

Clinical Reports

Hypoparathyroidism with extensive intracranial calcification: a case report

A. Goel, M.K. Bhatnagar, A. Vashishta and N.P.S. Verma

Department of Medicine, Lady Hardinge Medical College and Associated Dr Ram Manohar Lohia Hospital, New Delhi, India

Summary: We present a case of hypoparathyroidism, demonstrating extensive intracranial calcification, not only in basal ganglia, but also outside the extrapyramidal system. The patient presented with an unexplained epileptiform disorder, accompanied by extrapyramidal dysfunction in the form of choreoathetosis and hemiballismus. Hemiballismus is reported for the first time to our knowledge in association with hypoparathyroidism.

Introduction

Idiopathic hypoparathyroidism is uncommon.¹ A proportion of these patients demonstrate basal ganglia calcification.^{2,3} However, intracranial calcifications outside the basal ganglia have been reported only rarely.^{4,5} In addition, in spite of extensive basal ganglia calcifications, symptoms attributable to their involvement are an uncommon clinical presentation.^{6–8} The present report describes a case of hypoparathyroidism with extensive intracranial calcification and symptoms of basal ganglia involvement.

Case report

A 17 year old male was admitted with a history of generalized seizures since the age of 9 years and abnormal involuntary movements for 2 months prior to admission. He was admitted in status epilepticus. He was treated with intravenous phenytoin. The patient recovered from postictal state after 72 hours and demonstrated no neurological deficit. Examination on the fourth day of admission revealed a co-operative individual of low intelligence. He was short statured (138 cm), had hypoplastic dentition, thick dystrophic nails and an obvious goitre. The patient demonstrated tetany, a positive Chvostek's sign and generalized hyperreflexia. Systemic examination was normal. The patient had paroxysms of abnormal, involuntary, flinging, violent movements of the right hand

and leg lasting for 3–5 minutes, suggestive of hemiballismus. In addition, he demonstrated regular, slow, jerky movements of the right hand resembling choreoathetosis. Ophthalmological examination revealed reduced visual acuity in both eyes, right (6/36) more than left (6/12). Fundus examination was normal, however, slit lamp examination revealed posterior subcapsular lenticular opacities (Figure 1) in both eyes.

Investigations (Table I) revealed hypocalcaemia, hyperphosphataemia but normal alkaline phosphatase and renal function. A plain radiological survey did not reveal any metastatic calcification or bony abnormalities. However, a plain computed tomographic (CT) head scan demonstrated extensive bilateral calcification in the region of basal ganglia, cerebellum and cerebral cortex (Figures 2 and 3). The electroencephalogram did not reveal

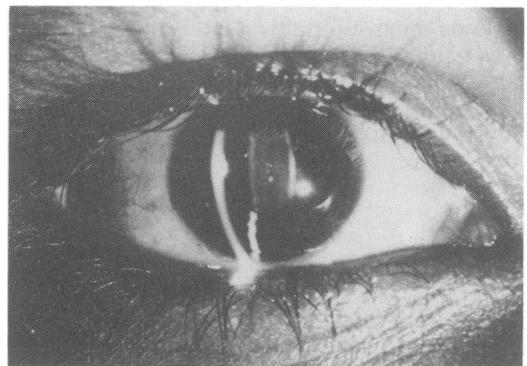


Figure 1 Slit lamp photograph of right eye showing posterior subcapsular opacities.

Correspondence: A. Goel, M.D., F15, Narain Vihar, New Delhi 110 028, India.

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Figure 2 Plan CT showing calcification in the region of the basal ganglia.



Figure 3 Plain CT showing extensive calcification bilaterally in the cerebral cortex.

Table I Investigations of the patient

Parameter	Patient
Serum calcium (mmol/l)	1.70
Serum phosphorus (mmol/l)	1.60
Serum magnesium (mmol/l)	1.00
Serum albumin (g/l)	45.00
Serum parathyroid hormone (ng/l)	Not detected

any abnormal seizure discharge. Serum magnesium was normal but serum parathyroid hormone was undetectable by radioimmunoassay.

The history, clinical examination and investigations were indicative of idiopathic hypoparathyroidism.

Discussion

Clinical manifestations in hypoparathyroidism result from hypocalcaemia, while biochemical abnormalities reveal hyperphosphatemia in addition to decreased detectable levels of serum parathyroid hormone and calcium. These patients demonstrate metastatic calcification, particularly

in basal ganglia, proposed to result from a degenerative vascular process in the extra-pyramidal system, initiated possibly by deposition of calcium crystals.⁹ This calcification is rarely symptomatic and may not be visualized on plain X-rays of the skull, although readily appreciable in a CT scan. This is possibly because of the thin layering of calcium along blood vessels in basal ganglia.⁹ Calcification may rarely extend intracranially beyond the basal ganglia, especially to the cerebellum and frontal lobes of the cortex.^{4,5,8}

The present case revealed extensive intracranial calcifications extending beyond the realms of basal ganglia. The patient displayed symptoms due to hypocalcaemia. He also had symptomatic basal ganglia involvement.

Symptoms attributable to basal ganglia involvement described previously include paroxysmal choreoathetosis⁶⁻⁸ and myoclonus.⁸ The present case suffered from hemiballismus in addition to choreoathetosis. During these movements and the intervening period, he remained conscious. The patient was treated with oral phenytoin, vitamin D supplements, oral calcium and haloperidol. All symptoms except hemiballismus improved. The patient continued to have convulsions, although with a decreased frequency after beginning medica-

tion implying calcification, and not hypocalcaemia as the aetiology.

Asymptomatic calcification outside basal ganglia has been reported previously only in seven cases. Three were a mother and two daughters, diagnosed to be suffering from autosomal dominant hypoparathyroidism.⁵ Of three cases reported by Mendelsohn *et al.*⁴ only one had true idiopathic hypoparathyroidism, whilst the others had pseudo-

hypoparathyroidism. The one case described by Barabas and Tucker⁸ had calcific involvement of dentate nuclei and frontal lobes. The present case also had calcification in the region of the cerebellum and widespread over the entire cerebral cortices. The present case combines certain interesting and rare aspects of idiopathic hypoparathyroidism.

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