SKIN FOCUS

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MORPHEA (LOCALISED SCLERODERMA)

Recently two patients with different types of morphea presented to the practice. The first patient was a 12-year-old girl who presented with indurated sclerotic hyperpigmented plaques involving the abdomen. On clinical assessment a diagnosis of morphea profundus (that extended to the deep subcutaneous fat) was made. Part of the lesion had ivory-white patches suggestive of lichen sclerosis atrophicus (Fig. 1).

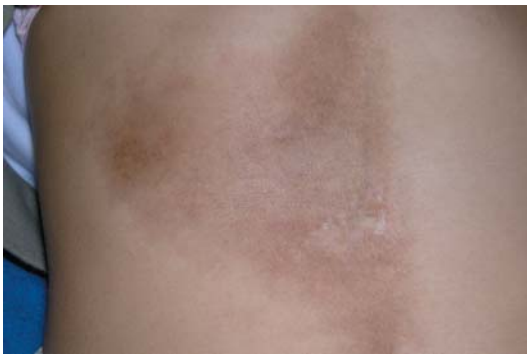

Fig. 1. Morphea profundus presenting as a sclerotic hyper- and hypopigmented plaque extending to the subcutaneous fat. The hypopigmented area is suggestive of lichen sclerosis atrophicus.

The second patient was a 16-year-old girl who presented with erythematous superficial atrophic plaques involving the left arm (Fig. 2). A diagnosis of plaque morphea was made.

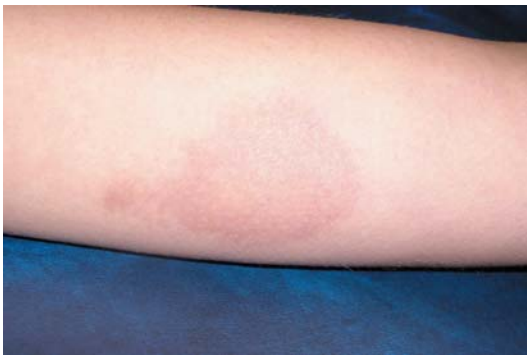

Fig. 2. Morphea presenting on the arm as an atrophic erythematous plaque in a 16-year-old girl.

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These clinical cases provided the impetus for the following review of the literature on morphea.

Morphea is a localised cutaneous sclerosis characterised by early erythematous plaques with a lilac border which evolves to ivory-coloured sclerotic areas. Eventually the plaques become hyperpigmented. The lesions of morphea may be localised plaques, linear or generalised. Although the pathomechanism of the sclerosis in morphea and systemic scleroderma may be similar, the two diseases are distinct entities and can easily be distinguished by clinical features. Compared to morphea, systemic scleroderma is a systemic disease with a poor prognosis while morphea usually has a good prognosis. Raynaud's phenomenon does not occur in morphea. Morphea never starts as symmetrical sclerosis of the fingers and never extends to involve internal organs.

Epidemiology

About 20% of affected individuals are children or adolescents and the disease is more prevalent in females than males (3:1). In children morphea is ten times more common than the systemic form of scleroderma. Also linear morphea is more frequently observed in children than adults. Morphea may be triggered by localised trauma to the skin. The role of infection with *Borrelia burgdoferi* has not been proven.

Pathogenesis

Sclerosis of the skin involves vascular damage, activated T-cells and altered connective tissue production by fibroblasts.

Vascular changes

Microvascular injury to capillaries is a primary event. This initial change includes expression of adhesion molecules and endothelial swelling followed by intimal hyperplasia.

T-lymphocytes modify collagen synthesis by fibroblasts via the cytokines they secrete. Production of collagen (types 1, 2, 3) is induced by two T-cell derived cytokines, interleukin 4 (IL-4) and transforming growth factor beta (TGF-β). Immune responses dominated by IL-4 and TGF-β-producing cells are involved in the development of skin sclerosis.

Clinical features

Morphea can be subdivided into plaque morphea, linear morphea and generalised morphea.

Plaque morphea

Plaque morphea is the most common type and starts insidiously. The initial lesion is an asymptomatic erythematous to purple patch. Later the lesion becomes indurated and ivory in colour with a lilac edge. Hairs are absent. The lesions are round, 3-15 mm in diameter, multiple and asymmetrical. They occur on the trunk and limbs. Sometimes the lesions may be preceded by pigmentation.

Guttate morphea presents as multiple 3 mm indurated papules.

Morphea profundus or deep morphea presents with scelerotic hard plaques which extend deep into the subcutaneous fat.

Nodular morphea presents as thick indurated nodules indistinguishable from keloids.

Linear morphea and Parry-Romberg syndrome

The clinical features of linear morphea are similar to plaque lesions. However they may be differentiated by the age of onset, distribution, clinical outcome and also serology. The lesions occur as linear areas of induration, mainly on the limbs. Occasionally this may extend to involve the underlying muscles and bones leading to disturbance in growth. Flexion deformities may also occur. The *en coup de sabre* type of linear morphea extends from the forehead to the scalp and may cause a linear zone of alopecia. The Parry-Romberg syndrome presents as hemifacial atrophy but shows no sclerosis. It affects the distribution of the trigeminal nerve, including the eye and the tongue. Parry-Romberg syndrome may be associated with neurological abnormalities including seizures and myopathy of the eye muscles.

Generalised morphea

Generalised morphea is due to plaques which coalesce until they affect the entire trunk. Constrictions lead to impaired thorax mobility and breathing problems may occur.

Disabling pansclerotic morphea of children

Although this is similar to generalised morphea, it causes severe disability as a result of atrophy of the underlying muscles and contractures of the involved joints. It starts before the age of 14 years and may involve the trunk and extremities with sparing of the fingertips. Raynaud's phenomenon is not found. Flexion contractures are frequent. This condition is progressive and treatment is usually ineffective.

Differential diagnosis

The diagnosis of morphea is a clinical one. This may be confirmed by skin biopsy. Areas of hyperpigmented atrophic plaques may resemble atrophoderma of Pasini and Pierini. The other considerations are lichen sclerosus et atrophicus, morphea-like lesions of sarcoid, eosinophilic fasciitis, acrodermatitis chronica atrophicans, pseodoscleroderma and scleroderma. Morphea is differentiated from systemic scleroderma by the fact that Raynaud's phenomenon is not present and there is no lung or renal involvement.

Laboratory findings

There are no abnormal laboratory findings in plaque morphea. However, generalised and linear morphea have positive anti-single-stranded DNA antibodies, antihistone antibodies and eosinophilia. The presence of these parameters may indicate disease activity. Procollagen type 1 carboxy-terminal propeptide is elevated in 30% of patients. The serum levels also correlate with the extent of skin involvement. Thermography is a sensitive new means of evaluating disease activity and risk of further tissue damage.

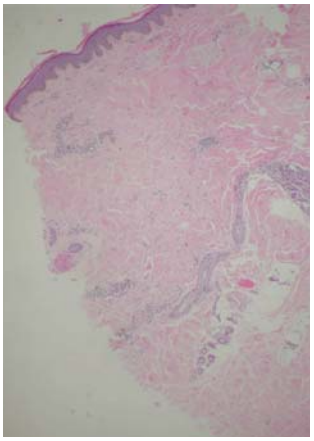

Fig. 3 Dermatopathology of patient No. 2 with plaque morphea. The epidermis is normal. The pathology is in the dermis which shows a superficial and deep perivascular infiltrate of lymphocytes. The dermis shows an increased amount of collagen bundles, which are thickened, hyalinised and eosinophilic, with an entrapment of eccrine sweat glands. The histology is in keeping with plaque morphea.

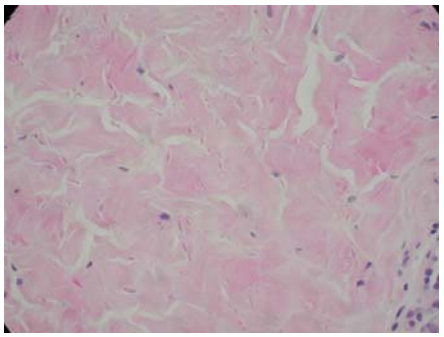

Fig. 4. Close-up featues of dense collagen bundles in the dermis characteristic of morphea.

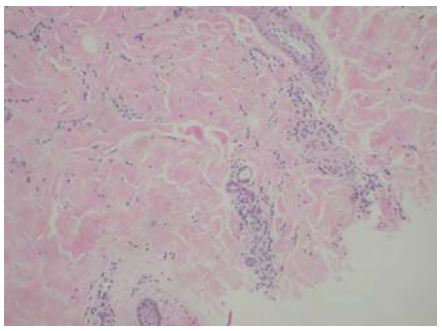

Fig. 5. Entrapment of adnexal structures including eccrine sweat glands seen in morphea.

Dermatopathology (Figs 3 - 5)

The epidermis is flattened and atrophic with loss of the rete ridges. There is dermal oedema and the collagen fibrils become eosinophilic. There is a perivascular infiltrate of lymphocytes, plasma cells or macrophages. Later, the dermis is thickened with dense collagen and the dermal appendages are lost.

Prognosis in morphea

Plaque-type morphea improves spontaneously within 3-5 years, but atrophy, induration and pigment changes

may persist. Linear morphea, including *en coup de sabre* and facial hemiatrophy, persists.

Treatment

Treatment of morphea is difficult and few controlled clinical trials have been published.

Topical therapy

Corticosteroids

Although ultra-potent topical steroids such as clobetasol propionate may be useful in superficial active lesions by reducing inflammation in plaque morphea, corticosteroid creams are ineffective in resolving sclerosis.

Vitamin D derivatives

Calcipotriol inhibits proliferation of cultured fibroblasts. Calcipotriol ointment has led to softening and repigmentation of morphea lesions.

Phototherapy

Phototherapy for morphea has been found to be effective and this has been confirmed by many clinical publications. Both bath PUVA and UVA 1 provide marked improvement in morphea. They both induce expression of matrix metallo proteinase 1, a collagenase that reduces procollagen within the skin. Phototherapy has been found to be effective in all types of morphea. Both PUVA and UVA 1 have been effective in resolving the scar-like sclerosis of advanced disease. The role of photodynamic therapy in treating morphea needs further investigation.

Systemic treatment

Immunosuppression

The combination of methotrexate and corticosteroids has been found to be helpful in the inflammatory stage of progressive and linear morphea. Oral steroids (1 mg/kg body weight of methyl prednisolone) is given for the first 3 months and methotrexate 15-25 mg weekly (plus folic acid) may be given initially and on an ongoing basis. Because of its side-effect profile, cyclosporine should be reserved for recalcitrant cases.

Penicillin and penicillamine

Penicillin and penicillamine have been found to be effective in cases of regression of morphea in some patients.

Oral vitamin D derivative

Oral calcitriol has shown improvement in children with linear scleroderma.

Vitamin A derivatives

Treatment with acitretin (20-50 mg daily) is effective in localised scleroderma/ morphea. Retinoids can inhibit TGF-β, the cytokine which promotes collagen synthesis by fibroblasts.

Physical therapies

Physiotherapy helps to mobilise joints which have been affected by linear morphea. Contractures require reconstructive surgery. Orthodontic devices may minimise the asymmetry of the lower face resulting from Parry-Romberg syndrome.

Figure credits

Figs 1 and 2 – Dr ME Docrat.

Figs 3 - 5 – Dr Nick van Diggelen, Pathcare.

Declaration of conflict of interest

The author declares no conflict of interest..

ACKNOWLEDGEMENTS AND FURTHER READING

Bolognia JL, Jorizzo JL, Rapini RP. *Dermatology*, vol. 2. Philadelphia: Mosby, 2003: 1503-1515.

Champion RH, ed. *Rook, Wilkinson, Ebling Textbook of Dermatology*, vol. 3. Oxford: Blackwell Science, 1998: 2502-2512.

Kane SK, Ryder JS, Johnson RA. *Color Atlas and Synopsis of Pediatric Dermatology*. New York: McGraw-Hill, 2002: 420-422.

Paller AS, Mancini AJ. *Herwitz Clinical Pediatric Dermatology (A textbook of skin disorders of childhood and adolescence*), 3rd ed. New York: Elsevier Saunders, 2006: 595-599.

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