MedicineToday 2011; 12(8): 38-42

PEER REVIEWED FEATURE
POINTS: 2 CPD/2 PDP

An approach to management of morphoea

VERNON PUA MB BS(Hons), MPH, FACD
STEPHEN LEE AM, MB BS(Hons), DDM, FACD

Key points

- **Morphoea is a benign dermatosis that has a good long-term prognosis.**
- **Diagnostic tests are not usually necessary for morphoea but it needs to be differentiated from systemic sclerosis, with which it is often confused.**
- **Small morphoeic plaques can be monitored without active treatment.**
- **Topical agents can be used in the early inflammatory stage.**
- **Reassurance of patients is the key to preventing needless worry.**

Reassurance and counselling for patients with morphoea may be sufficient as morphoea often resolves with time. It should, however, be differentiated from systemic sclerosis, which is a far more serious disease.

Morphoea is a benign dermatosis that usually develops as one or more circumscribed patches or indurated plaques on the trunk (Figure 1). These lesions vary in size from 0.5 to 30 cm in diameter. Typically, the onset is insidious, although some patients experience pruritus, mild tenderness and swelling. Dyspigmentation of the lesions is common and active lesions often have a violaceous or erythematous border. With time, the lesions develop a white or yellow hue, become indurated and feel firm on palpation. Upon resolution, there is often atrophy and varying degrees of dyspigmentation (Figure 2). Although morphoea usually presents on the trunk, the face and scalp may also be involved. Morphoea is two-and-a-half times more common in women than in men and affects all races equally.[1]

The aetiology of morphoea remains unclear but infections such as Lyme disease (*Borrelia burgdorferi*) and measles, and trauma, radiation and medications have been implicated. The pathogenesis of morphoea and systemic sclerosis is likely to be similar: endothelial cell damage, inflammation and excessive extracellular matrix produced by fibroblasts, resulting in sclerosis.[2] In morphoea, the process is usually restricted to the skin, whereas in systemic sclerosis, the respiratory, cardiac and gastrointestinal systems may be involved. There is no known

Dr Pua is in private practice at Sydney Dermatology, Eastern Suburbs Dermatology and Sutherland Shire Dermatology, Sydney. Associate Professor Lee is Clinical Associate Professor at Sydney Medical School, University of Sydney; Head of the Department of Dermatology at Concord Repatriation General Hospital; and Senior Visiting Dermatologist at Royal Prince Alfred Hospital, Sydney, NSW.

CUTANEOUS DISEASES WITH SCLEROSIS AS A FEATURE

- Morphoea
- Systemic sclerosis
- Chronic graft-versus-host disease
- Scleromyxoedema
- Radiation dermatitis
- Nephrogenic fibrosing dermopathy

genetic predisposition to either morphoea or systemic sclerosis.

There are a number of other diseases that cause fibrosis or hardening of the skin and which may be confused with morphoea (see the box on this page). Of these, the most familiar – especially among rheumatologists and clinical immunologists – would be systemic sclerosis and the CREST (calcinosis, Raynaud's, oesophageal dysmotility, sclerodactyly, telangiectasia) syndrome. However, the most common cause of cutaneous sclerosis is morphoea, a dermatosis that may be unfamiliar to nondermatologists.

Morphoea has too frequently been mistaken for systemic sclerosis: while both conditions manifest with cutaneous sclerosis, morphoea is primarily a skin disorder whereas systemic sclerosis often has extracutaneous involvement of the respiratory and gastrointestinal systems. Moreover, the distribution of lesions and the clinical course also differ between the two conditions. Although a skin biopsy may confirm the pathological process of sclerosis, it is unhelpful in differentiating between morphoea and systemic sclerosis. Therefore, it is vital to appreciate the differences and similarities between the two dermatoses, and an accurate diagnosis is imperative as the treatment and prognosis of two diseases are different.

CLINICAL VARIANTS OF MORPHOEA

Morphoea is not a homogeneous skin disease and there are several clinical variants. Plaque morphoea is the most common, comprising more than 50% cases of morphoea (Figure 3).[1]

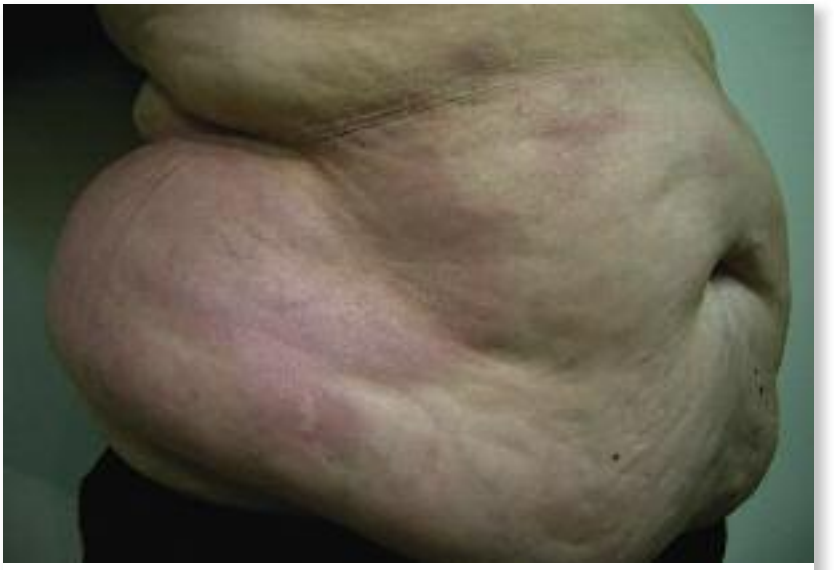

Figure 1. Early plaque of morphoea on the trunk.

In addition to plaque morphoea, which is usually seen in middle-aged adults, the other clinical variants include generalised, linear, subcutaneous, bullous, keloidal, guttate, en coup de sabre and progressive hemifacial atrophy. The latter is a severe and usually persistent form of morphoea. Occasionally, early lesions may appear urticarial-like (Figure 4). Generalised morphoea is defined as the development of multiple plaques of morphoea in more than two anatomical sites.[3] Linear morphoea is usually seen in children and young adults.

It is useful to recognise that although morphoea is active for about four years in many patients, spontaneous resolution may occur within a shorter time span. Chronic disease with relapses is more likely to occur in patients with generalised, subcutaneous or linear morphoea. Unfortunately, significant disfigurement may result in patients with en coup de sabre, progressive hemifacial atrophy or pansclerotic morphoea, in which underlying structures are involved. Furthermore, linear morphoea occurring over a joint can result in permanent contractures, causing functional impairment. As trauma can trigger morphoea, multiple biopsies or surgery should be avoided whenever possible.

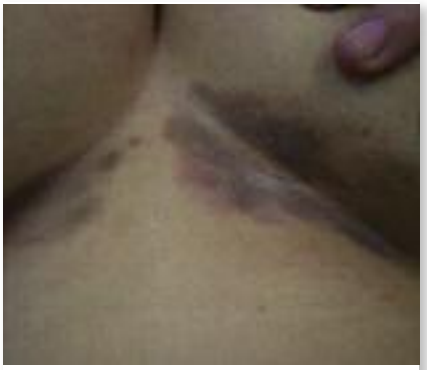

Figure 2. Postinflammatory hyperpigmentation of morphoea.

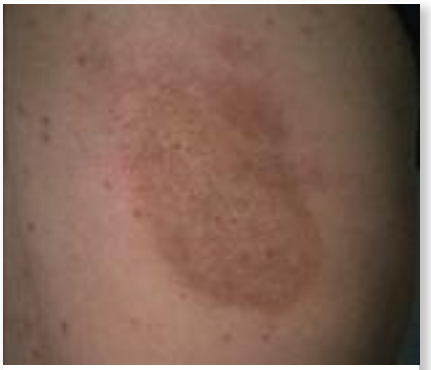

Figure 3. Plaque morphoea on the trunk.

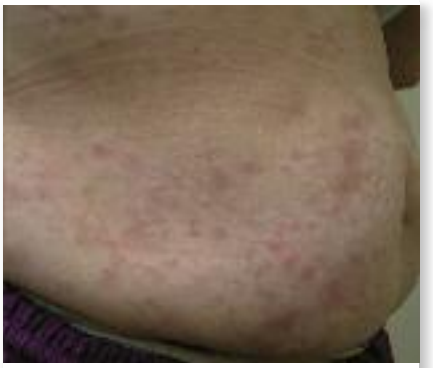

Figure 4. Early lesions of morphoea resembling urticaria.

CLINICAL DIFFERENCES BETWEEN MORPHOEA AND SYSTEMIC SCLEROSIS

Morphoea is a skin disease and extracutaneous involvement is uncommon. Arthralgia appears to be the most frequently reported extracutaneous symptom and this usually resolves with the improvement of the skin disease.

By contrast, in patients with systemic sclerosis, systemic complications such as dyspnoea, gastro-oesophageal reflux, neuropathy, hypertension and cardiac failure may develop. Also, it is important to note that Raynaud's phenomenon, sclerodactyly, acral and facial telangiectasia and narrowing of the mouth are rare in patients with morphoea (Figure 5). These latter features are clinical features of systemic sclerosis and should prompt the clinician to investigate and exclude active systemic disease. Finally, the truncal distribution of morphoeic plaques contrasts with the predominantly acral distribution in systemic sclerosis.[4]

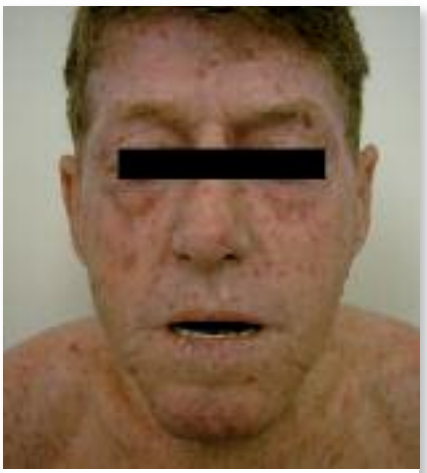

Figure 5. Systemic sclerosis with facial mat-like telangiectasia and narrowing of the mouth.

DIAGNOSIS

The correct diagnosis of morphoea is based on recognising its distinctive clinical features.[5,6] In most cases, a skin biopsy is unhelpful and unnecessary. Nevertheless, the histological features include a dense inflammatory infiltrate of lymphocytes, macrophages, plasma cells in the dermis during the early phase, followed by thickened, hyalinised collagen bundles in older lesions. The histological findings are similar for morphoea and systemic sclerosis.

There are no diagnostic blood tests as such for morphoea. In fact, a broad battery of tests, including chest CT and lung function tests, for a patient with classic morphoea is unnecessary and unlikely to enhance the management outcome. Symptoms suggestive of extracutaneous involvement should nonetheless be investigated with the appropriate tests; for example, a patient with rapidly progressing or widespread scleroderma warrants thorough investigation.

TREATMENT

Once the diagnosis of morphoea has been established, it is important to clearly explain the nature of the disease to the patient. Reassurance that the dermatosis will involute over the next few years in most instances, regardless of treatment, is vital. The exceptions to this are the deeper variants of morphoea such as en coup de sabre and progressive hemifacial atrophy.

In most cases of morphoea, the plaques on the trunk will resolve with some atrophy and dyspigmentation. Progression to systemic sclerosis is extremely rare.

A clear understanding by the caring practitioner will assist in the counselling of worried patients who may assume that morphoea is systemic sclerosis and therefore incorrectly believe that the disease has a poor prognosis.

Topical treatment

If the plaques of morphoea are small and do not trouble the patient, clinical monitoring without active treatment is an acceptable approach. In this situation, a topical camouflage cosmetic or a suitable moisturiser can be helpful. If more active treatment is desired, topical agents that have been useful in reducing discomfort

and inflammation include topical corticosteroids with or without occlusion, topical vitamin D analogues (calcipotriol) and topical tacrolimus.*

Intralesional corticosteroids can be useful in carefully selected patients but the treatment should be administered by experienced practitioners with realistic expectations. Topical treatments, particularly corticosteroids, should be used primarily at the early inflammatory stage and should not be prescribed once atrophy has developed.

Systemic agents and other treatments

The role of systemic agents such as oral corticosteroids in localised morphoea is limited. They are usually used when there is widespread involvement or in cases where there is a high risk of disfigurement, especially in darker-skinned patients. Furthermore, the potential side effects need to be considered on an individual basis.

Colchicine* has been used with some success in selected patients, particularly those with generalised morphoea.[7] This medication's most common side effect is gastrointestinal, including diarrhoea.

A form of photochemotherapy, psoralen combined with broad-band ultraviolet A (PUVA), may be of benefit but its accessibility is limited to specialised dermatological centres. Long-wave ultraviolet A (UVA-1) may also improve generalised morphoea but is not readily available in Australia at present. Short-term side effects of PUVA include photosensitivity, nausea and pruritus; long-term side effects include photoageing and skin cancers.

In the paediatric population, combination treatment with calcipotriol, prednisolone and methotrexate* has been useful in halting the progression of en coup de sabre.

If morphoea occurs over joints, physical therapy is necessary to improve limb function. Psychological support and counselling may be needed when the disease has resulted in functional impairment or disfigurement. In the subtypes of morphoea that are likely to cause permanent disfigurement, prompt referral is important as systemic agents such as hydroxychloroquine*, methotrexate* and prednisolone may halt disease progression.

CONCLUSION

The generally good long-term prognosis of morphoea contrasts with the poorer prognosis of systemic sclerosis. Failure to diagnose and differentiate between the two diseases may result in unnecessary worry, needless investigations and prolonged systemic treatment for the patient with morphoea. It is important to recognise that for many patients with morphoea, sympathetic counselling and reassuring support may be all that is required therapeutically. It should be remembered that a successfully managed patient with morphoea has no more fear. MT

* Off-label use of medication.

REFERENCES

1. Peterson L, Nelson AM, Su WP, Mason T, O'Fallon WM, Gabriel SE. The epidemiology of morphoea (localized scleroderma) in Olmsted County 1960-1993. J Rheumatol 1997; 24: 73-80.
2. Prescott R, Freemont AJ, Jones CJ, Hoyland J, Fielding P. Sequential dermal microvascular and perivascular changes in the development of scleroderma. J Pathol 1992; 166: 255-263.
3. Chung L, Lin J, Furst DE, Fiorentino D. Systemic and localized scleroderma. Clin Dermatol 2006; 24: 374-392.
4. Gupta RA, Fiorentino D. Localized scleroderma and systemic sclerosis: is there a connection? Best Pract Res Clin Rheumatol 2007; 21: 1025-1036.
5. Christen-Zaech S, Hakim MD, Afsar FS, Paller AS. Pediatric morphea (localized scleroderma): review of 136 patients. J Am Acad Dermatol 2008; 59: 385-396.
6. Kroft EB, de Jong EM, Evers AW. Psychological distress in patients with morphoea and eosinophilic fasciitis. Arch Dermatol 2009; 145: 1017-1022.
7. Kim Y, Lee S, Choi J. Primary biliary cirrhosis in a patient with generalised morphoea. Hong Kong J Dermatol Venereol 2009; 17: 30-34.

COMPETING INTERESTS: None.