Lupus erythematosus

Lupus is a chronic autoimmune disease with the potential for multiorgan disorder and prominent involvement of the skin. There is great variety in the clinical features and severity from one patient to the next. There are several distinct and recognizable patterns of skin involvement that are specific for lupus.

Patient History

Lupus preferentially affects young women but can occur at any age. The most common clinical presentation involves skin rashes and constitutional symptoms, with fatigue and musculoskeletal complaints predominating. The skin manifestations of lupus are classified as acute cutaneous lupus, subacute cutaneous lupus, and chronic cutaneous lupus. Chronic cutaneous lupus includes several subtypes, the most common being discoid lupus. Less common forms of chronic cutaneous lupus are lupus panniculitis, chilblain lupus, and tumid lupus. Some controversy exists about the relationship of tumid lupus to the rest of the lupus spectrum.

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Lupus is characterized by relapses and remissions. It is expected that the clinical pattern that develops early in the disease will predominate over the course of the illness. Recognition of the type of skin disease can help predict systemic disease because the subsets of cutaneous lupus relate to systemic disease differently (Fig. 1). Patients with lupus also present with skin findings that are not specific to lupus, and these include pernio, vasculitis, photosensitivity, alopecia, livedo, and bullous lesions.

It is important to carefully consider medications, because they can be a trigger for lupus. Subacute cutaneous lupus erythematosus (SCLE) is a form of lupus commonly attributed to medicines. More than 40 medications are reported to cause SCLE; of these, the most common are hydrochlorothiazide, diltiazem, angiotensin-converting enzyme inhibitors, and terbinafine. In recent years it has been recognized that the tumor necrosis factor (TNF) inhibitors cause multiple presentations of autoimmunity, including lupus. Patients often have systemic and cutaneous findings, although the systemic findings may be more prominent. In contrast with drug-induced lupus caused by procainamide, hydralazine, and minocycline, antihistone antibodies may not be present. Drug-induced forms often resolve when the offending medications have been stopped, although it may take months.

Clinical Findings

Acute cutaneous lupus

This condition presents as classic malar erythema, known as a butterfly rash (Fig. 2). It is present in about 40% to 50% of patients at the diagnosis of systemic lupus. Erythema spreading over the cheeks and nose, sparing the sun-protected areas like the nasolabial fold, is characteristic. There can be extension onto the forehead and chest. In a few patients a more generalized eruption accompanies this rash, involving the extensor arms and hands and often localized to the interphalangeal skin and sparing the skin over the knuckles. Erythema and small papules tending toward confluence are present. Small amounts of scale may be found. The clinical course of the rash can worsen with sun exposure or reappear with systemic disease flares.

Fig. 1. Relationship of lupus subsets to systemic disease. ACLE, acute cutaneous lupus erythematosus; BLE, bullous lupus erythematosus; CCLE, chronic cutaneous lupus erythematosus; HLE, hypertrophic lupus erythematosus; LEP, lupus erythematosus profundus; LET, lupus erythematosus tumidus; NLE, neonatal lupus erythematosus; SCLE, subacute cutaneous lupus erythematosus; SLE, systemic lupus erythematosus. (Courtesy of J. Callen, MD, Louisville, KY.)
Many of these patients have systemic lupus and this is reflected in their serologies. Positive antinuclear antibodies (ANA) are common and titers are generally greater than or equal to 1:160. Anti–double-stranded DNA and anti-Smith antibodies are both specific for systemic lupus, with anti-Smith being seen less frequently. Multiple other antibodies can be found that are beyond the scope of this article. This rash resolves in days to weeks without scarring, although marked postinflammatory hyperpigmentation is common.

**Subacute cutaneous lupus**

These patients have marked photosensitivity. The rash is characterized by erythematous, scaly, and polycyclic or ring-shaped macules and papules predominantly on the upper chest and back (Fig. 3). The rash can extend down the arms but rarely involves the legs. The central face is characteristically spared, although peripheral lesions do occur on the face. Up to 70% of patients have SSA (anti-Ro) antibodies, and although

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**Fig. 2.** Facial erythema in lupus, with redness over both cheeks, slightly over the nose. Rash does not extend over the nasolabial fold.

**Fig. 3.** Widespread classic skin lesions of subacute cutaneous lupus.
they may meet criteria for systemic disease they can be reassured that they are more likely to have a benign course. Again, postinflammatory pigment changes are common but scarring is usually avoided. Careful scrutiny for a causative medication is important.

**Discoid cutaneous lupus**

Discoid lupus starts out as a papule that gradually expands into an indurated round plaque. As it enlarges the center develops a depressed scar that is hypopigmented (Fig. 4). There is often scale, and follicular plugging may be visible. When the scalp is involved the follicular changes result in scarring and irreversible alopecia. The lesions can be triggered by sun exposure and also by trauma (Koebner effect). The most common areas involved are the head and neck, and, when only here, the disease is deemed localized. When it extends below the neck, it is described as generalized. When the presentation is predominantly discoid lesions, the patients have minimal risk for systemic involvement. However, up to 20% of patients with systemic lupus have discoid lesions at some point in the course of their disease.

**Variations of lupus skin findings**

There are several additional skin findings or variations that are less common but associated specifically with lupus. Overlaps occur between lichen planus and lupus, as well as lupus and erythema multiforme, known as Rowell syndrome. Patients can present

Fig. 4. Discoid lupus.
with lupus panniculitis, in which deep nodules are found in the skin, often with overlying discoid changes, most commonly on the head, neck, chest, and proximal arms. This condition is called lupus profundus. In addition to the mucosal ulcers often seen early in diagnosis, about 10% of patients with lupus have other mucosal findings. These mucosal findings are more common in patients with discoid type skin disease. Any mucosa can be affected. In the mouth it is most likely to involve the lips and buccal mucosa. The appearance may resemble oral lichen planus.

Tumid lupus presents with erythematous, almost urticarial, plaques. On skin biopsy, it is characterized by prominent mucin deposition. Usually there is little of the interface dermatitis that is characteristic of other lupus skin presentations. These patients are typically ANA negative and do not progress to develop systemic lupus. Tumid lupus is marked by extreme photosensitivity with prominent involvement in photoexposed areas, including the face. Extension to photoprotected sites only rarely occurs. Because of its various nonlupus features, experts debate whether tumid lupus should be considered part of the lupus spectrum.

**Treatment**

Treatment of cutaneous lupus is based initially on severity, with attention later to clinical response and escalation of therapy for resistant disease (Fig. 5). Large clinical trials evaluating therapies for skin disease are lacking, so treatment recommendations are often based on case reports, case series, and expert opinion. Topical therapy is a useful starting point and may have additive benefit even in patients on systemic treatment. For the treatment of skin disease, systemic therapy beyond hydroxychloroquine is reserved for patients with widespread skin lesions, disfiguring or scarring lesions, and refractory disease.

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**Fig. 5. Therapeutic considerations in cutaneous lupus.**
Hydroxychloroquine is an important therapy for lupus. This therapy is effective for treating musculoskeletal complaints, skin findings, and constitutional symptoms. It also prevents flares of disease, reduces thrombotic complications of lupus, and improves survival.\textsuperscript{9} It should be continued even when other therapy is added. Dosing is usually 200 to 400 mg per day up to a maximum of 6 mg/kg. Regular monitoring for eye toxicity is recommended. Patients may also develop blue-gray to brown pigment changes in the skin. When this occurs it is often subtle and can take years to develop. An alternative is chloroquine and in refractory cases quinacrine may be added.

Topical steroids of medium (ie, triamcinolone 0.1\%) to high potency (ie, clobetasol 0.05\%) are used up to 2 times per day. High-potency steroids are more effective and the risk of atrophy from therapy is often outweighed by the potential for disfiguring scars if left untreated. To minimize the risk of skin atrophy, use of steroids for 1 to 2 weeks then a 1-week break can be recommended. Alternating topical steroids with topical calcineurin inhibitors may avoid steroid-induced skin atrophy without interruption in therapy.\textsuperscript{8}

If skin disease remains uncontrolled after initiating the treatment described earlier, then consideration for treatment with additional systemic agents is reasonable. Initial choices include methotrexate, mycophenolate, and azathioprine.\textsuperscript{10} For SCLE and discoid lesions, thalidomide and lenalidomide are considered effective.\textsuperscript{11–13} The historical context for these therapies and the demonstrated severe risk of birth defects have resulted in a required monitoring program for providers and patients to comply with in order for these medications to be prescribed. In 2011, belimumab, an infusion therapy that modulates B cells, was approved for the treatment of lupus and results show improvement in skin disease.\textsuperscript{14} Rituximab has shown effect in case reports, but relapses are common and the role in lupus therapy is still being determined.\textsuperscript{15} Oral retinoids (isotretinoin and acitretin) are used for hyperkeratotic lesions or lesions on the palms and soles. Dapsone has been recommended for bullous lesions, urticarial vasculitis, and oral ulcers. Overall response rates with dapsone are low.\textsuperscript{3,10}

The inherent photosensitive nature of lupus along with routine recommendation for sun protection results in vitamin D deficiency,\textsuperscript{16–18} which has been documented in multiple studies and ongoing investigations are underway to examine the complex interplay of the immune system and vitamin D. Clinical monitoring and supplementation when necessary are recommended. In addition to sun protection with broad-spectrum sunscreens (recommended ingredients are titanium, zinc, and Mexoryl),\textsuperscript{19} it is also important to encourage smoking cessation. Smoking reduces the effectiveness of therapy and can be a risk factor for more severe disease.

**Future Therapy**

Future directions of therapy for lupus include anti-interferon therapies and clinical trials are currently underway.\textsuperscript{20} Although photoprovocation of lupus is well documented there is interest in modulation of lupus and clinical improvement with specific wavelengths of light (ultraviolet [UV] A1, 340–400 nm).\textsuperscript{21} The exact role in therapy is yet to be determined.

**DERMATOMYOSITIS**

Dermatomyositis is an autoimmune disease that classically affects skin and muscles. Vigilance for important associations with lung disease and malignancy can be aided by serology testing. Distinctive skin findings are readily identifiable and assist in diagnosis.
**Patient History**

Patients with dermatomyositis usually present in midlife and women are 2 times more often affected than men. The incidence of dermatomyositis is increasing worldwide, although this is attributed to better diagnosis. It is associated with geographic latitude, and regions with higher surface UV exposure have higher incidences. Photosensitivity is an important clinical feature of dermatomyositis.

Symptoms may come on quickly or develop over several months. Skin findings may precede or follow characteristic symmetric muscle weakness of the proximal muscle groups (classic dermatomyositis). It is important to recognize that skin findings can occur without ever developing muscle weakness (amyopathic dermatomyositis). Other subtypes have been described. Postmyopathic dermatomyositis follows classic dermatomyositis with persistent skin disease despite resolution of muscle symptoms. Hypomyopathic dermatomyositis is also described and these patients have very mild subclinical muscle disease.

Patients may report difficulty swallowing or changes to their voice caused by effects on muscles in the pharynx and upper esophagus. This condition may portend more severe disease. Interstitial lung disease can occur and can be rapidly progressive and fatal. Swallowing difficulty has been associated with lung disease.

**Clinical Findings**

The distinctive skin findings of dermatomyositis are periorbital erythema and swelling described as a heliotrope rash. Redness and papules overlying the knuckles are called the papules of dermatomyositis (Gottron papules) and are often seen in conjunction with ragged cuticles, dilated nail fold capillaries, and cuticle hemorrhage (Fig. 6). A less common hand finding is hyperkeratosis along the sides of the fingers and on the palms, sometimes with surrounding erythema known as mechanic’s hands (Fig. 7). Gottron sign is violaceous erythema over the elbows and knees (Fig. 8). The finding of erythema, hypopigmentation and hyperpigmentation, and telangiectasia extending over the shoulders (shawl sign; Fig. 9), in a V distribution of the neck, and lateral thighs (holster sign), is often described simply as poikiloderma associated with dermatomyositis. Scale is often present and the rash is difficult to distinguish from lupus or other papulosquamous disorders. Attention to the characteristic distribution and careful examination of the nail folds is helpful in these cases. Scalp involvement with erythema, scale, and often intense pruritus can persist even when other areas of skin involvement have come under control.

![Fig. 6. Nail fold changes in dermatomyositis.](image-url)
Cardiac involvement, with conduction abnormalities or arrhythmias, occurs infrequently.\textsuperscript{25} Calcinosis of the muscles and skin, common in children but unusual in adults, is a challenging complication to treat.

Skin biopsy is not specific, but can support the diagnosis of dermatomyositis. It shows similar features to lupus, with interface dermatitis and perivascular lymphocytic inflammation. Clinical findings remain the most distinguishing feature. Along with strength testing, detection of increased muscle enzyme levels in serum can indicate muscle involvement. Levels of creatine kinase, lactate dehydrogenase, aldolase, aspartate aminotransferase, and alanine aminotransferase are all potentially increased in muscle disease, and checking several is reasonable. Although the complete evaluation of muscle disease is not discussed here it is important to remember that once dermatomyositis has caused extensive damage to the muscles and atrophy has occurred, the muscle enzyme levels may return to normal despite significant muscle disease.

Fig. 7. Mechanics hands with dermatomyositis.

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Fig. 8. Violaceous skin changes over the knees, Gottron sign in dermatomyositis.
A positive ANA test occurs in 80% to 90% of patients. In addition, multiple myositis autoantibodies can be detected, and increasing relevance and clinical implications are being recognized for these antibodies (Table 1). Clinically important antibodies include Mi-2, MDA5, anti-p155/140, and antisynthetase antibodies. Mi-2 is most specific for dermatomyositis, but is not very sensitive. When present, it suggests treatment-responsive disease.

The constellation of rapidly progressive severe interstitial lung disease, myositis, arthritis, often mechanic’s hands, and antisynthetase antibodies, sometimes with SSA antibodies, is referred to as the antisynthetase syndrome. Fever and Raynaud phenomenon may be additional symptoms. There have been 8 distinct antisynthetase antibodies detected, and more are likely be to be found in the future. Of these, anti–Jo-1 is the most common. In some cases, lung disease is the initial presenting symptom and other features are less prominent.

The presence of MDA5 antibodies is also associated with severe interstitial lung disease. It is found in amyopathic forms of dermatomyositis and may be associated with punched-out–type ulcerations in the skin. In both of these scenarios the lung

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Frequency in DM (%)</th>
<th>Clinical Association</th>
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<tbody>
<tr>
<td>Mi-2</td>
<td>20–30</td>
<td>Most specific antibody for DM. More responsive disease</td>
</tr>
<tr>
<td>MDA5</td>
<td>50</td>
<td>Associated with amyopathic DM. Increased ILD and skin ulcers, levels may decrease in response to therapy</td>
</tr>
<tr>
<td>P155/140</td>
<td>20–25 DM</td>
<td>Cancer-associated myositis, severe skin disease, high negative predictive value for malignancy</td>
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<td></td>
<td>40–75 cancer-associated DM</td>
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</tr>
<tr>
<td>Antisynthetase (Jo-1 and others)</td>
<td>5–10</td>
<td>Antisynthetase syndrome high frequency of ILD and arthritis</td>
</tr>
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Abbreviation: ILD, interstitial lung disease.
disease is common, affecting more than 70% of patients, and mortality is significant as well (~30%).

Overlaps exist between dermatomyositis and lupus, as well as systemic sclerosis. In these presentations, a variety of autoantibody patterns can be seen, including positive SSA, anti-RNP, anti-sm, and SCL.

An association between dermatomyositis and malignancy is well recognized. The clinical findings of dermatomyositis may precede, coincide with, or follow the cancer diagnosis. Recent analysis has suggested a 5-fold greater occurrence of cancer in these patients than would normally be expected. The malignancies most represented in the setting of dermatomyositis are hematopoietic and lung. A variety of other cancers are also noted to be over-represented, including ovary, colon, bladder, breast, cervix, pancreas, and esophagus. Cancer screening recommendations are not standardized. Consequently appropriate clinical decision making includes symptom-directed screening and the usual age-appropriate screening. It is reasonable to repeat the screening annually for 3 years, because this is when the risk of cancer-associated dermatomyositis tends to decrease. Because routine screening is not very effective for ovarian cancer, lung cancer, and pancreatic cancer, vaginal ultrasonography and a computed tomography scan of the chest, abdomen, and pelvis is ordered by some experts. The impact of anti-p155/140 antibodies on screening for malignancy remains to be determined. In light of the high negative predictive value (93%) for malignancy, in the setting of a negative anti-p155/140, it may be reasonable to consider cancer screening tests at diagnosis of dermatomyositis and then return to the usual recommended schedule for the patient’s age.

**Treatment**

Although treatment algorithms do not exist for dermatomyositis, a common approach is to start oral prednisone at doses around 1 mg/kg/d for patients with evidence of muscle disease. This dose is tapered slowly, often over months. With long courses of steroids, side effects are a predicted complication. Concurrent addition of a steroid-sparing immunosuppressive drug may shorten the need for high-dose steroids and prevent steroid-associated side effects. Commonly used therapies are methotrexate (10–20 mg per week), mycophenolate (1000–1500 mg twice daily), or azathioprine. Often these can be started at the same time as prednisone. In cases resistant to standard therapy, intravenous immunoglobulin or rituximab are reasonable choices. For interstitial lung disease, a more aggressive treatment regimen is usually required, and is not discussed here.

Once therapy has been initiated it seems that exercise, both resistance and aerobic, may benefit patients with myositis. It is reasonable to recommend an exercise program after about 4 weeks of systemic treatment.

Skin disease in patients with dermatomyositis can be resistant and attention to skin-specific treatments is important. Photoprotection is recommended, with clothing and broad-spectrum sunscreen. Midpotency topical steroids and topical calcineurin inhibitors are also helpful for the rash and related itching. Addition of hydroxychloroquine or chloroquine can benefit patients significantly. Quinacrine can be added when patients do not respond to a single agent. In patients with amyopathic forms, escalation of therapy to other systemic agents, including methotrexate and mycophenolate, may be required to control symptoms.

**Future Therapy**

Although TNF cytokines play a role in the pathogenesis of dermatomyositis, use of TNF blockers has not been uniformly successful. Note that dermatomyositis has been
identified as an unusual autoimmune side effect of TNF blocker therapy. Further research with new agents may identify effective treatments. In 1 patient, inhibition of the JAK pathway with ruxolitinib resulted in improvement of dermatomyositis.

MORPHEA

Morphea is an idiopathic inflammatory skin disease that presents in a variety of ways, but ultimately causes skin hardening and in some cases loss of function. Although understanding of the pathophysiology is incomplete, it seems to share features with systemic sclerosis. Despite this common pathophysiology, morphea and systemic sclerosis are readily distinguished clinically and should be thought of separately. Morphea is largely confined to the skin and subcutaneous tissues, whereas systemic sclerosis is a multisystem disease. Distinctive findings on examination can help differentiate the two conditions and direct appropriate therapy.

Patient History

The onset of symptoms in morphea may be insidious, and studies have shown a delay in diagnosis of a year or longer. Morphea is nearly 3 times more common in women than in men. It affects children and adults equally. Patients may describe arthralgias, myalgias, and fatigue occurring along with morphea but the systemic symptoms of gastrointestinal reflux, pulmonary symptoms, and cardiac symptoms seen in systemic sclerosis are not characteristic of morphea.

In addition to the skin findings, there are some special clinical considerations in patients with morphea. Morphea that involves the scalp or occurs around the eyes can be associated with symptoms of uveitis, headaches, or seizures. Several reports show a strong association between morphea and genital lichen sclerosis. Frank discussion with patients about genital symptoms, including skin changes, itching, or burning, should be included in the history because patients may be reluctant to volunteer these concerns.

The course of morphea is one of remission and relapse. Ongoing monitoring is helpful to treat relapses early and avoid additional disability.

Clinical Findings

Plaque morphea

Several distinct patterns of morphea occur on the skin. Plaque-type morphea is the most common. The earliest lesions are often erythematous plaques with little induration; sometimes a lilac border is observed at the periphery of the lesion (Fig. 10). This condition progresses to central sclerosis with induration and smooth yellow to white scarring. There is often peripheral erythema or hyperpigmentation (Fig. 11). Over time, destruction of the hair and sweat glands occurs in the affected skin. The plaques can increase in size and number. Involvement in the inframammary area, the area around the hips, and on the low back together as a pattern is common.

Generalized plaque morphea

When the plaques are greater than 3 cm in size and there are more than 4 plaques involving 2 body areas it is classified as generalized plaque morphea. Patients can have superficial or deep involvement in the skin.

Pansclerotic morphea

In rare patients the course is rapid and progressive with multiple enlarging plaques that eventually result in widespread involvement of close to the entire skin, except
for sparing of the hands and feet. This presentation is referred to as panniculitis and requires aggressive treatment.

**Linear morphea**
The linear subtype of morphea occurs most commonly in children. In this type, plaques develop in a linear pattern, eventually coalescing into a single band of scarring that can extend the length of a limb. When it crosses joints it can cause loss of mobility.

![Fig. 10. Early morphea plaque with violaceous border.](image)

![Fig. 11. Morphea plaques on the low back.](image)
because of scarring. Linear morphea often involves deeper structures, including muscle and bone. When it involves the scalp and forehead it has been referred to as en coup de sabre (Fig. 12), which is a French term referring to the injuries sustained by soldiers hit on the head by a sword.

Patients often have a mixed presentation with both linear and plaque types. Several less common forms of morphea have been reported, including superficial morphea, guttate morphea, bullous morphea, and keloidal morphea.

The diagnosis of morphea is made based on history and clinical examination of the skin. Differentiating from systemic sclerosis is an important first consideration. In contrast with systemic sclerosis, which begins with sclerodactyly, morphea usually does not involve the hands. Raynaud phenomenon is characteristic of systemic sclerosis and is absent in morphea. Nail fold and serologic findings of systemic sclerosis are not present in morphea. Examination of the genital skin is recommended to detect asymptomatic lichen sclerosus and to direct treatment if present. The most likely clinical differential diagnoses to consider are eosinophilic fasciitis, lipodermatosclerosis, graft-versus-host disease, and nephrogenic systemic fibrosis (Box 1).

Skin biopsy is usually not required. When performed it shows an inflammatory pattern of lymphocytes and plasma cells in the dermis and subcutis, and in late lesions thickened collagen bundles are present. The histologic findings may be indistinguishable from systemic sclerosis.

There are no specific laboratory tests to confirm the diagnosis of morphea. Patients with morphea, especially the linear and deep subtypes, often have a positive ANA test. The clinical utility of this test is yet to be determined and routine testing is not clinically useful.

**Treatment**

For patients with early and limited skin involvement, topical therapy with high-potency steroids, topical calcineurin inhibitors, or calcipotriene is recommended. Patients with continued progression or generalized disease should be treated with either phototherapy (nbUVB [narrow band ultraviolet B], bbUVA [broad band ultraviolet A], or UVA1) or systemic treatment with either methotrexate or methotrexate combined with pulsed-dose steroids (1 g of Solu-Medrol intravenously daily for 3 consecutive days, repeated...
monthly for 3–6 months). Systemic therapy should be considered for linear morphea, which often is more aggressive and disabling. An alternative to methotrexate is mycophenolate using doses of 1000 to 1500 mg twice daily.49

Further progression after a period of quiescence is encountered. Treatment should be restarted in these cases. If the clinical findings are not clear and comparison photographs are not available a skin biopsy may be helpful. The finding of inflammation on the biopsy suggests clinical activity and may be compelling to restart therapy.

**Future Therapy**

Future therapeutic directions in morphea include studies to better understand the comparative effectiveness of systemic treatments and phototherapy. The timing of therapeutic intervention may also become an important part of treatment. As awareness increases it may be possible to treat early, when the disease is thought to be more responsive.

Ultrasonography and MRI studies have been able to detect the skin and deeper inflammatory and tissue changes associated with morphea. Ultrasonography or MRI may be useful for monitoring response to therapy, surveillance for recurrence, and to guide clinical decision making regarding the use of systemic therapy in this disease.38

**REFERENCES**


