Sarcoidal alopecia mimicking discoid lupus erythematosus biology essay

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Dermatologica SinicaManuscript DraftManuscript Number: DSI-D-12-00070literatureArticle Type: Case reportKeywords: Sarcoidal alopecia; discoid lupus erythematosusAbstract: Sarcoidal alopecia is a subtype of plaque-form cutaneous sarcoidosis that may resemblediscoid lupus erythematosus (DLE). The clinical appearance of the two lesions is similar, therebyleading to diagnostic confusion. Because of the systemic involvement and progressive nature ofsarcoidosis, it is important to differentiate sarcoidal alopecia from DLE so that proper treatment can beinitiated, thereby avoiding the potential long-term sequelae. We herein present a case of Taiwanesewoman with scalp sarcoidal alopecia mimicking DLE.*Manuscript without author detailsSarcoidal alopecia mimicking discoid lupus erythematosus: Report of a case and review of theliteratureChia-Fen Tsai, M. D., Hsing-Chuan Lee, M. D., Chia-Yu Chu, M. D., Ph. D. AbstractSarcoidal alopecia is a subtype of plaque-form cutaneous sarcoidosis that may resemble discoid lupuserythematosus (DLE). The clinical appearance of the two lesions is similar, thereby leading to diagnosticconfusion. Because of the systemic involvement and progressive nature of sarcoidosis, it is important todifferentiate sarcoidal alopecia from DLE so that proper treatment can be initiated, thereby avoiding thepotential long-term sequelae. We herein present a case of Taiwanese woman with scalp sarcoidal alopeciamimicking DLE. 1IntroductionSarcoidosis is an idiopathic systemic granulomatous disease in which non-caseating granulomatousinflammation can occur in any organ. The skin is affected in around 25% of cases, and the majority ofthem are African-American. 1 Involvement of the scalp is rare and may lead to cicatricial alopecia as aresult of the destruction of the hair follicles by the

granulomatous formation. 1 Clinically, sarcoidosis maypresent as papules, nodules, or plaques, and in some cases resembling discoid lupus erythematosus (DLE)or necrobiosis lipoidica. 2-6Case ReportThis 57-year-old female patient was quite well in the past. This time, she suffered from alopecia on thefronto-parietal scalp for 4-5 years (Figure 1). There are several bean to coin-sized depressed ulcers withsurrounding violaceous to erythematous hue and telangiectasia on the right fronto-parietal scalp. Anincisional biopsy was performed under the suspicion of DLE. The pathology showed non-caseatinggranulomatus inflammation involving superficial and deep dermis (Figure 2). Therefore, the diagnosis ofsarcoidal alopecia was confirmed. The immunologic profiles including antinuclear antibody, anti-extracted nuclear antigen (ENA) antibody, C3 and C4 were within normal range but chest plain filmand computed tomography revealed pulmonary sarcoidosis with lymphadenopathies (Figure 3). The lungfunction tests showed normal spirometry and diffusion capacity. Other blood tests such as complete bloodcell counts, serum aspartate aminotransferase, alanine aminotransferase, creatinine, blood urea nitrogen, sodium, potassium and calcium levels were all within normal limits. Because the patient refused2treatment with systemic corticosteroids, she was treated with oral hydroxychloroquine 400 mg daily andintralesional injections of triamcinolone 10 mg monthly. A total of 9 intralesional triamcinoloneinjections were performed from October 2009 to December 2011 in a monthly base except the 6 monthsfrom April to September of 2011. Because of significant improvement and hair regrowth, wediscontinued the local corticosteroid treatment since January 2012. The

dosage of oralhydroxychloroquine was also tapered to 200 mg daily since February 2012 and the disease showed nodeterioration after 3 months (Figure 4). Serial computed tomographic examinations of the chest from2009 to 2012 showed stable of the disease and the lung function tests remaining normal. DiscussionSarcoidal alopecia is rare and predominantly affects African American women. 1 It is a form ofsecondary scarring alopecia and can have several morphologies. Most commonly, sarcoidal plaqueslocated on the fronto-parietal facial region may extend into the scalp, thus leading to hair loss. Suchplaques are the type of sarcoidal alopecia that most closely resembles DLE. 1-4On the scalp, sarcoidosis can start as an atrophic, red, scaling or ulcerative area of alopecia. 1 Thetypical lesion of classic DLE is a well-circumscribed, erythematous, slightly scaly, atrophic plaque andmay be ulcerative occasionally. 2 Follicular plugging can be appreciated under dermoscopy, in bothsarcoidal alopecia and DLE. 5 Differential diagnosis of sarcoidal alopecia versus DLE could be made byhistopathologic examination. Sarcoidal alopecia shows classic sarcoidal granulomas in the dermis. 1-4 Incontrast, DLE is characterized by follicular plugging, epidermal basal cell vaculopathy, as well as3superficial and deep perivascular and periadnexal lymphocytic infiltrate. 2In patients with sarcoidal alopecia, the patient almost has other cutaneous lesions, and the vastmajority of cases will demonstrate systemic involvement. 1 About 30% of patients with the initial form ofcutaneous sarcoidosis will develop its systemic form within months to several years of diagnosis. 6Therefore, it is recommended that any patient with cutaneous sarcoidosis be screened for systemic lesions, even if there are no clinical complaints of systemic involvement at initial visits. Several

diagnostic studiescan be performed in the workup of sarcoidosis, including chest x-ray, chest computed tomography (CT)and pulmonary function tests. 1, 7-17Review of the English revealed a total of 47 reported cases of scalp sarcoidosis, including ours. A female predominance was noted among these patients (35/40). Where race was mentioned, 23 of 36patients were African-American and 3 of them were Chinese. Many of them (32 of 37) exhibitedextracutaneous involvement and lung was the most frequent involved site. This review documents diversemorphologies of scalp sarcoidosis. Most of these cases showed scarring alopecia, but cases ofnon-scarring alopecia have also been reported. 1, 10 Sarcoidal alopecia may resemble DLE, lichenplanopilaris, pseudopelade of Brocq, necrobiosis lipoidica, morphea or alopecia neoplastica, of whichDLE is the most confusing one. A comparison of sarcoidal alopecia, DLE, lichen planopilaris, andpseudopelade of Brocq was given as Table 1. The standard treatment for sarcoidosis is systemic corticosteroids. Topical corticosteroids are oftenineffective in treating sarcoidal alopecia because of their inadequate depth of penetration into the skin. Combination of antimalarial therapy (hydroxychloroquine plus quinacrine or chloroquine plus quinacrine)4can be of value after single-agent antimalarial therapy has failed. 1 The rationale of using antimalarialtherapy in cutaneous sarcoidosis is based on the ability of these agents to inhibit antigen processing andpresentation by antigen presenting cells to CD4+ T cells. 20 One of the initial steps in granuloma formationis antigen processing and presentation, and antimalarials may function to raise the pH within lysosomes, thus preventing assembly of MHC-peptide complexes and transport to the cell surface. Without

antigenprocessing and presentation via MHC-peptide complexes, T cells are not activated to propagategranuloma formation. 20 These agents have a relatively long history of use in the treatment of sarcoidosisand are considered standard therapy, typically in conjunction with corticosteroids or for patients in whomcorticosteroids are neither desirable nor necessary for long-term treatment. Based on reported clinicalexperience, the primary benefit of antimalarials appears to be their ability to suppress cutaneous lesions. 21Due to the disfigurement of the scalp lesions and the patient��s hesitance to the systemic corticosteroidstreatment, oral hydroxychloroquine and intralesional corticosteroids were administered simultaneously. DLE and several other diseases that resemble cutaneous sarcoidosis are relatively benign diseases. Because of its characteristic clinical appearance and low association with systemic lupus erythematosus, therapy for DLE is often administered without histologic confirmation. 2 This precludes identification ofits close clinical stimulant, cutaneous sarcoidosis. We stress the importance of a skin biopsy to confirmthe diagnosis of DLE on the scalp and to exclude sarcoidal alopecia. Because of the systemic andprogressive nature of sarcoidosis, it is critical that this distinction is made and the patient treatedaccordingly. 2 Even though our patient did not receive any systemic corticosteroid treatment due topersonal reasons, the identification of sarcoidosis did help disease monitoring and appropriate treatment if5disease progression in the future. ConclusionCicatricial alopecia presents a diagnostic challenge to clinicians. In particular, lesions of cutaneoussarcoidosis of the scalp may resemble DLE. Pathological

examination is required to make the correctdiagnosis, which leads to effective treatment.