Idiopathic calcification of the basal ganglia

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Summary: Idiopathic calcification of the basal ganglia is a rare disorder characterized by neuropsychiatric abnormalities, a movement disturbance of parkinsonian and/or choreoathetoid type and dense calcification of the basal ganglia. We report a 60 year old patient diagnosed as having delusional disorder and tardive dyskinesia who was subsequently found to be suffering from idiopathic calcification of the basal ganglia.

Introduction

Idiopathic calcification of the basal ganglia (ICBG) is a rare disorder characterized by neuropsychiatric abnormalities, a movement disturbance of parkinsonian and/or choreoathetoid type and dense calcification of the basal ganglia. Its aetiology is unknown but it is familial in some cases. Other causes associated with calcification of the basal

References


Acknowledgements

We wish to thank Dr J. Reidy, F.R.C.R., consultant radiologist, and Dr A.M. Hanby, M.R.C.Path., consultant histopathologist, Guy’s Hospital for help in compiling this case report.

Mr W.R. O’Flynn died suddenly and unexpectedly whilst this manuscript was in preparation. We (P.B. and J.P.) believe this case to be a celebration of his diagnostic and professional skill.
ganglia include hypoparathyroidism, pseudohypoparathyroidism, pseudosyndrome, hyperparathyroidism, tuberous sclerosis, various inflammatory and infectious conditions, such as cytomegalic inclusion disease, toxoplasmosis, encephalitis, toxic and anoxic conditions, such as lead intoxication and carbon monoxide poisoning. Tardive dyskinesia is an involuntary movement disorder associated with prolonged antipsychotic drug use and its prevalence is around 20% in psychiatric patients. We report a patient with ICBG presenting with neuropsychiatric manifestations and movement disorder misdiagnosed as delusional disorder with tardive dyskinesia.

Case report

A 60 year old married Chinese man, born in Malaysia, had no formal education and came to work as a labourer in Hong Kong around 30 years ago. He lost contact with his family in the subsequent years, hence we had no details of his family history. His psychiatric history began at age 55 when he was hospitalized for a brief psychotic episode. He presented with a 2 week history of delusion of reference and delusion of persecution. As a result of his psychotic symptoms, he hit a stranger and then attempted suicide for fear of being arrested by police. After admission into a mental hospital, a diagnosis of delusional disorder was made and he was put on chlorpromazine 50 mg daily. His symptoms subsided after a few weeks and he was subsequently discharged. He attended out-patient treatment irregularly and his compliance to drug treatment was poor.

A year later he manifested delusion of persecution and was hospitalized again. After admission, he was noticed to be dull and slow in mentation, and harboured a delusion that his neighbour would kill him. He was slightly depressed but had no sleep, appetite or weight disturbance. After receiving thioridazine 50 mg daily for a few weeks, his psychotic symptoms subsided. A year later he manifested buccal-linguomasticatory syndrome and choreoathetoid movements of his limbs and a diagnosis of tardive dyskinesia was made. Anti-psychotic drugs were discontinued but the dyskinesia persisted for 3 years.

He was subsequently referred to us for management because of the authors' special interest in movement disorder. On examination, he was slow and retarded. He had no psychotic features but his cognitive function was impaired with poor attention and concentration. He could recall two out of three items after 5 minutes. His clinical picture was consistent with a mild dementia but he refused any formal psychometric tests. Physical examination revealed moderate buccolinguodyskinesia and choreoathetoid movements of his limbs with bradykinesia and rigidity. A computed tomographic (CT) scan of the brain showed extensive bilateral calcification of the globus pallidus (Figure 1). Clinically he had no evidence of hypoparathyroidism, pseudohypoparathyroidism or pseudosyndrome. Routine laboratory studies including serum calcium and phosphorus levels, thyroid function tests and caeruloplasmin level were normal. A diagnosis of idiopathic basal ganglia calcification with dementia was made.

Discussion

Dementia, a schizophrenic-like psychosis and organic mood disorder have been described as neuropsychiatric manifestations in ICBG. Cummings et al. described two distinct manifestations of ICBG. An early-onset type appeared between the ages of 20 and 40 years in which a schizophrenic-like psychosis was noted at initial presentation. Late-onset ICBG presented between the ages of 40 and 60 years with dementia and a movement disorder of the parkinsonian and/or choreoathetoid type. König suggested that time of occurrence of the calcification might be important as depositions in early life may produce oligophrenia while at later dates, mood changes occurred and at still later dates, dementia syndromes predominated. In many cases of ICBG, the globus pallidus was a prominent area of involvement but no correlation of symptomatology with localization or extent of calcification was found.

Our patient had bilateral calcification of the globus pallidus, a movement disorder in the form of parkinsonian and choreoathetoid type and a

Figure 1 CT scan of brain showing basal ganglia calcification.
picture of brief psychotic episodes evolving into a picture of late-onset ICBG described by Cummings et al.\textsuperscript{1}

He had exposure to antipsychotic drugs for nearly 2 years but these were already discontinued for 3 years when he was referred to us. As his psychotic illness and treatment with antipsychotic drugs preceded the abnormal movements, it remains possible that part of his movement disorder can be attributed to tardive dyskinesia. Furthermore, excessive susceptibility of ICBG patients to the extrapyramidal effects of antipsychotics has been noted by various authors\textsuperscript{1,2} as some of these patients had also received antipsychotic treatment prior to the onset of their movement disorder. However, the patient's parkinsonian features could not be accounted for by tardive dyskinesia and is likely to be related to ICBG.

We hope that this report will heighten clinicians' awareness of this rare entity and we recommend that ICBG should be considered as a differential diagnosis in psychiatric patients with a movement disorder.

References


Pulmonary alveolar microlithiasis presenting with chronic cough

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Summary: A case of a 10 year old boy with pulmonary alveolar microlithiasis is presented. Although most children with this disease are asymptomatic, our patient had persistent cough for more than 3 years. It is likely that his chronic cough was a direct consequence of the disease.

Introduction

Pulmonary alveolar microlithiasis (PAM) is a rare disease of unknown etiology characterized by the presence of multiple microscopic stones within the pulmonary alveoli. Most patients with PAM have no symptoms and many others have minor or questionable symptoms when their disease is first diagnosed. The pattern in our patient follows that described in many cases in the literature but chronic cough has been seldom reported.\textsuperscript{1,2}

Case report

A 10 year old boy had been complaining of dry cough for 6 months and his chest X-ray showed bilateral dense miliary mottling. He was diagnosed as having miliary tuberculosis by a local physician. The cough persisted following 6 months of antituberculosis therapy and the chest X-ray remained unchanged. He was referred to our hospital in
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Postgrad Med J 1993 69: 68-70
doi: 10.1136/pgmj.69.807.68

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