

# LoCa LoPa myelopathy: is prevention better than cure?

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## Summary

**Manifestations of primary hypoparathyroidism are produced by neuromuscular irritability or by extraosseous calcifications. We present a patient of primary hypoparathyroidism who had extensive calcification of brain parenchyma, and was suffering from chronic, generalised and progressive stiffness of body due to cervical compressive myelopathy, caused by calcification of posterior longitudinal ligament and ligamentum flavum. By presenting this case we wanted to emphasize the usefulness of meticulous clinical examination to differentiate the stiffness caused by myelopathy from that which is caused by possible coexisting extrapyramidal disorder. This case presentation also builds the hypothesis that early diagnosis and institution of early and appropriate treatment has potential to prevent the complications arising from extraosseous calcifications in patients with primary hypoparathyroidism.**

*KEY WORDS:* hypoparathyroidism; myelopathy; extraosseous calcification.

## Introduction

Primary hypoparathyroidism has variable presentations, mostly produced by neuromuscular irritability due to the resulting hypocalcaemia. These include perioral and digital paresthesias, muscle cramps, tetany, laryngospasm and bronchospasm. Other manifestations like parkinsonism, dystonia, ataxia and spastic paraplegia are caused by extraosseous calcifications. As an example, basal ganglia calcifications due to hypoparathyroidism can present as parkinsonism (1, 2). We present a patient of primary hypoparathy-

roidism who had spastic quadriplegia due to cervical compressive myelopathy caused by calcification of posterior longitudinal ligament and ligamentum flavum.

## Case report

A 48-year-old ethnic Kashmiri, who worked as a labourer in ammunition depot since 1992, presented to us, in December 2015, with progressive stiffness of body for the past six years. Initially he had insidious onset stiffness of the back, bent posture, and gait difficulty. Onset was also remarkable for generalized tonic clonic seizures, which occurred on three to four occasions. He took anticonvulsants for a short duration of several months only, and then remained seizure free, without anticonvulsants. His stiffness of limbs and back increased progressively and, over the past nine months to a year, he attained a 'stiff and curled up' posture, and got totally bedridden, and dependent. Before falling ill he used to play cricket, volleyball, and kabaddi, regularly. His past history was remarkable for bilateral cataract surgery, done some years before the onset. His father had gout, one of the maternal uncles, who was 80 years of age, had tremor disorder since 60 years of age, and one of his paternal uncles had leprosy. Review of his previous medical records revealed: persistent low serum calcium (LoCa), low serum parathormone (LoPa), high serum phosphorus, and extensive parenchymal calcification of brain on head CT scan (Figure 1 A, B). With the diagnosis of primary hypoparathyroidism, he had received cholecalciferol in the dose of 200 to 500 units per day and elemental calcium in the dose of 400 milligram per day, along with muscle relaxants and levodopa, with no relief. On examination, his higher functions, speech, and cranial nerves were normal. Motor power and cerebellar functions could not be assessed formally because of great stiffness (spasticity versus rigidity), that was more in lower than upper limbs. Small muscles of his hands were weak and wasted. Vibration sense was reduced at ankles and position sense was impaired at great toes. His planter responses were extensor. Rest of his general and systemic examination was unremarkable. CT scan of cervical spine showed calcification of the posterior longitudinal ligament and ligamentum flavum causing spinal canal stenosis (Figure 1 D). MRI of cervical spine, in addition, revealed signal change in the spinal cord at the site of compression by hypertrophied ligaments (Figure 1 C). His blood biochemistry was remarkable for hypocalcemia, with serum calcium of 5.2 mg/dL, hyperphosphatemia with serum phosphorus of 5.8 mg/dL, and very low parathormone level 2.5 pg/mL. He had normal serum magnesium level of 1.9 mg/dL, and normal 25(OH) Vit D<sub>3</sub> level of 43ng/mL. Rest of his blood tests were within normal range. We made a diagnosis of primary hypoparathyroidism with cervical myelopathy due to calcification of posterior longitudinal ligament and ligamentum flavum, with extensive brain calcification. We administered, orally, calcitriol

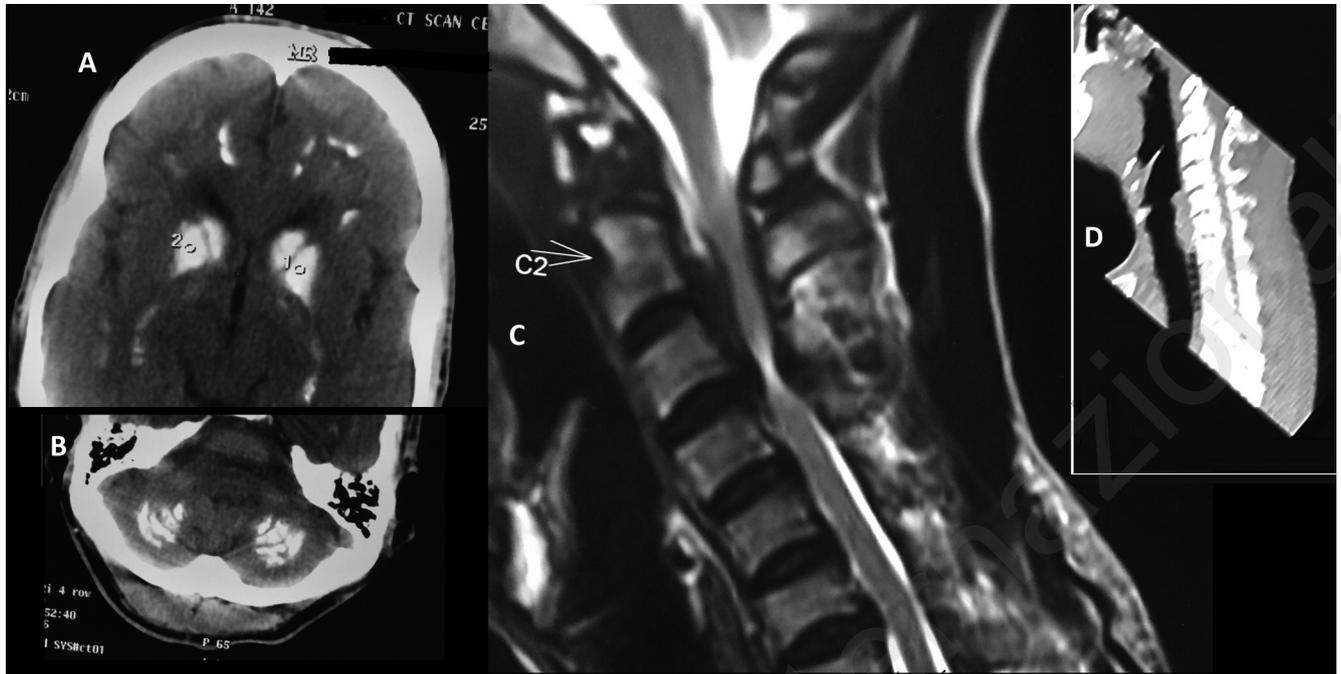


Figure 1 - A, B) CT scan of brain showing extensive parenchymal calcification of brain. C) MRI of cervical spine showing calcification of the posterior longitudinal ligament and ligamentum flavum causing spinal canal stenosis and signal change in the spinal cord. D) CT scan of cervical spine showing calcification of the posterior longitudinal ligament and ligamentum flavum causing spinal canal stenosis.

in dose of 1.0 to 1.5 microgram per day, cholecalciferol in the dose of 800 units per day, along with elemental calcium of 1000 mg per day, and tried to maintain serum levels of calcium between 8.0 to 8.5 mg per decilitre. On the advice of neurosurgeon he underwent laminectomy and decompression of second to sixth cervical vertebrae. After the decompression surgery, there was some improvement of power in the upper limbs, and of stiffness in lower limbs. Although, with optimization of medical therapy and with surgical decompression, there was significant improvement in well being as reported by the patient and his family members, still he was left with a lot of disability.

## Discussion

In our patient, two errors, those contributed to the poor outcome, had occurred before presenting to us. One, the European society of endocrinology (ESE) clinical guideline (3) for treatment of primary hypoparathyroidism, had not been adhered to. During this period, he had been receiving vitamin D in doses much lower than the recommendations, and had never reached the recommended goals. Second, the widespread calcification of brain parenchyma had served as a red herring. Generalized and marked stiffness along with this widespread calcification of brain had possibly been taken as extrapyramidal manifestation of hypoparathyroidism. It was the more detailed and finer examination which helped us to find out the clues about the presence of cervical myelopathy, as well. These clues included differential stiffness in upper and lower limbs (upper limbs >> lower limbs), wasting of small muscles of hands, loss of vibration and position sensations in lower limbs, extensor planter responses, and normal cranial nerve examination. Thus, subtle pointers to the pres-

ence of myelopathy, those we alluded to above, had been easily overlooked.

On reviewing the literature, we found only two cases of myelopathy due to calcification of posterior longitudinal ligament and ligamentum flavum, in patients with primary hypoparathyroidism (4, 5).

ESE guidelines are silent about the prevention and treatment of delayed complications of primary hypoparathyroidism, namely those due to the widespread calcifications, as were seen in our patient. Studies need to be done to find out whether early identification and early treatment to achieve the goals as recommended by the guidelines can prevent the occurrence of these ectopic calcification-related complications. In other words, is prevention of LoCa LoPa myelopathy better than its cure?

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