Seizure, tetanic spasm and primary hypoparathyroidism

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Abstract

Primary hypoparathyroidism is not a common disease, and it is not a common cause of seizures. Here we present a 21-year-old female with hypoparathyroidism who presented with tetanic spasm and seizure. Her parathyroid hormone level was low, Calcium level was low and Phosphorus was increased. Electroencephalogram showed sharp spike and Computed Tomography scan showed multiple symmetrical calcification in the basal ganglia, thalamus, cerebellum, cerebral hemisphere and periventricular region. With these features, our provisional diagnosis was Fahrs syndrome. Patient was treated with calcium gluconate and anticonvulsant. Patient improved and was discharged on medication.

Key words: Basal ganglia, Hypocalcaemia, Hypoparathyroidism, Tetanic spasm

INTRODUCTION

Hypoparathyroidism is an uncommon endocrine-deficiency disease characterized by low serum calcium levels, elevated serum phosphorus levels, and absent or inappropriately low levels of parathyroid hormone (PTH) in the circulation[1, 2]. It is caused as a result of congenital disorders, iatrogenic causes (e.g., drugs, removal of the parathyroid glands during thyroid or parathyroid surgery, radiation), infiltration of the parathyroid glands (e.g., metastatic carcinoma, Wilson's disease, sarcoidosis), suppression of parathyroid function such as in hypomagnesemia, Human immunodeficiency virus/ Acquired immunodeficiency syndrome (HIV/AIDS), or idiopathic mechanisms[3]. Pseudohypoparathyroidism is a condition that is primarily associated with resistance to the parathyroid hormone presented with low serum calcium and high phosphate, but the parathyroid hormone level (PTH) is actually appropriately high (due to the hypocalcemia). Its pathogenesis has been linked to dysfunctional G Proteins (in particular, Gs alpha subunit)[4]. Neurological signs of hypoparathyroidism includes symptomatic or latent tetany, seizure, hemiparesis, difficulty in speaking and ischemic attack. In basal ganglia calcification presentation may be choreathetosis and parkinsonism[5].

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CASE REPORT

A 21-year-old female from Tanahu presented to Out Patient Department of College of Medical Sciences in neurology department with a tetanic spasm. She came with history of three episodes of tetanic spasm followed by seizure which she was unaware of. On the next day of admission she had a similar episode in the hospital.

According to her past record she was apparently well six years back when she developed seizure and tetany while climbing downstairs. For about four years she had frequent episodes of tetanic spasm but she continued to live with that. During that time she had undergone cataract surgery. Two years back when she had a tetanic attack she was admitted in Intensive Care Unit .

On examination, Chvostek's and Trousseau's signs were positive. Fundoscopy was normal. Investigation report shows parathyroid hormone - <2.5pg/ml (14 to 72pg/ml), 1, 25 Dihydroxy Vitamin D3- 22.37pg/ml (25 to 80pg/ml), Calcium -4.1mg/dl (9 to 11mg/dl), Phosphorous -4.9mg/dl(2.4 to 4.1 mg/dl). Computed Tomography scan showed multiple symmetrical calcification in the basal ganglia, thalamus, cerebellum, cerebral hemisphere and periventricular region: features suggestive of Fahr's syndrome. Electroencephalogram reported - high voltage generalized slowing of delta range with sharp and spike wave activity, and demonstrated that hyperventilation accentuated the abnormalities. She was diagnosed as a case of Primary hypoparathyroidism and treated with Calcium glugonate, Magnesium,

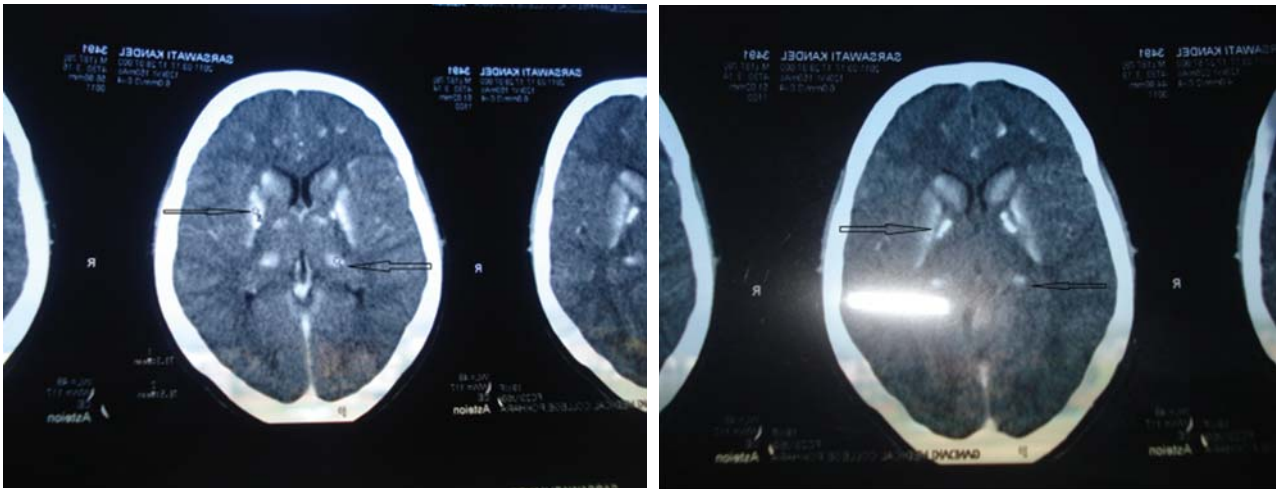

Figure1(a, b): Calcification of bilateral basal ganglia and thalamus

Vitamin D and Levetiracetam. Patient improved and medication was continued after discharge.

DISCUSSION

In this particular case, a 21-year-old female presented with repeated tetanic spasm and seizure. Tetanic spasm was attributed to recurrent hypocalcaemia, for which there was no history of surgical procedure for thyroid gland or parathyroid gland. In this case hypocalcaemia was due to low parathyroid hormone level. Parathyroid hormone, a molecule composed of 84 amino acids secreted by the parathyroid glands, is the main hormone in control of circulating calcium levels[2, 6]. It is well known that low parathyroid hormone can be due to damage to the glands which includes thyroid or parathyroid surgery. Apart from this, developmental defects like agenesis of the parathyroid glands in infants with the DiGeorge syndrome (closely related velocardiofacial syndrome), autoimmune hypoparathyroidism: polyglandular syndrome type 1, defects in the hormone molecule: familial hypoparathyroidism, defective regulation of hormone secretion: autosomal dominant hypercalciuric hypocalcaemia,[6] also lead to hypocalcaemia.

In our study we found that there was calcification in different parts of brain structure as shown in figure 1 (a, b). Calcification was localised in basal ganglion and thalamus. Basal ganglion calcification, also referred to as Fahr's disease, nonarteriosclerotic calcification and striato-pallido-dentate-calcinosis, can be associated with more than 24 other illnesses such as type-1 diabetes, Acquired Immunity Deficiency Syndrome and osteopetrosis[7-9]. The pathogenic mechanisms of Basal ganglion calcification in most of these, including hypoparathyroidism, are not known, and the association continues to attract attention even now[10]. Its occurrence in the hypocalcaemic milieu of hypoparathyroidism is believed to be due to high serum calcium–phosphorus product and poor calcium control. However, Basal ganglion calcification can occur without any alteration in mineral homoeostasis in familial and incidentally detected Basal ganglion calcification[9, 11]. So for this case the better option would be Injection parathyroid.

CONCLUSION

Hypoparathyroidism is not a common cause for seizure disorder. A 21-year-old female with recurrent seizure was found to have primary hypoparathyroidism, low serum calcium with extensive calcification in basal ganglia, thalamus and dentate nucleus.

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