

Extensive Intracranial Calcification and Choreoathetoid Movements in a case of Idiopathic Hypoparathyroidism

Pradeep Sreekumar^a, Calwin David Singh^a, K Umakanthan^a

a. Department of Medicine, Coimbatore Medical College, Coimbatore*

ABSTRACT

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Idiopathic hypoparathyroidism rarely presents with choreoathetoid movements and extensive intracranial calcification, though neurological manifestations like seizures and tetany are more common. We report a case idiopathic hypoparathyroidism with both choreoathetoid movements and extensive cerebral calcification.

Keywords: Extensive intracranial calcification, Idiopathic hypoparathyroidism, Choreoathetoid movements

*See End Note for complete author details

INTRODUCTION

Hypoparathyroidism is an inherited or acquired deficiency of the parathyroid hormone (PTH) or its action. PTH secretion by the parathyroid glands (prime regulators of serum calcium concentration) maintains serum calcium within a strict range. Biochemical hallmarks of PTH insufficiency are hypocalcemia and hyperphosphatemia.

Basal ganglion calcification is a common manifestation of hypoparathyroidism, but calcification of the cerebral cortex is extremely rare.

We present a rare case of idiopathic hypoparathyroidism with extensive intracranial calcifications (including thalamus, cerebellum) and choreoathetoid movements.

CASE HISTORY

Forty two year old male patient presented to the medical ward with history of involuntary movements involving both upper limbs and lower limbs on and off for the past 2 months. These movements are present for forty five minutes to one hour duration and disappears during sleep. He has a past history of seizures for which he is on phenytoin. On examination patient had chorea, positive chovstek sign and trousseau sign. There were no focal neurological deficits. Investigations revealed serum calcium of 8.5 mg/dl (9 to 11 mg/dl) and phosphate of 6.5 mg/dl (3.5 to 5.5). His parathyroid

level by radioimmunoassay was 2.0 pg/ml (12 – 55pg/ml). Magnesium results and renal function tests were normal. CT scan showed bilateral basal ganglia, thalamus, cerebellar calcifications. He was treated with clonazepam and his involuntary movements subsided. He was treated with vitamin D and calcium supplementation.

DISCUSSION

Intracranial calcifications commonly basal ganglia calcifications are common. But extensive intracranial calcifications are very rare but reported¹ in hypoparathyroidism. Other causes of intracranial calcifications include² neoplasms like craniopharyngioma, glioma,

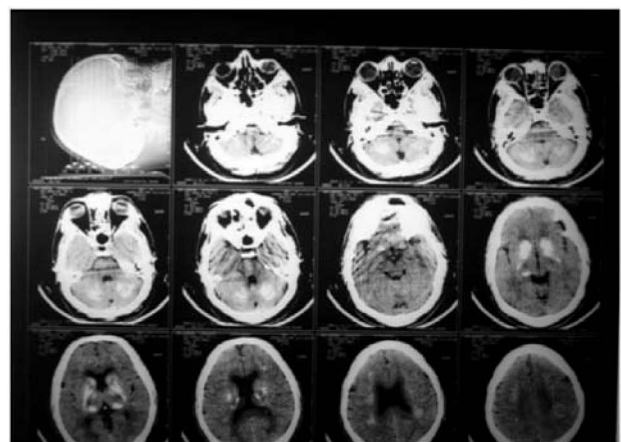


Figure 1. CT Brain-Note the extensive intracranial calcifications involving both cerebellum, thalamus, basal ganglia.

Corresponding Author:

Dr. Pradeep Sreekumar, Junior Resident, Department of Medicine, Coimbatore Medical College, Coimbatore.
Phone: 09994361627. Email: pradeepsreekumar@gmail.com

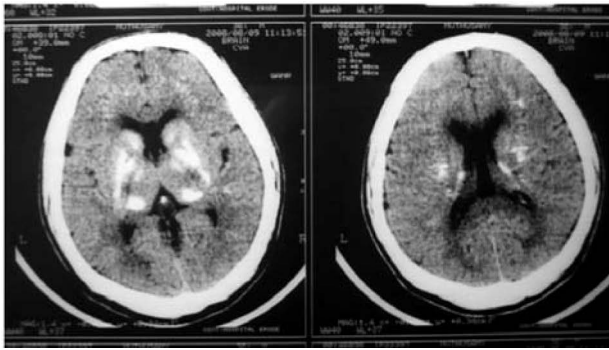


Figure 2. Basal ganglia and Thalamic calcifications-a closer view

vascular like angiomas, aneurysms, infections like toxoplasmosis, herpes, rubella and metabolic include Fahr's syndrome, idiopathic basal ganglia calcification, pseudohypoparathyroidism, Tuberous sclerosis, Neurofibromatosis., Lissencephaly. All possible other causes of intracranial calcification was ruled out with appropriate investigations.

Hypoparathyroidism with basal ganglia calcification (in about 50% of patients) and frontal lobe calcification has been reported.³ Caudate, lenticular, thalamic and frontal lobe calcifications have also been reported.⁵

Usual neurological manifestations include muscle spasms called tetany (can lead to spasms of the larynx, causing breathing difficulties), convulsions (seizures). Choreo athetoid movements have been rarely reported in hypoparathyroidism.⁴ Our patient had choreoathetoid movements which subsided with clonazepam. He also developed episodes of hypocalcemia with low s.calcium during attacks, positive trosseus sign, chvosteks sign which responded to IV calcium gluconate.

Severe hypocalcemia, a potentially life-threatening condition, should be treated as soon as possible with intravenous calcium (e.g. as calcium gluconate). Long-term treatment of hypoparathyroidism is with calcium and Vitamin D3 supplementation (D1 is ineffective in the absence of renal conversion). Teriparatide, might become the treatment of choice for PTH supplementation. In the event of a life-threatening attack of low calcium levels or tetany (prolonged muscle contractions), calcium is administered by intravenous (IV) infusion. Precautions are taken to prevent seizures or larynx spasms.

The mechanism of calcification in hypoparathyroidism has not been completely elucidated. Participation of PTH receptor 2 in the brain and superoxide production by mitochondria in hypoparathyroidism should be explored with reference to intracerebral calcification and neurodegenerative diseases. Our patient was advised to take lifelong supplementation of oral calcium and to continue clonazepam for control of choreo athetoid movements.

END NOTE

Author Information

1. Dr. Pradeep Sreekumar, Junior Resident, Department of Medicine, Coimbatore Medical College, Coimbatore.
2. Dr. Calvin David Singh, Assistant Professor, Department of Medicine, Coimbatore Medical College, Coimbatore.
3. Dr. K Umakanthan, Professor, Department of Medicine, Coimbatore Medical College, Coimbatore

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