Comment

The clinical features of tetany, convulsions, laryngeal spasm, Parkinsonian tremor, mental changes, irritability, and cataracts all suggested the diagnosis of hypoparathyroidism. This was confirmed by the low calcium and raised phosphate levels. The alkaline phosphatase was mildly raised initially but was subsequently normal, and x-ray examination showed no evidence of rickets or osteomalacia. The blood urea was 62 mg/100 ml but there was no evidence of chronic renal failure. The initial faecal fat collection was a little high but hypocalcaemia may cause steatorrhoea (Harrison, 1970), and after the serum calcium had returned to normal the faecal fat was well within physiological limits. The patient was therefore thought to be suffering from idiopathic hypoparathyroidism. Additional evidence in support of this was the failure to detect any parathyroid hormone.

The hypocalcaemic type of cataract is said to be slightly different from the usual senile type (Kopin and Rosenberg, 1960). In most reports, however, as in this case, this was not recognized at the time of cataract extraction, and it is suggested that routine estimation of the serum calcium on all patients with cataract would disclose a larger number of hypocalcaemic patients in the older age groups. A similar search among irritable or demented patients might also show a greater incidence of hypocalcaemia. Mental changes are recorded in up to 25% of patients with idiopathic hypoparathyroidism (Bronsky *et al.*, 1958; Aurbach and Potts, 1964). These changes include anxiety, depression, increased irritability, psychoses, hallucinations, delusions, mental dulling, and dementia (de Mowbray *et al.*, 1954; Robinson *et al.*, 1954; Treusch, 1957).

The aetiology of the hypoparathyroidism in this case was not clear. There were no grounds for suspecting an autoimmune

process and no signs of Addison's disease or pernicious anaemia were evident (Hung *et al.*, 1963; Blizzard *et al.*, 1966). Antibodies to parathyroid, thyroid, adrenal, and gastric mucosa were not detected.

It is suggested that in the elderly idiopathic hypoparathyroidism may be either a primary degenerative process or secondary to arteriosclerosis and diminished blood supply and that it is more common than is at present suspected.

I am grateful to Dr. Kenneth MacLean, under whose care the patient was admitted, for permission to make this report. I should like to thank him and also Professor Morris Lessof for their help in the preparation of this paper, and Dr. R. Buckle for the parathyroid hormone assay.

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Cerebrovascular Moyamoya Disease

A. M. HOARE, A. J. KEOGH

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"Moyamoya" disease was first described in the Japanese literature between 1956 and 1963 and later in the Western literature by Kudo (1965, 1968) and Nishimoto and Takeuchi (1968). It is characterized by bilateral intracranial internal carotid artery occlusion of unknown aetiology, non-filling of the circle of Willis, and development of collaterals including a carotid rete (Kudo, 1968). The first cases described were all in Japanese patients resident in Japan or America (Leeds and Abbott, 1965). It has now been reported in patients from America (Taveras, 1969; Solomon *et al.*, 1970) and Europe (Simon *et al.*, 1968; Lepore *et al.*, 1969; Galligioni *et al.*, 1971). In 1967 Hawkins and Scott described a bilateral carotid rete and hypoplasia of the internal carotid arteries in an English patient. The report on the cerebral angiograms suggested that this patient might have had moyamoya disease. This condition has been recognized in British patients (Bickerstaff, 1972) but to our knowledge no case histories have been published.

Brook General Hospital, London SE18 4LW

A. M. HOARE, M.B., M.R.C.P., Medical Registrar (Present appointment: Senior Registrar, Queen Elizabeth Hospital, Birmingham B15 2TH)
A. J. KEOGH, M.B., F.R.C.S., Surgical Registrar, (Present appointment: Senior Registrar, Department of Neurological Surgery, Royal Infirmary, Sheffield S6 3DA)

We present two cases in which air encephalography showed gross cerebral atrophy; this is a new finding.

Case 1

A 23-year-old white British woman was well until the age of 13, when she developed numbness of the left side of her body and occasional weakness of the left hand. This persisted intermittently for two years. She was able to attend a normal school, however, and after leaving at the age of 15 worked as a punch-card operator, machine operator, and shipping clerk.

In December 1970, when 10 weeks pregnant, she fell downstairs without injury to her head, and two days later her first convulsion occurred. The fits were repeated and characterized by jerky movements of the left arm and occasionally the left leg. These attacks were thought to be focal in origin. They settled in six weeks after treatment with diazepam but recurred four weeks after the birth of her child and again in October 1971, associated this time with left-sided weakness. For the first time she developed headaches; these were right-sided, intermittent, and very severe, causing her to lie down until they improved, which was usually within one to six hours.

In December 1971 she was admitted for investigation. Examination showed a left homonymous hemionopia, slight left-sided pyramidal weakness, left-sided astereognosis, and loss of two-point discrimination. Wechsler adult intelligence tests showed full-scale I.Q. 87, verbal scale I.Q. 102, and performance scale I.Q. 70. Brachial artery pressure as measured by Doppler was 116 mm Hg, and supraorbital artery pressure was reduced at 44 mm Hg for both right and left arteries. Lumbar puncture showed nothing abnormal. E.E.G. showed flattening of all activity over the right hemisphere, and a brain scan showed increased uptake in the right posterior parietal and occipital regions. Four-vessel cerebral angiography was performed (figs. 1 and 2). There was bilateral occlusion of the internal carotid artery at the level of the syphon and of the middle, anterior, and posterior cerebral arteries. The anterior cere-

FIG. 1—Case 1. Left carotid angiogram.

FIG. 2—Case 1. Vertebral angiogram.

bral arteries and the left posterior cerebral filled via collaterals, but neither the middle cerebral nor the right posterior cerebral arteries were visualized. There was a mass of perforating vessels in the base of the brain. Collaterals had formed between the internal carotid system and branches of the external carotid, including the ophthalmic artery and middle meningeal artery. Air encephalography showed marked cerebral atrophy.

Subsequently she was followed as an outpatient. Clinically there was a left homonymous hemianopia and she had severe apraxia, with inability to do housework or to look after her baby. There was a slight improvement after treatment with cyclandelate, and her supraorbital artery pressure increased to 56 mm Hg (for both arteries) with a brachial artery pressure of 110 mm Hg. She remained severely incapacitated.

Case 2

A white British man aged 49 had had measles at the age of 2 years, after which he developed a right hemiparesis and later epilepsy. He remained mentally subnormal and spent most of his life in institutions.

In November 1971, during a grand mal fit, he hit his head. Subsequently mental deterioration and an increase in his right hemiparesis led to his admission to hospital. He was transferred to the neurosurgical unit at Brook General Hospital for investigation of a possible subdural haemorrhage. On admission he was found to be aphasic and withdrawn though he responded to sharp commands. He had a dense right hemiplegia with wasted, under-developed limbs on the right. Lumbar puncture showed a raised protein level. On plain x-ray films the middle meningeal artery groove was seen to be enlarged. Bilateral carotid angiograms (figs. 3 and 4) showed obstruction to both internal carotid arteries and filling of the anterior cerebral arteries via collaterals but no middle cerebral arteries were seen. Gross cerebral atrophy was seen on the air encephalogram.

FIG. 3—Case 2. Right carotid angiogram.

FIG. 4—Case 2. Left carotid angiogram.

He died in January 1972. At necropsy there was gross thickening of the leptomeninges over both hemispheres. The area of brain supplied by the left middle cerebral artery was atrophied. A large butterfly glioblastoma multiforme replaced much of the right basal ganglia, extending to the mid brain and depressing the anterior horn of the left ventricle. The glioma was not thought to be connected with the angiographic findings.

Comment

Suzuki and Takaku (1969) first named this disease "moyamoya," which is Japanese for something hazy, like a puff of smoke—the appearance of the carotid rete on angiography. The clinical course in those whose first symptoms occur in childhood is different from those in whom symptoms develop in adult life. The dominant symptom in children is weakness, which is usually of sudden onset, has a rapid recovery, and often recurs. Eye symptoms, speech disturbance, fits, headache, and intellectual deterioration are common. Examination often shows a hemiparesis or, less commonly, monoplegia or diplegia. Cranial nerve palsies and disturbances of gait and speech are also found. Subarachnoid haemorrhage is uncommon in children; however, this is the commonest presentation in adults. Headaches are common but paresis and intellectual deterioration are less common in adults (Nishimoto and Takeuchi, 1968).

Lumbar puncture gives a normal picture in the absence of other lesions. E.E.G. appearances are usually abnormal but non-specific. Four-vessel angiography provides the diagnosis. There is occlusion of the internal carotid arteries at the level of the syphon, and occlusion of the middle and anterior cerebral arteries and sometimes of the posterior cerebral arteries. The cerebral circulation is maintained by collaterals. A mass of perforating vessels arise in the base of the brain. Further anastomoses occur between the internal and external carotid systems via leptomeningeal vessels and rete mirabile. The rete is a tightly-packed network of arterial vessels joining the internal and external carotid systems directly across the subdural space. It occurs in some animals. In the Western literature air encephalography has been performed only twice, and showed only minimal cerebral atrophy. One of these patients developed a subdural haemorrhage after the examination, and this was reported as possibly a result of rupture of a rete vessel (Taveras, 1969). Both of the present patients showed marked cerebral atrophy, which to our knowledge has not been previously reported, though intellectual deterioration is common in those whose symptoms first appear in childhood.

There is no known effective treatment. Galligioni *et al* (1971) used cerebral vasodilators in four patients with apparent improvement. The long-term prognosis is unknown. Most patients appear to deteriorate initially and then remain static (Kudo, 1968; Taveras, 1969; Galligioni *et al.*, 1971). In Nishi-

moto and Takeuchi's series 66 out of 73 children had recurring hemiplegia without deterioration, 4 deteriorated, and 3 died at the ages of 7, 9, and 15 years. None of the 23 adults deteriorated.

The aetiology is unknown. There is no reported evidence of atherosclerosis, autoimmune disease, temporal arteritis, cerebral emboli, or abnormal coagulation in the cases studied (Taveras, 1969). Most workers believe that it is a progressive arterial disease (Kudo, 1968; Nishimoto and Takeuchi, 1968; Suzuki and Takaku, 1969; Taveras, 1969), and some serial arteriograms have shown progressive occlusion of the circle of Willis (Suzuki and Takaku, 1969). The cause of the arterial disease is unknown, though bacterial toxins have been suggested (Taveras, 1969). Others believe that it is a congenital abnormality (Galligioni *et al.*, 1971). In some series there have been cases reported in siblings (Suzuki and Takaku, 1969). It seems probable that it is not a single disease entity but a group of diseases, in which the normal collateral circulation via the circle of Willis is occluded. Therefore, other collaterals develop, and their appearance is the same regardless of the aetiology of the circle of Willis occlusion.

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