Lupus Pernio or Chilblain Lupus? : Two Different Entities

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Lupus Pernio or Chilblain Lupus?

Two Different Entities

To the Editor:

In a recent issue of CHEST (February 2009), an interesting article was published about the treatment of lupus pernio in 54 patients. We would like to draw attention to the possible confusion that may exist, from a clinical point of view, between this type of patients. We would like to draw attention to the possible confusion between this type of patients and other skin lesions of discoid lupus erythematosus, defined by Hutchinson in 1888.

Like lupus pernio, CL is characterized by erythematous-purple plaques located in acral areas (most often, the nose and ears) but that are induced by exposure to cold or a drop in temperature (Fig 1), unlike other lesions of lupus erythematosus, which worsen with sun exposure, and sarcoidosis lesions, which remain unaltered with changes in temperature.

CL can cause mild pain or itching, and can be associated with hyperhidrosis. During the evolution of the disease, the presence of atrophic scarring and residual pigmentation are common findings. Like others forms of lupus lesions, this is more common in women, and although it is sporadic, two families with autosomal-dominant inherited CL have been reported.

The main difference with the cutaneous variety of sarcoidosis is the histopathologic study findings. In patients with lupus pernio, we observe granulomas without caseous necrosis but with few inflammatory infiltrates on the periphery; however, in patients with CL, epidermal atrophy, the degeneration of the basal layer, perivascular inflammatory infiltrates are found, and other uncommon findings, such as dyskeratosis, increased mucin in the dermis, and the presence of granular deposits of IgG and complement in basement membrane, have been described.

To establish a proper diagnosis, Su et al suggested using the Mayo Clinic diagnostic criteria. These comprise two major criteria (skin lesions in acral locations induced by exposure to cold or a drop in temperature and evidence of lupus erythematosus in the skin lesions, as determined by histopathologic examination or indirect immunofluorescence study) and four minor criteria (coexistence of systemic lupus erythematosus or other skin lesion of discoid lupus erythematosus, response to anti-lupus therapy, and negative results of cryoglobulin and cold agglutinin studies). Both major criteria and one minor criterion need to be present to diagnose CL.

Recently, a treatment review of CL was conducted that stressed the importance of protection from cold by physical measures as a trigger factor and the use of topical or oral antibiotics if the lesions are infected. Therapy with topical corticosteroids or calcium channel blockers get a good response from patients. In patients with severe cases, systemic corticosteroids and mycophenolate may be used. However, according to these authors, therapy with antimalarial agents has a minor effect. Conversely, therapy with infliximab appears to be superior to therapy with systemic corticosteroids, with or without additional agents, for the treatment of lupus pernio. Instead, infliximab does not seem useful in the treatment of CL; even a case of CL induced by this drug has been described. In any case, it is essential to make a correct diagnosis; although clinically the two entities are similar and have in common their association with systemic disease (lupus erythematosus or sarcoidosis), the prognosis and treatment differ considerably.

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Response

To the Editor:

We appreciate the comments of Arias-Santiago and colleagues on our recently published article in CHEST (February 2009).1 We agree that there is confusion regarding various skin conditions that include the term “lupus” in their nomenclature. “Lupus pernio” (a form of cutaneous sarcoidosis), “lupus vulgaris” (a form of cutaneous tuberculosis), “lupus miliaris disseminata faciei” (a form of rosacea), and “chilblain lupus” are but a few examples.

Specifically, the authors expressed concern that chilblain lupus could be confused with lupus pernio. Although this is an obvious problem in terms of nomenclature, we feel that in clinical practice these entities are distinct and are unlikely to be confused for many of the reasons cited by Arias-Santiago and coauthors. Chilblain lupus is usually associated with systemic lupus erythematosus. It is extremely rare, with only 70 cases reported in a 2008 review.2 It involves primarily the toes and fingers; involvement of the ears or nose is rare.2 The lesions generally occur first during cold or damp periods. They are usually pruritic and later painful; such symptoms are extremely unusual with lupus pernio. Pathologically, chilblain lupus reveals vascular thrombosis and not granulomatous inflammation.2,3 Furthermore, although both systemic lupus erythematosus and sarcoidosis are systemic diseases, we disagree with Arias-Santiago and coauthors that they are difficult to distinguish clinically.

The problem with this nomenclature most probably stems from the word “pernio,” which refers to a localized inflammatory lesion of the skin resulting from an abnormal response to cold.4 Lupus pernio skin lesions have no relationship to cold exposure, but it is this unfortunate antiquated description that we believe has led to the confusion. It is probably more appropriate to refer to lupus pernio as “disfiguring facial sarcoidosis.” Nonetheless, we disagree that this confusion in nomenclature may lead to problems in distinguishing these entities clinically.

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The American College of Chest Physicians Evidence-Based Educational Guidelines for Continuing Medical Education Interventions

Estimating Effect Size

To the Editor:

We appreciate the commentary of Norman1 regarding the methodology of our evidence-based review of the effectiveness of continuing medical education (CME), and agree with him that reporting effect size can be extremely useful in reporting results.2 Synthesizing the results of educational interventions represents one of the methodological challenges to performing systematic reviews in health care.3 The studies in our review differed in many important ways (used nonstandardized definitions of CME and targeted multiple types of objectives across vastly different audiences and content areas), and often were flawed in the metrics they used and in how those metrics were reported.4 These limitations in the primary literature led to a qualitative synthesis of the evidence, as an aggregate estimate of effect size could have implied greater confidence in the results than would have been appropriate.

In conclusion, we strongly lend our voice to the importance of estimating effect size in systematic reviews of educational interventions and recommend that original studies of CME give more attention to using valid measures of effectiveness that would allow such estimates.

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