

Extensive intracranial calcification in idiopathic hypoparathyroidism : rare presentation in an adolescent.

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Key words : -Extensive intracranial calcification, Idiopathic hypoparathyroidism

Introduction:

Hypoparathyroidism is an endocrine disorder caused as result of congenital disorders, iatrogenic causes (eg, drugs, removal of the parathyroid glands during thyroid or parathyroid surgery, radiation), infiltration of the parathyroid glands (eg, metastatic carcinoma, Wilson's disease, sarcoidosis), suppression of parathyroid function, HIV/AIDS, or idiopathic mechanisms. Idiopathic Hypoparathyroidism is diagnosed when all the possible causes of hypoparathyroidism are ruled out. Hypoparathyroidism is well known to cause detectable basal ganglia calcification in most of the patients. It is also well known that extensive intracranial calcification caused by hypoparathyroidism is rare. Here we would like to present a case of idiopathic hypoparathyroidism which presented with extensive intracranial calcification.

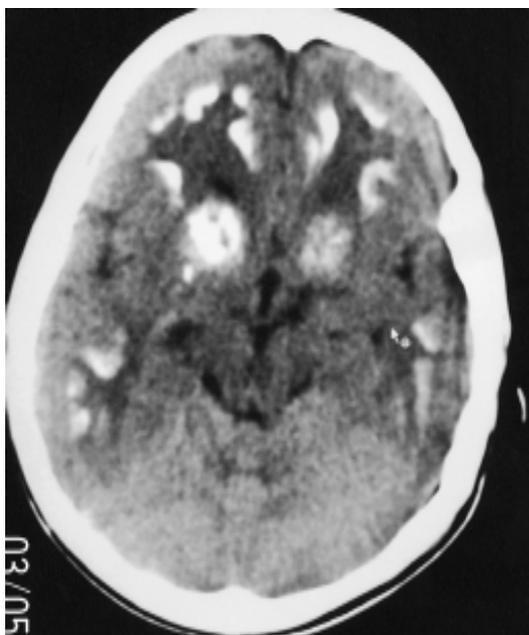


Fig 1: Spiral CECT Head shows dense calcification in the head of caudate nucleus, basal ganglia and the sub cortical white matter region including the cingulated gyrus.



Fig 2: Spiral CECT Head shows dense calcification in the sub cortical white matter region and the periventricular areas

Case Report

A 14-year-old male patient presented as status epilepticus with history of seizures from the age of 9 yrs and was on treatment with anticonvulsants. Since three months the frequency of the seizures had increased, with no evidence of psychosis or altered mental status. On examination Positive Chvostek or Trousseau sign were present. Contrast enhanced spiral CT done showed bilateral extensive nonenhancing hyper dense lesions (calcifications) involving the thalamus, dentate nuclei, putamen, globus pallidus, caudate nucleus and sub-cortical white matter (fig 1,2,3). Investigations revealed reduced serum levels of calcium and phosphorus. Serum

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magnesium levels and renal function tests were normal. Radio immunoassay (RIA) revealed reduced PTH levels. With these investigations and after ruling out other causes of hypoparathyroidism we suggested the possibility of idiopathic hypoparathyroidism.

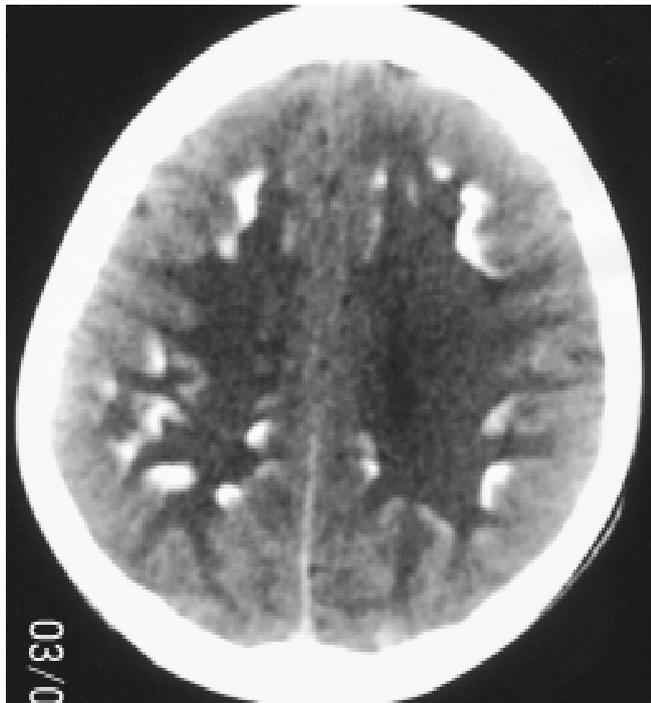


Fig 3: Spiral CECT Head shows dense calcification in the corona radiata and sub cortical white matter region .

Discussion

Hypoparathyroidism is an endocrine disorder caused as result of congenital disorders, iatrogenic causes, infiltration of the parathyroid glands, suppression of parathyroid function, or idiopathic mechanisms(3). In these cases either there will be apparent deficiency of PTH secretion or end-organ failure(4) .Idiopathic hypoparathyroidism is an uncommon condition of unknown etiology . Acquired and congenital hypoparathyroidism will have PTH levels which are either normal or undetectable, with hypocalcaemia whereas in idiopathic hypoparathyroidism both calcium and PTH levels are low(4). In pseudo hypoparathyroidism PTH levels will be high with hypocalcaemia. In pseudopseudohypoparathyroidism with increased PTH level both serum calcium and phosphorus levels are normal(4). Prevalence of hypoparathyroidism is equal in men and women and occurs in all age groups. Classical clinical presentation is with features of hypocalcaemia (Chvostek or Trousseau sign, tetany, seizures, cardiac failure)(1).

Radiologically hypoparathyroidism causes calcification most often in bilateral basal ganglia (1). The most common site is often globus pallidus (2). Calcification can also occur in cerebellum, sub cortical white matter, corona radiata and the thalamus. In skeletal survey calvarial thickening, soft tissue calcification, and premature closure of the epiphysis may or may not be present in all types of hypoparathyroidism (4). Hypoparathyroidism commonly has mild or diffuse oostoesclerosis(4). Pseudo hypoparathyroidism, pseudo pseudo hypoparathyroidism classically will have metacarpal/metatarsal shortening and exostosis formation (4).Most commonly involved are the 1st and 4th meta carpal/tarsal bone. In idiopathic hypoparathyroidism none of the skeletal manifestations may be present at early stages.

The immediate treatment for all types is immediate calcium supplement with supplementation of PTH in cases of acquired hypoparathyroidism. Also one has to treat symptomatically for the seizures and other changes. This disease has a good prognosis if detected early and treated.

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