Answer to Dermacase *continued from page 353*

1. Dermatomyositis

Laboratory tests revealed the following results: hemoglobin, 117 g/L; creatine phosphokinase, 10 173 U/L; lactate dehydrogenase, 1719 U/L; aspartate transaminase, 428 U/L; and alanine transaminase, 389 U/L. Electromyogram of the deltoid muscle revealed a myopathic pattern. Endoscopy revealed an infiltrative ulcerated gastric lesion; histology of the lesion confirmed diagnosis of gastric adenocarcinoma. Deltoid muscle biopsy showed vacuolar degeneration of the basement membrane, with lymphohistiocytic infiltration of the dermis, striated atrophy of the muscle layer, and partial muscle fibre atrophy. Computed tomography of the thoracolumbar fascia revealed 2 metastatic lesions to the liver. The patient started treatment with 1 mg/kg of prednisone daily, which improved his skin and muscle symptoms. He died 6 months later.

Polymyositis (PM) and dermatomyositis (DM) are uncommon systemic rheumatic diseases characterized by inflammatory and degenerative changes, either in the muscles (PM) or in the skin and muscles (DM). The female-to-male incidence ratio is 2:1. These diseases can appear at any age but occur most commonly in those between 40 and 60 years or in children aged 5 to 15 years.[1] Etiology of DM is unknown; however, some studies have reported associations with histocompatibility antigens, environmental agents (eg, virus, drugs), and autoimmunity.[2]

In up to 40% of patients, dermatologic symptoms can be the sole manifestation of DM at onset. Muscle degeneration can occur simultaneously or precede or follow the dermatologic symptoms by weeks to years.[3] The characteristic and possibly pathognomonic cutaneous features of DM are heliotrope rash and Gottron papules. Other cutaneous features (although not pathognomonic) include malar erythema, poikiloderma in a photosensitive distribution, violaceous erythema on the extensor surfaces, and periungual and cuticular changes. Onset of PM might be acute or insidious, particularly in adults. The myopathy produces symmetric weakness of the limb-girdle muscles and anterior neck flexors with or without muscle tenderness. This weakness progresses over weeks to months with variable involvement of the pharynx or upper esophagus, manifesting as dysphonia or dysphagia. Other possible symptoms include respiratory muscle weakness, visual changes, and abdominal pain.

Diagnosis

Dermatomyositis should be suspected in patients with heliotropic rash or Gottron papules (even without PM), and in those with PM and skin findings compatible with DM.

Diagnosis requires as many as possible of the following[4]: progressive symmetric muscle weakness of the inferior and superior extremities of the girdle, with or without dysphagia or with respiratory muscle involvement; muscle biopsy confirming myositis; increase in muscle enzyme serum levels; electromyographic abnormalities indicating primary muscle damage; and characteristic skin lesions.

Paraneoplastic dermatomyositis

The association between DM (and possibly PM) and cancer has long been recognized.[3,5] The most common malignancies are ovarian and gastric cancers and lymphoma; others include lung, male genital organ, and nonmelanoma skin cancers, Kaposi sarcoma, mycosis fungoides, and melanoma.[6,7] Skin changes do not differ between patients with or without malignancies; careful investigation for malignancy should be initiated at the time DM is diagnosed.

Screening should entail a physical examination that includes breast, pelvis, and rectum (with fecal occult blood testing); complete blood count; biochemical profile; mammogram; carcinoembryonic antigen check; urinalysis; chest x-ray scan; and other appropriate tests based on the patient's symptoms. Some authorities recommend computed tomography of the chest, abdomen, and pelvis.[7] Younger patients without symptoms of cancer do not need to undergo screening. As the skin condition might precede or follow the development of myositis by weeks to years, careful follow-up should be ensured.

Treatment

Corticosteroids are the best choice initially.[8] Adults with acute disease should take 0.5 to 1 mg/kg/d of prednisone. Once enzyme levels return to normal, prednisone can gradually be reduced. If muscle-enzyme levels rise, the dose is increased. If a patient does not respond to corticosteroids or depends on a high dose, immunosuppressors such as methotrexate, azathioprine, cyclosporine, and intravenous immunoglobulin should be considered. Paraneoplastic DM is usually more refractory to corticosteroids. This will, however, subside if the tumour is removed. Other possible emerging therapies include anti-tumour necrosis factor agents and rituximab.[9,10]

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Competing interests
None declared

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