Hypertensive encephalopathy- Atypical presentation of Liddle's syndrome in an elderly male

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Abstract
Liddle's syndrome is a rare cause of secondary hypertension. Identification of this disorder is important as treatment differs from other forms of hypertension. We report a case of a 68 year old male who presented with acute confusion state and hypertensive urgency with features of encephalopathy. Investigations confirmed Liddle's syndrome.

Keywords: Hypertension, Confusion, Encephalopathy.

Introduction
When hypertension is associated with hypokalemia, the investigations are very specifically addressed to rule out secondary hyperyension. Liddle's syndrome is one of them. It is associated with hypertension, hypokalemia, metabolic alkalosis and hyporeninemic hypoaldosteronism.[1,2] Treatment is usually with a potassium sparing diuretic along with control of hypertension.

Case Report
A 68 year old male was brought by relatives to our hospital with the complaints of irrelevant talk and confusion since I day. He was a known case of Hypertension since 3 years on irregular medications. There was no history of weakness, headache, vomiting, seizures, abnormal speech, chest pain, fever. He was non diabetic.

On evaluation, his BP was 220/120 mm of Hg. The rest of his general and systemic examination was normal. Central nervous system examination revealed confusional state. There was no speech abnormality. Pupils were bilaterally normal size and were reacting to light. She was moving all 4 limbs. Her deep tendon reflexes were normal and bilateral plantars were flexor.

On investigations; Complete blood count was normal. His serum potassium was 1.8 mEq/L (hypokalemia). Twenty four hour urine potassium was 38 mEq/L.The arterial blood gas revealed a pH of 7.543, HCO 3 = 28 mEq/L. Serum creatinine (1.1 mg %) and sodium (139 mEq/L) were within normal limits. Chest X ray was normal. ECG showed left ventricular hypertrophy. Fundus examination revealed hypertensive retinopathy changes in form of exudates and hemorrhages without papilledema. Serum cortisol levels were within normal limits.In view of metabolic alkalosis, hypertension and hypokalemia, further investigations were advised. His serum aldosterone level was <9.2 pg/ml and plasma renin was < 0.01 ng/ml/hr. Computed tomography (CT) of brain and abdomen was normal.

In view of hypokalemia, metabolic alkalosis, hypertension and associated hyporeninemic hypoaldosteronism, a diagnosis of Liddle's syndrome was made.

He was treated with iv bolus Labetolol 10 mg over 2 mins, which was again repeated after 10 mins. The BP came down to 178/110 in half hour. Then he was started on tablet Labetolol 50 mg twice a day, tablet amlodipine twice a day, and spironolactone 25 mg twice daily. His blood pressure was controlled to 140/88 mm of Hg in 48 hours and his mentation was normal. He was discharged with the above per oral medications.

During 2 months follow up, his serum potassium improved to about 4.5 mmol/L, blood pressure was 138/88 mm of Hg. Serum creatinine was 1.2 mg%.

Discussion
The differential diagnosis of hypertension with associated hypokalemia are; hyperaldosteronism, diuretic therapy, Cushing's syndrome, licorice ingestion, Liddle's syndrome and rare renin secreting tumors.[1,2] Our patient had no history of diuretic therapy or licorice ingestion. The serum cortisol levels were normal so, the possibility of Cushing's syndrome was ruled out. CT abdomen was negative for adrenal enlargement or masses and our patient had low renin and aldosterone levels ruling out Conn's syndrome.

Liddle's syndrome is an autosomal dominant condition characterized by primary increase reabsorption of sodium and potassium in the collecting tubules of the nephron.[2,3] These patients have low renin and aldosterone levels and there is conservation of sodium and excretion of potassium in the absence of mineralocorticoid excess. Genetic studies have revealed that the basic cause of this syndrome is due to mutations affecting cytosolic tail of the ß subunit of the epithelial sodium channel (ENaC).[3-5] These mutations also cause activation of the epithelial sodium channel of the luminal membrane of the collecting tubules that leads to excessive absorption of sodium leading and volume expansion. This resultant increase sodium absorption and volume expansion causes hypertension with inhibition of renin and aldosterone secretion.

Liddle's syndrome presents with hypertension, hypokalemia and metabolic alkalosis, akin to that seen in

mineralocorticoid excess.[3,4] The usual presentation of cases are mostly in younger individuals which triggers the evaluation of secondary hypertension. Presentation of Liddle's syndrome in elderly beyond 6th decade is rare as in our case.Our patient presented with hypertensive urgency which is still rarer.[6,7] In our case spironolactone was chosen because amiloride was not available as a single agent, it is usually available in India with combination of thiazide diuretics, which may precipitate hypokalemia.

To conclude; It is important to screen for Liddle's syndrome in patients with hypertension, hypokalemia and metabolic alkalosis, as the treatment of this condition differs from other forms of essential or secondary hypertension.[2,3] Potassium-sparing diuretics like amiloride and triamterene which directly close the sodium channels are effective in Liddle's syndrome whereas mineralocorticoid antagonist spironolactone is ineffective since the increase in sodium channel activity is not mediated by aldosterone in this disorder.

Conflict of Interest: None.

References

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How to cite this article: Acharya S, Shukla S, Gagneja S. Hypertensive encephalopathy- Atypical presentation of Liddle's syndrome in an elderly male. *Ann Geriatrics Educ Med Sci* 2019;6(1):18-9.