Chorea and seizures in iatrogenic hypocalcaemia caused by accidental parathyroidectomy

Introduction

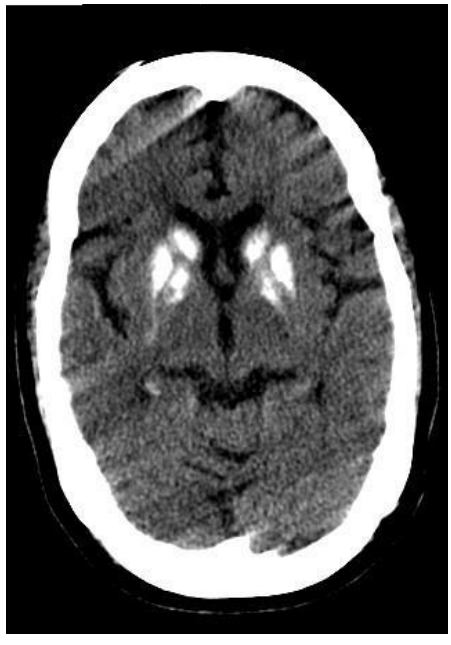
Chorea, derived from the Greek word choreia meaning 'dance-like', is a hyperkinetic movement disorder presenting with irregular, involuntary and brief fleeting movements that seem to flow from one body part to another. Chorea may involve different body parts such as the face, limbs, upper torso and lower extremities, although in rare cases other body parts may also be affected. Patients with chorea appear restless, fidgety and unable to sit still.

Chorea is caused by a variety of acquired, metabolic and hereditary factors. One of the rare causes of chorea is hypocalcaemia. This article describes a patient who developed chorea, seizures and tetany as a result of accidental removal of parathyroid glands during total thyroidectomy that lead to hypoparathyroidism, and eventual hypocalcaemia. The authors stress the importance of metabolic dysfunction, clinical presentation and behavioural symptoms in its differential diagnosis from Fahr's disease and DiGeorge syndrome, all three of which produce basal ganglia calcification and choreiform movement.

Discussion

This case presents an interesting relationship between accidental parathyroidectomy-induced hypocalcaemia and chorea. Since basal ganglia calcification is also observed among other disorders with neurological symptoms, this case presents a challenge of distinguishing between iatrogenic hypocalcaemia, Fahr's disease and DiGeorge syndrome. It is also important to keep in mind that 0.3–1.5% of the population can exhibit physiological basal ganglia calcification but they are asymptomatic and the calcifications are not extensive (Basak, 2009). *Table 1* shows other causes of basal ganglia calcification.

Figure 1. Computed tomography scan of the brain showing mild generalized atrophy and heavy bilateral calcification of basal ganglia involving caudate, putamen and globus pallidus.



Hypocalcaemia is a known complication of thyroid surgery when total thyroidectomy is performed, since the parathyroid glands are located posterior to the thyroid glands. Parathyroid glands secrete parathyroid hormone which is very important in calcium homeostasis. Lack of parathyroid hormone decreases serum calcium and increases serum phosphate levels. These high phosphate levels promote precipitation of calcium phosphate in different organs of body, including the basal ganglia in the brain.

Basal ganglia play an important role in smoothing fine motor movements. Any damage to the basal ganglia will result in either poverty of movements or involuntary movements. Calcification of basal ganglia is also observed in disorders other than hypoparathyroidism with other accompanied features. Fahr's disease is an autosomal recessive disorder resulting in calcification of basal ganglia and dentate

Case Report

A 79-year-old woman with goitre and hyperthyroidism was treated with total thyroidectomy 15 years ago and developed generalized seizures postoperatively in an overseas hospital. She also developed abnormal movements after 3 weeks which were choreiform in nature, would get worse with stress, and disappear during sleep. She was started on phenytoin and calcium replacement therapy post surgery; this lead to significant control of seizures and abnormal movements. She stopped taking calcium replacement therapy several days before presenting to the authors' hospital.

On physical examination she had abnormal movements involving frequent raising of eyebrows, jaw grinding, neck turning and abnormal fleeting movements of extremities. Other findings included hyperreflexia and presence of Trousseau and Chvostek sign. She had no family history of Huntington's disease or any other neurological condition and denied any previous episodes of encephalitis, lupus or infections, as well as exposure to any toxins or antipsychotic medication. Suspecting hypocalcaemia as a possibility, serum calcium levels were observed and found to be markedly below normal. Brain computed tomography scan showed calcification of basal ganglia, specifically the caudate nuclei (*Figure 1*). Interictal electroencephalogram was normal. There was no caudate atrophy present. A diagnosis of chorea secondary to iatrogenic hypocalcaemia caused by parathyroidectomy during total thyroidectomy resulting in the calcification of basal ganglia was considered. The patient was started on calcium replacement therapy which resulted in significant resolution of her choreiform movements.

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Table 1. Other causes of symmetrical basal ganglia calcification

Acquired immunodeficiency syndrome (especially in children)
Birth anoxia
Carbon monoxide intoxication
Cockayne's syndrome
Down syndrome
Familial encephalopathies
Hyperparathyroidism
Kearns–Sayre syndrome and other mitochondrial diseases
Lead poisoning
Methotrexate therapy
Postinfectious
Pseudohypoparathyroidism
Radiation therapy
Tuberous sclerosis

nuclei, and has the following clinical features: parkinsonism, dementia, dysarthria, visual problems, athetosis (Mittal et al, 2010) and psychosis (Sieberer et al, 2005; Srivastava et al, 2010). Some reports map the disorder to the IBGC1 gene on 14q (Oliveira et al, 2007; Lemos et al, 2010) while others do not (Brodaty et al, 2002).

The only symptoms in common between this patient and a patient with Fahr's disease are bilateral and symmetrical calcification of the caudate and choreoathetotic movements, but this patient had no other clinical features characteristic of Fahr's disease.

Hypocalcaemia resulting from genetic deficits can also result in basal ganglia calcification. This phenomenon is seen in 22q11.2 deletion syndrome or DiGeorge syndrome (Cao et al, 2010). This patient did not present with the classic signs of DiGeorge syndrome (symptoms after birth of tetany, abnormal facies, immunocompromised and cardiac abnormalities) nor did she have any family history of DiGeorge syndrome, so it was ruled out.

Basal ganglia damage is also a feature of Wilson's disease but this patient did not have any asterixis, her liver function tests were normal, there were no Kayser-Fleischer rings present, she did not exhibit signs of psychiatric disease, cognitive dysfunction, abnormal facies, or wing beating tremor characteristic of Wilson's disease.

Calcium supplemental therapy in hypoparathyroidism significantly improves choreiform movement disorder (as was the case in this patient). There may be a physiological basis for this improvement. The calcification of basal ganglia which is more of an anatomical problem nevertheless persists. How it is possible to have improvement of the chorea without an improvement of the basal ganglia calcification remains unknown.

The overarching cause of choreiform movements in all three disorders presented herein is the calcification of basal ganglia. However, this case report shows calcification could be caused by various factors. Metabolic dysfunction, clinical presentation and behavioural symptoms are key information when considering a differential diagnosis of chorea which may be secondary to iatrogenic hypocalcaemia caused by total thyroidectomy from Fahr's disease and 22q11.2 deletion syndrome, among others. BJHM

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LEARNING POINTS

- Calcium precipitation in the face of low plasma calcium levels seems paradoxical at first; logically precipitation is expected with high calcium levels. It seems that the high phosphate levels (as a result of low parathyroid hormone) have a more potent effect in precipitating calcium than does a high calcium level alone.
- Basal ganglia calcium precipitation is a very rare event in hypocalcaemic states but still occurs.
- This case shows how one should go about carefully differentiating the other more common causes of basal ganglia damage.
- The authors believe basal ganglia damage caused by an endocrinological abnormality (e.g. hypoparathyroidism) to be extremely rare.