

An Unusual Presentation of Conn's Myopathy: Surgical Remedy For Medical Illness: A Case Report

*Dr. Udipta Ray, Dr. Souvik Saha

Corresponding Author: *Dr. Udipta Ray, Dr. Souvik Saha

Abstract: Acute onset paraparesis, though a very common entity, has a wide variety of etiologies including spinal and electrolyte disorders. One of the unusual causes of Paraparesis includes Primary Hyperaldosteronism (Conn's syndrome) and despite being a medical illness, the myopathy is readily reversible with surgical intervention. We encountered a case of 42-year old lady with hypertension and acute paraparesis with normal MRI Spine. Investigations revealed Primary Hyperaldosteronism due to unilateral Adrenal tumor. Surgical intervention by laparoscopic Adrenalectomy was carried out that relieved her symptoms. Histopathology confirmed Adrenal adenoma.

Keywords: Adrenal adenoma, Conn's Syndrome, Hyperaldosteronism

Date of Submission: 19-08-2017

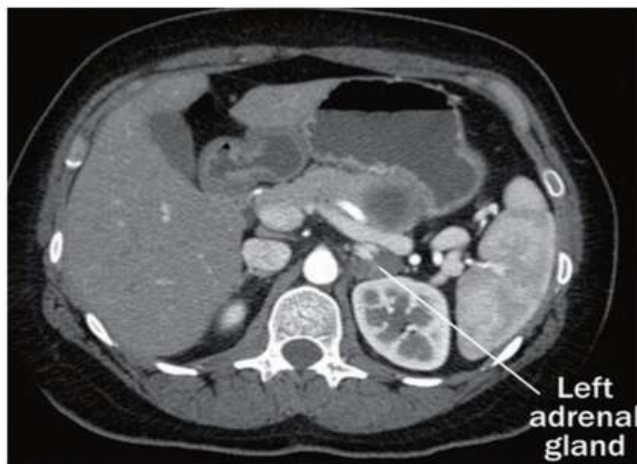
Date of acceptance: 22-09-2017

I. Introduction

Primary hyperaldosteronism is characterized by hypertension, hypokalemia, suppressed renin activity and increased aldosterone excretion and was first described by J. W. Conn in 1955 [1]. Other than resistant hypertension, paralytic myopathy in association with hypokalemia is a recognized yet unusual feature of Conn's syndrome and is seen more commonly in Asian patients [2]. Involvement of proximal muscles is more often reported but distal myopathy can occur as well [3]. Therefore, due to its wide presentations, often it creates a diagnostic dilemma resulting in unnecessary wastage of time and health-cost and undue delay in treatment as well. We report such a case of Conn's syndrome presenting with bilateral paraparesis that is not only atypical but also curable with surgery

II. Case Report

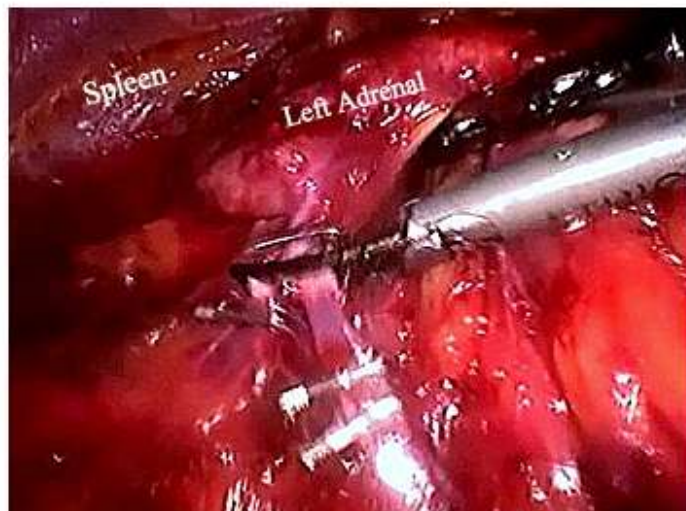
A 42-year old lady, hypertensive for last two years on oral Amlodipine, presented with non-traumatic acute onset gradually progressive weakness of bilateral lower limbs without any bladder or bowel involvement. Examination findings revealed Hypertension and decreased motor function of both lower limbs without any sensory deficit. X-ray and MRI spine found to be normal. Blood investigations revealed hypernatremia and hypokalemia. Furthermore, increased serum Aldosterone and decreased plasma renin activity alarmed a contrast-enhanced CT abdomen which revealed a mass of 4 cm x 5 cm size in left adrenal gland [Figure.1].



CECT showing left adrenal mass

Fig.1: CECT showing left adrenal mass

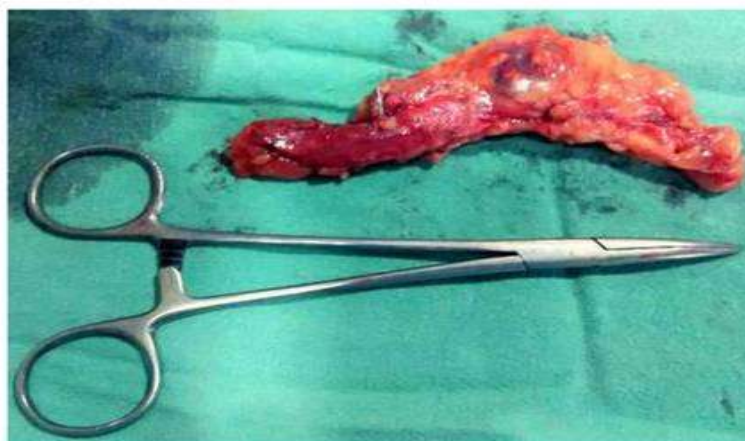
After ruling out pheochromocytoma and hypercortisolemia, she underwent Laparoscopic Left Adrenalectomy [Figure. 2] with proper pre-operative optimization.



Laparoscopic Left Adrenalectomy

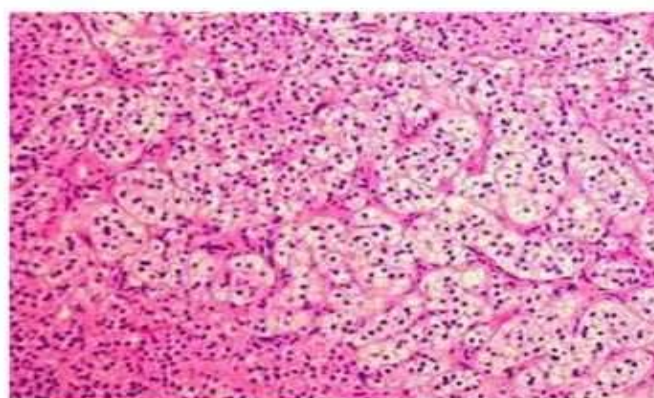
Fig.2: Laparoscopic picture of left adrenal gland

The specimen [Figure. 3] sent for histopathology revealed adrenal adenoma [Figure. 4]



Specimen of Left Adrenal gland

Fig.3: Specimen of excised left adrenal gland



Adrenal Adenoma with Spironolactone bodies

Fig.4: Histopathology of excised specimen showing adrenal adenoma with plenty of Spironolactone bodies

Post-operatively, her serum sodium and potassium was normal. Her paraparesis gradually improved over few weeks and hypertension was well-controlled without any medication over a 1 year follow-up period.

III. Discussion

Primary hyperaldosteronism occurs most commonly due to aldosterone-producing adenoma (Conn's syndrome) or bilateral idiopathic adrenal hyperplasia and less commonly due to primary (unilateral) adrenal hyperplasia, aldosterone-producing adrenocortical carcinoma or familial hyperaldosteronism [4]. It is usually common in females in their 3rd to 5th decades. Hypertension, Hyponatremia and Hypokalemia are its usual presentations but paraparesis or myopathy secondary to hypokalemia can often pose a challenge to diagnosis.

Our patient also presented with an acute onset paraparesis which also called for investigations (i.e. MRI spine, NCV) to exclude spinal or neuro-muscular etiology but hypertension and hypokalemia pointed towards hyperaldosteronism. Her serum aldosterone and urinary aldosterone were also increased along with a decrease in plasma renin activity. So, CECT abdomen was done and a 4x5 cm mass found in left adrenal gland. Therefore in a case of non-traumatic paraparesis, after exclusion of neuro-muscular etiology, blood investigations should be done to exclude primary hyperaldosteronism [5]. Surgical removal of diseased adrenal gland is the preferred treatment for Conn's. Our patient underwent laparoscopic adrenalectomy of left side and histopathology revealed adrenal adenoma. Her post-operative recovery was well with gradual improvement of lower limb weakness over few weeks. She did not need any medications at all to keep her blood pressure normal during this one year follow-up period.

Surgery is the mainstay of treatment of Conn's syndrome even if it may present like a medical entity. Unilateral adrenalectomy is usually associated with improvement in all patients and a cure in 30–60% of hypertensive patients. Correction of hypokalemia and marked reductions in aldosterone levels have been noted after surgery [6]. Resolution of hypertension after adrenalectomy may be independently associated with younger age, lack of family history of hypertension, a short duration of hypertension, use of no more than two antihypertensive medications during the preoperative period, a higher preoperative ratio of plasma aldosterone concentration to PRA, and a higher urine aldosterone level [7].

IV. Conclusion

Non-spinal paraparesis though a medical entity, whenever associated with Hypokalemia and hypertension, alarms for a rare presentation of Conn's syndrome. Serum investigations for hyperaldosteronism should be done in the earliest period along with an abdominal imaging preferably CT scan. After pre-operative optimization, adrenalectomy by laparoscopic approach usually is the intervention of choice for its minimal access and early recovery. Adrenal adenoma, which is the commonest, needs no further intervention unlike its malignant counterpart. Hypertension usually requires no or minimal medication and myopathy gradually improves over time.

References

- [1]. Conn JW: Primary aldosteronism, a new clinical syndrome. *J Lab Clin Med*; 1955;45:3-17.
- [2]. Huang YY, Hsu BR, Tsai JS. Paralytic myopathy: A leading clinical presentation for primary aldosteronism in Taiwan. *J Clin Endocrinol Metab* 1996;81:4038-41.
- [3]. Chi Chui, Wei-Hsi Chen, Hsin-Ling Yin. Periodic Drop Thumb, Hypokalemia and Adrenal Adenoma in *Med Princ Pract* 2014;23:80–82.
- [4]. Hari Kumar K, Jha S, Jha R, Modi KD. Conns' syndrome - atypical presentations. *Saudi J Kidney Dis Transpl*. 2009 Mar;20(2):278-81. PubMed PMID: 19237819.
- [5]. Chuang, T. H., Wang, C. H., Tseng, B. Y., Hsu, Y. H., Tsai, J. P., Hsu, B. G., & Fang, T. C. (2008). Conn's Syndrome with an Unusual Presentation of Rhabdomyolysis Secondary to Severe Hypokalemia. *Tzu Chi Medical Journal*, 20(4), 327-331. DOI: 10.1016/S1016-3190(08)60060-9.
- [6]. W Butt, M Butt, T Gauhar, A Khawaja, A Afzal, A Ahmed, K Azim. Conn's Syndrome: A Diagnostic Dilemma. Case Report. The Internet Journal of Surgery. 2009 Volume 24 Number 1.
- [7]. Sawka AM, Young WF, Thompson GB, et al. Primary aldosteronism: factors associated with normalization of blood pressure after surgery. *Ann Intern Med* 2001;135:258–61.

*Dr. Udipta Ray, Dr. Souvik Saha. "An Unusual Presentation of Conn's Myopathy: Surgical Remedy For Medical Illness: A Case Report." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)* 16.9 (2017): 20-22