Case Report

Sarcoidosis Mimics Lepromatous Leprosy: A Case Report

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A 34-year-old Thai man presented with a two years history of progressively enlarged lepromatous leprosy like nodules and plaques on his back, chest, and scalp. Skin biopsy showed diffuse nonnecrotizing granulomatous inflammation with numerous multinucleated giant cells, lymphocytes, and plasma cells infiltration. The missed diagnosis of leprosy was made and was treated with antilepromatous drugs for one year. After repeated skin biopsy, the diagnosis was compatible with sarcoidosis. He was treated with prednisolone 40 mg per day for two weeks. The lesions gradually decreased in size and were controlled with prednisolone 10 mg per day.

Keywords: *Systemic sarcoidosis, Leprosy, Cutaneous manifestation*

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Sarcoidosis is a multisystem chronic lymphocytic poor noncaseating granuloma of unclear etiology. Approximately 20 to 35 percent of patients have cutaneous involvement. Although cutaneous lesions may present at any time during the course of the disease, they often present early after disease onset. The primary lesions most typically found are papules, plaques, nodules, infiltrations of scars, lupus pernio and a chronic form present with violaceous plaques[1-3]. Because of the multiple manifestations of sarcoidosis, the disease has become known as one of the great imitators, masquerading as a wide range of disorders from benign appendageal growths to malignant Kaposi sarcoma[4]. Therefore, delayed or missed diagnosis often occurs because it may resemble leprosy, tuberculosis, lymphoma cutis (pseudolymphoma) and other granulomatous diseases. Herein, the authors described the distinctive clinicopathologic feature that initially presented with skin lesions that looked like lepromatous leprosy.

Case Report

A 34-year-old healthy man came to Rajavithi Hospital with asymptomatic widespread red papules, nodules and plaques at his scalp, back, chest, and a few at both legs for two years. One year ago, he was diagnosed as cutaneous leprosy from one famous private hospital in Bangkok. He was treated with dapsone, ofloxacin, and rifampin, but the lesions progressively increased in number and size.

The patient's medical history was unremarkable. He had no systemic illness, no fever but had slight weight loss. Physical examination revealed multiple nontender erythematous to bluish red shiny smooth surface papules and plaques with a diameter of 0.5 to 3 centrimeters on his back (Fig. 1), chest, arms, and both legs. There was also a few lymph nodes enlargement with a diameter of 4 centimeter at both axillas. Heart, lung, and abdomen were normal.

Skin biopsy from his arm was done for histopathologic evaluation. Routine staining with hematoxylin and eosin (H&E) revealed noncaseating epithelioid granulomatous inflammation with numerous multinucleated giant cells, interspersed lymphocytes, and plasma cells (Fig. 2). Special stains for acid-fast bacilli using both the Ziehl-Nielsen and Fite-Faraco methods were negative; no fungal elements were seen by using Gomori methenamine silver staining technique. Fungal, bacterial, and acid-fast bacilli tissues cultures were all negative. Diagnosis was compatible with sarcoidosis.

Laboratory evaluation including complete blood count with differential, electrolyte, liver function

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studies, human immunodeficiency virus, and urinalysis were all within normal limits. Chest x-ray showed paratracheal and bilateral hilar lymphadenopathy. Bronchoscope for bronchial washing and biopsy were negative for malignancy and tuberculosis but showed chronic granulomatous lymphadenitis.

The treatment for sarcoidosis was started with prednisolone 40 mg. per day (approximately 0.5 mg/kg per day). At the patient's 2-week follow-up visit, all lesions had slightly decreased in size. His treatment was gradually tapered to prednisolone 10 mg per day. His condition has been maintained successfully for one year on prednisolone 10 mg per day. Discontinuation of the prednisolone resulted in flaring of his skin lesions.

Discussion

Sarcoidosis is a multisystem disorder and lesions may occur in almost any tissue or organ of the body. Because symptoms are due primarily to local tissue infiltration and to injury from pressure and displacement by sarcoid lesions, the clinical manifestations depend largely on the organ or system involved. The organs most commonly involved are the lungs, lymph nodes, eyes, skin, liver, spleen, and phalangeal bones. Less frequent involvement has been noted in the kidneys, central nervous system, myocardium, and endocrine glands. Radiological evidence of bilateral hilar lymphadenopathy with or without parenchymal involvement is the hallmark of the disease in almost all cases of sarcoidosis in preadolescents or adolescents of both sexes between the ages of 20 and 40, but it may occur at any age. Two thirds of the patients are less than 40 years old at the time of diagnosis. The prevalence is slightly higher in females than in males and is more common in black people than in white. Diagnosis is made by the demonstration of granulomatous inflammation in involved organs and with the exclusion of other causes[5].

While the cause of sarcoidosis is still unknown, it is understood that exaggerated cellular immunity plays a key role, namely a T-cell-mediated response to an antigen as a primary step. Sarcoidosis is dependent on TH1-type lymphocytes and interferon acts to increase TH1 and decrease TH2[6]. The proposed antigens are ; the first is infectious causes the most common of which is Mycobacterium tuberculosis, atypical mycobacterium, herpes simplex virus, Epstein-Barr virus, Mycoplasma species, Corynebacteria species, Propionibacterium acnes, spirochetes, Borrelia burgdorferi, cytomegalovirus, coxsackievirus, rubella virus and systemic fungal infections; the second is environmental antigens which include metals (e.g., aluminum, beryllium, zirconium), organic dusts (e.g., pine, pollen) and inorganic dusts (e.g., soil, talc, clay); the third is autoantigens. Serum angiotensin converting enzymes and active vitamin D may play a role as mediators that increase the inflammatory response[7].

The initial reports by Besnier in 1889[8] and Hutchinson in 1989[9] were published. After that, many incidence rate reports have been observed in many parts of the world[10-22]. In Thailand, the incidence of sarcoidosis is unknown. The first clinical case of sarcoidosis in Thailand was reported in 1959[23], since then there have been several reports which were all demonstrated in 2002 by Bovornkitti S[24] and the last papulonecrotic tuberculid-like lesions of sarcoidosis was reported in the year 2004[25].

Approximately one third to one half of patients with chronic sarcoidosis have skin lesions. Skin manifestations can be either specific or nonspecific. Common specific skin lesions contain granuloma and manifest as macules, papules, nodules, plaques, subcutaneous nodules, infiltrative scars, and lupus pernio, which consist of persistent violaceous lesions on the nose, cheeks and ears, occur in 4 to 9 percent of sarcoidosis patients. In general, specific skin lesions have no prognostic significance, and do not show any correlation with the extent of systemic form of diseases. A nonspecific skin lesion is erythema nodosum, which has been shown to have a good prognosis because of its association with sarcoidosis that resolves spontaneously. Other nonspecific changes seen with sarcoidosis are calcification, prurigo, and erythema multiforme[7]. The uncommon manifestations may include hypopigmentation, psoriasiform, folliculitis, lichenoid, ulceration, acquire ichthyosis, verrucous, erythroderma, lupus like, scarring alopecia and granulomatous cheilitis[26].

Although cutaneous lesions may present at any time during the course of the disease, they often present early after disease onset. The primary lesions most typically found are papules, plaques, nodules, infiltrations of scars, and lupus pernio, a severe chronic form of cutaneous sarcoidosis with violaceous plaques on the central face[2]. The tumoral nodular and other atypical lesions are also described. They warrant consideration of several other diseases in the clinical differential diagnosis such as leprosy, granulomatous rosasea, lymphoma, histiocytoses, lupus vulgaris, leishmania, and other infectious granuloma[1,27].

As cutaneous sarcoidosis often masquerades as many other disease entities, the authors describe the case of a 34-year-old Thai man with a two year history of progressively enlarging nodules and plaques of the scalp, chest, back and both legs resulting in lepromatous leprosy appearance. A missed diagnosis was made and the patient was treated with antilepromatous drugs for one year. However, skin lesions resembling lepromatous leprosy are rare. There was, however, a case reported of a 37-year old African American woman who had a long history of facial, neck and arm lesions, which recently had become more prominent and pruritic. She also had the diffuse scaling of the scalp with small patches of scarring alopecia in the bilateral temporal regions. Her face also had leonine facies[28]. Her diagnosis was sarcoidosis by histopathological examination as the presented case report showed diffuse nonnecrotizing granulomatous inflammation with numerous multinucleated giant cells and interspersed lymphocytes and plasma cells. Therefore, the final diagnosis may be through the exclusion of other skin diseases and by histopathological examination.

Glucocorticoids are the first line treatment of sarcoidosis. In chronic cases, nonsteroidal immunosupressive agents are used to avoid side effects of steroid[29]. They are antimalarials, azathioprine, chlorambucil, methotrexate, cyclophosphamide, and cyclosporine. The presented case was treated with prednisolone 40 mg per day for two weeks. The dosage of prednisolone was gradually tapered off until the lesions had disappeared and the maintenance dosage was 10 mg per day.

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ผื่นผิวหนังซาร์คอยด์ดูคล้ายผื่นโรคเรื้อน: รายงานผู้ป่วย 1 ราย

ยุพิน ไทยพิสุทธิกุล, ไพรัช เกตุรัตนกุล

ผู้ป่วยชายไทย อายุ 34 ปี มีผื่นผิวหนังเป็นก้อนและปื้นแดงนูนดูคล้ายผื่นผิวหนังโรคเรื้อน พบที่บริเวณหลังหน้าอกและศีรษะเป็นมาประมาณสองปี ผู้ป่วยได้รับการวินิจฉัยว่าเป็นโรคเรื้อนด้วยการตัดชิ้นเนื้อไปตรวจทางพยาธิวิทยาจากโรงพยาบาลเอกชนแห่งหนึ่ง และได้รับยารักษาโรคเรื้อนมาประมาณ 1 ปี อาการผื่นเป็นมากขึ้น ผู้ป่วยจึงมาตรวจซ้ำและถูกตัดชิ้นเนื้อครั้งที่สองที่โรงพยาบาลราชวิถี และได้รับการวินิจฉัยใหม่ว่าเป็นโรคผื่นซาร์คอยด์ ผู้ป่วยได้รับการรักษาด้วยยาเม็ดเพรดนิโซโลนขนาด 40 มิลลิกรัมต่อวันประมาณ 2 สัปดาห์ ผื่นยุบลงดีและได้ลดขนาดยาเพรดนิโซโลนลงจนสามารถควบคุมโรคได้ด้วยขนาดยา 10 มิลลิกรัมต่อวัน