Striopallidodentate calcifications in a Chinese elderly woman with parkinsonism and dementia: a case report

CASE REPORT

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INTRODUCTION

Striopallidodentate calcification (SPDC), also known as Fahr's disease, is a rare disorder characterised by bilateral symmetrical calcification in the basal ganglia with or without involvement of the cerebellar dentate nucleus.[1] The exact aetiology is still unknown. Movement disorder is the most common clinical presentation.[2] We report on an elderly Chinese woman with a history of parkinsonism and dementia who was subsequently found to have marked SPDCs in her computed tomographic (CT) scan.

CASE REPORT

In May 2005, a 79-year-old Chinese woman living in a nursing home with a history of parkinsonism, dementia, and duodenal ulcer presented to the Queen Mary Hospital with sudden onset of fever and confusion. She had a history of a fall with minor head injury 1 week before. She was diagnosed as having a urinary tract infection with *Escherichia coli* and was treated with a course of cefuroxime. In view of the acute onset of confusion and the history of head injury, an urgent CT scan of the brain was performed. The CT scan showed cerebral atrophy and marked bilateral symmetrical basal ganglia and cerebellar calcifications compatible with SPDC (Figure). She had not had any previous CT scans so there were no films available for comparison. She had a normal calcium level (adjusted calcium level, 2.45 mmol/L) and parathyroid hormone level (39 pg/mL; normal range, 11-54 pg/mL). Other blood tests, including renal and liver function tests, were unremarkable. Clinically she had a mask-like face, severe lead-pipe rigidity and cogwheel rigidity but no tremor. Her rigidity did not improve with upward titration of L-dopa during her admission. She developed dysphagia and was assessed as unfit for oral feeding using a video fluoroscopy swallowing study. Nasogastric tube feeding was started and a gastrostomy insertion planned. All along she remained chair-ridden and totally dependent in her activities of daily living, showing limited rehabilitation potential. She had no close relatives and all her welfare issues were handled by the social worker. During the latter period of hospitalisation she developed severe pneumonia which did not respond well to antibiotics and died 2 months after admission.

DISCUSSION

Seldom in the field of medicine does a disease have such varied nomenclature.[1] Previously known as Fahr's disease (a misnomer), most clinicians prefer to call it SPDC yet there are actually more than 30 names in the literature describing the same condition. The more commonly used names

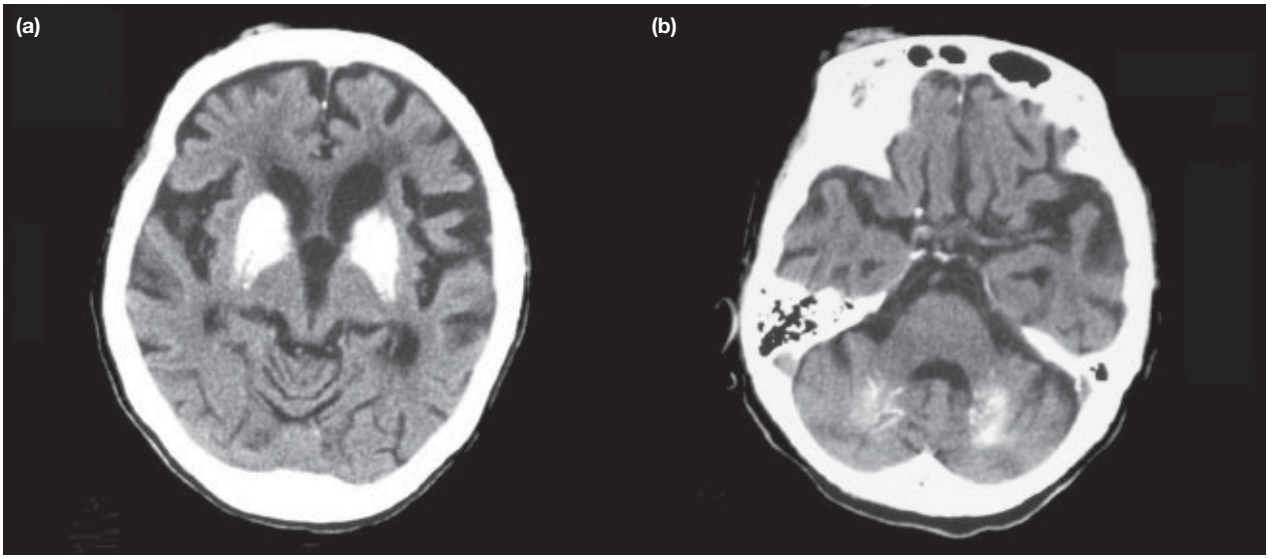

Figure. Computed tomographic scans showing (a) bilateral symmetrical basal ganglia calcifications, and (b) symmetrical calcifications in the cerebellum

include idiopathic basal ganglia calcification, nonarteriosclerotic cerebral calcification, striopallidodentate calcinosis, and cerebrovascular ferrocalcinosis.

The existence of so many different names reflects the limited understanding of this disease. To date, pathological studies have demonstrated the presence of other elements apart from calcium including mucopolysaccharides, arsenic, cobalt, aluminium, copper, iron, lead, magnesium, manganese, phosphorus, zinc, and silver.[3] The exact reason why calcium and other substances are deposited in the basal ganglia is still a mystery.

Striopallidodentate calcifications can be classified as primary or secondary.[1] The primary form can be autosomal dominant, familial, or sporadic. Secondary SPDC may be associated with diseases such as hypoparathyroidism, pseudohypoparathyroidism, pseudo-pseudohypoparathyroidism, or hyperparathyroidism.[4,5] It can also occur in some developmental conditions, lead and carbon monoxide poisoning, inflammatory and infective disorders such as acquired immunodeficiency syndrome, and in systemic lupus erythromatosis.[1] Our patient had normal calcium and parathyroid hormone levels so an association with parathyroid hormone disorder was unlikely in this case. A lack of close relatives also made it difficult to determine whether her condition was hereditary.

Our patient had parkinsonism and dementia. In the series reported by Manyam et al,[2] movement disorders, especially parkinsonism, were the most common neurological association. Other neurological associations reported were cognitive disorders, speech disorders, cerebellar signs, pyramidal signs, abnormal gait, and psychiatric manifestations.[2,6] However, about two thirds of SPDC patients were asymptomatic.

Striopallidodentate calcifications is underreported, partly because many cases are asymptomatic and partly because not all patients with parkinsonism have brain CTs performed, especially in places where CT scanning is not easily available. However, it is suggested that when parkinsonism, dementia, and cerebellar signs develop, a CT scan is helpful for ruling out SPDC.[1] If there is SPDC, calcium and parathyroid hormone levels should be checked to exclude disorders of calcium metabolism.[1] Hitherto, no successful treatment has been found for SPDC per se when there are no treatable secondary causes such as parathyroid hormone disorders.

Many questions concerning SPDC remain unanswered. We do not know whether there are ethnic differences in the prevalence and clinical manifestations of SPDC. To date, only one Chinese case report has been found in the literature.[6] Information about the prognosis and natural

progression of SPDC is lacking, especially in the elderly population. Future study of SPDC is needed to address these questions.

References

1. Manyam BV. What is and what is not 'Fahr's disease'. *Parkinsonism Relat Disord* 2005;11:73-80.
2. Manyam BV, Walters AS, Narla KR. Bilateral striopallidodentate calcinosis: clinical characteristics of patients seen in a registry. *Mov Disord* 2001;16:258-64.
3. Smeyers-Verbeke J, Michotte Y, Pelsmaeckers J, Lowenthal A, Massart DL, Dekegel D, et al. The chemical composition of idiopathic nonarteriosclerotic cerebral calcifications. *Neurology* 1975;25:48-57.
4. Illum F, Dupont E. Prevalences of CT-detected calcification in the basal ganglia in idiopathic hypoparathyroidism and pseudohypoparathyroidism. *Neuroradiology* 1985;27:32-7.
5. Margolin D, Hammerstad J, Orwoll E, McClung M, Calhoun D. Intracranial calcification in hyperparathyroidism associated with gait apraxia and parkinsonism. *Neurology* 1980; 30:1005-7.
6. Chiu HF, Lam LC, Shum PP, Li KW. Idiopathic calcification of the basal ganglia. *Postgrad Med J* 1993;69:68-70.