VA to Treat Any Veteran With ALS, Regardless

Presumption of service connection means vets won't have to waste time collecting claims evidence.

BY MARY ELLEN SCHNEIDER
New York Bureau

Veterans with amyotrophic lateral sclerosis can now receive services and equipment from the Department of Veterans Affairs without having to prove that their condition is connected to their military service.

The VA established a presumption of service connection for amyotrophic lateral sclerosis (ALS) for any veteran who develops the disease at any time after their separation from service. Veterans must have 90 days or more of continuous active service in the military to qualify for the benefits, according to an interim final regulation issued on Sept. 23.

Researchers have been studying the link between military service and ALS for years, and the evidence has been suggestive of an association but a definitive link has not been made. In 2001, the VA issued a policy giving "special consideration" to ALS claims made by veterans of the 1991 Gulf War, regardless of when the disease manifested itself.

VA Secretary James B. Peake said he based the decision to presumptively connect ALS to military service primarily on a November 2006 report from the Institute of Medicine that concluded that "there is limited and suggestive evidence of an association between military service and later development of ALS." The IOM conclusion was based on a review of available evidence. The IOM committee that issued the study recommended further research to establish a definitive association, but Dr. Peake said he thought it was unlikely that further research would show the link given the rarity of the disease.

"There simply isn't time to develop the evidence needed to support compensation claims before many veterans become seriously ill," Dr. Peake said in a statement. "My decision will make those claims much easier to process, and for them and their families to receive the compensation they have earned through their service to our nation."

The new regulation will apply to all applications for benefits received by VA on or after Sept. 23, as well as those currently pending before the VA, the U.S. Court of Appeals for Veterans Claims, and the U.S. Court of Appeals for the Federal Circuit. VA officials are also planning to contact veterans with ALS whose claims were previously denied.

For neurologists working outside of the VA who are seeing veterans with ALS, its critical to get patients connected with a VA physician, said Dr. Carlayne E. Jackson, professor of neurology and otolaryngology at the University of Texas at San Antonio, and director of the South Texas ALS Clinic at the University Health Science Center. Once patients are connected to the VA system, a physician there will be able to help them get coverage for necessary equipment and services, she said.

"It's an opportunity to dramatically improve the services and equipment that our patients have access to," Dr. Jackson said.

For example, veterans will have greater access to wheelchairs, speech devices, specialized mattresses, and home health care services that may not have been covered or were only partially covered through Medicare disability benefits, she said.

The new designation by the VA will also ease the burden on veterans who previously had to prove that their disease was connected to their military service, said Pat Wildman, director of public policy for the ALS Association. Because of the progressive nature of the disease, some veterans were dying before they had the chance to prove their service connection, he said.

At press time, the ALS Association was developing frequently asked questions and answers about the VA benefits that will be posted on the organization's Web site (www.alsa.org).

It is estimated that ALS affects about 20,000-30,000 individuals in the United States. Though the number of veterans with the condition is unknown, recent studies indicate that the disease strikes military veterans at about double the rate of the general population, according to the ALS Association.

But researchers and patients may get more information about the disease prevalence thanks to the passage of the ALS Registry Act (S. 1382). The legislation passed Congress in late September and was signed into law by President Bush.

The bill will establish the first national patient registry of individuals with ALS. The registry, which will be run by the Centers for Disease Control and Prevention, could yield clues about the cause, treatment, and cure for the disease, including why vets appear to be at greater risk. ■

Skin Involvement Is Key in Dermatomyositis Diagnosis

BY DAMIAN McNAMARA
Miami Bureau

MONTREAL — The cutaneous features of dermatomyositis often hold the key to making its definitive diagnosis, without having to resort to a muscle biopsy.

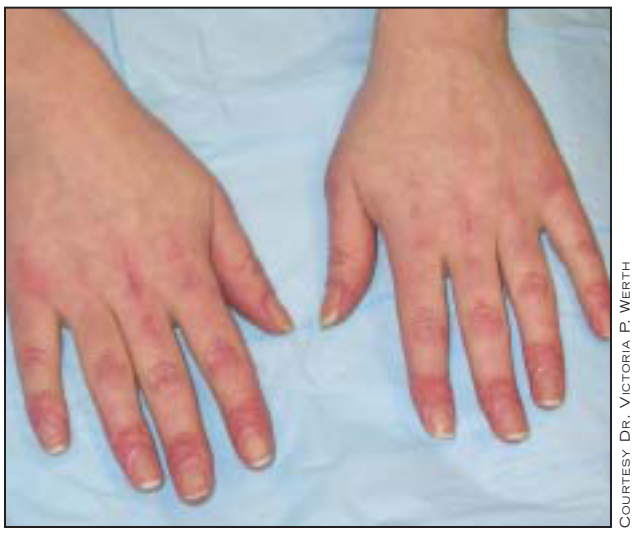
So key are the cutaneous manifestations that dermatologists can be a useful resource for rheumatologists faced with a possibly affected patient. "We play a critical role in diagnosing dermatomyositis—we know the visual diagnosis," Dr. Victoria P. Werth said. The incidence is increasing and the early presentation is often subtle. "We are seeing more of these patients, and they are confusing our rheumatology colleagues."

Patients can present differently to the dermatology and rheumatology departments at the University of Pennsylvania, Philadelphia, according to a study by Dr. Werth and colleagues (J. Am. Acad. Dermatol. 2007;57:937-43). The degree of skin and/or muscle involvement was the primary distinction among 131 patients who consulted dermatology, 58 who saw rheumatologists, and 13 seen by both specialties.

A correct diagnosis is crucial because dermatomyositis can affect many organ systems beyond the skin, including lung, heart, and muscle. In addition, patients need to be monitored regularly over the long term for malignancies (Ann. Intern. Med. 2001;134:1087-95). Researchers found a relative risk of 2.4 for malignancy among dermatomyositis patients, compared with those who had polymyositis in this retrospective study of 537 patients with biopsy-proven myopathy.

"The adjusted relative risk for malignancy was higher in the first 3 years after diagnosis with dermatomyositis, including for lung, ovary, pancreatic, and other cancers," Dr. Werth said at the annual conference of the Canadian Dermatology Association. "Risk remains high for years." She recommended evaluation at least annually.

Cutaneous features include scale, pruritus, and Gottron's papules. A violaceous erythema in a photodistribution pattern or confluent erythema is possible. Also look for Gottron's sign, a symmetrical, macular erythema with or without edema located on the dorsal hand, interphalangeal joints, elbow, and/or medial ankle. Subepidermal vesicles or blisters, vasculopathy, and poikiloderma are among the secondary skin features.



COURTESY DR. VICTORIA P. WERTH

Cutaneous features such as violaceous erythema and Gottron's papules can hold the key to diagnosing dermatomyositis.

A meeting attendee commented that many patients have extensive scalp pruritus. "Scalp dermatomyositis is very resistant [to treatment]. I have patients where everything else gets better," Dr. Werth said. "Pruritus is a very big problem. And some patients with only scalp involvement early on get misdiagnosed as seborrheic dermatitis," added Dr. Werth, who is on the dermatology and medicine faculties at the university.

Dermatomyositis patients often get nasolabial fold involvement, a distinction from systemic lupus erythematous patients. In addition, "nail fold findings are critical," she said. "It is very important to look at the hands of all our dermatomyositis patients." Periungual infarcts and telangiectasias are among the characteristic signs.

A definitive dermatomyositis diagnosis includes the typical skin rash and at least three of the following criteria: an abnormal muscle biopsy, abnormal electromyogram (EMG), elevated skeletal muscle–derived enzymes, or symmetric proximal weakness with or without dysphagia or respiratory muscle involvement.

Physicians often ask Dr. Werth if a muscle biopsy is required. "By and large, we can make our diagnosis with a skin biopsy," she said. Skin findings and an abnormal creatine kinase, EMG, or MRI result are generally sufficient. Make sure to document the muscle disease if a patient is symptomatic, she added. Another attendee asked how to determine if muscle weakness is caused by the disease or by steroid treatment. "You sometimes have to do an MRI," Dr. Werth replied. "You can also taper the steroid and see if they get better. EMGs can also be helpful. If the patient lets you do a [skin] biopsy, you at least know what you are dealing with," Dr. Werth said. "We had one patient who we thought had psoriasis—but it was dermatomyositis that had targeted his hand joints."

Systemic involvement can be quite extensive, Dr. Werth said. Interstitial lung disease, for example, is present in about a quarter of patients. "Cardiac involvement is less common, but still worth looking for," said Dr. Werth, who is a consultant for Centocor Inc., Aegis Pharmaceutical Ltd., Celgene Corp., Kemia Inc., and Teva Pharmaceutical Industries Ltd.

Topical antipruritic agents and/or corticosteroids are often prescribed as first-line agents for localized skin dermatomyositis, whereas systemic therapy is typically reserved for more widespread disease, according to a review article by Dr. Werth and Rhonda D. Quain, who is now a resident at Mount Sinai School of Medicine, New York (Am. J. Clin. Dermatol. 2006;7:341-51). Antimalarial agents are often used to treat significant skin disease, as are other anti-inflammatory agents, systemic corticosteroids, corticosteroid-sparing immunosuppressants, and more recently, biologics and intravenous immunoglobulin (IVIG).

Dr. Werth cautioned about IVIG in dermatomyositis, however. She cited the case of a hyperviscous dermatomyositis patient who experienced a stroke while on IVIG therapy. ■

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