CASE REPORT

IDIOPATHIC HYPOPARATHYROIDISM PRESENTING AS STATUS EPILEPTICUS AND CHOREOATHETOSIS

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ABSTRACT

Idiopathic hypoparathyroidism presenting as convulsions along with choreoathetosis simultaneously is a distinctly rare scenario. We present a case of Idiopathic hypoparathyroidism who presented with status epilepticus along with extrapyramidal dysfunction in the form of choreoathetosis. Clinical diagnosis was confirmed by the demonstration of basal ganglia calcification in Non Contrast Computed Tomography (CT) brain.

Keywords: Idiopathic hypoparathyroidism, basal ganglia calcification, choreoathetosis, convulsions

INTRODUCTION

Idiopathic hypoparathyroidism is uncommon.[1] These patients may demonstrate basal ganglia calcification.[2] But in spite of extensive basal ganglia calcifications, symptomsattributable to their involvement are an uncommon clinical presentation.[2] We describe a case of hypoparathyroidism with calcification and symptoms of basalganglia involvement.

CASE REPORT

A 23 year old male was rushed to the emergency in a state of status epilepticus. There was no history of head trauma or of any suicidal or accidental ingestion of any poisons. No history of fever, headache, projectile vomiting was there. But there was a history of occasional seizures since the age of 9 years which was however controlled on medications. He had developed abnormal involuntary movements in the upper extremities for 2 months prior to admission. He was treated with intravenous lorazepam and phenytoin along with general supportive treatment. The patient recovered from postictalstate after 48 hours with no residual neurological deficit. Examination during this time revealed a conscious, co-operative individualof low intelligence. He was short statured (129 cm) with hypoplastic dentition and dystrophic nails. The patient demonstrated features of tetany in the form of a positive Trousseau sign and Chvostek's sign and generalized hyperreflexia during stay in hospital. He demonstrated regular, slow, jerky movements of the both hands resembling choreoathetosis. Ophthalmologicalexamination revealed reduced visual acuity in botheyes with slit lamp examination revealed posterior subcapsular lenticular opacities in both eyes. Fundus examination was normal, however. Examinationof other systems was normal.Investigations revealed hypocalcaemia - ionised Ca 3.2mg/dl (N: 4.4-5.5mg/dl), total Ca 6.8 mg/dl (N: 8.5-10.2mg/dl); hyperphosphataemia- PO4 1.4mg/dl (N: 2.3-4.5mg/dl) but normal alkaline phosphatase and renal function. Rest haematological and biochemical parameters were normal. A plain radiologicalsurvey did not reveal any metastatic calcification or bony abnormalities. Computed tomographic (CT) scan brain demonstrated extensive bilateral calcification in the region of basalganglia (Figure 1). The electroencephalogram (EEG) did not reveal any abnormal finding. Serum magnesiumwas normal but serum parathyroid hormone wasundetectable by radioimmunoassay.The history, clinical examination and lab findings were indicative of idiopathic hypoparathyroidism. The patient was treated with oral phenytoin, vitamin D supplements and oral calcium. Follow-up at 3 months demonstrated both clinical and biochemical improvement

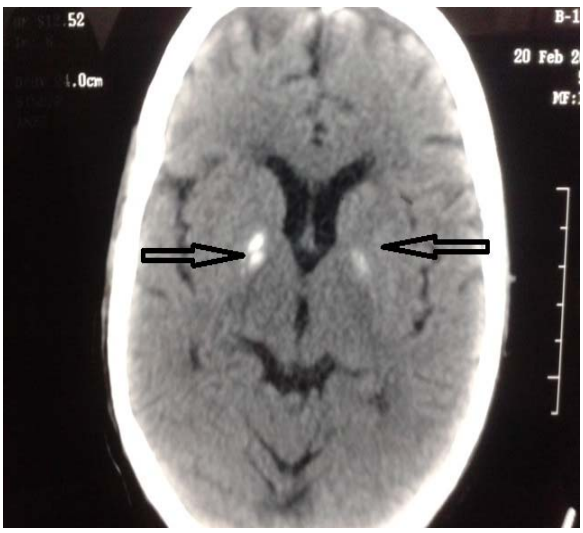

Figure 1: bilateral basal ganglia calcification

DISCUSSION

Clinical manifestations in hypoparathyroidism result from hypocalcaemia, while biochemical abnormalities reveal hyperphosphatemia and decreased detectable levels of serumparathyroid hormone and calcium. Most common radiological finding is basal ganglia calcification. Calcification may rarely extend beyond the basal ganglia, especially to the cerebellum and frontal lobes of the cortex[3,4]. Calcification, particularly in basal ganglia, is proposed to result from adegenerative vascular process in theextrapyramidal system, initiated possibly by deposition of calcium crystals.[5] This calcification is rarelysymptomatic and may not be visualized on plainX-rays of the skull. It is usually noted in a CT scan. This may be because of the thinlayering of calcium along blood vessels in basalganglia [5] . Our patient displayed symptoms due tohypocalcaemia. He also had symptomatic basalganglia involvement. Symptoms attributable to basal ganglia involvement described previously include paroxysmal choreoathetosis and myoclonus [6] The present case also suffered from choreoathetosis.

CONCLUSION

This case highlights the importance of keeping in mind the possibility of idiopathic hypoparathyroidism in appropriate clinical scenario as appropriate diagnosis and treatment might lead to complete resolution of the potentially fatal condition and untreated state is highly dangerous.

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