series of Winkelmann *et al.* (1968) of 279 patients with dermatomyositis/polymyositis indicates that the prognosis may be considered by thirds. One-third die, one-third remain ill and one-third achieve complete remission or cure. (It is of interest that of the 87 fatalities in their series only 8 were due to malignancy.) Recurrences of dermatomyositis, separated by such long periods of remission, are rare. Over the course of 33 years our patient has had three attacks of dermatomyositis. Between attacks she has enjoyed good health and there have been no permanent sequelae.

This case serves to demonstrate the variability of morphology and natural history of this disease.

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Reference

Winkelmann R K, Mulder D W, Lambert E H, Howard F M (jr) & Diessner G R
(1968) *Mayo Clinic Proceedings* **43**, 545–556

Progressive hemifacial atrophy with scleroderma and ipsilateral limb wasting[1] (Parry-Romberg syndrome)[1]

P K Lakhani MB MRCP
T J David MD MRCP
Department of Child Health
University of Manchester

Progressive hemifacial atrophy (Parry-Romberg syndrome) is a very rare disorder characterized by a slowly progressive unilateral atrophy of the face affecting variably the skin, subcutaneous fatty tissue, muscle, connective tissue and bone (Rogers 1963). We present a 13-year-old boy in whom progressive hemifacial atrophy was accompanied by wasting of the ipsilateral arm and leg, and scleroderma.

[1]Case presented to Section of Paediatrics, 26 November 1982. Accepted 26 July 1983

Case report

The patient, a 13-year-old Caucasian boy, is the youngest of eleven children of unrelated parents; his birth followed a normal full-term pregnancy and normal delivery. There was no history of birth trauma. The family history was unremarkable.

He was admitted to hospital at the age of one year following full-thickness scalds caused by hot water; these involved the dorsum of the left shoulder and scapula region extending to the axilla, for which skin grafting was required. However, at 7 years there was persistence of a raised, red, hypertrophic scar behind the left shoulder. This was treated by a single injection of triamcinolone into the scar. At the age of 8 years hair loss was noted over the left side of the scalp, and this was followed a few months later by the appearance of a linear area of scleroderma in the left parietal area extending forwards on to the forehead. At 9 years atrophy of the left side of the face was first noticed, as was an area of scleroderma on the left anterior chest wall. Earlier photographs confirm the prior absence of facial atrophy. At the age of 12 he was noted to be limping and to have wasting and slight shortening of the left arm and the leg; he had complained of cramp in the left leg for three years. During the last four years the facial appearance has progressively worsened (Figure 1). There is no history of headaches, facial pain, or convulsions.

Extensive haematological and biochemical investigations at the age of 13 years revealed normal values. An autoantibody profile was negative. Chromosomes (with banding) were normal. X-rays showed asymmetry of the skull vault which was hypoplastic on the left side. The ramus of the left mandible was thinner, the left femur 0.25 cm shorter and the left tibia 1.5 cm shorter and more slender. The bones of the hand and foot were marginally smaller on the left. An electroencephalogram and computerized axial tomogram were normal. Electromyography showed normal motor and sensory nerve conduction velocities, with distal velocities similar on both sides and normal latencies in the facial nerve on the left. Skin biopsy from the left parietal and the left pectoral area showed the histological changes of scleroderma.

Discussion

In some cases of progressive hemifacial atrophy (including the present one) the atrophic process extends beyond the face, usually to the ipsilateral parts of the body. Contralateral atrophy occurs rarely, and exceptionally the disease is bilateral (Rogers 1963). Linear localized scleroderma may occur (Wartenberg 1945, Lewkonia & Lowry

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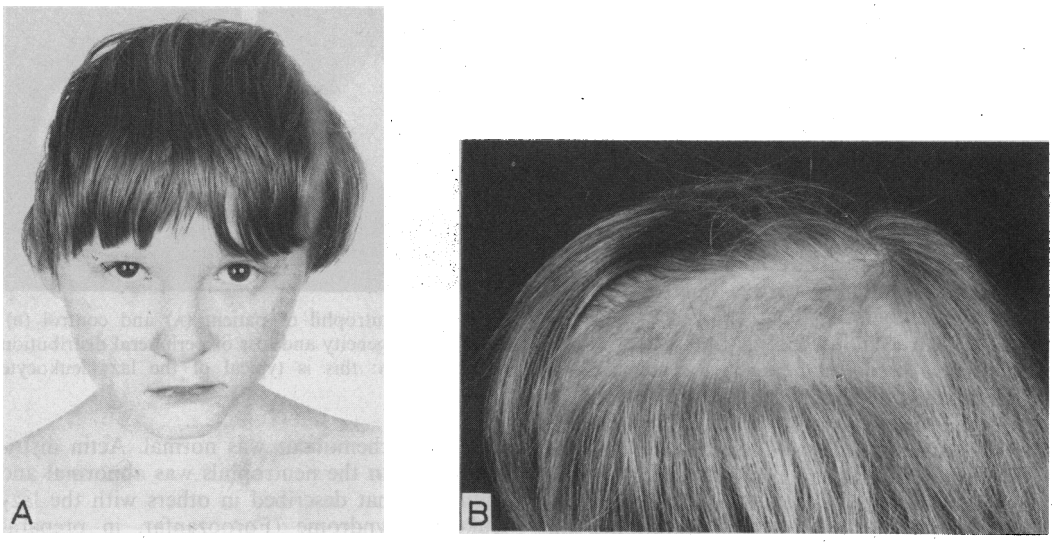

Figure 1. Patient aged 13 years with progressive hemifacial atrophy. A: showing atrophy of left side of face and left mandible. B: showing linear scleroderma of left side of scalp

1983), as in the present case. The disease appears in the first or second decade of life, cases beginning in the second decade having less interference with overall growth of the facial bony skeleton (Wartenberg 1945). Most cases are sporadic, and the aetiology of the condition is unknown. Both sides of the face can be affected with equal frequency and the disorder is more common in females with a F:M ratio of 3:2 (Rogers 1963). The disease progresses for a variable time, usually between two and ten years. The final degree of deformity varies widely, as the disease may 'burn itself out' at any stage.

The progression of linear scleroderma and facial atrophy in our patient, observed over a five-year period, is typical. The accompanying ipsilateral limb wasting is a recognized feature of the disorder, occurring in less than 10% of cases (Archambault & Fromm 1932, Rees 1976). Ocular manifestations (Rees 1976) (e.g. enophthalmos) and neurological abnormalities (Rogers 1963) (e.g. ipsilateral hemiatrophy of the brain, epilepsy) were not present in our patient.

The aetiology and pathogenesis of this condition, and the relationship between progressive hemifacial atrophy and scleroderma, are obscure. It would appear that the preceding scalds in the present case are unrelated to the subsequent development of the disorder, although this view is not shared by the mother of the boy who has always blamed herself and regarded the scalds as the cause.

Treatment of progressive hemifacial atrophy with cartilage or bone grafts, free dermis fat grafts or silicone implants has in the past been unsatisfactory. However, the introduction of free tissue transfer with microvascular anastomosis promises better results in the future (Harashina & (Fujino 1981, Shintomi *et al.* 1981).

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References

Archambault L & Fromm N K
(1932) *Archives of Neurology and Psychiatry* **27**, 529–584
Harashina T & Fujino T
(1981) *Annals of Plastic Surgery* **7**, 289–294
Lewkonia R H & Lowry R B
(1983) *American Journal of Medical Genetics* **14**, 385–390
Rees T D
(1976) *Clinics in Plastic Surgery* **3**, 637–646
Rogers B O
(1963) Transactions of the 3rd International Congress of Plastic Surgery (International Congress Series No. 66). Ed. T R Broadbent. Excerpta Medica, Amsterdam; pp 681–689
Shintomi Y, Ohura T, Honda K & Iida K
(1981) *British Journal of Plastic Surgery* **34**, 398–409
Wartenberg R
(1945) *Archives of Neurology and Psychiatry* **54**, 75–96