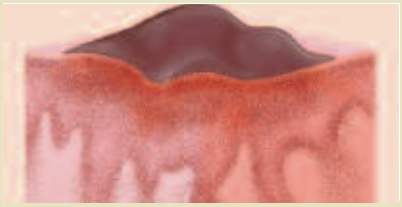
abstract



The skin can be a window to certain internal diseases. Notable internal diseases with a prominent skin component include systemic lupus erythematosus, dermatomyositis, scleroderma, psoriasis, and sarcoidosis. This article will review some of the common skin manifestations of these diseases.

Key words: *skin disease, internal disease, systemic lupus erythematosus, dermatomyositis, scleroderma, psoriasis, sarcoidosis*

Skin Manifestations of Internal Disease

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Introduction

It is a generally accepted concept that a patient's skin can show signs of internal disease.[1] In fact, clues on the skin can often be a key to diagnosing complex systemic illnesses.[2] The recognition of this fact may allow some diseases to be quickly diagnosed and treatment to be initiated early in the course of the illness.[2] Skin conditions are common and represent approximately seven percent of all office visits.[3] However, the sheer number of internal diseases with a skin component[1] precludes a systematic overview in this article. This article will focus on a few of the notable internal diseases with a prominent skin component, including systemic lupus erythematosus, dermatomyositis, scleroderma, psoriasis, and sarcoidosis.

Systemic Lupus Erythematosus

The American Rheumatology Association has suggested 11 criteria for the diagnosis of systemic lupus erythematosus (SLE).[2,4] Four of these criteria manifest on the skin. These include malar rash, photosensitivity, oral ulcers, and discoid lesions.[2,4] Acute cutaneous lupus erythematosus is represented by the classic malar butterfly rash. This lesion is acute, erythematous, and often edematous. It is situated on the face—overlying the cheekbones—and is often provoked by sunlight or artificial light. These lesions often worsen during flares of the disease.[5] More than 95% of patients with this rash will have a positive antinuclear antibody test.[2] In contrast, chronic discoid lupus lesions begin as erythematous plaques covered by scale with central scarring often occurring on the scalp, ear, face, or neck, but may be more generalized.[2,5] Only 10% of patients with discoid lupus meet the classic criteria for SLE.[2] An immunofluorescence study of involved skin shows deposition of immunoglobulin or complement at the basement membrane.[2]

Lupus can be associated with a number of underlying conditions in various organs of the body, including pericarditis, myocarditis, endocarditis, coronary arteritis, pleuritis, pleural effusions, pulmonary embolism, oral ulcers, esophageal motility disorders, serositis, pancreatitis, mesenteric artery occlusion, renal involvement, stroke, and transient ischemic attacks.[4] This list is by no means exhaustive but is included to illustrate the fact that lupus is, in reality, an internist's playground.

Dermatomyositis

Polymyositis and dermatomyositis are idiopathic inflammatory myopathies.[2,6] Dermatomyositis is suggested by proximal muscle weakness with elevations of serum muscle enzyme levels, as well as abnormal electromyographic and muscle histology findings.[2,6] Characteristic skin findings include a heliotrope rash—a purple to red patch around the eyes associated with some periorbital edema.[2,4] Also characteristic are Gottron's papules, which are flat-topped, violet-coloured papules found predominantly over the knuckles.[2,4]

Interestingly, dermatomyositis is twice as common in women. There are two distinct forms—a juvenile form and an adult form.[2] The adult form is associated with an increased incidence of internal malignancy.[7,8] The prevalence of malignancy in dermatomyositis has ranged from 3–60% in various series.[7,8] However, these results may be biased by increased cancer

surveillance in these patients.[8] The conditions are often diagnosed within two years of each other; either the malignancy or the dermatomyositis may come first.[7] Patients with newly diagnosed dermatomyositis should undergo a thorough physical examination and screening workup including urinalysis, liver chemistry profiles, and chest radiography, as well as age- and gender-appropriate cancer screening (e.g., sigmoidoscopy, fecal occult blood test, prostate-specific antigen test, mammography, PAP smear, or pelvic examination). In addition, some experts suggest that computed tomography of the chest, abdomen, and pelvis should also be considered.[7]

Scleroderma

Scleroderma, literally "hardening of the skin," can be seen in both systemic and cutaneous forms.[2] Systemic scleroderma can involve multiple organs, including the skin, blood vessels, synovium, skeletal muscle, gastrointestinal tract, heart, and especially the lungs and kidneys.[2,9] Patients initially complain of Raynaud's phenomenon: swelling and puffiness of the fingers or hands. The typical skin lesion follows these complaints by several months and is an ill-defined, indurated patch that often occurs peripherally and gradually involves the forearm.[2,9]

Gastrointestinal tract involvement occurs in the majority of patients with distal esophageal motor dysfunction, leading to dysphagia for solids and food sticking in the substernal region. Lung involvement also occurs in the majority of patients, and has recently surpassed renal disease as the leading cause of death in patients with systemic scleroderma.[2,9] Chest x-ray reveals bibasilar linear or nodular interstitial fibrosis. Pulmonary function testing reveals a restrictive lung disease.[9]

The kidneys are also commonly affected. The primary targets are the arcuate and interlobular arteries via severe constriction and mucoid intimal hyperplasia.[9] However, the use of angiotensin converting enzyme inhibitors has improved the course of scleroderma renal crisis, which had commonly been fatal.[2]

Psoriasis

Psoriasis is a relatively common dermatological condition, affecting one to two percent of the population.[2] The lesions of psoriasis are classical papulosquamous lesions that typically affect the elbows, buttocks, knees, and scalp. There are a variety of forms, including plaque-type, guttate, pustular, and erythrodermic. Erythrodermic psoriasis is a generalized exfoliating lesion that can be severe and lead to metabolic complications, such as hyperthermia, hypothermia, high-output heart failure, hypoalbuminemia, anemia, and dehydration.[2,10]

Importantly, erythrodermic psoriasis can be exacerbated by a variety of agents, such as the discontinuation of corticosteroids, infection, beta-blocking agents, lithium, antimalarials, or severe sun exposure.[2]

Psoriatic arthritis is an HLA-B27-associated, seronegative inflammatory arthropathy that can occur in up to six percent of patients with psoriasis.[2,11,12] Most patients (70%) have asymmetric oligoarthritis of the small joints of the hand.[2,11] An important systemic involvement includes ocular involvement, occurring in 30% of patients,[11] which takes the form of eye inflammation (conjunctivitis, iritis, episcleritis, and keratoconjunctivitis sicca).[11,13] Aortic insufficiency has also been reported in these patients.[11]

Sarcoidosis

Sarcoidosis is a multisystemic disorder of unknown origin characterized by noncaseating granulomas.[2,14] There are theories that sarcoidosis may represent an immune reaction to an unrecognized antigen, such as a mycobacterium.[2,15] Lungs, lymph nodes, skin, and eyes are the most common organs involved.[2] Skin lesions can be nonspecific, although erythema nodosum may be seen in up to 30% of patients. The Lofgren syndrome, which consists of erythema nodosum, bilateral hilar adenopathy, uveitis, and arthritis, is diagnostic for acute sarcoidosis and is associated with a good prognosis.[2,16]

Conclusion

This article has given a brief overview of some of the many internal diseases that can have a characteristic skin component.

No conflicts of interest declared.

References

1. Bolognia JL, Braverman IM. Skin Manifestations of Internal Disease. In: Kasper DL, Braunwald E, Fauci A, et al, editors. Harrison's Principles of Internal Medicine, 16th edition. New York, McGraw-Hill Companies, 2005:296–311.
2. Kirsner RS, Federman DG. Cutaneous clues to systemic disease. Telltale skin signs enable early diagnosis. Postgrad Med 1997;101:137–50.
3. Stern RS, Johnson ML, Delorizier J. Utilization of physician services for dermatological complaints: the United States, 1974. Arch Dermatol 1977;113:1062–6.
4. Tan EM, Cohen AS, Fries JF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1982;25:1271–7.
5. Asherson RA, Hughes GRV. SLE-clinical features. In: Primer on rheumatic diseases, 9th edition. Atlanta: The Arthritis Foundation, 1988:100–6.
6. Plotz PH, Dalakas M, Leff RL, et al. Current concepts in the idiopathic inflammatory myopathies: polymyositis, dermatomyositis, and related disorders. Ann Intern Med 1989;111:143–57.
7. Federman DG, Brescia G, Horne M, et al. Cutaneous manifestations of malignancy. Post Grad Med 2004; www.postgradmed.com/issues/2004/01_04/federman.htm.
8. Maoz CR, Langevitz P, Livneh A, et al. High incidence of malignancies in patients with dermatomyositis and polymyositis: an 11-year analysis. Semin Arthritis Rheum 1998;27:319–24.
9. Medsger TA. Systemic sclerosis and localized scleroderma. In: Primer on rheumatic diseases, 9th edition. Atlanta: The Arthritis Foundation, 1988:111–7.
10. Shuster S. Internal Disorders due to diseases of the skin. Manitoba Medical Review 1967;47:258–9.
11. Vasey FB. Psoriatic arthritis. In: Primer on rheumatic diseases, 9th edition. Atlanta: The Arthritis Foundation, 1988:151–2.
12. Baker H. Epidemiological aspects of psoriasis and arthritis. Br J Dermatol 1966;78:249–61.
13. Lambert JR, Wright V. Eye inflammation in psoriatic arthritis. Ann Rheum Dis 1976;35:354–6.
14. Spilberg I. Sarcoidosis. In: Primer on rheumatic diseases, 9th edition. Atlanta: The Arthritis Foundation, 1988:190–2.
15. Saboor SA, Johnson NM, Mcfadden J. Detection of mycobacterial DNA in sarcoidosis and tuberculosis with polymerase chain reaction. Lancet 1992;339:1012–5.
16. Kerdel FA, Moschella SL. Sarcoidosis: an updated review. J Am Acad Dermatol 1984;11:1–19.