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CONTENTS

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ORIGINAL ARTICLES

Bilateral HIV related ocular surface squamous neoplasia: a paradigm shift?.....	R Masanganise, A Mukome, J Dari, R Makunike-Mutasa.....	23
Use of cotrimoxazole prophylaxis in HIV infected in-patients at a referral hospital.....	S Khoza, V Mkudu, J Mthethwa, R Bulaya-Tembo, CFB Nhachi.....	26
Annual distribution of births and births outcomes at Harare Maternity Hospital, Zimbabwe.....	S Feresu.....	30
A potentially treatable cause of dementia.....	PL Katsidzira, T Machiridza, A Ndlovu.....	41

CASE REPORTS

Post operative fatal hypothermia in hydranencephaly with pre-operative hypothermia and a 7th nerve palsy: A case report.....	A Musara, KKN Kalangu.....	44
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NOTES AND NEWS

<i>Central African Journal of Medicine</i>	48
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CASE REPORTS

A potentially treatable cause of dementia

L KATSIDZIRA, T MACHIRIDZA, A NDLOVU

Abstract

Idiopathic hypoparathyroidism is an uncommon medical condition, which may present with dementia. This may be misdiagnosed as 'senile dementia' in elderly patients. We report such a case, made more remarkable by associated extensive cerebral calcification and we review the background literature.

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*Department of Medicine
University of Zimbabwe College of Health Sciences
P O Box A178
Avondale, Harare
Zimbabwe*

Corresponding to:

*Dr Leolin Katsidzira
Department of Medicine
University of Zimbabwe College of Health Sciences
P O Box A178, Avondale, Harare
Zimbabwe
E-mail: lkatsidzira@hotmail.com*

Introduction

Dementia is under-diagnosed in routine clinical practice in the developing world. Symptoms are often attributed to normal ageing, even though they may be a result of underlying medical conditions. We report a case of a patient with extensive cerebral calcification due to hypoparathyroidism who was being managed for dementia. While cases of extensive calcification in parathyroid hormone disorders have been reported sporadically in the literature over the years, the current case highlights the potential diagnostic pitfalls when patients present with dementia.¹⁻⁵

Case Report

A 77 year old woman presented to a referral hospital in Harare, Zimbabwe, with a 4 month history of progressive memory impairment and confusion. She had developed auditory and visual hallucinations in the 2 months preceding presentation. Her mental state had rapidly deteriorated over this period; she would occasionally forget her children and had developed paranoia. She had not been communicating in the 3 days prior to presentation. She was being managed for 'senile dementia' and was on amitryline 25mg and haloperidol 5mg daily. She had no history of hypertension, diabetes mellitus or previous strokes and she did not smoke or use alcohol. On examination she was afebrile and her blood pressure was 110/70mmHg. She had a Glasgow coma scale of 10 but there were no focal signs. There were coarse crepitations over the right lung base.

The results of investigations done were as follows; white cell count $14.88 \times 10^3/\mu\text{L}$ (neutrophils 83.6%, lymphocytes 9.3%, monocytes 6.9%, eosinophils 0.1%, and basophils 0.1%), haemoglobin 12.4g/dl, platelets $279 \times 10^3/\mu\text{L}$, sodium 143mmol/L, potassium 3.4mmol/L, urea 13.8mmol/L, creatinine 108mmol/L and glucose 8.3 mmol/L. She was commenced on intravenous fluids and antibiotics for the pneumonia. Computerised tomography scan of the brain (Fig 1) demonstrated extensive symmetrical calcification involving the dentate nuclei in the cerebellum, the centrum semi ovale, basal ganglia and peri-ventricular areas. Subsequent investigations revealed hypocalcemia and hyperphosphataemia [calcium 1.04 mmol/L(2.1-2.55), albumin 26g/l and phosphate 2.42mmol/L]. Intravenous calcium gluconate was commenced but she continued to deteriorate, and died 11 days after admission. The final diagnosis was hypoparathyroidism with cerebral calcification and pneumonia.

Figure 1a: CT scan demonstrating bilateral symmetrical calcification of the dentate nuclei in the cerebellum.

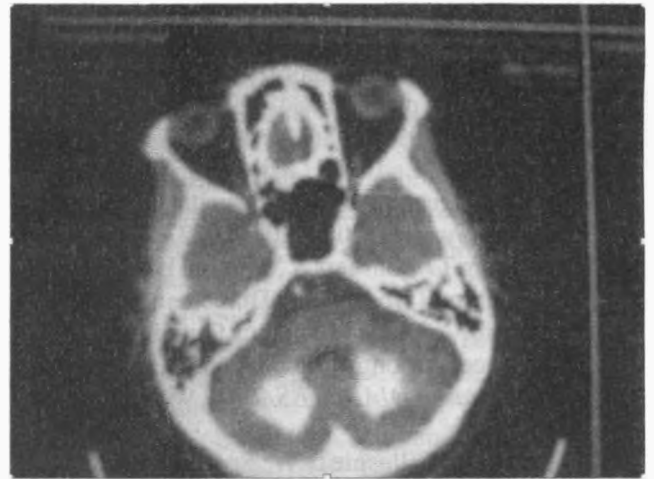


Figure 1b: Extensive symmetrical basal ganglia and peri-ventricular calcification.

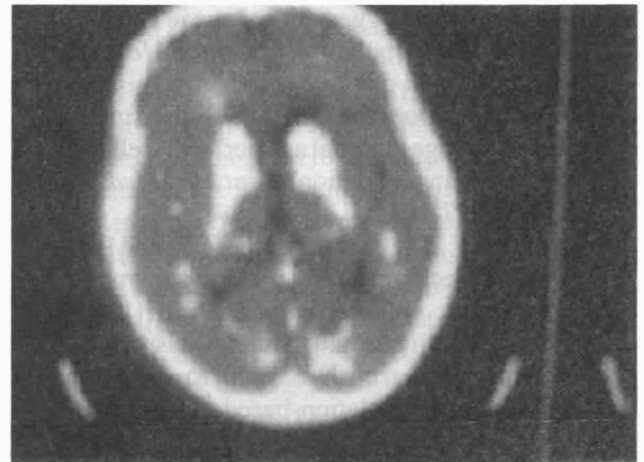
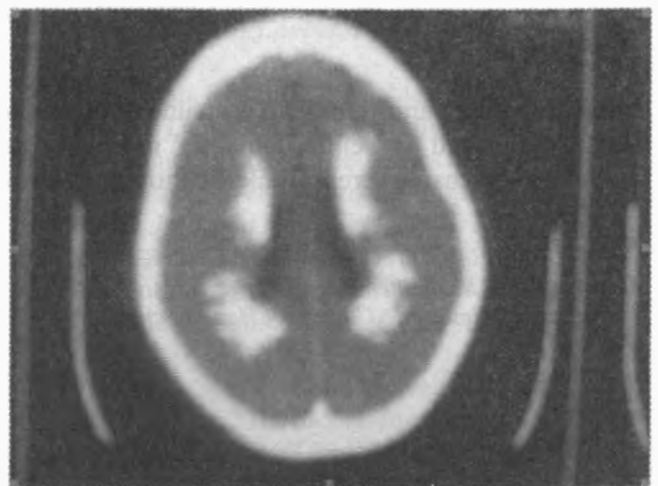


Figure 1c: Peri-ventricular calcification extending to involve the centrum semi-ovale bilaterally.



Discussion

Cerebral calcification is associated with a diverse group of conditions including neurocysticercosis, toxoplasmosis, healed tuberculous granulomas, cryptococcomas, haemochromatosis, tuberous sclerosis, myotonic muscular dystrophy, tumours, aneurysms and Fahr's disease.⁶ However, symmetrical calcification involving the dentate nuclei and the corona radiata is considered pathognomonic of hypoparathyroidism or pseudohypoparathyroidism². The current case serves not only as a reminder of this little known fact, but more importantly, amply illustrates the dictum that primary dementia should be a diagnosis of exclusion.

Hypocalcaemia, which is usually caused by hypoparathyroidism is one of the many conditions which can present with dementia.^{3,7,8} The dementia is characterised by "poor concentration, impaired memory, disorientation and apathy"⁹. Dementia in elderly patients can also be due to nutritional deficiencies (e.g vitamin B12 deficiency, pellagra), thyroid disease, chronic subdural haemorrhage, normal pressure hydrocephalus, HIV infection or intracranial tumours among other causes. More commonly, it is a result of the primary dementia syndromes such as Alzheimer's disease, vascular dementia, Lewy body dementia and frontotemporal dementia. An attempt should be made to differentiate them in clinical practice as management strategies may differ.

The proper evaluation of patients presenting with dementia includes meticulous history taking, including collateral history from close family members and a comprehensive physical and mental state examination. Whenever possible, the following investigations should be performed; full blood count, erythrocyte sedimentation rate, urea and electrolytes and thyroid function tests.¹⁰ A lumbar puncture should be carried out if clinically appropriate. Computed tomography scan of the brain may be considered, especially when the history and examination suggest secondary causes such as a chronic subdural haemorrhage or intra-cranial tumours. This is often unavailable or costly in resource limited settings and history taking becomes even more crucial, as it allows investigations to be targeted.

The unavailability of more extensive investigations was apparent in the management of our patient, as it was not possible to perform a parathyroid hormone assay. However, the presence of hypocalcaemia and hyperphosphataemia with normal renal function, and the pathognomonic CT findings virtually confirms the diagnosis. Historically, the criteria for diagnosing idiopathic hypoparathyroidism has been a low serum calcium in the presence of high serum inorganic phosphorus level and normal renal function.¹¹ The major differential diagnosis of the radiological findings in our patient would be bilateral striopallidodentate calcinosis, commonly known as Fahr's disease¹². However, this rare familial disease is not associated with calcium and phosphate abnormalities despite

causing extensive cerebral calcification.

There are two ways through which hypoparathyroidism give rise to dementia. Firstly, hypocalcaemia may present with symptoms of dementia, which resolve after correction.¹⁹ Less recognised, is the fact that chronic hypoparathyroidism is associated with long term cognitive decline, even if it is well controlled.¹³ This is often associated with the presence of cerebral calcification. Interestingly, motor and movement disorders in hypoparathyroidism are also associated with cerebral calcification. This suggests that cerebral calcification in hypoparathyroidism plays a role in the pathogenesis of the various neurological abnormalities seen in the condition.^{7,13}

It was not possible to distinguish between hypoparathyroidism and pseudohypoparathyroidism in our patient. Curiously, calcification is more common in the rarer pseudohypoparathyroidism.⁵ However, pseudohypoparathyroidism has other associated phenotypic features such as short stature, brachydactyly, and a round face apart from heterotopic calcification.¹⁴ Our patient certainly had idiopathic hypoparathyroidism, a condition which is often diagnosed late in Africa.¹⁵

Conclusion

It is crucial to rule out secondary causes of dementia in elderly patients presenting with cognitive impairment because correction some of these factors, such as hypocalcaemia can arrest or even reverse the symptoms.

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