Cutaneous Manifestations of Chronic Graft-versus-Host Disease

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ABSTRACT
Cutaneous chronic graft versus host disease has traditionally been classified into lichenoid and scleroderma-like forms. However, the initial presentation is sometimes subtle and a variety of less common cutaneous manifestation may be prevalent. This clinical review focuses on the lesional morphology of chronic graft versus host disease, and presents a classification system that may prove useful in early diagnosis. In addition, this approach may help to facilitate the correlation of different morphologic entities with outcome and response to therapy.

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KEY WORDS
Stem cell transplant • GVHD • Dermatology

INTRODUCTION
Hematopoietic stem cell transplantation (HSCT) using peripheral blood, cord blood, or bone marrow is used to treat a wide variety of genetic and immunologic disorders and hematologic and solid organ malignancies. Significant advances in technology and immunology have improved the prognosis, rate of engraftment and quality of life for many stem cell recipients. Chronic graft-versus-host disease (cGVHD) is a multisystem disease often presenting with prominent skin involvement.

Traditionally, two types of cutaneous GVHD have been described: the acute form that occurs <100 days after transplantation and the chronic form that occurs >100 days. These rigid time-related criteria should serve only as a guide, especially as HSCT practices change. Since the availability of new, less intensive preparative regimens and the use of donor lymphocyte infusions, it is not uncommon to see a delayed or late acute GVHD (aGVHD; ie, >100 days after transplantation) and, in some cases, overlapping aGVHD and cGVHD, or other “atypical” forms of cGVHD. Acute GVHD and cGVHD in the skin are more accurately diagnosed by clinical and, less frequently, histopathologic features [1]. Based on similarities with other dermatologic diseases, we describe and summarize the diversity of cutaneous manifestations of cGVHD in Table 1. In our patients, the diagnosis of cutaneous GVHD was confirmed by a combination of elements including the clinical course and the presence of GVHD in other organ systems. All of our patients also had pathologic confirmation. By specifically describing lesional morphology, we hope to achieve a common, systematic, and consistent terminology for cutaneous cGVHD. This approach may also facilitate the correlation of different morphologic entities with outcome and response to therapy.

CLINICAL DESCRIPTION
Early on, the skin lesions of cGHVD are often subtle and the progression is insidious, characterized by the development of marked xerosis (skin dryness), follicular prominence (Figure 1), and ichthyosis (fish scale-like skin) (Figure 2). Papulosquamous lesions may simulate the appearance of keratosis pilaris (Figure 3) or present with pityriasisform (annular plaques demonstrating a branny scale), eczematous, or psoriasiform plaques (Figure 4).