A 62-year-old woman presented with a four-week history of edema in both of her upper extremities, as well as changes in skin pigmentation. She had alopecia involving the frontal area of the scalp. She had dyspnea on exertion, which had worsened over several weeks.

Physical examination showed firm skin with nonpitting edema in the forearms, hands, and fingers. She also had pigmentation changes on her face, neck, and upper back (Figures 1 through 3).

Laboratory results included the following: white blood cell count, 10,000 per mm$^3$ (10.0 × 10$^9$ per L); hemoglobin, 8.5 g per dL (85 g per L); erythrocyte sedimentation rate, 115 mm per hour (normal, 0 to 30 mm per hour); sodium, 137 mEq per L (137 mmol per L); potassium, 4.4 mEq per L (4.4 mmol per L); blood urea nitrogen, 23 mg per dL (8.2 mmol per L); creatinine, 1.4 mg per dL (124 µmol per L); urinalysis, normal; antinuclear antibody, positive (1:640) with a nucleolar pattern; and rheumatoid factor, 24.6 IU per mL (normal, 0 to 13.9 IU per mL). Chest radiography showed cardiomegaly with evidence of vascular congestion.

Question
Based on the patient’s history, physical examination, and laboratory findings, which one of the following is the most likely diagnosis?

- A. Dermatomyositis.
- B. Rheumatoid arthritis.
- C. Scleroderma.
- D. Systemic lupus erythematosus.
- E. Vitiligo with repigmentation.

See the following page for discussion.
Discussion

The correct answer is C: scleroderma. Non-pitting edema may appear months or years before the characteristic changes of scleroderma develop. These changes may include tightly bound skin, with ulcer formation, tightness of the facial skin with fine periorial wrinkling, and an inability to fully open the mouth. The skin changes tend to affect fingers and hands initially, with gradual proximal spread to the forearms. The lower extremities tend to be relatively spared.

“Salt-and-pepper” pigment changes are characteristic of scleroderma with perifollicular sparing of pigment. This pattern is most common on the scalp, upper back, and chest. Dermal sclerosis may obliterate hair follicles, resulting in hair loss. Pulmonary involvement may include interstitial lung disease and pulmonary artery hypertension. Nucleolar pattern antinuclear antibodies are associated with antiribonucleoprotein, which is characteristic of scleroderma and rare in other immune disorders.

Dermatomyositis often presents as proximal muscle weakness and a heliotrope rash, commonly involving the eyelids. Rheumatoid arthritis is typically associated with symmetrical joint involvement, morning stiffness, synovial thickening, and a family history of the disease.

Systemic lupus erythematosus can manifest in a variety of ways. Dermatologic manifestations include nonpruritic, erythematos to violaceous plaques, often on sun-exposed areas. Telangiectasias, alopecia, urticaria, and Raynaud phenomenon are also common.

Vitiligo with repigmentation can appear similarly to this patient’s presentation, with areas of repigmentation beginning in the perifollicular area. Although vitiligo has been associated with several other conditions (e.g., hypothyroidism, Graves disease, Addison disease, pernicious anemia, diabetes mellitus), most patients with vitiligo have no other associated findings.

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REFERENCES