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Review Article

A CASE OF PRIMARY ALDOSTERONISM WITH REFRACTORY HYPOKALEMIA AS THE MAIN MANIFESTATION AND LITERATURE REVIEW

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ABSTRACT

Primary Aldosteronism is a disease with clinical manifestations such as hypertension and hypokalemia due to the excessive secretion of aldosterone by the adrenal cortex. The most common cause is adrenocortical adenoma., which is rare in children. The earliest symptom of primary aldosteronism is generally hypertension, and severe hypokalemia is the main manifestation. This case reports a case of primary aldosteronism caused by adrenal adenoma, including the pathogenesis, clinical manifestation, laboratory examination and treatment of severe hypokalemia and metabolic alkalosis, so as to provide reference for clinical practice.

Keywords: Primary aldosteronism ; Severe hypokalemia ; Metabolic alkalosis ; Adrenal adenoma

CASE REPORT

A 12-year-old child, without any significant past history, was hospitalized with weakness. A year before admission, there was no obvious inducement to feel weakness, but still stand and engage in light labor. The attack occurred once every half a month, and each attack lasted about 4-5 days, could be recovered after rest. Three days later, general weakness appeared again, the degree was worse than before, and it was impossible to walk, polydipsia, polyuria,nocturia. At presentation her temperature-36.2 ℃, pulse rate-84 beats/per min, respiratory rate-24 breaths/min, blood pressure-122/78 mmHg, weight-44kg. The muscle strength of the limbs was grade IV, the muscle tension was normal, tendon reflexes were not elicited, and pathological reflexes were negative.

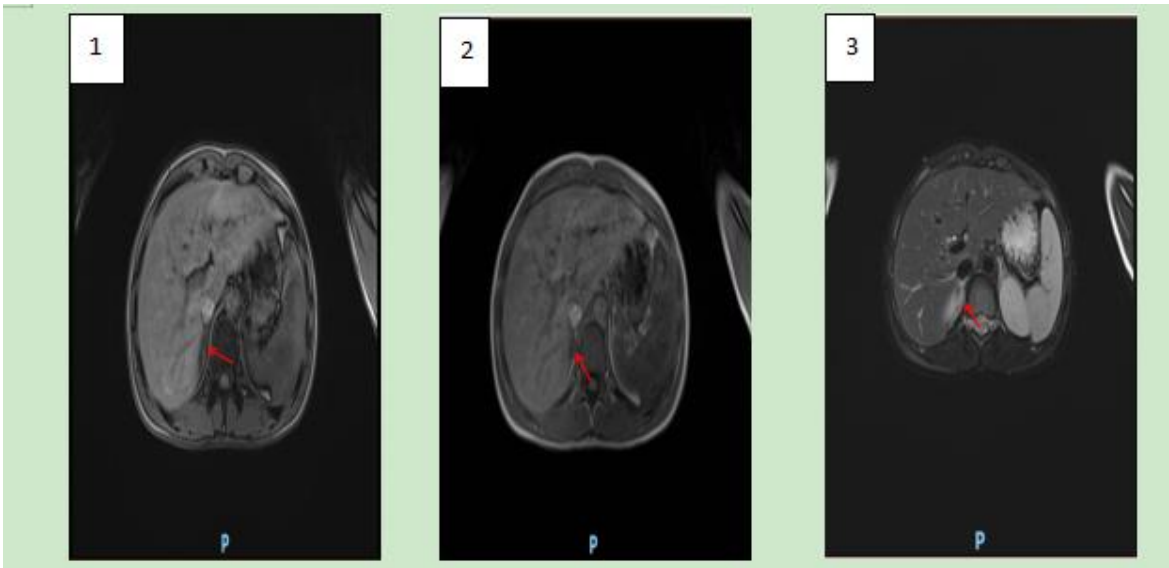
Her investigation profile showed that Electrolyte: K 2.15 mmol/L↓, Na 142.3 mmol /L; ECG: QT interval prolongation, U wave change; Endocrine-related tests: aldosterone 234.40pg/ml ↑; Thyroid function: TSH 2.2866μIU/mL, TG 446.57IU/ ml↑, TPO 181.64IU/ml↑; cortex Alcohol 6.80μg/dl; three items of bone metabolism: total parathyroid hormone (iPTH) 239.5pg/ml↑, serum calcitonin (CT) <2.000pg/ml, 25-hydroxyvitamin D 6.89ng/ml↓. Hypertension 4 items (standing): Angiotensin I (4°C) 1.5100ng/ml/h, Angiotensin I (37°C) 1.6300ng/ml/h, Angiotensin II 72.1700. Hypertension 4 items (recumbent position): Angiotensin I (4°C) 1.3130ng/ml/h, Angiotensin I (37°C) 1.3830ng/ml/h, Angiotensin II 64.6500. 24h urine potassium 67.91mmol/24h (potassium 3.57mmol/L in the same period). Adrenal gland MRI examination: The right adrenal gland was thickened and partially oval, with a size of about 13x8mm, T1 showed low signal, no obvious abnormal changes in positive and negative phase signals, and T2 showed slightly high signal shadow with clear boundary; the shape, size and signal of the left adrenal gland showed no obvious abnormality. Right adrenal nodule, adenoma? . The head MRI showed no obvious abnormality; the pituitary gland was plump (Figure 1).

Diagnosis:

Primary aldosteronism, Adrenal adenoma, Severe hypokalemia, Arrhythmia, Metabolic alkalosis, Vitamin D deficiency tetany.

Treatment:

After admission, potassium chloride was taken orally and intravenously to supplement potassium, Calcium gluconate and potassium magnesium aspartate were taken orally and intravenously to supplement magnesium. Spironolactone inhibited aldosterone secretion, electrolytes returned to normal, tetany symptoms disappeared, and then the child was transferred to the urological department for surgical treatment. At present, the children have completed adrenalectomy. Postoperative pathological diagnosis considered right adrenal cortical adenoma. Postoperative monitoring showed that blood potassium was normal, blood pressure was normal, and the re examination of aldosterone and renin levels was basically normal.



Figures: 1, 2 - MRI of the right adrenal gland showed low signal in T1 positive and negative phase; 3 - MRI T2 image of adrenal gland shows slightly high signal shadow with clear boundary, about 13x8mm in size

DISCUSSION

Primary aldosteronism is considered to be the most common cause of endocrine hypertension, accounting for 6% in the etiological diagnosis of hypertension[1], and can be cured. Primary aldosteronism is characterized by a low plasma renin level and a high plasma aldosterone level, which is marked by the autonomic secretion of aldosterone by one or two adrenal glands, rather than being regulated by angiotensin II, hyperkalemia and adrenocorticotropic hormone (ACTH) [2-3].

Normal physiological conditions, the secretion of aldosterone is regulated by three main factors: angiotensin II, extracellular hyperkalemia and ACTH. The decrease of blood volume and glomerular filtration rate is perceived by reducing the sodium and chloride transported to the compact spot, which can make the paraglomerular cells release renin, which catalyzes the decomposition of angiotensinogen into angiotensin I, and angiotensin converting enzyme further converts it into angiotensin II, which is an important stable peptide of blood pressure and blood volume, and can stimulate the synthesis and secretion of adrenal aldosterone. Therefore, the aldosteronism caused by the increase of intravascular volume is a physiological "renin dependent aldosteronism", and its role is to expand the intravascular volume and maintain blood pressure homeostasis [4-5].

When aldosterone binds to the mineralocorticoid receptor of the distal nephron main cell, it induces sodium reabsorption through epithelial sodium channel (ENaC), and excretes potassium ions or hydrogen ions accordingly. ENaC mediated sodium reabsorption induces osmotic pressure changes, thus driving water reabsorption, leading to tissue edema, increased glomerular filtration rate, and inhibition of renin and angiotensin II. Since angiotensin II is an important medium for sodium reabsorption in the proximal nephron,

inhibition of angiotensin II will lead to more sodium delivery to the distal nephron, thereby amplifying the sodium reabsorption and tissue edema driven by aldosterone, as well as the excretion of potassium and acid. When one or two adrenal glands secrete aldosterone autonomously, clinical manifestations such as hypertension, hypokalemia and metabolic alkalosis will eventually occur through a series of renal and hemodynamic effects [6-7].

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The diagnosis of primary aldosteronism mainly depends on screening tests, namely the aldosterone to renin ratio (ARR). The definition of positive screening is that ARR is>30 ng/dL/ng/mL/h, and the serum aldosterone level is>15 ng/dL [8-10]. The higher the aldosterone level, the lower the plasma renin activity, and the more obvious the diagnosis. In addition, it is also necessary to determine whether there is a primary lesion, such as unilateral or bilateral adrenal diseases. For patients diagnosed as primary aldosteronism, surgical resection can cure hyperaldosteronism and hypokalemia, and cure about 60% of hypertension[11]. For patients with bilateral diseases, spironolactone therapy can reduce blood pressure and aldosterone secretion [12-14].

To sum up, in this case report, the patient was admitted to the hospital due to severe hypokalemia and metabolic alkalosis. Through a series of examinations, we found that the level of aldosterone in the patient was on the high side. Later, we improved the examination of renin, angiotensin, and aldosterone system related hormones. We found that the patient's ARR was up to 575.6 ng/dL/ng/mL/h in upright position and 648.2857 ng/dL/ng/mL/h in supine position, which was far greater than the positive screening criteria, Combined with adrenal MRI, it indicates right adrenal nodule and adenoma?, We finally considered a series of clinical manifestations such as severe hypokalemia and metabolic acidosis caused by primary aldosteronism caused by adrenal adenoma. The patient was a unilateral adrenal nodule, which was diagnosed as adenoma by surgical resection and pathological examination. After surgery, the clinical symptoms of the child disappeared, the hypokalemia was corrected, and the aldosterone level returned to normal, which in turn confirmed the diagnosis. Therefore, the clinical manifestations of hypokalemia and metabolic alkalosis should consider the possibility of primary aldehyde disease. Proaldosis is mostly seen in adults, and rarely in children. The main manifestation of Proaldosis is hypertension. In clinical practice, cases with severe

hypokalemia, especially in children, are very rare. We report this case for your reference.

REFERENCES

1. Monticone S, Burrello J, Tizzani D, Bertello C, Viola A, Buffolo F, Gabetti L, Mengozzi G, Williams TA, Rabbia F, Veglio F, Mulatero P. Prevalence and Clinical Manifestations of Primary Aldosteronism Encountered in Primary Care Practice. J Am Coll Cardiol. 2017 Apr 11;69(14):1811-1820.
2. Hundemer GL, Vaidya A. Primary Aldosteronism Diagnosis and Management: A Clinical Approach. Endocrinol Metab Clin North Am. 2019 Dec;48(4):681-700.
3. Rossi GP. Primary Aldosteronism: JACC State-of-the-Art Review. J Am Coll Cardiol. 2019 Dec 3;74(22):2799-2811.
4. Zennaro MC, Boulkroun S, Fernandes-Rosa FL. Pathogenesis and treatment of primary aldosteronism. Nat Rev Endocrinol. 2020 Oct;16(10):578-589.
5. Vaidya A, Carey RM. Evolution of the Primary Aldosteronism Syndrome: Updating the Approach. J Clin Endocrinol Metab. 2020 Dec 1;105(12):3771–83.
6. Stavropoulos K, Imprialos K, Papademetriou V, Faselis C, Tsioufis K, Dimitriadis K, Doumas M. Primary Aldosteronism: Novel Insights. Curr Hypertens Rev. 2020;16(1):19-23.
7. Pillai PR, Griffith M, Schwarcz MD, Weiss IA. Primary Aldosteronism: Cardiovascular Risk, Diagnosis, and Management. Cardiol Rev. 2020 Mar/Apr;28(2):84-91.
8. Funder JW, Carey RM, Mantero F, Murad MH, Reincke M, Shibata H, Stowasser M, Young WF Jr. The Management of Primary Aldosteronism: Case Detection, Diagnosis, and Treatment: An Endocrine Society Clinical Practice Guideline. J Clin Endocrinol Metab. 2016 May;101(5):1889-916. doi: 10.1210/jc.2015-4061.
9. Hundemer GL, Baudrand R, Brown JM, Curhan G, Williams GH, Vaidya A. Renin Phenotypes Characterize Vascular Disease, Autonomous Aldosteronism, and Mineralocorticoid Receptor Activity. J Clin Endocrinol Metab. 2017 Jun 1;102(6):1835-1843.
10. Mulatero P, Stowasser M, Loh KC, Fardella CE, Gordon RD, Mosso L, Gomez-Sanchez CE, Veglio F, Young WF Jr. Increased diagnosis of primary aldosteronism, including surgically correctable forms, in centers from five continents. J Clin Endocrinol Metab. 2004 Mar;89(3):1045-50.
11. Carey RM. Primary aldosteronism. J Surg Oncol. 2012 Oct 1;106(5):575-9.
12. Institute of Medicine, National Academy of Sciences Standards for developing trustworthy clinical practice guidelines. Accessed January 8, 2015， Updated August 19, 2015
13. Steichen O. Hypertension cure rate after adrenalectomy for unilateral primary aldosteronism. J Surg Oncol. 2013 Apr;107(5):561.
14. Williams TA, Reincke M. MANAGEMENT OF ENDOCRINE DISEASE: Diagnosis and management of primary aldosteronism: the Endocrine Society guideline 2016 revisited. Eur J Endocrinol. 2018 Jul;179(1):R19-R29.