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REFRACTORY SEIZURES AND CHOREA IN A CASE OF HYPOPARATHYROIDISM RESPONDING TO PARATHORMONE (PTH) SUPPLEMENTATION

Neurology

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ABSTRACT

Hypoparathyroidism is decreased function of the parathyroid glands with underproduction of parathyroid hormone. The condition can be inherited, but it is also encountered after thyroid or parathyroid gland surgery, and it can be caused by immune system related damage as well as by a number of rarer causes. The main symptoms of hypoparathyroidism are the result of the low blood calcium level which can lead to various neuromuscular, cardiac and skin manifestations. Long term treatment of hypoparathyroidism is with vitamin D analogs and calcium supplementation but in some recombinant Parathyroid hormone(rPTH) has to be given. Here we report a case of 18 year old male patient who presented with wide array of symptoms related to hypocalcemia and responded only to the parathyroid homone replacement therapy.

KEYWORDS

Hypoparathyroidism, Hypocalcemia, Recombinant Parathyroid Hormone, Seizures, Chorea.

INTRODUCTION

Hypoparathyroidism is a rare condition that occurs when the parathyroid glands don't produce enough parathyroid hormone (PTH). The major function of PTH is to regulate the level of calcium in the body. It also controls the level of phosphorus and has a role in the production of the active form of vitamin D. All of these activities are required to maintain calcium balance. Low PTH levels result in excessive urinary calcium losses, decreased bone remodeling, and reduced intestinal calcium absorption[1]. Hypocalcemia can range in severity from being asymptomatic in mild cases to an acute life-threatening crisis [1].

CASE REPORT

An 18 years old male presented with abnormal dance like movements of the left upper and lower limbs distally which were episodic in occurrence, used to remain for two to three minutes and then used to subside by themselves from last one month with frequency of one episode after every three to four days. These episodes were not associated with loss of consciousness, tongue bite, uprolling of eyes or urinary or faecal incontinence. He also had complaints of imbalance while walking and he used to walk like drunked person but there was no history of tremulousness of the limbs while reaching for objects. There was history of drowsiness with decreased responsiveness to verbal commands from same duration with increasing irritability and decreased sleep. There were no complaints of weakness of any limb, cranial nerve involvement. There were no preceding complaints of fever, athralgias, raynauds phenomena, pain abdomen, loss of weight or appetite. He was on antiepileptics(phenytoin, clobazam and sodium valproate) for episodes of tonic posturing of the body from last two years but inspite of appropriate doses and compliance for medications patient kept on having these episodes twice or thrice a week. There was no significant family history.

On examination vitals of the patient were stable. On neurologic examination patient was in confused state, not responding to verbal commands appropriately. Cranial nerves and motor system examination was normal. Reflexes were diminished in the lower limbs bilaterally with flexor planters. Gait was ataxic and he was not able to walk without support. There were no meningeal signs and skull and spine were also normal. Keeping the possibility of phenytoin toxicity initially, phenytoin was replaced with leviteracetam. But there was no improvement in the complaints and on second day of admission patient had urinary retention alongwith eczematous lesions on the skin involving trunk and limbs(Figure1) with increased irritability and increase in the frequency of the abnormal dance like movements. On observing tetanic spasm like movements, second possibility of chronic hypocalcemia was kept.

CT head was done which was suggestive of bilateral basal ganglia calcification(Figure 2). Abnormal biochemical parameters included serum calcium levels of 4.8 meq/l and phosphorus levels of 6.7 meq/l. All other parameters were within normal limits including Haemoglobin-9.6 g%, Total Leucocyte count- 6270/microlitre, Platelet count-3,30,000/microlitre, Serum Bilirubin-0.3mg%, Serum SGOT-36 IU/L, Serum SGPT- 17IU/L, Blood Urea- 33mg/dl, Serum Creatinine- 0.6mg/dl, Serum sodium- 136 mmol/L, Serum potassium- 4.0 mmol/L and CSF studies(total cell count- 02 cells/cumm, all lymphocytes, proteins- 41mg/dl, sugar- 52mg/dl, no gram stain organism, no acid fast bacilli, no cryptococci, ADA- 3.5 U/L). Serum parathyroid hormone levels were low suggestive of hypoparathyoridsm (<0.23pg/ml).

Patient was started on oral calcium and vitamin D but he did not responded to treatment. After two days he was started on injection calcium gluconate infusion with vitamin D and magnesium supplementation but still he was having paresthesias alongwith tetanic episodes though with less frequency and they further used to increase when the rate of infusion was decreased. Multiple attempts to stop the infusion failed as patient used to have reccurrence of symptoms. Subsequently patient was started on injectable recombinant PTH in dose of 40 micrograms subcutaneously once daily which was reduced to 20 micrograms once daily after the symptoms got subsided. The serum calcium and phosphorus levels came in normal range and the patient was withdrawn from the injectable calcium. He was discharged in stable condition with continued rPTH.

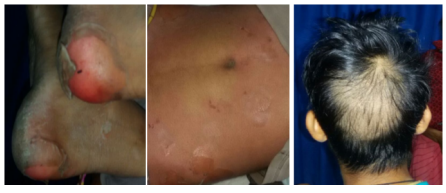

FIGURE 1-Eczematous and petechial lesions involving feet, abdomen and head with exfoliation

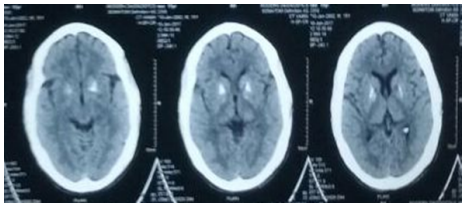

FIGURE 2- Axial section of CT head showing hyperdensity in bilateral basal ganglia

DISCUSSION

The main symtoms of hypoparathyroidism results due to underlying hypocalcemia. The normal range is 2.1– 2.6 mmol/L (8.8–10.7 mg/dL, 4.3–5.2 mEq/L) with levels less than 2.1 mmol/L defined as hypocalcemia[2]. The main symtoms involve the neuromuscular, cardiac and skin manifestations. The neuromuscular symptoms of hypocalcemia are caused by a positive bathmotropic effect due to the decreased interaction of calcium with sodium channels. Since calcium blocks sodium channels and inhibits depolarization of nerve and muscle fibers, reduced calcium lowers the threshold for depolarization[3]. The symptoms include paresthesias over the limbs and face, spasms, myalgias, wheezing due to bronchospasm, voice changes due to laryngospasm, tetany, carpopedal spasm. Hypocalcemia has also been mentioned as a cause for detrusor atony leading to functional urinary retention. Neurologic or neuropsychiatric symptoms include anxiety, depression or excessive irritability, seizures, movement disorders in form of chorea or paroxysmal dyskinesias and ataxic gait and impairment of intellectual ability. Hypocalcaemia is a rare cause of dystonia or choreoathetosis. Usually the aetiology is idiopathic hypoparathyroidism[4-6]. Patients may present with the abnormal movements, which may be asymmetric, are usually paroxysmal and, rarely, kinesigenic[7,8]. The same type of movement abnormality alongwith ataxic gait, altered levels of consciousness and urinary retention was seen in this patient. Cardiovascular symptoms include prolonged QT interval, hypotension and congestive heart failure. Dermatologic manifestations in form of eczema, hyperpigmentation, psoriasis and petechiae which appear as spots, then later become confluent, and appear as purpura (larger bruised areas, usually in dependent regions of the body). Similar lesions appeared in our patient also, over feet and back.

As far as the treatment for this disorder is concerned the main emphasis is on the calcium and calcitriol supplementation. Only recently rPTH has been approved by the US FDA for management of hypopathyroidism associated hypocalcemia but still its not approved in European countries. The recommended dose is minimum dose at which the serum calcium comes in the lower normal range and the clinical manifestations subside, the usual dose is 25-50 micrograms per day as subcutaneous injection once daily.

This patient reported here presented with whole array of hypocalcemic symptoms who was previously being treated as a seizure disorder. Also when treatment was started he did not respond to oral calcium and calcitriol supplementation alongwith magnesium supplementation. After starting intravenous calcium we were not able to stop it till we started with recombinant PTH.

CONCLUSION

Its important to take meticulous history in every patient of refractory seizures who is not responding to the treatment appropriately despite appropriate doses and compliance. Chorea is a rare manifestation of hypocalcemia. High degree of suspicion is required to diagnose hypoparathyroidism when refractory seizures and movement disorder coexist. Also role of PTH supplementation as primary treatment for idiopathic hypoparathyroidism including its effects on the natural history, skeletal dynamics and renal functions and the safety profile needs to be further studied in our set up as in most studies and textbooks the primary treatment mentioned is calcium and vitamin D analog supplementation.

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