Case Report

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Bilateral Symmetrical Basal Ganglia Calcification with Atypical Presentation: A Case Series

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Physiological intracranial calcification is asymptomatic and is detected incidentally by neuroimaging. Pathological basal ganglia calcification (BGC) is caused by various causes such as metabolic disorders, infectious and genetic diseases, and others. The most common causes of pathological BGC are hypoparathyroidism and pseudohypoparathyroidism. We present three cases of bilaterally symmetrical BGC associated with hypoparathyroidism. All of them presented with seizures as the only presentation without any signs of hypocalcemia and without extrapyramidal features. One should not rule out hypoparathyroidism in the absence of other signs of hypocalcemia and extrapyramidal features. Biochemical analysis pertaining to hypoparathyroidism must be done. Timely treated patients can have a good prognosis.

Keywords: Basal ganglia calcification, Extrapyramidal, Hypoparathyroidism, Seizures

INTRODUCTION

In 1855, Virchow and Bamberger first described calcification in basal ganglia.[1] Physiological basal ganglia calcification (BGC) can be found incidentally in approximately 0.3-1.5% of computed tomography scans.[2] Such calcifications are usually benign, especially in patients over 60 years of age but must be suspected for an underlying disorder in children and young adults.[3]

Pathological BGC is most commonly caused by hypoparathyroidism and pseudohypoparathyroidism. Other causes include pseudopseudohypoparathyroidism, hyperparathyroidism, hypothyroidism, birth anoxia, Fahr's syndrome (ferrocalcinosis), carbon monoxide poisoning, Hastings-James syndrome, lead intoxication, Tuberose sclerosis, toxoplasmosis, cysticercosis, encephalitis caused by measles, chicken pox, parkinsonism, vascular disease, radiation, methotrexate therapy, and Cockayne's syndrome.[4,5]

We report three cases of bilateral BGC presenting only with seizures. We studied their clinical and biochemical profile, and all of them were found to have hypoparathyroidism.

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CASE REPORT

Case 1

A 24-year-old male patient presented in the Emergency Department of Netaji Subhash Chandra Bose Medical College and Hospital, Jabalpur with convulsions. There was a history of seizure disorder since 8 years for which he never took any proper treatment. The patient did not have a history of electrolyte disturbance, psychiatric disorders, or obvious cognitive impairments. On examination, Chvostek's and Trousseau's signs were positive. Fundoscopy was normal. He had no dysmorphic features. Neurological examination was normal (Table 1, Figure 1).

Other investigations were normal. The patient was treated with anticonvulsants, calcium, and Vitamin D supplements. On follow-up, the patient is taking treatment regularly and has no seizure/neurological manifestations.

Case 2

A 45-year-old female patient presented in the Emergency Department of Netaji Subhash Chandra Bose Medical College and Hospital, Jabalpur with multiple episodes of generalized tonic-clonic seizures and difficulty in speech since 6 days. This was associated with motor aphasia, generalized motor weakness in all limbs, bilateral plantar extensor, hypertonia, and left optic atrophy. She was also a

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Table 1: Investigations of Case 1

CECT brain (Figure 1a and b)	Bilateral symmetrical calcifications in supra and infra tentorium regions of cerebellar folia, thalami, basal ganglia, and in periventricular region
MRI brain	Bilaterally almost symmetrical calcific lesion involving both cerebellar tonsils, both basal ganglion, thalami, and periventricular parenchyma
CECT scan brain of patient's father	Small both sided hypodense foci in periventricular areas. Looks like foci of (ischemic) demyelination
Serum parathormone level	4.2 pg/ml (N-15-65 pg/ml)
Serum calcium	7.82 mg/dl (N-9-11 mg/dl)
Serum phosphorus	7.38 mg/dl (N-2.5-48 mg/dl)

CECT: Contrast enhanced computed tomography, MRI: Magnetic resonance imaging

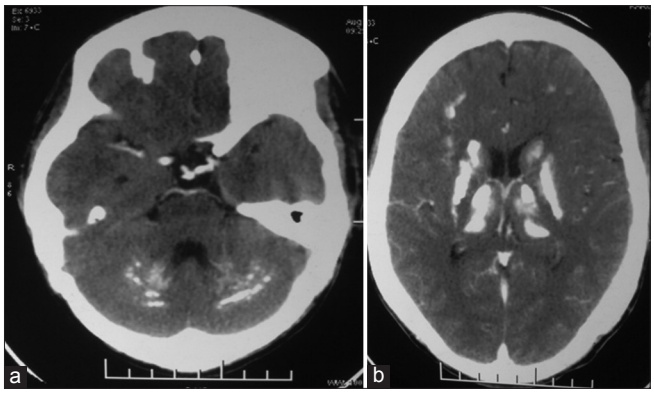

Figure 1: 1.1 ,1.2 CT brain of patient mentioned in case 1

known case of hypertension. The patient was immediately hospitalized and thoroughly investigated and treated in intensive care unit. The patient was brought with very poor general condition with aspiration pneumonitis and succumb to illness in spite of all available resuscitative measures (Table 2, Figure 2).

Case 3

A 22-year-old right-handed female patient known the case of seizure disorder since last 10 years came with multiple episodes of generalized tonic-clonic seizures associated with loss of consciousness, tongue bite, and multiple injuries due to fall during seizure episode. She had loss of eyebrows and depressed nasal bridge. The patient was diagnosed as having leprosy and put on anti-leprosy drugs since August 2013 without slit skin smear from outside. Neurological examination of patient does not reveal significant abnormality. Rest of the physical examination was unremarkable (Table 3, Figure 3).

DISCUSSION

This is the first case reporting of incidental intracranial calcification in our part of the world - Madhya Pradesh,

Table 2: Investigations of Case 2

Serum phosphorus	Normal
Serum parathormone	6.67 pg/ml (N-15-65 pg/ml)
Serum calcium	6.3 mg/dl (N-9-11 mg/dl)
Serum magnesium	1.2 mg/dl (N-1.8-2.4 mg/dl)
CT scan brain (Figure 2)	Multiple bilateral symmetrical basal ganglia, corona radiata, paraventricular calcifications
MRI brain	Extensive bilateral BGC with chronic ischemic foci in bilateral centrum semiovale
MRI spine	Mild disc protrusions at L1-2, L3-4, L4-5 levels. Ligamentum flavum thickening at D 9-10 levels markedly compressing cord

CT: Computed tomography, MRI: Magnetic resonance imaging, BGC: Basal ganglia calcification

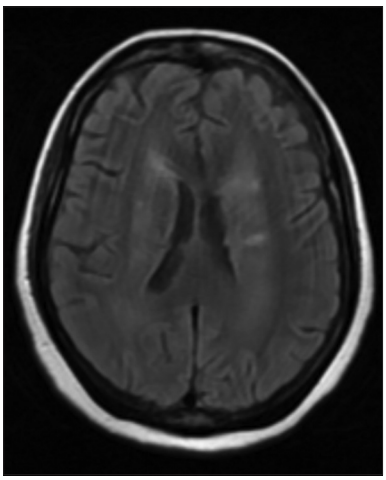

Figure 2: MRI Brain of patient mentioned in case 2

India. All the patients presented to us with a seizure disorder. Two of them were without other signs of hypocalcemia such as carpopedal spasms, facial twitching, laryngospasm, bronchospasm, and abdominal pain. On imaging studies, they were found to have bilateral symmetrical basal ganglia and intracerebral calcification. Investigations revealed hypoparathyroidism. Interestingly, none of the three cases had extrapyramidal features. One of the patients had thoracic compressive myelopathy due to ligamentum flavum calcification - a feature that can occur in hypoparathyroidism.[6]

In 1939, Eaton *et al*. described the association of BGC with hypoparathyroidism.[7] Hypoparathyroidism is one of the most common treatable causes of BGC. Prevalence of hypoparathyroidism is equal in men and women and occurs in all age groups.[8] The most common site is often globus pallidus.[9] Calcification can also occur in the cerebellum, sub cortical white matter, corona radiata, and the thalamus.[10] The presence of calcification in basal ganglia usually suggests chronic hypocalcemia.[11,12] Pathologically, it is characterized by hyalinization and calcification of the media and adventitia of small cerebral blood vessels.[13]

Table 3: Investigations of Case 3

Serum calcium	8.69 mg/dl
Ionic calcium	0.38 mmol/l (normal - 1.16-1.32 mmol/l)
Serum phosphorus	3.35 mg/dl (N-2.5-48 mg/dl)
Intact PTH	<1.2 pg/ml (N-15-65 pg/ml)
CT brain (Figure 3a and b)	Bilateral symmetrical calcifications involving basal ganglia, caudate nucleus, thalamus, and cerebral cortex
Serum magnesium	2.2 mg/dl (N-1.8-2.4 mg/dl)

CT: Computed tomography, PTH: Parathyroid hormone

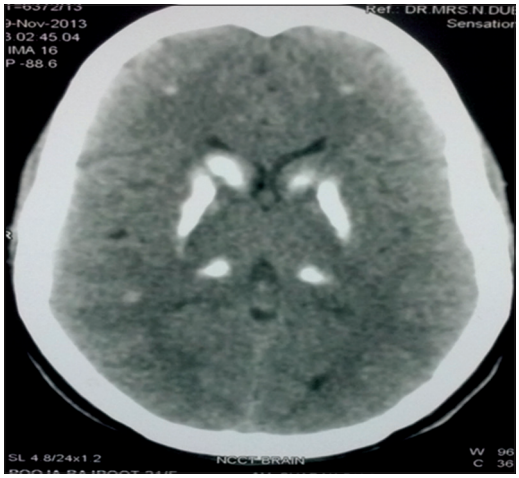

Figure 3: CT scan brain of patient mentioned in case 3

It has been reported in the old literature about the different neurological presentation of BGC in hypoparathyroidism which mainly includes symptoms of hypocalcemia such as tetany, seizures, extrapyramidal symptoms such as parkinsonism, dementia, and cerebellar dysfunction.[14] Such extrapyramidal features are refractory to treatment with levodopa.[15]

Pseudohypoparathyroidism must be differentiated from hypoparathyroidism as there can be the presence of obesity, skeletal abnormalities, and mental retardation along with high parathyroid hormone (PTH) levels in the former while PTH levels are low or undetectable in acquired and congenital causes of hypoparathyroidism.[15] Pseudopseudohypoparathyroidism is a different entity and characterized by increased PTH level but normal serum calcium and phosphorus levels.[8]

Hypomagnesaemia, Vitamin-D resistance, Vitamin-D deficiency, and renal failure must be excluded as the cause of the biochemical abnormalities.[15]

Treatment includes calcium and Vitamin D supplementation as well as symptomatic treatment. PTH replacement has not yet been approved. Adequate treatment of hypoparathyroidism can result in marked clinical improvement and prevent occurrence of life-threatening complications of severe hypoparathyroidism like spontaneous intracerebral bleed.[16]

CONCLUSION

Hypoparathyroidism as a cause of basal ganglia calcification has a good prognosis if timely diagnosed and treated.[17] Patient of hypoparathyroidism with BGC can present with seizures as the only presentation without other signs of hypocalcemia like tetany as well as without extrapyramidal features. Thus, one should not rule out hypoparathyroidism in the absence of other signs of hypocalcemia and extrapyramidal features. Biochemical analysis pertaining to hypoparathyroidism must be done.

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