Case Report

Conn Syndrome –A Rare Entity Presenting As Periodic Paralysis – An Even Rarer Presentation.

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Abstract:

Conn's syndrome presents with hypertension with hypokalemia. Hypokalemic Periodic Paralysis is another rare disease with genetic defect resulting in loss of function mutation in SCN4A sodium channel in muscle(1). Between the attacks of weakness , potassium values are normal in patients in hypokalemic periodic paralysis but remain low in secondary hypokalemia.(2) Here we report a case of primary aldosteronism where disease presented with hypokalemic periodic paralysis.

Keywords:

Hypokalemic periodic paralysis, conn syndrome , primary hyperaldosteronism, adnenal adenoma .

Introduction

Hypokalemic periodic paralysis is a rare disorder characterized by transient attacks of flaccid paralysis of varying intensity and duration with normal consciousness.(3) The condition can be potentially life-threatening hence early detection and rapid diagnosis are crucial. It usually occur in second decade of life. This article reports the case of a 38 year oldwoman who presented as quadriparesis due to hypokalemia as a result of Primary hyperaldosteronism.

Case Report

A 38-year-old female presented in emergency department with chief complaint of weakness of all four limbs of 2 days duration. She had history of hypertension for 2 years and was on irregular treatment for the same. Weakness first appeared in bilateral lower limbs, was symmetrical in onset and in next four to five hours it progressively involved the both upper limbs. She had no respiratory or swallowing difficulty and was able to move her neck and facial muscles. She had no history of such an episode in past. There was no history of upper respiratory tract infection or diarrhea or vigorous exercise or heavy carbohydrate meal.

On physical examination, the patient's pulse rate was 96/min, regular and blood pressure was 180/104. There was no jugular venous distension, goitre or lymphadenopathy. Cardiac examination revealed regular rhythm and no murmurs. Examination of the chest and abdomen was unremarkable. There were no deformities or edema of the extremities and distal pulses were present and equal bilaterally. Neurologic examination revealed flaccid paralysis of all extremities which involved both proximal and distal muscles. Powers in all limbs was 0/5 and there was areflexia of all deep tendon reflexes. All sensations –posterior column and spinothalamic tracts were intact and there were no cerebellar features. Cranial nerve function was grossly intact. There was no history of bowel or bladder involvement.

Investigations revealed dangerously low potassium levels (1.2mEq/L) with normal renal function, liver function and thyroid function tests. ECG revealed flattening of T wave with prolongation of PR interval and QRS duration suggestive of hypokalemia. Patient was managed with potassium supplementation and her weakness improved dramatically over 12 hrs.

In view of co existent stage 2 hypertension with hypokalemia, suspicion of patient having hyperaldosteronism was kept .As per literature,

commonest cause of hypertension with hypokalemia is adrenal adenoma.CECT of whole abdomen was performed which showed a well-defined hypodense lesion (MAV ~6 HU) of size 2.3cm(AP) × 1.4cm(T) ×1.9cm(CC) in right adrenal gland, suggestive of adrenal adenoma and a final diagnosis of Conn's syndrome with a rare presentation of quadri paresis due to right adrenal adenoma was made.

Discussion

Hypokalemic periodic paralysis is classified as primary and secondary; primaryhyokalemic periodic paralysis presents in teenage and adolescence whereas secondary presents in adulthood.Serum Potassium levels in Primary form rarely go less than 2.0 meq/l. The various causes of secondary form include gastrointestinal potassium wasting disorders, licorice ingestion, barium poisoning, renal tubular acidosis, thyrotoxicosis and primary hyperaldosteronism(4).Patients with first attack of hypokalemic periodic paralysis in adulthood should be screened carefully for a secondary cause.In Conn's syndrome, the hyperaldosteronism promotes intracellular potassium shifting by increasing the activity of Na/K-adenosine triphosphatase (ATPase) pump in skeletal muscle [5]. In the present case, there was no family history of such illness and patient had first episode at age of 38 years making us to have strong suspicion of secondary cause for hypokalemic periodic paralysis.

Etiology of Conn's syndrome was confirmed by presence of right adrenal adenoma on CECT of abdomen.

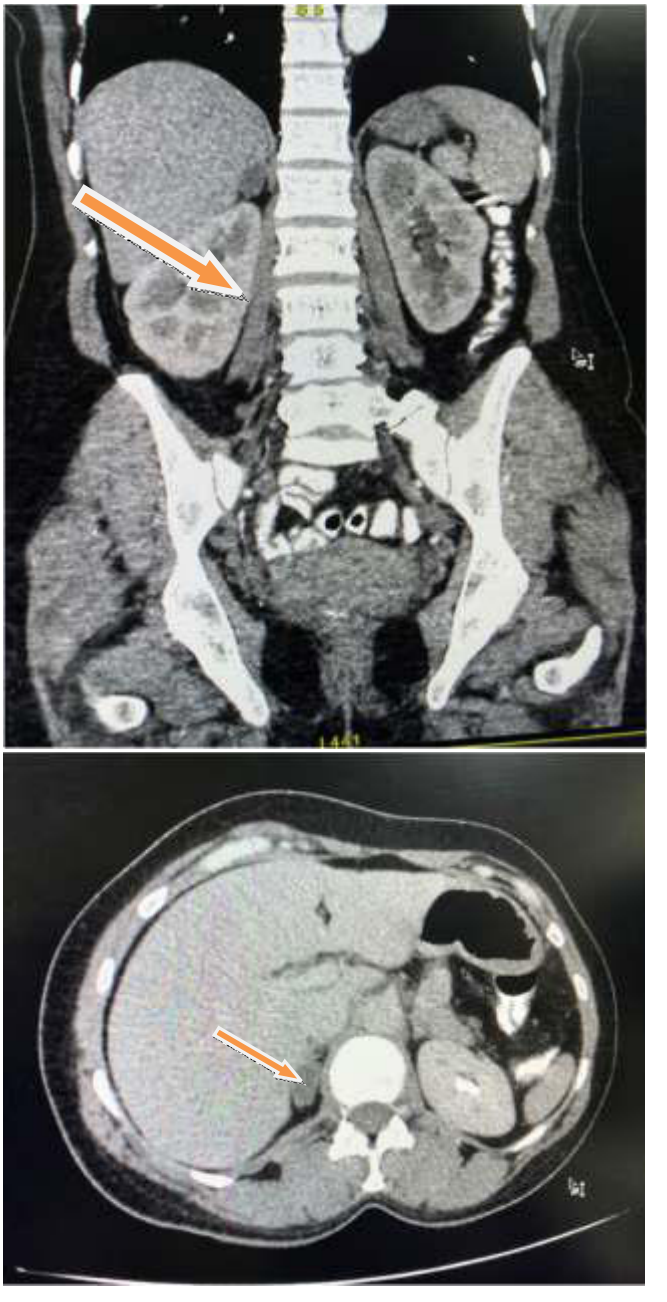
Hypokalemic periodic paralysis as a presenting manifestation of Conn's syndrome is extremely rare with only few cases reported in the literature.

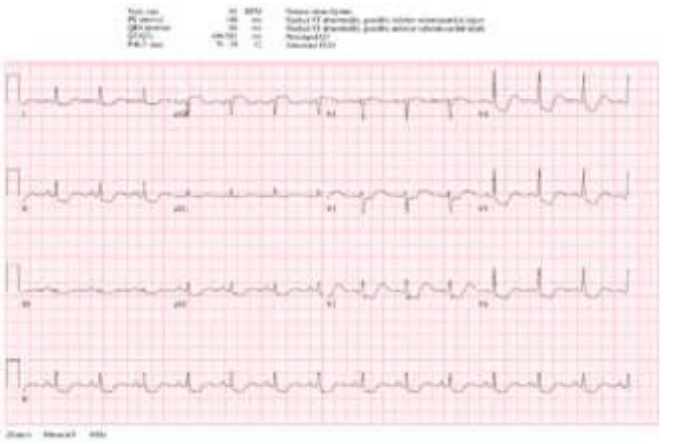
Conn's syndrome is characterized by suppressed plasma renin activity, elevated and non-suppressible plasma aldosterone and hypokalemia(6). Hypertension in primary aldosteronism is mostly due to persistent hypervolemia(7).

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(aldosterone producing adenoma) and BHA(Bilateral hyperaldosteronism) has been reported at 85% or higher[8]. In our case, the CECT of abdomen revealed a well-defined mass over the right adrenal gland and normal appearance of the left 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.Since patient refused to undergo any surgery,patient is on regular follow up and is being managed with spironolactone and her blood pressure is being maintained at 130/80 with normal potassium levels, the latest levels being 3.8mEq/L.

Conclusion:

High clinical index of suspicion of Conn's syndrome should be kept in every hypertensive and hypokalemic patients to make its early diagnosis. Since aldosterone is detrimental to multiple tissues, an early treatment, surgically (adrenalectomy) or medically (spironolactone) will effectively relieve these adverse events and potentially prevent permanent end organ damage.





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