Idiopathic Hypoparathyroidism in a Cerebral Palsy Child Presenting with Extensive Intracranial Calcifications: A Rare Case Report

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ABSTRACT

Introduction: Hypoparathyroidism is a rare endocrine disorder characterized by low serum calcium and parathyroid hormone levels. The most common cause is iatrogenic surgical removal. However, idiopathic primary hypoparathyroidism is to be kept in mind especially in children as a cause of hypoparathyroidism. Since PTH is important in calcium homeostasis, this condition may cause ectopic calcifications, including intra cerebral calcifications, though rare.

Case report: 11 year old MCH,with history of birth asphyxia and global developmental delay was presented to emergency with seizures. The child had seizures since the age of 5years, and wasn't on regular treatment. The patient was admitted with a provisional diagnosis of Cerebral Palsy with seizure disorder with polymorphic seizures was made and treated conservatively. Neuroimaging showed bilateral symmetric calcification in basal ganglia,thalami and capsule. A complete workup showed low serum calcium, high serum phosphorus and low parathormone. Calcium supplementation was given and seizures was made under control.

Conclusion: Idiopathic hypoparathroidism, though rarely, as it was in our case, may cause exuberant cerebral presentations with extensive intracranial calcifications and extreme hypocalcemia which may or may not correlate with severity of symptoms. Not only this is a treatable disorder that may have dangerous implications if untreated, but also its symptoms may be completely reversed with prompt treatment.

Keywords: Idiopathic Hypoparathyroidism, Cerebral Palsy Child, Intracranial Calcifications

INTRODUCTION

Hypoparathroidism is an endocrine disorder caused as a result of congenital disorders,iatrogenic causes,infiltrative causes or idiopathic. Hypoparathyroidism by any cause is well known to cause basal ganglia calcification in most patients.The mechanism of intracranial calcification in hypoparathyroidism[1], has not been completely elucidated. It may be related more to the duration of hypocalcemia and hyperphosphatemia than parathyroid hormone itself. Hyperphosphatemia promotes ectopic calcification in brain tissue in hypoparathyroidism.[2] Though calcification is expected,extensive intracranial calcification is rarely reported and hence,we are reporting this case,which presented with extensive basal ganglia calcification and idiopathic hypoparathyroidism.

CASE REPORT

11 year old MCH,with history of birth asphyxia and global developmental delay was presented to emergency with seizures. The child had seizures since the age of 5years,and was on homeopathic treatment since then and was started on valproate therapy for the past 6 months by a local practitioner. The compliance to the drug intake was poor and the child presented to ER with generalized tonic clonic seizures and was stabilized. But the mother told that the child used to throw tonic seizures as well as complex partial seizures while at home. And a provisional diagnosis of Cerebral Palsy with seizure disorder with polymorphic seizures was made and was admitted. The child was also found to have attention deficit,hyperactivity and aggressive behavior on recovery from post ictal state. Routine blood investigations revealed no abnormality. Neuroimaging showed bilateral symmetric calcification in basal ganglia,thalami and capsule. These findings evoked the diagnosis of Fahr syndrome, hypoparathyroidism.

Sr calcium, Phosphate, and PTH was sent and the reports were as follows.Sr calcium 2mg/dl. Sr, PTH was 8 pg/ml.

The diagnosis of fahr disease couldn't be established because of the non progressive course, early presentation,absence of family history and because of the evidence of metabolic abnormalities pertaining to hypoparathyroidism.

As the child improved considerably during hospital stay, the child was discharged with Levetiracetam, calcium supplements and risperidone.

DISCUSSION

Physiological intracranial calcifications occur in about 0.3-1.5% of cases.[1] Its asymptomatic and detected incidentally by neuro imaging. Pathological Basal Ganglia calcification is due to various causes, such as metabolic diseases,infections and genetic causes. Hypoparathyroidism is known to cause Basal Ganglia calcifications and in itself can be caused by

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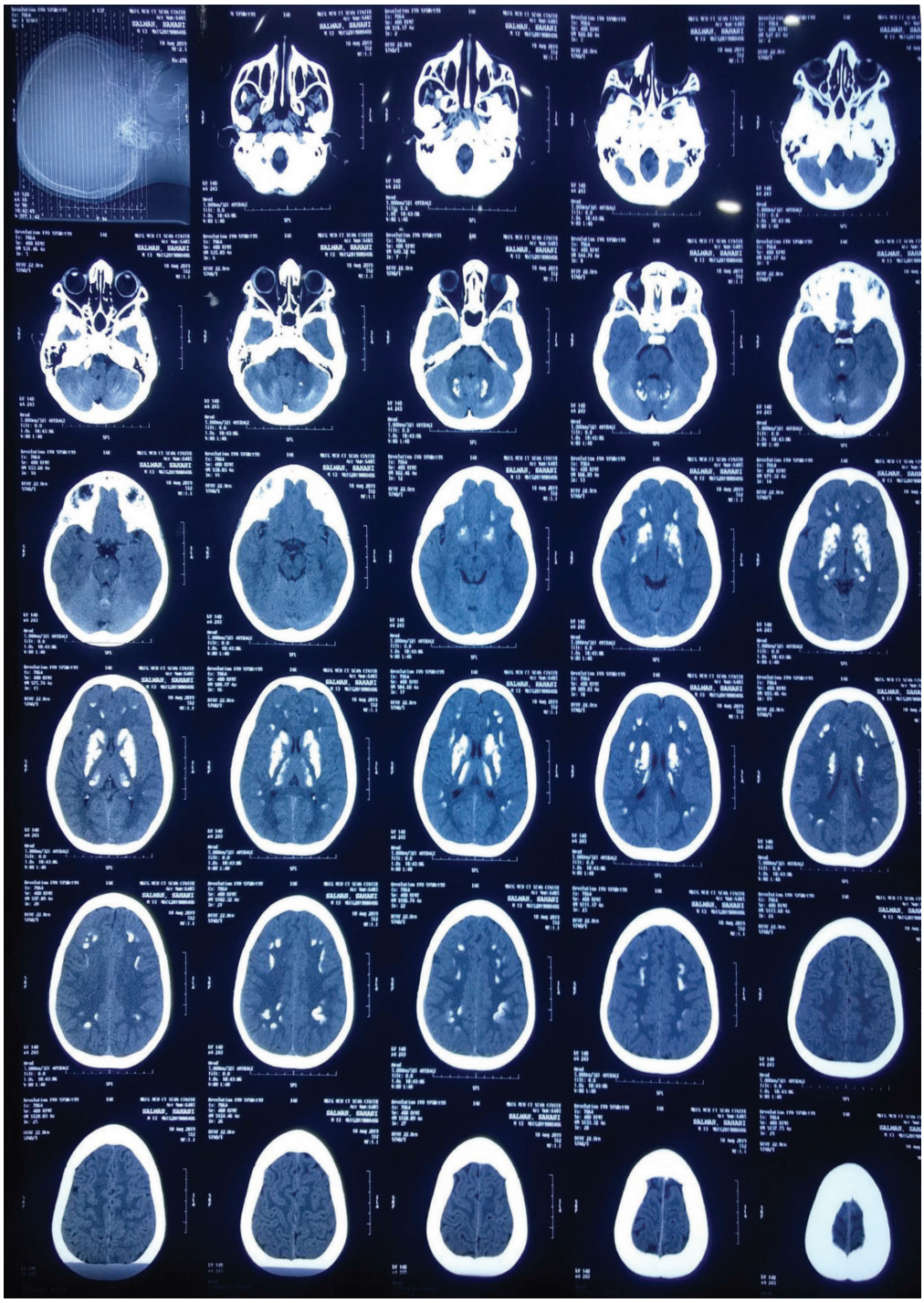

Figure-1: Showing NCCT Brain revealing abnormal symmetrical calcific deposits predominantly in B/L basal ganglia, thalami, pons, cerebellar lobes and subcortical white matter

congenital disorders,iatrogenic causes like surgery,radiation, or could be idiopathic. Acquired and congenital causes would have either normal or undetectable PTH levels with hypocalcaemia wheras in idiopathic variety,both Calcium and PTH is low.[2] In pseudohypoparathyroidism,PTH is high with low sr.calcium. In pseudopseudohypoparathyroidism,PTH is raised,but both calcium and phosphorus is low.

Radiologically, hypoparathyroidism causes calcification most often in bilateral basal ganglia,most common site being globus pallidus.[3] Calcification can also occur in cerebellum, thalamus, corona radiata etc.

Other causes of symmetrical calcification in Basal Ganglia include Fahr syndrome (familial idiopathic BG calcification), lead poisoning,Cockayne syndrome, AIDS, Mitochondrial encephalopathies etc.[4]

This can present with diverse presentations,the most common being seizures,mental deterioration and disorders of cerebellar or extrapyramidal function.[5] Delayed teeth eruption is also reported in hypoparathyroidism as was seen in this patient (fig 2) The treatment is directed towards severity of symptoms and level of Sr.Calcium. mostly, calcium supplements and vit D analogues are

Figure-2: Showing non eruption of teeth in the 11 year old.

used.

CONCLUSION

Since it's a treatable condition,its always wise to rule out idiopathic hypoparathyroidism in a patient presenting with symmetric basal ganglia calcifications.

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