis of PA. In view of elevated PAC/PRA ratio, aldosterone suppression test with saline infusion test was done. Plasma aldosterone was found to be non-suppressible with level of 69 ng/dL (N < 5 ng/dL) after saline infusion test. To find out its cause, CECT of abdomen was performed. CECT of abdomen showed a well-defined hypodense lesion (-10 HU to -20 HU) of size 1.3 × 1.1 cm in lateral limb of left adrenal gland and on contrast mild en-

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hancement (up to 20 HU) was present, suggestive of adrenal adenoma and a final diagnosis of Conn's syndrome with a rare presentation of quadriparesis due to left adrenal adenoma was made.

Patient was managed pre-operatively with spironolactone 100 mg and amlodipine 20 mg. Patient responded very well to treatment and underwent total left adrenalectomy. Histopathological findings were consistent with diagnosis of adrenal adenoma. Postoperatively patient's BP and serum K^+ levels were well controlled without any drug. Now patient is doing well and drug free.

Discussion

Hypokalemic periodic paralysis is a rare disorder with an estimated prevalence of 1 in 100,000 and is characterized by transient attacks of flaccid paralysis of varying intensity and duration which if not recognized in time has the potential to be life-threatening [1]. It is classified as primary and secondary hypokalemic periodic paralysis. The primary form is usually familial in origin with autosomal dominant mode of transmission, whereas secondary form which is much rarer is caused by different etiologies and several such sporadic cases had been reported in the literature. The various causes of secondary form include gastrointestinal potassium wasting disorders, licorice ingestion, barium poisoning, renal tubular acidosis, thyrotoxicosis and PA [2]. The secondary hypokalemic periodic paralysis is less common and the clues indicating a secondary cause include the lack of family history and the time of onset of symptoms. Patients who have their first attack of hypokalemic periodic paralysis in adulthood should be screened carefully for a secondary cause. In the present case, there was no family history of such illness and patient had first episode at age of 45 years making us to have strong suspicion of secondary cause for hypokalemic periodic paralysis. The patient history is not suggestive of any gastrointestinal, renal or thyroid disease and his renal function, liver function and thyroid function tests are within normal limit. Apart from hypokalemia, patient's ABG suggests metabolic alkalosis. The differential diagnosis of hypokalemia with alkalosis includes Conn's syndrome, i.e. PA, Bartter, Gitelmanand Liddle syndrome, syndrome of apparent mineralocorticoid excess, glucocorticoid remediable aldosteronism, hypomagnesemia, cystic fibrosis and diuretic use [3]. The presence of urinary chloride level > 10 mEq/L in present case ruled out extra renal causes of potassium wasting like cystic fibrosis while presence of hypertension rules out other causes like hypomagnesemia, Bartter syndrome and Gitelman syndrome.

In our case, patient presented with history of sudden onset flaccid quadriparesis and on clinical examination had blood pressure of 200/110 mm Hg with hypokalemia and metabolic alkalosis. Patient was initially managed with potassium supplementation and was given antihypertensive drugs for control of blood pressure. His weakness completely improved over a period of 24 - 36 h and on the basis of clinical triad of hypertension, hypokalemia and metabolic alkalosis, a presumptive diagnosis of PA was made and screened by measuring plasma aldosterone and plasma rennin activity ratio which was found to be high. Although plasma renin activity in this patient is very low, high ratio of aldosterone to rennin activity alone cannot ensure the diagnosis of primary aldosteronism and it has to be confirmed by aldosterone suppression test. The aldosterone suppression test is done by giving parenteral infusion of isotonic saline and aldosterone was found to be non-suppressible in the present case. Further the etiology of Conn's syndrome is confirmed by presence of left adrenal adenoma on CECT of abdomen.

PA or Conn's syndrome is characterized by suppressed plasma renin activity, elevated and non-suppressible plasma aldosterone and hypokalemia [4]. The primary function of aldosterone is regulation of extracellular volume and control of potassium homeostasis and inappropriate excess of it will result in hypervolemia state by the increasing sodium reabsorption at expense of potassium loss and this increased sodium reabsorption will suppress the renin production which results in lowering of plasma renin activity. Hypertension in primary aldosteronism is mostly due to persistent hypervolemia [5]. The various recognized etiologies for PA include unilateral aldosterone producing adenoma (APA), bilateral hyperplasia aldosteronism (BHA), adrenocortical carcinoma, and glucocorticoid-remediable aldosteronism (GRA) [5]. Out of these, solitary aldosterone producing adenoma is the most common cause of primary aldosteronism and accounts for approximately 65% of the cases. Conn's syndrome presentation is highly variable and the diagnosis of primary aldosteronism is usually made in patients who are in the third to sixth decade of life and having clinically hypertension with hypokalemia and metabolic alkalosis. High blood pressure is often the only sign and it is usually refractory in nature. Hypokalemia is not as common as we would expect in PA patients. Kuo et al [6] found that the plasma level of potassium was < 3.3 mEq/L in only 18% of all patients with confirmed PA.

Imaging study for the differential diagnosis of a unilateral adenoma or bilateral hyperplasia includes a CT scan or MRI. The sensitivity of CT to distinguish solitary APA and BHA has been reported at 85% or higher [7]. In our case, the CECT of abdomen revealed a well-defined mass over the left adrenal gland and normal appearance of the right adrenal gland. However, if the CT scan or MRI is normal, or if bilateral nodular hyperplasia cannot be ruled out, adrenal vein aldosterone sampling may be considered as a next step but it was not required in our patient. Before surgery patient switched on to aldosterone antagonist spironolactone 50 mg twice a day for control of blood pressure and he responded very well. Patient underwent total left adrenalectomy and histopathological report was suggestive of adrenal cortical adenoma.

Apart from thyrotoxicosis, Conn's syndrome is causing hypokalemic periodic paralysis; however, hypokalemic periodic paralysis as a presenting manifestation of Conn's syndrome is rarely reported in the literature. Although hypokalemia responded very well to surgical treatment, good number of patients required antihypertensive medications although less in number and dosage for control of blood pressure in postoperative period. Persistent hypertension after adrenalectomy is correlated directly with older age, increased serum creatinine level, use of more than two drugs in preoperative period, and duration of hypertension [8]. In the present case after operation on the same day in the evening, patient blood pressure and

potassium levels were returned back to normal and metabolic alkalosis was also subside. At present, patient blood pressure and serum levels are well controlled and he is totally drugs free. Patients who are not willing for surgery or where solitary adenoma cannot be recognized preoperatively can be managed with the help of aldosterone antagonist like spironolactone or eplerenone.

To conclude with a high clinical index of suspicion of Conn's syndrome should be kept in every hypertensive and hypokalemic patients to make its early diagnosis. More importantly, the detrimental effect brought about by aldosterone in multiple tissues may go far beyond a pure complication from hypertension, and early treatment, surgically (adrenalectomy) or medically (spironolactone) will effectively relieve these adverse events and potentially prevent permanent end organ damage.

Disclosure

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