Actas Dermosifiliogr. 2014;105(7):675–682

ACTAS Dermo-Sifiliográficas

Full English text available at www.actasdermo.org

ORIGINAL ARTICLE

Paraneoplastic Dermatomyositis: A Study of 12 Cases☆

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Received 11 July 2013; accepted 24 November 2013
Available online 24 July 2014

KEYWORDS
Amyopathic dermatomyositis;
Paraneoplastic;
Cancer;
Necrosis;
Heliotrope rash

Abstract
Introduction and objectives: Adult dermatomyositis presents as a paraneoplastic syndrome in up to 25% of cases, but no clinical, histologic, or laboratory markers completely specific for paraneoplastic disease in dermatomyositis have been identified to date. Furthermore, studies on adult dermatomyositis do not usually report the frequency of cutaneous features of dermatomyositis in patients with associated cancer. Our aim was to review the characteristics of paraneoplastic dermatomyositis in patients seen at our hospital.
Material and methods: We studied 12 cases of paraneoplastic dermatomyositis and recorded patient age and sex, associated cancer, time between onset of dermatomyositis and cancer, emergent cutaneous manifestations, muscle involvement, dysphagia, lung disease, and levels of creatine phosphokinase and circulating autoantibodies.
Results: The mean age of the patients was 61 years and the 2 most common malignancies were ovarian cancer and bladder cancer. The mean time between the diagnosis of cancer and dermatomyositis was 7 months and in most cases, the cancer was diagnosed first. Seven patients had amyopathic dermatomyositis. The most common cutaneous signs were a violaceous photodistributed rash sparing the interscapular area and a heliotrope rash, followed by Gottron papules and cuticle involvement. Superficial cutaneous necrosis was observed in 3 cases. Myositis-specific autoantibodies were not detected in any of the 6 patients who underwent this test.
Conclusions: Paraneoplastic dermatomyositis is often amyopathic. There are no specific cutaneous markers for malignancy in dermatomyositis. Myositis-specific antibodies are not associated with paraneoplastic dermatomyositis.

☆ Please cite this article as: Requena C, Alfaro A, Traves V, Nagore E, Llombart B, Serra C, et al. Dermatomiositis paraneoplásica: estudio de 12 casos. Actas Dermosifiliogr. 2014;105:675–682.

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1578-2190/$ - see front matter

PALABRAS CLAVE
Dermatomiositis amiopática;
Paraneoplásica;
Cáncer;
Necrosis;
Rash heliotropo

Dermatomiositis paraneoplásica: estudio de 12 casos

Resumen

Introducción y objetivos: La dermatomiositis del adulto es paraneoplásica hasta en una cuarta parte de los casos. Hasta la fecha no existe ningún rasgo clínico, histológico o analítico absolutamente específico de paraneoplasia en dermatomiositis. Además, los estudios sobre dermatomiositis del adulto no suelen hacer referencia a la frecuencia de aparición de los distintos signos cutáneos propios de la dermatomiositis en los casos asociados a cáncer. Por todo ello decidimos revisar nuestros casos de dermatomiositis paraneoplásicas.

Material y métodos: Estudiamos 12 casos de dermatomiositis paraneoplásicas en los cuales se recogió la edad, el sexo, el cáncer asociado, el tiempo entre el inicio de la dermatomiositis y el cáncer, 9 signos cutáneos, afectación muscular, disfagia, enfermedad pulmonar, niveles de creatinfosfocinasa y de autoanticuerpos circulantes.

Resultados: La media de edad fue de 61 años y los 2 cánceres asociados más frecuentes fueron el de ovario y el de vejiga. El tiempo medio transcurrido entre el diagnóstico del cáncer y el de la dermatomiositis fue de 7 meses, y en la mayoría el diagnóstico de cáncer precedió al de dermatomiositis. Siete pacientes tuvieron dermatomiositis amiopática. De los signos cutáneos, los más frecuentes fueron una erupción violácea fotodistribuida que respetaba la región interescapular y el *rash* heliotropo, seguidos de las pápulas de Gottron y la afectación de cutículas. Se encontró necrosis cutánea superficial en 3 casos. Los autoanticuerpos específicos de miositis resultaron negativos en los 6 casos estudiados.

Conclusiones: La dermatomiositis paraneoplásica es muchas veces amiopática. No existe ningún rasgo cutáneo específico de paraneoplasia en la dermatomiositis. Los anticuerpos específicos de miositis no se asocian a la dermatomiositis paraneoplásica.

Dermatomyositis, a rare inflammatory disease possibly of autoimmune origin, produces a characteristic skin rash and symmetrical proximal myopathy. Amyopathic dermatomyositis is a variant in which muscle involvement is absent or is very mild. The incidence of dermatomyositis shows a bimodal distribution, with 2 forms of presentation according to the age at onset of the disease and called respectively juvenile dermatomyositis and adult dermatomyositis. Juvenile dermatomyositis is not typically a paraneoplastic disease, and very few cases of juvenile dermatomyositis associated with cancer have been published.[1] In fact, the frequency is so low that the association between juvenile dermatomyositis and cancer is considered anecdotal. In contrast, adult dermatomyositis is a paraneoplastic condition in 15% to 25% of cases.[2–5] The pathogenic relationship between dermatomyositis and cancer is not fully understood.[6] It would appear that the regenerating cells that appear in muscles with myositis express high levels of the specific antigens of myositis,[7] and that these are the same as those expressed in various cancers associated with inflammatory myopathies. The link between cancer and dermatomyositis would thus appear to be the expression of antigens common to the cancer and to muscle tissue in some patients with dermatomyositis.

The first problem that we encountered on reviewing the literature on paraneoplastic dermatomyositis was that most of the studies published until relatively recent indistinctly included dermatomyositis and polymyositis, giving the risks of cancer for the two diseases together. However, it is ever more widely accepted that dermatomyositis and polymyositis are distinct entities that show differences not only in their pathogenic mechanisms[8] but also in their epidemiology and risk of associated cancer.[2] The results of many of those studies must therefore be interpreted with caution. Furthermore, the majority of the largest published series of paraneoplastic dermatomyositis were population-based series and did not accurately define the cutaneous manifestations.[2,5,9,10] In view of this situation, in the present study we have reviewed those patients with paraneoplastic dermatomyositis diagnosed in our hospital, paying special attention to the description of the dermatologic manifestations.

Materials and Methods

We performed a retrospective observational study of all patients diagnosed with paraneoplastic dermatomyositis in our hospital between January 1994 and January 2013. A total of 12 cases were included. The data sources for all the parameters studied were the medical histories of the patients, the biopsy library of the pathology department, and the image archive of our own department.

The following inclusion criteria were applied: presence of a skin rash that was clinically and histopathologically compatible with dermatomyositis, with or without the presence of a symmetrical proximal myopathy (defined as proximal weakness with or without elevation of the creatine kinase [CK]); and concomitant cancer (excluding basal cell carcinoma and cutaneous squamous cell carcinoma) first diagnosed or presenting relapse within a maximum of 2 years before the onset of the dermatomyositis. The characteristic skin rash was considered to be a violaceous rash in sun-exposed areas and/or a heliotrope rash and/or