1 Evaluation of the Integumentary and Musculokeletal Systems: An Approach to the Interdisciplinary Examination for Overlap Diseases

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Introduction

A comprehensive skin examination is an integral component to the evaluation of patients with, or suspected to have, an inflammatory or autoimmune musculoskeletal disease. Integumentary findings, inclusive of the skin, nails and hair, often provide specificity when clinical features otherwise yield an indeterminate diagnosis. These findings, although sometimes subtle (i.e., periungual swelling and erythema), may be detected through an attentive awareness of these signs and proper approach. While a full description of the dermatological lexicon and the comprehensive examination of the integument is beyond the scope of this chapter, the most important aspects of the examination that will support the evaluation and management of patients with connective tissue conditions are described herein. We conclude with a summary of elements of the musculoskeletal and joint exam that we recommend synthesizing with skin examination findings, particularly in the case of evaluating patients with rheumatologic skin disease.

General Recommendations

The examination of the integument should be performed in a well-lit room, ideally with natural sunlight, as this type of light is least likely to alter the perception of erythema color. A penlight is helpful in examining areas such as the mouth or the ears in which there is less natural light exposure. The dermatoscope, a hand-held magnifier and specialized light source, may be helpful in visualizing small structures such as capillary loops in the nailfolds. Dermoscopy is particularly helpful in examining the nailfolds of patients with systemic sclerosis, dermatomyositis, systemic lupus erythematosus, mixed connective tissue disease, and Raynaud's phenomenon. Several studies suggest that the use of dermoscopy can replace the need for nailfold capillaroscopy in many instances (Bergman, JAMA Derm 2003).

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The patient should be undressed at least to undergarments and donning a proper hospital gown. The patient should remove eyeglasses, dentures when feasible, jewelry including the watch, as well as make-up and nail polish, so that relevant findings are not hidden. It is important to be mindful of respecting the patient's modesty when undressed. Asking the patient's permission to move the gown or undergarments to expose the skin during the examination and informing the patient when examining sensitive areas helps to alleviate any unease.

A systematic approach that is used for every patient's examination will help ensure all relevant areas of integument are thoroughly evaluated. In patients with connective tissue disease, take note of several commonly affected areas, including the scalp, conchal bowls of the ears, face including the eyelids, oral mucosa, neck, upper chest, upper back, dorsal hands, nails and nail folds. It is important to examine all affected areas to determine the exact distribution of an eruption. In addition, making note of the eruption's color, shape, configuration, secondary changes such as scale, and palpability complete the morphologic assessment.

In addition, a complete and appropriately tailored rheumatologic/musculoskeletal examination can complement a thorough skin exam, informing the diagnosis and management for patients with rheumatologic-dermatologic overlap conditions. We focus here on the overview of assessing joint pathology; a thorough examination of neuromuscular disease is outside the scope of this introduction.

Distribution

The most important distribution pattern to recognize among patients with autoimmune and connective tissue diseases is the **photodistributed** eruption (Fig. 1.1). Photodistributed eruptions occur in areas usually not covered by clothing, which thereby receive the most direct ultraviolet exposure. These areas namely include the forehead, bilateral cheeks, nose, lower chin, lateral neck, a triangular area corresponding to the opening of a V-neck shirt on the anterior upper chest,

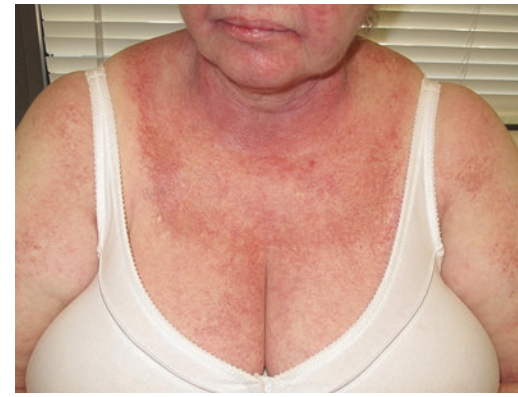

Fig. 1.1 Photodistributed. Erythema and poikiloderma most prominent over the cheeks, neck, and upper chest in a patient with dermatomyositis

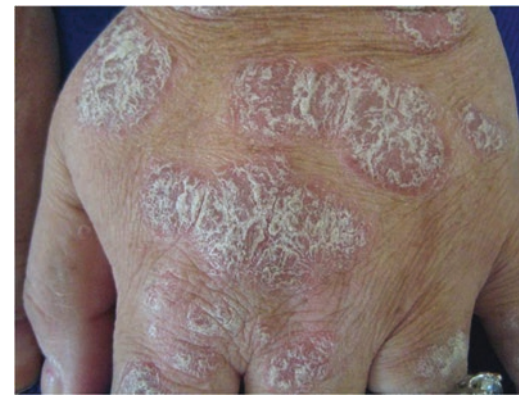

Fig. 1.2 Extensor. Well demarcated red erythematous plaque of psoriasis with white to silver colored scale on the dorsum of the hand

the upper back, flexor forearms, and dorsal hands including the phalanges. Typically, there is also relative sparing of skin fully protected from the sun. Photosensitive eruptions occur in systemic and cutaneous lupus erythematous, Sjogren's syndrome, and dermatomyositis.

Plaques of psoriasis among patients with psoriatic arthritis are classically **extensor** (Fig. 1.2) in their distribution pattern. These plaques occur over the scalp, buttocks and extensor surfaces of all four extremities, including the elbows and knees. This is in contrast to atopic dermatitis (eczema) which is most often apparent on flexural surfaces. Psoriasis plaques are also commonly **bilateral and symmetric**, particularly on extensor surfaces. In some cases, psoriasis

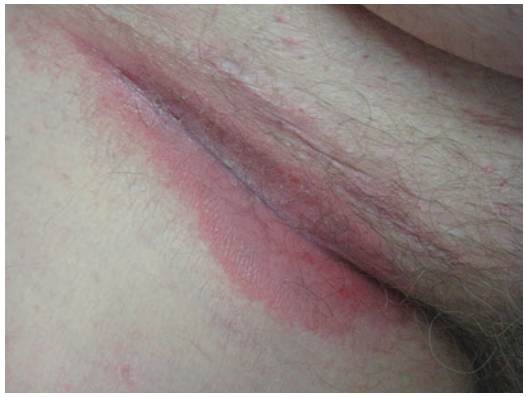

Fig. 1.3 Inverse. Well demarcated red erythematous plaque of psoriasis without scale in the inguinal crease

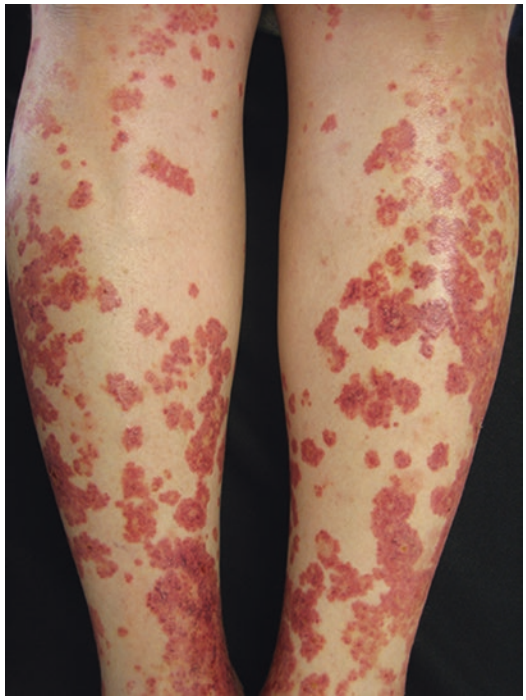

Fig. 1.4 Dependent. Purpuric (non-blanching) red to purple patches, papules and plaques over the lower extremities and more prominent distal to the knees

patients may present with an **inverse** distribution (Fig. 1.3), in which well-demarcated pink erythematous non-scaly plaques will appear in intertriginous folds such as the axillae, inframammary folds, and inguinal creases.

In leukocytoclastic vasculitis, palpable purpura is noted on **dependent** areas of the body (Fig. 1.4), such as the ankles and lower legs, and occasionally the buttocks and back in bedridden patients.

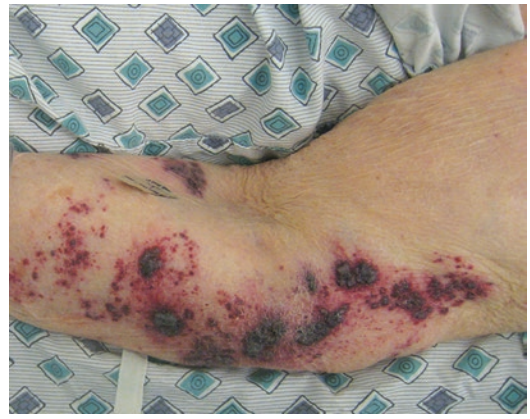

Fig. 1.5 Dermatomal. Grouped hemorrhagic vesicles and plaques following the C6 dermatome in this patient with herpes zoster

A **dermatomal or zosteriform** (Fig. 1.5) distributed eruption is in the distribution of a single spinal afferent nerve root (dermatome). It is unilateral and does not cross the midline of the body. The classic dermatomal eruption is herpes zoster, which commonly afflicts patients with chronic disease, immunotherapies, or advanced age.

Color

Perhaps the most important additional feature of an eruption, other than its distribution, is color. Color, which is often the first visual assessment made, is reliably reproducible with particular types of pathologies, including connective tissue diseases. As such, color provides meaningful insight into pathologic processes of the skin and facilitates clinical diagnosis.

Erythema represents the blanchable pink to red color of skin or mucous membrane. It exists in different colors, and to call a primary lesion *erythematous* alone is incomplete. Describing erythema with the color it most closely resembles provides a meaningful clue to diagnosis. For example, violaceous erythema of the periorbital area (heliotrope, Fig. 1.6), and in particular the lid margin, is highly suggestive of dermatomyositis. We note the same color of erythema in other

dermatoses of connective tissue disease which also involve an interface dermatitis and dermal-epidermal junction. As another example, lilac colored erythema surrounding a slightly whitish and firm plaque is suggestive of morphea.

Red blood cells that extravasate from cutaneous vessels into skin or mucous membranes result in reddish-purple patches referred to as purpura. The application of pressure with two glass slides or an unbreakable clear lens (diascopy) on a reddish-purple lesion is a simple and reliable method for differentiating redness due to vascular dilatation (erythema) from redness due to extravasated erythrocytes or erythrocyte products (purpura). If the redness is non-blanching under the pressure of the slides, the lesion is purpuric. As extravasated red blood cells decompose over time, the color of purpuric lesions changes from bluish-red to yellowish-brown or green. Petechiae are tiny, pinpoint purpuric macules. Ecchymoses are larger, bruise-like purpuric patches. These lesions correspond to a non-inflammatory extravasation of blood. If a lesion is purpuric and palpable ("palpable purpura"), the suggestion of an inflammatory insult to the vessel wall as a cause of extravasation of blood and inflammatory cells exists. The classic histopathological correlate to palpable purpura is leukocytoclastic vasculitis (Fig. 1.4).

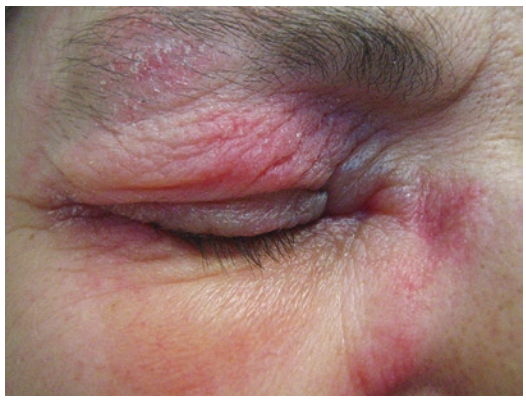

Fig. 1.6 Violaceous color. Intensely pink to purplish erythema, swelling and scale in the periorbital region representing the heliotrope in this patient with dermatomyositis

Erythema of any type is difficult to detect in darker skinned patients. Erythema in these instances may appear subtle or may appear more violaceous in color even when the true color is red or pink.

Shape or Configuration

Accurate appreciation of the shape or configuration of lesion(s) will facilitate narrowing of the differential diagnosis and specificity in diagnosis. There are several shapes and configurations of relevance to patients with inflammatory and autoimmune connective tissue diseases.

A linear configuration describes a lesion which resembles a straight line. This configuration may apply to a single lesion or to the arrangement of multiple lesions. Its appearance may suggest that the Koebner phenomenon (Fig. 1.7), defined as the appearance of the same lesion on previously normal appearing skin, has occurred in response to scratching or trauma. Psoriasis and Behcet's are two examples of conditions that exhibit koebnerization.

Round to oval, or coined-shaped, lesions that are uniform from the edges to the center of the lesion may be termed discoid (Fig. 1.8). Discoid lupus erythematous typically presents with coin-shaped plaques involving the scalp, face, and conchal bowls of the ears.

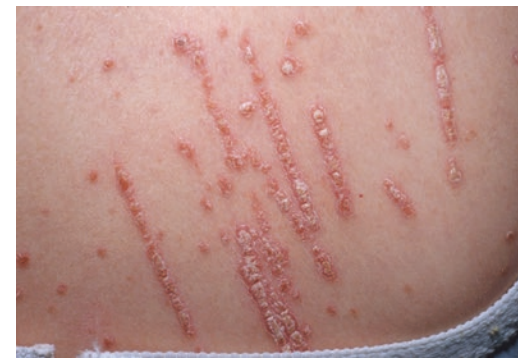

Fig. 1.7 Linear configuration. Well demarcated red erythematous linearized plaques on the back over areas that have been stimulated by scratching in a psoriasis patient

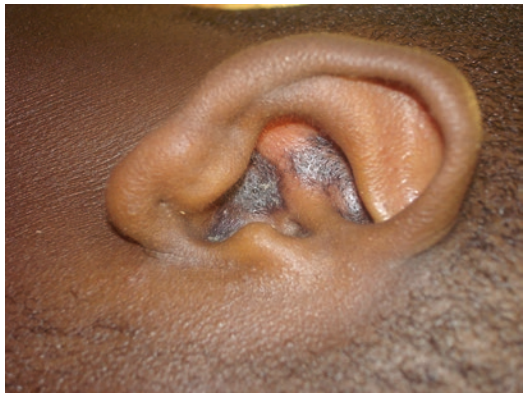

Fig. 1.8 Discoid shape. Rounded brown and slightly erythematous think plaques with follicular plugging in the conchal bowl of this patient with discoid lupus

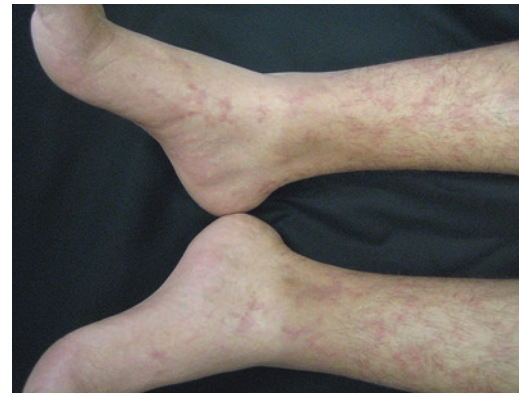

Fig. 1.10 Reticular. Light purple patches forming irregular, broken networks on the lower extremities and feet of this patient with cutaneous polyarteritis nodosa

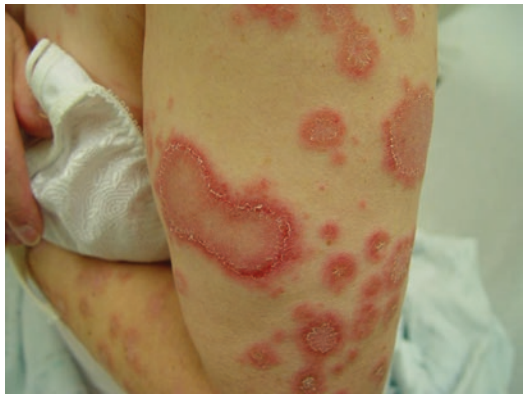

Fig. 1.9 Annular. Red erythematous annular plaques with peripherally located scale on the upper arms in this patient with terbinafine-induced subacute cutaneous lupus

In annular or ring-shaped lesions, the edge differs from the center, either by being flatter, more raised, scaly, or differing in color. Annular lesions that are incomplete form an arcuate shape. Annular lesions may coalesce to form circles or rings, in a polycyclic configuration. The annular, arcuate, and polycyclic patterns represent one form of subacute cutaneous lupus erythematosus (SCLE) (Fig. 1.9). Urticaria also commonly presents with the same shapes and configurations.

The reticular configuration is represented by net-like or lacy patches with somewhat regularly spaced rings or partial rings and sparing of intervening skin. The netlike pattern is more regular in livedo reticularis, whereas the rings in livedo racemosa tend to be more broken up and jagged (i.e., cutaneous polyarteritis nodosa, Fig. 1.10).

Scaling

When epidermal differentiation is disordered, accumulation and casting of stratum corneum become apparent as scale, which ranges in size from fine dust-like particles to extensive parchment-like sheets. Not all scales are similar, and the expert dermatologist with a well-trained eye can obtain diagnostically useful information from close examinations of the type of scale present.

Scale may be fine or thick, dry or greasy, loose or adherent, and may range in color from silvery-white to yellow or brown. In some instances, the diagnosis is based on specifying the type of scale present. For example, SCLE and psoriasis may both present with rounded well demarcated red erythematous plaques with scale. However, the scale in SCLE may be finer and more prominent around the periphery, whereas the silver-white colored scale in psoriasis will cover the plaque. Although layered, the scale in psoriasis is adherent and leads to pinpoint hemorrhage when peeled away. Scale in SCLE easily flakes off without bleeding. Plaques in discoid lupus erythematous also have an adherent scale which extends into the orifices of dilate hair follicles.

When the scale on these plaques is lifted, keratotic spikes formed with hair follicles under the surface of the scale may be visualized. These spikes are said to resemble carpet tacks, and hence this finding specific to Discoid lupus erythematosus (DLE) is known as the as "carpet tack" sign. Complete absence of scale is also helpful in differentiating eruptions. For example, urticaria and urticarial vasculitis may present as erythematous annular and polycyclic plaques, similar to SCLE. However, the former eruptions have no scale.

Consistency

Palpation of lesions is an important part of the physical examination. Plaques that are thick or firm may suggest fibroses of the skin, such as in morphea or systemic sclerosis. When the pannus of the skin in inflamed, as with erythema nodosum, the presentation typically includes rubbery to firm nodules on the lower extremities.

A similar, more diffuse firmness along with tiny dimples in the skin can be appreciated with deep fascial inflammation noted in eosinophilic fasciitis. "Rock" hard whitish plaques or nodules may represent calcinosis, seen in patients with dermatomyositis who have had an aggressive course, delay in treatment, or undertreatment, or in juvenile patients with the disease. Similar nodules can be seen in patients with limited scleroderma.

Scalp/Hair/Nails/Oral Mucosa

In addition to the skin examination, evaluation of the hair, nails, and oral mucosa offers important clues in the diagnosis of connective tissue disorders. Patients with Sjogren's syndrome have decreased salivary pools in the mouth, and in longstanding cases, the dorsum of the tongue may appear lobulated. In systemic lupus erythematosus, patients may have painful oral ulcers, most frequently observed on the roof of the mouth at the junction of hard and soft palates. In Behcet's disease, patients may present with multiple, painful, large aphthous ulcers on the mucosal lips, buccal mucosa and tongue.

The nail unit is comprised of the nail plate, nail bed, nail matrix, lunula, eponychium (cuticle), proximal nail folds (skin at base of nail plate), perionychium (skin at sides of nail plate), and the hyponychium (skin under free edge of nail plate). In connective tissue disorders, one or several of these structures may be altered. In psoriasis, for example, involvement of the nail matrix results in pitting of the nail plate. This finding, however, is not specific to psoriasis and can be seen in several other conditions, including alopecia areata and eczemas, and may also be present in otherwise healthy individuals. In psoriasis, patients will typically have multiple pits (>10) involving several fingernails. Psoriasis patients also commonly have yellow colored "oil spots" at the distal portions of the nail (Fig. 1.11). The discoloration occurs when the nail plate separates from the involved nailbed. The cuticles in patients with dermatomyositis become hypertrophic and appear ragged, a finding known as Samitz sign (Fig. 1.12). The proximal nailfold is also typically erythematous and edematous with dilatations in capillary loops, as it is in patients with systemic lupus erythematosus (Fig. 1.13). Capillary loops are also altered in patients with lupus erythematous and systemic sclerosis. In patients with systemic sclerosis, capillary loss ("dropout") often alternates with dilated capillary loops, forming a distinc-

Fig. 1.11 Oil spot. Yellowish discoloration of the distal fingernails due to separation of the nail plate from the nail bed (onycholysis) in psoriasis

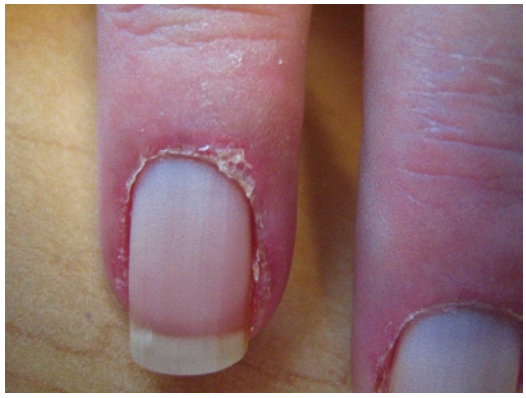

Fig. 1.12 Ragged cuticles. Periungual erythema and hyperkeratosis of the proximal and lateral nail folds (Samitz sign) in this patient with dermatomyositis

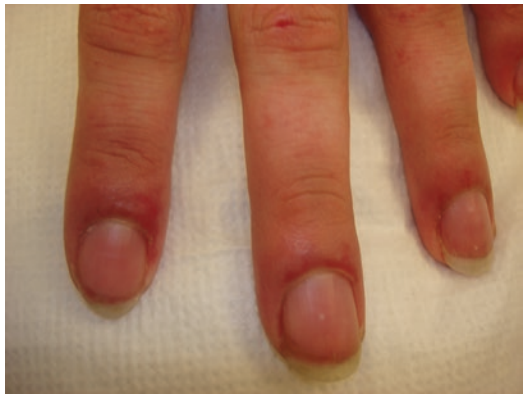

Fig. 1.13 Periungual erythema. Edema and periungual erythema in patients with systemic lupus erythematosus

tive pattern. The capillary abnormalities in systemic lupus erythematosus are more subtle and less specific but can include capillary dropout, alterations in capillary length (either sorter or longer) and alterations in capillary morphology (including tortuous or meandering capillaries); capillary microhemorrhages may also be present. (Fig. 1.14).

The scalp examination is particularly important in patients suspected of having psoriasis or dermatomyositis. Not infrequently, the only manifestation of psoriasis among patients with psoriatic arthritis is scalp involvement. Psoriasis plaques in the scalp tend to be well marginated, often involving the hairline of the scalp, and have a characteristic silver-colored scale. Patients with dermatomyositis frequently demonstrate diffuse scalp erythema of a similar color to the heliotrope sign involving the periorbital area.

Alopecia involving the scalp is a frequent finding in patients with lupus erythematosus. Patients with systemic disease commonly have non-scarring alopecia, i.e., hair loss without damage or destruction of the follicle. They may experience excessive diffuse shedding of scalp hairs, with the potential for hair to regrow. In discoid lupus erythematosus, however, inflammatory injury of the follicular epithelium leads to scarring alopecia. Once lost, these hairs do not regrow, even when the inflammatory process has regressed with or without treatment. Along with

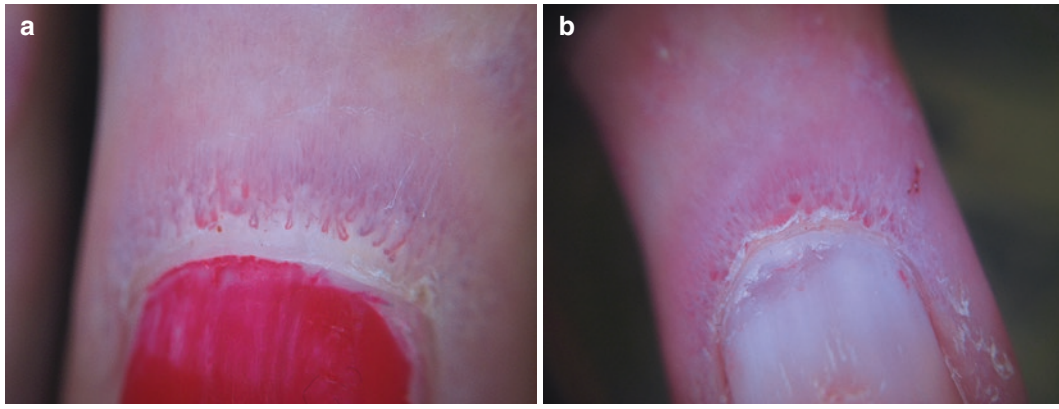

Fig. 1.14 Altered capillary loops. (**a**) sclerosis of proximal nail fold and dilatation of capillary loops in a patient with systemic sclerosis. (**b**) Advanced sclerosis of proximal nail fold with dilatation and further obliteration of capillary loops in a patient with systemic sclerosis

atrophy and discoloration of the skin, scarring alopecia results in significant morbidity. As such, this condition should be managed early and appropriately to minimize the occurrence of permanent hair loss.

Rheumatologic Musculoskeletal Examination

A targeted joint and musculoskeletal examination should be synthesized with the findings of a skin examination, particularly for patients with psoriasis who are at-risk for psoriatic arthritis, hidradenitis suppurativa (where increased prevalence of peripheral and/or axial spondyloarthritis occurs), connective tissue disorders, and other overlap conditions. Herein, we will review the hallmarks of the joint and muscle examinations that are most relevant for overlap diseases.

Joint Examination

Just as for the skin examination, a high-quality joint examination begins with establishing the optimal setting. Lighting should allow for visualization of the joint and any gross changes such as swelling, protrusions, overlying skin changes. The patient should be comfortable and in a gown or other non-restrictive clothing; this is important for both observation of the joints and the range of motion. Socks and shoes must be removed to properly assess all lower extremity joints.

A systematic and consistent approach to the examination will ensure a comprehensive and efficient evaluation. One commonly employed order includes evaluation of distal interphalangeal finger joints, proximal interphalangeal finger joints, metacarpal joints, wrist joints, elbow joints, shoulder joints, hip joints, knee joints, ankle joints, midtarsal joints, subtalar joints, metatarsal joints, proximal interphalangeal toe joints, distal interphalangeal toe joints, and spine (cervical to sacral), in succession.

Examination should begin with observation of each joint while the joint is at rest in order to assess for overlying skin changes [i.e., erythema, scale, papules, plaques, nodules), swelling, or gross deformities. Joint swelling is soft tissue swelling surrounding the joint, which is detectable along the joint margins. When a synovial effusion is present, it invariably means the joint has swelling.

After a joint is observed, it should be palpated. The objective of joint palpation is to assess for *warmth*, which may suggest inflammatory, infectious or crystal-induced arthritis; *tenderness,* and *swelling*. While the specific technique for joint palpation varies with each joint, the examiner should press into the fluid-filled bursa between the overlying muscle and bone. Sufficiently deep palpation of the joint bursa ensures that the examination is assessing the underlying joint, rather than the overlying muscle. Palpation is also necessary to confirm suspected swelling and to rule out bony swelling or deformity. Fluctuation is a characteristic feature of swollen joints.

Joint tenderness is pain in a joint under defined circumstances. Tenderness may occur at rest with pressure (i.e., MCPs and wrist joints); on movement of a joint (i.e., shoulder and tarsal joints); or may be assessed through questioning about joint pain (i.e., hips and cervical spine). Pain with physical manipulation of the bursa cushioning a specific joint may also suggest bursitis. When assessing for tenderness through palpation, pressure should be exerted by the examiner's thumb and index fingers to a sufficient degree to cause "whitening" of the examiner's nail bed, blanching about one-third to half-way down on the thumb to achieve adequate standard pressure.

Of particular utility is the assessment of 'entheseal' points; that is, points of tendon or ligament insertion into bone. Tenderness at these sites may suggest enthesitis, inflammation at the enthesis insertion, which is of particular relevance to the seronegative inflammatory arthritides such as psoriatic arthritis and other spondylo-athritis variants. Typical sites of examination include those noted in the Leeds Enthesitis Index, at the lateral epicondyles of the humerus, medial femoral condyles, and bilateral Achilles tendon insertions. The additional presence of widespread soft tissue tenderness might suggest a central sensitization syndrome such as fibromyal-

gia with referral to differentiating from inflammatory entheseal disease. To elicit diffuse soft tissue pain, palpation with similar pressure as noted above, to soft tissue areas of the upper back, upper arms and forearms, away from tendon insertion points or joints is applied. The finding of diffuse and often severe pain as experienced by the patient might suggest a concurrent or isolated pain syndrome such as central sensitization / fibromyalgia; it is worth noting that there is a relatively high co-prevalence of fibromyalgia with other inflammatory arthritides which means they may not be mutually exclusive in nature.

Finally, range of motion testing should be assessed for each joint. The patient's joints may be moved as tolerated to assess passive range of motion, or the patient can be observed performing active range of motion. Range of motion testing is important to assess restriction of movement, particularly in active range of motion testing. Range of motion testing can also be useful in determining presence of underlying swelling. For example, decreased dorsiflexion of the wrist and decreased elbow extension may suggest swelling of the involved joints.

The number of involved joints and overall distribution of joint tenderness and/or swelling is important for an examiner to synthesize. Joint diseases can be divided into monoarticular (one joint), oligoarticular (2–5), or polyarticular (>5) disease. The number of affected joints can inform the differential diagnosis of a patient's joint pain. For example, a monoarticular joint pain may be caused by crystal-induced disease (gout, calcium pyrophosphate deposition), septic arthritis, or a traumatic hemarthrosis; whereas, oligo- or polyarticular disease raises the possibility of rheumatoid arthritis, psoriatic arthritis, systemic lupus erythematosus (SLE), and viral infection (i.e., parvovirus B19), psoriatic arthritis, for example.

Distributions of disease may be described as symmetrical or asymmetrical, or may be small joint-predominant (MCPs, PIPs, DIPs) or large joint-predominant (knees, hips, shoulders, spine). Absence of disease in key joints may help to distinguish inflammatory joint diseases. For example, rheumatoid arthritis and osteoarthritis both involve joints of the upper and lower extremities. Rheumatoid arthritis tends to involve the MCPs and PIPs symmetrically while sparing the DIPs, while osteoarthritis involves the DIPs and can spare the MCPs. Involvement of the base of the thumb would be most typical of osteoarthritis.

Finally, an examination of the joints should include an assessment of the spine and chest. As with other joints, examination should begin with observation of overlying skin and any present deformities or swelling, and then proceed to palpation of the joint spaces of the cervical to sacral spine. A wide variety of specialized techniques exist that may be used to assess for presence of joint disease involving the spine, the details of which are beyond the scope of this chapter. However, a brief look at cervical neck range of motion and consideration of maneuvers such as the modified Schober test are useful in considering axial involvement of disease and can be successfully performed by the non-rheumatologist. The FABER (leg flexed, thigh abducted, externally rotated) test can also be helpful in eliciting hip, lumbar spine or sacroiliac pathology.

Other specialized assessments may be applied when specific conditions are uspected. For example, dactylitis of the fingers and toes is a common feature in psoriatic arthritis, and other peripheral / axial spondyloarthritides. A simple count of affected digits is relevant. For a more comprehensive entheseal evaluation, the Leeds dactylitis instrument can be utilized.

Muscle Examination

The muscular examination is comprised of both palpation and strength testing. Deep palpation of the muscle is unnecessary, in contrast to when joint palpation is performed. Tenderness upon palpation of muscle may suggestan underlying inflammatory myositis, such as dermatomyositis, polymyositis, or anti-synthetase syndrome.

Finally, a full-strength examination should be completed by assessing the force of each of the joint movements under resistance. Procedures for the strength examination vary with the muscle

group being assessed, but it should always be conducted with resistance. For example, abduction of the elbow may be assessed with the examiner applying an adducting force to the forearm. Similarly, extension at the knee may be assessed against force applied to the anterior shin. Of note, the finding of reduced strength on examination is a non-specific sign, and may point to disease processes at the level of the bone, muscle, joint, or the nervous supply.

Conclusion

This chapter has reviewed the fundamentals of the skin and musculoskeletal examinations relevant for overlap diseases. The astute clinician, attentive to the myriad of integumentary disorders that may afflict patients with these diseases, will have an opportunity to add specificity to the overall evaluation with the goal of early and accurate diagnosis.