Successful Treatment of Rosai–Dorfman Disease with Low-Dose Oral Thalidomide

Rosai–Dorfman disease (RDD) is a rare idiopathic histiocytic proliferative disorder.1 The skin is the most frequently involved extranodal organ. Nevertheless, primary cutaneous manifestation of RDD (CRDD) without systemic involvement is rare.2 We present herein a case of CRDD showing excellent remission after low-dose oral thalidomide therapy.

Report of a Case | A 43-year-old man (height, 178 cm; weight, 80 kg) was referred to our department for evaluation of a 12-month history of painless, enlarging papules and crusts involving the facial and pectoral areas and upper extremities. The papules were moderately tender and occasionally pruritic (Figure 1A). Findings of physical examination, comprehensive laboratory tests, and whole-body radiography were all within normal limits. Two skin biopsy specimens were taken, one from the right mandibular region and the other from the right upper arm. Histopathologic examination revealed that intact lymphocytes, plasma cells, and even neutrophils were readily found within the cytoplasm of histiocytes (emperiplois). Immunohistochemically, the histiocytes were positive for S-100, CD68, and CD20 and negative for CD1a, CK, and CD21. Periodic acid–Schiff and Giemsa staining results were negative. Based on the clinical and histopathologic findings, a diagnosis of CRDD was made.

The patient had been treated with 36 mg/d of methylprednisone for 3 months and received 10 fractionated local radiation doses of 2 Gray with a total dose of 20 Gray to the face. One lesion in the neck was intralesionally injected once with combination betamethasone dipropionate/betamethasone disodium phosphate (Diprospan; Merck Sharp & Dohme [Malaysia] Sdn Bhd).3,4 All treatments failed to demonstrate clinical efficacy. After providing his informed consent, the patient was given oral thalidomide, 50 mg/d for 2 weeks with slight improvement and no significant adverse effects. Therefore, the dose of thalidomide was increased to 100 mg/d. After 8 months of treatment, the nodules on his face and limbs had nearly re-

dermatitis, and vitiligo do not retain gentian violet. Thus, in vivo Gram staining has potential to be a rapid diagnostic test, especially given the prevalence of tinea versicolor in the developing world, as well as a potential therapeutic agent. Further studies of the antifungal activity of gentian violet are indicated.

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A. The patient had a history of tinea versicolor but was uncertain if he had been successfully treated. B. To determine whether he still was colonized with tinea versicolor, topical gentian violet (crystal violet) was applied to his back; note the accentuation of infected patches by gentian violet. C. Confirmation of infection was noted by positive potassium hydroxide preparation, revealing classic “spaghetti and meatball” conformation of fungus (original magnification ×100).
solved, and most of the erythematous plaques had lightened and partially cleared (Figure 1B). At 2-year follow-up, the lesions had not recurred.

Discussion | Various treatment options, including administration of high-dose thalidomide, steroids, chemotherapy, or surgical excision, have been reported with variable success. 3-8 Prior to thalidomide treatment, our patient received systemic steroid treatment, local radiotherapy, and an intralesional injection of steroid in a nodule without clinical improvement. Instead, the lesions progressively enlarged. Our treatment with oral low-dose thalidomide for this benign proliferative disorder resulted in excellent clinical outcome. The most challenging adverse effect of thalidomide is neuropathy, which might be persistent at a cumulative dose of over 20 g. 9 Although the cumulative dose of thalidomide was greater than 20 g in our patient, there were no signs of peripheral neuropathy or other adverse effects.

Both successful and failed treatments of RDD with oral thalidomide have been reported. 5,6,10 suggesting that there may exist different genetic and etiologic factors in the disease. Those patients with RDD who responded to thalidomide were given doses ranging from 200 to 300 mg/d, and amenorrhea occurred after 3 weeks of treatment (cumulative dose, approximately 6 g). 5,10 The present case demonstrates the effectiveness of low-dose oral thalidomide in treatment of CRDD.

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A Dreadful Infestation

Pediculus humanus capitus, the head louse, is a blood-sucking, obligate parasite that can lead to significant infestation and both direct and indirect medical costs. Treatment can become complicated with outbreaks and increasing resistance.

Report of a Case | A 52-year-old, wheelchair-bound, paraplegic man with a history of pemphigus foliaceus well controlled by azathioprine presented for scalp pruritus. The patient had extensive dreadlocks, which he had grown for decades, since the accident that had paralyzed him (Figure, A). On examination, he also had adherent white nits along his dreadlocks and multiple visible mobile insects close to his scalp. He was diagnosed with a florid infestation of P. humanus capitus (Figure, B).