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Case report

Schmidt syndrome; Poly Glandular AutoImmune Syndrome type II (PGA II): Presenting as Hyperkalaemic Paralysis

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ABSTRACT

A young women who was a known diabetic presented with recurrent episodes of periodic muscular weakness, which was ascending in nature and was labelled to have hyperkalaemic periodic paralysis. During current presentation patient was fully evaluated for the cause for hyperkalaemia as hyperkalaemic periodic paralysis is very rare. On evaluation patient had multiple endocrine abnormalities probably autoimmune in etiology and after ruling out pseudohypoaldosteronism, patient was diagnosed to have an autoimmune hypothyroidism and Isolated Hyporeninemic Hypoaldosteronism presenting as hyperkalaemic paralysis and was treated with fludrocortisone. Patient improved significantly and there was no recurrence of weakness over a year of follow up.

1. Introduction

A 32 year old lady presented with weakness of both the legs which started in foot and progressed to involve the entire lower limbs over a period of 2 days. There was no history of sensory abnormalities and bowel and bladder involvement. Patient had similar history of muscle weakness which ascended to involve respiratory muscles and was found to have Diabetes Mellitus and refractory hyperkalaemia, patient was subjected to haemodialysis following which she improved significantly and was discharged in a peripheral centre as a probable channalopathy with T2DM (Hyperkalaemic Periodic Paralysis).

Since hyperkalaemic periodic paralysis is very rare and the diagnosis of channalopathy is after the exclusion of other underlying diseases, a thorough clinical and laboratory evaluation was done.

Clinical Examination:

Patient was conscious, oriented with hoarseness of voice. There was no coarse facial feature.

BMI: 23

Pulse rate: 62/minutes, regular, normal in character and all peripheral pulses were equally felt.

B.P: 90/60 mm of Hg

CNS:

- Higher Mental Functions was normal
- Cranial Nerves were intact
- Flaccid Quadriparesis with intact neck muscle power with intact reflexes was present and there was no respiratory insufficiency.
- plantars were bilaterally flexor
- autonomic nervous system was normal
- sensory system was intact.

Other organ systems were normal.

Investigations:

Time	Serum Sodium (meq/L)	Serum Potassium (meq/L)	ECG
At admission	130	8.5	Absent P waves with tall tented peaked T waves
With medical management	132	8.1	Absent P waves with tall tented peaked T waves
After haemodialysis	138	5.5	Normal
3 days after Fludrocortisone therapy	142	5.0	Normal
At discharge	144	4.8	Normal

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Complete blood counts and peripheral smear study was normal.

Blood Urea: 30 mg/dl

Serum Creatinine; 1.0mg/dl

Free T3 = 1.8 pmol/L

Free T4 = 3.04 pmol/L

TSH = 108 mIU/L

Anti thyroid antibodies were positive (>5IU/ml)

Serum calcium = 9.4mg/dl

Serum phosphate = 2.8mg/dl

FSH & LH suggestive of follicular phase 8.0u/L and 10.0u/L respectively.

FBS = 140mg/dl

PPBS = 210mg/dl

HbA1C = 7.5

Anti GAD antibodies >5 lakhs (24000 – 200000 u/ml)

Serum cortisol (8a.m = 10 µg/dl) = normal

Serum aldosterone in upright with salt restriction = 2.1ng/dl (expected 5 fold increase from baseline value i,e. 2-9ng/dl with supine and normal sodium diet)

Serum renin level was 2.0µIU/ml (3.3 - 41µIU/ml)

CT abdomen = normal study.

Diagnosis: After analysing the investigation reports, the cause for hyperkalaemic paralysis was found to be Poly Glandular Autoimmune Syndrome Type II (Schmidt syndrome) which constituted of Latent Autoimmune Diabetes of Adults (LADA), Autoimmune hypothyriodism and Isolated Hyporeninemic Hypoaldosteronism.

Treatment:

Patient was started with medical line of management for hyperkalaemia, there was no significant improvement and since the weakness was progressive and the fall in serum potassium was marginal, patient was considered for an emergency haemodialysis via right internal jugular vein. After a 4 hours of haemodialysis patients weakness started improving significantly and the post haemodialysis serum potassium was 5.5meq/L. ECG changes of hyperkalaemia subsided.

Patient was treated with OHA's, thyroxine supplementation and fludrocortisone.

Patient was observed over a period of one year, blood sugars, TFT and serum electrolytes remained within acceptable limits and there was no recurrent episode of muscle weakness and hyperkalaemia.

Discussion:

There are various causes of hyperkalaemia and hypoaldosteronism remains one of the rarest possible cause.

Hyposecretion of aldosterone is not commonly encountered in our clinical practice. suppress excess ACTH stimulation and hence symptoms.

Hypoaldosteronism is of 2 types:

1. Hypo-reninemic hypoaldosteronism
2. Hyper-reninemic hypoaldosteronism

Hypo-reninemic hypoaldosteronism occurs due to inherited biosynthetic defect of renin secretion, post operative state following removal of aldosterone secreting adenomas, during protracted heparin therapy, due to pre-tectal disease of CNS and in severe postural hypotension. These patients may have hyperkalemia and they show inability to increase aldosterone secretion in response to salt restriction.

Hyper-reninemic hypoaldosteronism is seen in case of adrenal necrosis, prolonged ACTH stimulus, Aldosterone biosynthetic defect or when there is a selective unresponsiveness to Angiotensin II and rarely salt wasting forms of congenital adrenal hyperplasias (21- hydroxylase deficiency).

Mutation in ENa channels can present with features of hypoaldosteronism but the renin and aldosterone levels remain high, this is called as pseudo- hypoaldosteronism.

Before proceeding with evaluation of hypoaldosteronism one should rule out the possibility of pseudohyperkalaemia as a result of hemolysis and thrombocytosis.

After assessing a normal response to ACTH stimulation test, the serum renin and aldosterone levels are estimated with salt restriction. The levels of renin and aldosterone determines the type of hypoaldosteronism.

Treatment of hypoaldosteronism is based on etiology, generally fludrocortisone 0.05 to 0.15mg restores electrolyte balance if salt intake is adequate. In congenital adrenal hyperplasias daily administration of glucocorticoids can

PolyGlandular Autoimmune (PGA) Syndrome

When immune dysfunction affects two or more endocrine glands and other nonendocrine immune disorders are present, the polyglandular autoimmune (PGA) syndromes should be considered. The PGA syndromes are classified as two main types:

The Type I syndrome starts in childhood and is characterized by mucocutaneous candidiasis, hypoparathyroidism, and adrenal insufficiency.

The type II, or Schmidt, syndrome is more likely to present in adults and most commonly includes adrenal insufficiency, thyroiditis, or type 1 diabetes mellitus.

PGA I	PGA II
Epidemiology	
Autosomal recessiveMutations in APECED gene	Polygenic inheritanceHLA-DR3 and HLA-DR4 associated
Childhood onsetEqual male: female ratio	Adult onsetFemale predominance
Disease Associations	
Mucocutaneous candidiasis	Adrenal insufficiency
Hypoparathyroidism	Hypothyroidism
Adrenal insufficiency	Graves' disease
Hypogonadism	Type 1 diabetes
Alopecia	Hypogonadism
Hypothyroidism	Hypophysitis
Dental enamel hypoplasia	Myasthenia gravis
Malabsorption	Vitiligo
Chronic active hepatitis	Alopecia
Vitiligo	Pernicious anemia
Pernicious anemia	Celiac disease

Abbreviation: APECED, autoimmune polyendocrinopathy-candidiasis-ectodermal dystrophy.

PGA type I usually is recognized in the first decade of life and requires two of three components for diagnosis: mucocutaneous candidiasis, hypoparathyroidism, and adrenal insufficiency.

PGA type II is characterized by two or more of the endocrinopathies listed in Table. Most often these endocrinopathies include primary adrenal insufficiency, Graves' disease or autoimmune hypothyroidism, type 1 diabetes mellitus, and primary hypogonadism. Because adrenal insufficiency is relatively rare, it is used frequently to define the presence of the syndrome. Among patients with adrenal insufficiency, type 1 diabetes mellitus coexists in 52% and autoimmune thyroid disease occurs in 69%.

The clinical manifestations of adrenal insufficiency often develop slowly, may be difficult to detect, and can be fatal if not diagnosed and treated appropriately. Thus, prospective screening should be performed routinely in all patients and family members at risk for PGA types I and II. The most effective screening test for adrenal disease is a cosyntropin stimulation test. A fasting blood glucose level can be obtained to screen for hyperglycemia. Additional screening tests should include measurements of TSH, luteinizing hormone, follicle-stimulating hormone, and, in men, testosterone levels.

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