

### Discussion

Acute exacerbation of atopic eczema with weeping red skin is usually caused by secondary infection and requires prompt treatment with appropriate systemic antibiotics. Hyponatraemia and hypoalbuminaemia are rare complications of atopic eczema and occur if weeping (exudation of serum) is chronic and persistent.

Recently the number of parents who seek alternative forms of treatment has increased with the growing popularity of 'natural' products. While homeopathic medicine has relieved some diseases,<sup>2</sup> in our opinion there is

little evidence to suggest it is helpful in atopic eczema. Thus caution is needed in the use of homeopathic medicines as the sole treatment for this condition. Severe exacerbations should be recognised and treated early by conventional medicines to avoid the risk of potentially life threatening complications.

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## Growth failure secondary to moyamoya syndrome

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### Abstract

**We describe a boy who presented at the age of 7 years with short stature due to hypopituitarism. Six months after starting appropriate hormone replacement treatment at the age of 8 he suffered his first generalised convulsion. Further neuroradiological investigation led to the diagnosis of moyamoya syndrome.**

Moyamoya is the arteriographic appearance of a filmy network of small vessels at the termination of the internal carotid artery. We describe a boy with hypopituitarism associated with moyamoya.

### Case report

The boy presented initially to our hospital at 18 months of age with obesity and developmental delay. He was the fifth child of unrelated, white parents (maternal height 0 SD, paternal height -0.9 SD) and was born at term by spontaneous vaginal delivery. The birth weight was 3450 g and there were no perinatal problems. The developmental delay was thought to be due to lack of parental stimulation and a physiotherapy programme was devised with support for the family. At 3 years of age he was admitted to hospital for investigation of persistent lower limb weakness but biochemical investigation excluded a myopathy. The family subsequently failed to attend hospital appointments.

At 7 years he was referred to the endocrine clinic with short stature, small genitalia, and headaches. On examination he was moderately obese with weight on the 25th percentile and height well below the 3rd percentile (-3.1 SD). There were no dysmorphic features. Blood pressure was 95/60 mm Hg. Fundoscopy and visual fields to confrontation were normal. He was alert and cooperative but clumsy with reduced power and tone in the lower limbs with tendon reflexes preserved and flexor plantaris. His penis was small and the testes were less than 1 ml volume.

Investigation showed a TW2 bone age of 3.4 years at chronological age 7.1 years. Anterior

pituitary function was assessed after intravenous injection of thyrotrophin releasing hormone (200 µg) and luteinising hormone releasing hormone (100 µg) with oral clonidine 150 µg/m<sup>2</sup>. Fasting venous samples were taken at 0, 20, 75, 90, 120, and 150 minutes. Plasma cortisol concentrations were 456, 440, 155, 215, 347, and 469 nmol/l respectively. Peak plasma growth hormone concentration was 1.6 mIU/l at 150 minutes and the peak plasma luteinising hormone and follicle stimulating hormone concentrations were 1.3 IU/l. Plasma thyroxine was 44 nmol/l with an abnormal, sustained rise in thyroid stimulating hormone (2.0, 13.0, 20.0 mIU/l at 0, 20, and 75 minutes) indicative of hypothalamic pathology. The basal prolactin concentration was raised at 642 mIU/l. These results confirmed hypopituitarism. Posterior pituitary function was not affected.

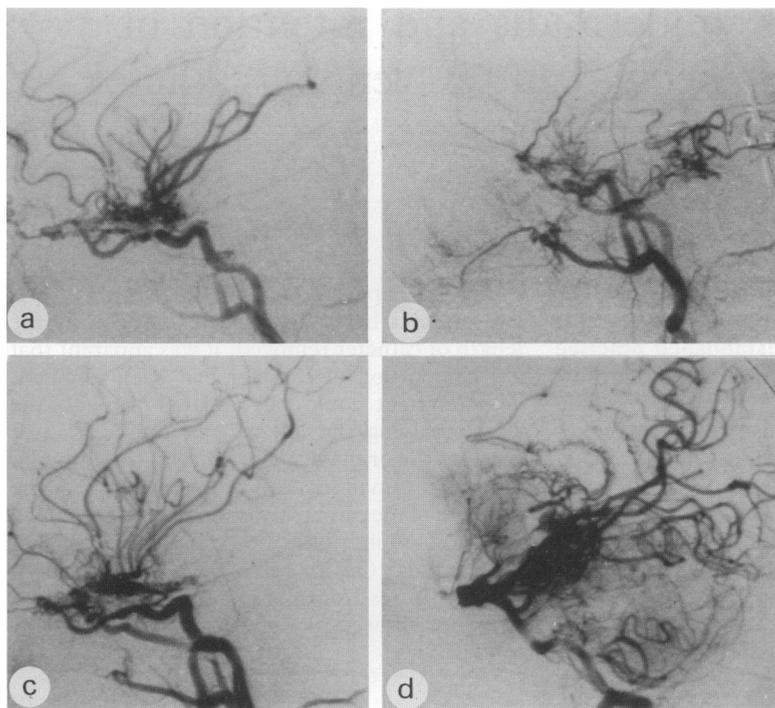
Treatment with growth hormone and thyroxine was started at the age of 7.4 years resulting in a rise in growth velocity from 2.3 cm year in the four years before treatment to 9.8 cm in one year on treatment. At 7.9 years he suffered his first generalised seizure and an electroencephalogram confirmed epileptiform activity. A radiograph of the skull showed amputation of the dorsum sellae; computed tomography showed a shallow pituitary fossa and suprasellar calcifications with obliteration of the cisternae but no well defined soft tissue mass. The third ventricle was deviated slightly to the left side. The radiological findings were not considered to be sufficient for diagnosis but the possibility of a craniopharyngioma was discussed. The seizures were controlled with carbamazepine and a magnetic resonance scan requested to allow better definition of the suspected tumour. Subsequently the child developed worsening headaches and at 8.4 years of age was admitted to hospital with blurred vision, ataxia, and increased seizure frequency. A repeat computed tomogram confirmed suprasellar calcification with highly attenuating tumour like tissue in the suprasellar region, now also extending to the ambient cisterns with

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Cerebral angiography (DSA) showing bilateral intradural carotid occlusion with moyamoya type of collateral circulation: lateral series of (a) left internal carotid, (b) left external carotid, (c) right carotid, and (d) left vertebral injections. Vascular territories recruited for collateral circulation include ophthalmic, posterior thalamoperforating, and posterior pericallosal as well as multiple meningeal vessels.

indentation of the posterior part of the third ventricle and obstructive hydrocephalus. The impression was that of a slowly spreading lesion in the cerebrospinal fluid spaces and in view of the apparently poor prognosis, growth hormone treatment was discontinued.

During this admission a ventriculoperitoneal shunt was inserted, and cerebrospinal fluid and plasma tumour markers were measured and all were normal ( $\beta$  human chorionic gonadotrophin, placental alkaline phosphatase, and  $\alpha$  fetoprotein). Exploratory surgery at the age of 8.6 years showed a collection of abnormally prominent blood vessels in the sylvian fissure with a collection of tortuous arteries and arterIALIZED veins. Subsequent cerebral angiography (figure) showed bilateral internal carotid artery occlusion distal to the origin of the ophthalmic arteries with a very extensive collateral circulation situated in the basal subarachnoid cisterns—so called moyamoya syndrome. The vascular nature of the 'tumour' and the absence of any other mass has subsequently been demonstrated by magnetic resonance imaging. The boy has been restarted on his growth hormone treatment and the thyroxine and carbamazepine continued. The family have been reassured of the absence of any malignant disease and advised that he should avoid contacts sports and extreme physical exertion as subarachnoid haemorrhage is a recognised complication.

### Discussion

Growth failure as a consequence of hypopituitarism is not uncommon,<sup>1</sup> but we believe our patient is the first reported case of hypopituitarism associated with a moyamoya syndrome. Cerebrovascular moyamoya is the descriptive term applied to an abnormal blood vessel pattern on the base of the brain as a result

of stenosis or occlusion of the internal carotid artery,<sup>2</sup> usually bilateral, with development of a prolific collateral vascular network.<sup>3</sup> It is the angiographic appearance that lead to Japanese authors referring to this as 'moyamoya', meaning 'puff of smoke'.

Similar neuroradiological appearances may be encountered with a variety of associated pathological entities. The syndrome has been reported in phakomatoses, septic infection of the head and neck, and tuberculous meningitis. Autoimmune diseases, peripheral vascular occlusive disease, head trauma, sickle cell disease, Fanconi's anaemia, intracranial aneurysm, arterial hypertension, brain irradiation, and chromosomal aberration have also been associated in reported cases.<sup>2-6</sup> However, despite various hypotheses the aetiology of moyamoya remains unclear. The vast majority of cases, as in our case reported here, have no obvious cause for the vascular occlusion and the prognosis therefore can not be assessed on a firm pathological basis.

The exact association between the moyamoya syndrome and our patient's growth failure remains unclear but in view of the delayed bone age and pretreatment SD score of  $-3.1$  it must be longstanding. As the initial computed tomogram showed a shallow pituitary fossa with suprasellar calcification, it seems likely that the space occupying vascular mass responsible for those appearances was also responsible for the pituitary dysfunction and growth failure.

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