- AMP in response to parathyroid hormone. *Journal of Clinical Investigation*, **48**, 1832.
- CRANIN, A.A. & KATZ, H.E. (1967) Pseudohypoparathyroidism—Report of a case. *Journal of the American Dental Association*, **74**, 741.
- CROFT, L.K., WITKOP, C. J. JR. & GLAS, J. (1965) Pseudohypoparathyroidism. *Oral Surgery*, **20**, 758.
- DENT, C.E., RICHENS, A., ROWE, D.E.J. & STAMP, T.C.B. (1970) Osteomalacia with long-term anticonvulsant therapy in epilepsy. *British Medical Journal*, **4**, 69.
- ELLSWORTH, R. & HOWARD, J.E. (1934) Studies on the physiology of the parathyroid glands. VII. Some responses of normal human kidneys and blood to intravenous parathyroid extract. *Bulletins of Johns Hopkins Hospital*, **55**, 296.
- GREENBERG, M.S., BRIGHTMAN, V.J., LYNCH, M.A. & SHIP, J.I. (1969) Idiopathic hypoparathyroidism, chronic conditions and dental hypoplasia. *Oral Surgery*, **28**, 42.
- HUNTER, J., MAXWELL, J.D., STEWART, D.A., PARSONS, V. & WILLIAMS, R. (1971) Altered calcium metabolism in epileptic children on anticonvulsants. *British Medical Journal*, **4**, 202.
- KRUSE, R. (1968) Osteopathien bei antiepileptischer langzeittherapie (Vorläufige Mitteilung). *Mtschr Kinderheilk*, **116**, 378.
- MANN, J.B., ALTERMAN, S. & HILL, A.G. (1962) Albright's hereditary osteodystrophy comprising pseudohypoparathyroidism and pseudopseudohypoparathyroidism. *Annals of Internal Medicine*, **56**, 315.
- RASMUSSEN, H. (1968) In: *Textbook of Endocrinology* (Ed. by Robert H. Williams), p. 920. W. B. Saunders Company, Philadelphia, London, Toronto.
- RICHENS, A.S. & ROWE, D.J.E. (1970) Disturbance of calcium metabolism by anticonvulsant drugs. *British Medical Journal*, **4**, 73.
- STONES, H.H. (1966) In: *Oral and Dental Disease* (Ed. by E. D. Farmer and F. E. Lanton) p. 869. E. and S. Livingstone.
- SUNDE, D.E. & HALS, E. (1961) Dental changes in a patient with hypoparathyroidism. *British Dental Journal*, **111**, 112.
- WITKOP, C.J. (1968) Gardner's syndrome and other osteognathodermal disorders with defects in parathyroid function. *Journal of Oral Surgery*, **26**, 639.

Postgraduate Medical Journal (October 1972) **48**, 636–639.

Calcification of the basal ganglia apparently presenting as a schizophreniform psychosis

PETER HALL

M.B., Ch.B., Ph.D., M.R.C.Psych., D.P.M.

Powick Hospital, Worcester

Summary

A patient with gross basal ganglia calcification is described, whose condition was associated with severe psychotic symptoms and in whom the clinical picture was not typical of hypoparathyroidism, pseudohypoparathyroidism or pseudo-pseudohypoparathyroidism.

Calcification of the basal ganglia may occur in a variety of infections, toxic and metabolic disorders, particularly hypoparathyroidism, pseudohypoparathyroidism, toxoplasmosis and some anoxic conditions (Moskowitz, Winickoff & Heinz, 1971).

Though there is doubt as to the precise diagnosis in the present case, familial calcification appears to be the most probable.

Case history

Mrs X, a 41-year-old widow, wrote to the Medical Officer of Health in 1955 in the belief that her three children had venereal disease and were also suffering from cancer. She believed that their condition was infectious, that things had gone too far for anything to be done for them and that the local people grinned and whispered behind their hands when they passed and that she and the children would suffer an early, lonely and agonizing death because of her wickedness in having had two illegitimate children. She had heard voices at night commenting on her 'pink

eyes' (which she regarded as a sign of venereal disease) and also had visual hallucinations of 'worms everywhere'. A diagnosis of schizophrenia was made and she was treated as an in-patient by electroconvulsive therapy with some improvement. She was re-admitted in September 1963 in a cachectic, dehydrated and pitiable state; whimpering, grimacing and protruding her tongue. Her auditory hallucinations this time were of voices saying 'act normal' and she was treated with thioridazine and further ECT and was discharged, apparently much improved after some weeks.

She was again admitted in April 1965 when she was disorientated in time and place, grossly dysarthric, apparently demented and with marked parietal lobe dysfunction. She improved with anti-Parkinsonian drugs but not sufficiently to cope with her home and was placed in a Local Authority Home but was once again admitted in December 1970 after she had become very restless and paranoid. On this occasion she showed extreme dysarthria and oral dyskinesia, an upper motor neurone right-sided facial palsy and moderately severe right-sided hemiparesis. She also again believed that she was suffering from venereal disease. Psychological testing (WAIS) showed her to have a verbal IQ of 82, a performance IQ of 69 and marked impairment of visuo-spatial

performance. An electroencephalogram showed dominant activity at 12 cycles/sec, blocking on eye opening and with some central theta (3–6 cycles/sec) activity and also some fast (24 cycles/sec) activity frontally. Skull X-rays (Figs. 1–3) showed gross calcification of the caudate and dentate nuclei and excretion urography a double left renal pelvis and ureter but no renal calcification. X-rays of her chest, abdomen and hands were normal, as was an electro-

FIG. 1.

FIG. 2.

FIG. 3.

cardiogram, liver function tests, serum proteins and cholesterol, blood urea (30 mg/100 ml) and blood sugar. Serum electrolytes and serial calcium levels were normal (9–9.4 mg/100 ml) as were serial phosphorus levels (4–4.6 mg/100 ml) and serum alkaline and acid phosphatases. Wassermann reaction was negative and haematology normal, as was protein-bound iodine (8 μ g/100 ml). Serum creatinine was 1.4 mg/100 ml, while urinary creatinine was 38 mg/100 ml (creatinine clearance 34 ml/min).

In view of her skull X-rays and a history of a partial thyroidectomy for multiple cystic thyroid adenomata on 3 August 1954, Mrs X was transferred to the Metabolic Research Unit, East Birmingham Hospital (Professor J. Hardwicke). At surgery there had been no clinical signs of toxicity and histology had shown 'simple nodular colloid goitre'. Further investigations were carried out at the Metabolic Research Unit, where Mrs X's hands, feet and metacarpals were found to be quite normal and the previous clinical and other signs confirmed. Further serum calcium, phosphate, magnesium and alkaline phosphatase levels were normal, as was an amino acid chromatogram. A further PBI was also normal, as was a triosorb test (26%) of thyroid function and serum iron, folate and iron-binding capacity were all within normal limits. Urine calcium, phosphate and phosphate/creatinine clearance ratio (0.3) were normal but it proved impossible to do a parathormone infusion test because of her occasional incontinence and the consequent inability to ensure complete collections of urine. It was concluded unlikely that Mrs X suffered from any gross disorder of calcium metabolism nor was she thought to show the physical

defects (other than her short stature) of patients with pseudo, or pseudo-pseudohypoparathyroidism.

Attempts to investigate Mrs X's relatives revealed a complex story. Mrs X herself was illegitimate; her parents unknown and her only relative—a much older 'sister' (?) lives in the depths of rural 'Border country' 50 miles away. She failed to answer several letters but, according to Mrs X's son (F) she is well. Mrs X's own first two children were also illegitimate (by a farmer whose wife was currently in a mental hospital) and the eldest girl (A) who, again, did not reply to any letters, has herself a son and (according to F) is well. The youngest girl (B) apparently eventually graduated at a northern University and was treated for (?) schizophrenia in Wales and again at an East Anglian hospital with electroconvulsive therapy and apparently improved. Her electroencephalogram was said to be normal and psychological testing to show no organic impairment although, like her mother, she had apparently suffered from visual hallucinations and her condition had been attributed to the strains of her work and background and a preceding attack of influenza. Quite understandably, no further investigations (including skull X-rays) had been carried out and Miss B had subsequently moved to Bristol and her whereabouts were unknown.

Mrs X had married a much older widower—an intellectually dull and bad-tempered farm labourer—in her late 20s and thus acquired two mentally defective step-sons (C and D) who had been in a colony since early childhood. Her husband died of carcinoma in 1954 but had fathered two further children—a daughter (E—a dull girl with an IQ of 64) and a son (F). E had married a Serviceman who was posted overseas but (according to F, who remained single and kept in touch with his mother) she was well and also had some children. The son (F), aged 22, was therefore the only member of the family accessible to investigation and he had no complaints other than a slight tremor and some other anxiety symptoms. His skull X-ray, electroencephalogram and serum calcium and phosphorus levels proved to be quite normal.

Comment

This patient presented with a schizophreniform psychotic illness for which (like that of her daughter—B) there was, at first sight, more than adequate psychodynamic cause. The clinical picture, however, gradually developed into an increasingly obvious one of organic cerebral disease with marked extrapyramidal features and severe calcification of the basal nuclei. Although she had been treated with a variety of psychotropic drugs over the years 1955–70, it seems very doubtful indeed if she took them regularly and the only likely culprit for her Parkinsonism

was chlorpromazine, of which she was supposed to be taking 100–200 mg/day (a modest dose) between 1962 and 1964. It is possible that she suffers from cerebral arteriosclerosis as she is moderately hypertensive but, despite her previous thyroidectomy, Mrs X never showed any convincing evidence of the hypoparathyroidism which may produce basal nuclei calcification and is well known to sometimes present with psychiatric symptoms. Other than for her short stature (4' 10") she also shows no clinical evidence of pseudohypoparathyroidism—the familial syndrome of end-organ unresponsiveness to parathyroid hormone—or of pseudo-pseudohypoparathyroidism. Whilst the exact diagnosis must remain in doubt, due in part to the extreme difficulty in obtaining a reliable family history and follow-up in this well-nigh totally inarticulate patient, it is possible that Mrs X suffers from familial calcification of the basal ganglia (Moskowitz, Winickoff & Heinz, 1971) and that her daughter (B), who seems to have presented with a somewhat atypical schizophreniform illness closely resembling Mrs X's original presentation, may also suffer from the same condition. A recent study (Moskowitz, Winickoff & Heinz, 1971) of the condition reports that various psychiatric symptoms were noted in five of twenty-eight patients but not apparently as psychotic or presenting ones. The syndrome is rare and the few families described by previous workers (Roberts, 1959; Mathews, 1957) apparently varied considerably in the precise genetics of the condition. Another possibility is that she may, after all, be a case of pseudo-pseudohypoparathyroidism, and a recent review (Dudley & Hawkins, 1970) suggests that occasional patients with this condition develop mineralization of the globus pallidus and dentate nuclei with not only calcium deposited but also iron and trace elements such as phosphorus, zinc and cobalt. In this condition one sees the physical and mental changes of pseudohypoparathyroidism, i.e. a significant family history, short stature, round face, obesity, mental deficiency and short metacarpal bones—but with normal serum calcium, phosphorus concentrations (Albright, Forbes & Henneman, 1952). Calcification of the basal ganglia is therefore much rarer than in hypoparathyroidism or pseudohypoparathyroidism. This case illustrates the importance of early skull X-ray and other physical investigation of atypical psychoses, particularly those where (as in Mrs X's case) there are features of possible clouding of consciousness or symptoms such as visual hallucinations which are, in fact, extremely rare in functional psychoses.

Acknowledgments

I am indebted to several colleagues for their help and advice with the investigation of this patient, notably Dr

J. Young, Dr D. J. Beale and Dr A. Vickers and Mr Ian Malcomson.

References

- ALBRIGHT, F., FORBES, A.P. & HENNEMAN, P.H. (1952) Pseudo-pseudo hypoparathyroidism. *Transactions of the Association of American Physicians*, **65**, 337.
- DUDLEY, A.W. & HAWKINS, H. (1970) Mineralisation of the central nervous system in pseudo-pseudo hypoparathyroidism. *Journal of Neurology, Neurosurgery and Psychiatry*, **33**, 147.
- MATHEWS, W.B. (1957) Familial calcification of the basal ganglia with response to parathormone. *Journal of Neurology, Neurosurgery and Psychiatry*, **20**, 172.
- MOSKOWITZ, M.A., WINICKOFF, R.N. & HEINZ, R.E. (1971) Familial calcification of the basal ganglia. *New England Journal of Medicine*, **8**, 72.
- ROBERTS, P.D. (1959) Familial calcification of the cerebral basal ganglia and its relation to hypoparathyroidism. *Brain*, **82**, 599.

Postgraduate Medical Journal (October 1972) **48**, 639–642.

Slow recovery from carbon monoxide poisoning

D. F. ABBOTT
M.B., M.R.C.P.

Addenbrooke's Hospital, Cambridge

Summary

A case of accidental carbon monoxide poisoning due to the incomplete combustion of natural gas is reported. The patient had prolonged coma and decerebrate rigidity, but eventual recovery of neurological and intellectual function was almost complete.

Introduction

Poisoning with carbon monoxide as a result of accidental or suicidal exposure to coal gas is a frequent occurrence. In 1963 there were 1193 deaths due to accidental poisoning (Registrar General's Statistics). Since that time there has been a fall in the incidence of coal gas poisoning, so that in 1969 there were 316 deaths. This is partly due to the use of natural gas (*British Medical Journal*, 1970). Natural gas consists of methane and minor proportions of other gases and is non-toxic. However, its use may lead to carbon monoxide poisoning as a result of incomplete combustion (Report of the Inquiry into the Safety of Natural Gas as a Fuel). There has been a report of such a case in this neighbourhood (Anderson, 1970).

Carbon monoxide poisoning may result in diverse neurological and psychiatric features. The incidence of different syndromes varies from series to series. A recent report of 138 patients (Smith & Brandon, 1970) showed prolonged delirium in 20%, pyramidal signs in 19.6% and extra-pyramidal signs in 2.5%. There were no cases of decerebrate rigidity. Five patients (3.8%) developed permanent defects, of which two had dementia alone, one had dementia with Parkinsonism, and two patients had spastic hemiplegia which may have preceded exposure.

Present address: Geriatric Unit, Whittington Hospital and University College Hospital, 4 St Pancras Way, London, N.W.1.

In a series of 290 cases of severe carbon monoxide poisoning from Paris (Bour, Pasquier & Bertrand-Hardy, 1966), 33.7% developed decerebrate rigidity. The usual duration of rigidity was a few hours. The average duration of coma in their series was 16 hr and in only two patients did it exceed 24 hr. One patient was in coma for 4 days and another for 5 days. Six patients had residual neurological manifestations and twenty-three had residual psychiatric syndromes.

Case report

The patient was an 18-year-old undergraduate. He was found lying deeply comatose on the floor of his room on the first day of the Lent term, 1971. Another undergraduate in the room was found to be dead and had not been seen for the preceding 24 hr.

On arrival at Addenbrooke's Hospital the patient was stuporous but responded to painful stimuli and turned his head in response to his name being called. Deep tendon reflexes were symmetrically very brisk; both plantar reflexes were equivocal. There was marked rapid eye movement. The pupils were of normal size and reacted briskly to light. The fundi were normal. There was no neck stiffness.

There was a tachypnoea of 34/min, BP 120/90 mmHg, pulse 110/min regular, good volume. Both hands were cyanosed but there was no cyanosis elsewhere and no evidence of abnormal pinkness. There were blisters on the left thigh and the buttocks had signs of early epidermal necrolysis (Fig. 1). There was no evidence of intravenous injections. The patient's clothing was covered in vomit.

There was a strong suspicion that drugs had been used and drug overdose was initially considered to be the most likely possibility.