Use Skin Changes to Identify Scleroderma Mimics

BY PATRICE WENDLING

CHICAGO — Few physicians would be fooled nowadays by gadolinium-induced nephrogenic systemic fibrosis, but there are other diseases that can masquerade as scleroderma.

The precise diagnosis of scleroderma-like illnesses is important because even though many of them are called scleroderma, they are different from systemic sclerosis in their treatments and outcomes, Dr. Virginia Steen said at a symposium sponsored by the American College of Rheumatology.

The diagnosis is most often based on the distribution and clinical characteristics of skin findings, as biopsies don't always differentiate types of scleroderma. She recommended watching for the following four conditions:

► **Lipodermatosclerosis** is one condition that physicians often fail to think of as a scleroderma mimic. Also known as hypodermatitis sclerodermaformis, it refers to localized chronic inflammation and fibrosis of the skin and subcutaneous tissues of the lower leg. In the acute stage, the leg is inflamed and warm, the skin is very tight, and cellulitis may be present. The ankle and toes are not involved.

In its chronic stage, there is induration, contraction of the skin and subcutaneous tissues, and irregular depressions that can look almost identical to lower-leg scleroderma, she said.

The leg eventually resembles an inverted champagne bottle, in which the upper half remains edematous and has a much greater circumference than does the lower sclerotic portion.

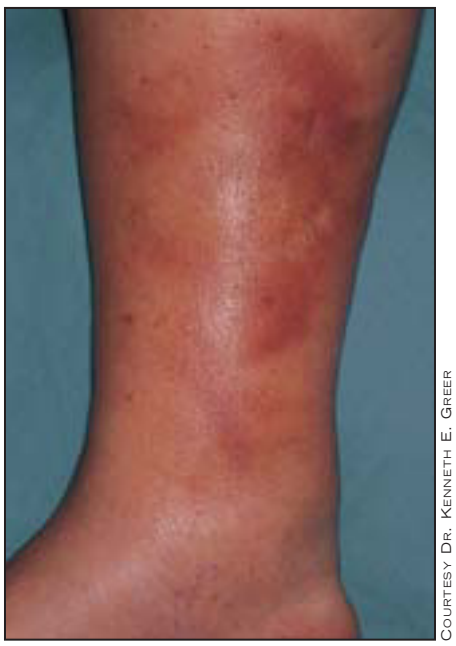
Lipodermatosclerosis is sign of severe end-stage venous insufficiency, and should be differentiated from scleroderma, cellulitis, superficial thrombophlebitis, and erythema nodosum. The diagnosis is made from clinical observation, but direct immunofluorescence of early and late lesions has been used to show dermal pericapillary fibrin deposits.

If left untreated, lipodermatosclerosis can progress to ulceration, atrophy blanche, or shortening of the Achilles tendon. Treatment involves weight loss, controlling the underlying disease, and emphasis on support stockings that may need to be specially made, said Dr. Steen, who is professor of medicine and director of the rheumatology fellowship program at Georgetown University in Washington.

Topical steroids are useful if the skin is inflamed, and antibiotics are recommended for cellulitis.

► **Scleredema** tends to target the upper body without affecting the lower extremities. The skin on the neck and face thicken and harden; severely affected patients are unable to wrinkle their foreheads or open their mouths. In most patients, the shawl sign is present, with skin involvement over the chest and arms, she said.

Pathologic features include swollen collagen with clear spaces and accumulation of hyaluronic acid and glycosaminoglycans. Although scleredema is commonly associated with diabetes, it can also occur after a viral illness.



COURTESY DR. KENNETH E. GREER

The sclerotic plaque on this patient's lower leg is lipodermatosclerosis.

► **Eosinophilic fasciitis** is a rare disorder characterized by symmetrical and painful inflammation and swelling of the extremities, leading to induration and the characteristic peau d'orange configuration. The palms may be involved, but typically the fingers and toes are spared. Contractures demonstrating the groove sign commonly evolve as a result of induration.

Eosinophilic fasciitis is slightly more common in middle-aged men, but can occur in women and children. It was initially distinguished from systemic sclerosis by the absence of Raynaud's phenomenon, autoantibodies, and visceral involvement, and—unlike systemic sclerosis—it responds to corticosteroids, Dr. Steen said.

Histologically, there are marked eosinophilia and inflammatory infiltrates in the fascia. The extent of the histologic changes depends on the stage of the disease, and thus is not a consistent component of the disease. Aside from marked peripheral eosinophilia, other laboratory features to watch for include an increased erythrocyte sedimentation rate, increased gamma globulin, and an increased aldolase, with a normal creatinine phosphokinase.

Aggressive physical therapy is a key component of treatment, because this and most scleroderma mimics discussed here can cause joint contractures. In eosinophilic fasciitis, low- to moderate-dose prednisone (20-30 mg)—and, if needed, methotrexate as a steroid-sparing agent—can be given, Dr. Steen said.

► **Diabetic cheiroarthropathy** is a syndrome of limited joint mobility in the hands. It is characterized by thickened, tight, waxy hands with sclerosis of the palmar tendon sheaths that noticeably restricts mobility in the proximal interphalangeal joints and metacarpophalangeal joints.

The "prayer sign," in which the patient will be unable to fully oppose the palmar surfaces of the digits, is a clue to this diagnosis when it is present in a patient with diabetes, said Dr. Steen, who disclosed no relevant conflicts of interest. ■

Periocular Skin Cancers Tend To Be Basal Cell Carcinoma

BY ALICIA AULT

AUSTIN, TEX. — A chart review aimed at quantifying the incidence and type of periocular skin cancers showed that the vast majority were basal cell carcinomas, and that there was a slight predominance of the cancers in men.

The study was undertaken because there has been an increase in eyelid malignancies that is thought to be due to a lack of protection from ultraviolet radiation, Dr. Jens Thiele said at the annual meeting of the American College of Mohs Surgery.

This is the largest U.S.-based study of periocular cancers ever conducted, said Dr. Thiele, a dermatologist in private practice in Birmingham, Ala. He and his colleagues reviewed charts at a single center from 553 consecutive Mohs surgery patients from January 2005 to September 2008.

All of the patients were white (Fitzpatrick skin types I, II, and III). There were 346 men and 207 women. Interestingly, 61% of the tumors were in men.

Of the tumors, there were 435 basal cell carcinomas (BCCs), 105 squamous cell carcinomas (SCCs), 10 melanomas, and one of each of the following: sebaceous carcinoma, trichoepithelioma, and dermatofibrosarcoma protuberans.

The investigators also quantified location and pre- and postoperative defect sizes. Most often, BCCs were located on the lower eyelid (246, or 57%). They were also found on the medial canthus (28%), upper eyelid (10%), and lateral canthus (6%).

The squamous cell tumors also were found most frequently on the lower eyelid (64 or 61%), followed by the medial canthus (17%), the upper eyelid (15%), and the lateral canthus (7%). Six of the 10 melanomas were also on the lower eyelid; 8 of the tumors were in females.

For BCCs, the pre- and postoperative sizes were smallest on the upper eyelids, while the largest tumors were found on the medial canthus. The mean number of Mohs layers needed for BCC clearance ranged from 1.33 in the lateral canthus to 1.42 in the medial canthus.

SCCs had larger pre-op and postop sizes, but the number of layers needed for clearance was lower. The mean number for SCC clearance was 1.5 in the medial canthus and 1.1 in the lateral canthus, Dr. Thiele said.

Dr. Thiele reported no conflicts. ■

Unexplained Erythema May Be Associated With Cancer

BY CHARLES BANKHEAD

PRAGUE — Unexplained erythema should raise suspicion about a possible underlying malignancy, according to a review of cases as one Asian institution.

In an effort to determine the clinical implications of idiopathic erythema, Dr. Steven Thng and colleagues at the National Skin Center in Singapore reviewed the records of patients evaluated for erythema from 2001 to 2005 and compared those patients with published cases series as well as with data from the Singapore Cancer Registry.

Dr. Thng and colleagues identified 218 patients evaluated for erythema during the study period—108 cases (50%) were classified as idiopathic. Among patients with an identified cause of erythema, preexisting dermatoses (30%) and drug reaction (15%) were the most common diagnoses.

On follow-up, the researchers found idiopathic erythema was associated with visceral malignancy in 18% of patients and with cutaneous T-cell lymphoma in 5%.

"We recommend close follow-up with re-evaluation for malignancy even if the initial investigation had been negative," they said in a poster at the International Congress of Dermatology.

Few investigators have attempted to examine the natural history and potential clinical consequences of unexplained erythema, the researchers wrote. Moreover, previous studies primarily involved white populations.

Most patients with idiopathic erythema were men (73%) and idiopathic erythema tended to occur at an older age (69 years) when compared with erythema of known cause (62 years). Patients with idiopathic erythema tended to experience a slow onset of disease and an average duration of 22 days. They also experienced more episodes of disease (average of 1.75 episodes), compared with patients who had erythema of known cause (average of 1.32 episodes).

When compared with age-standardized cases, patients with idiopathic erythema had more than a threefold greater risk of visceral malignancy. Dr. Thng and colleagues reported no disclosures. ■