

Sarcoidosis Coexisting With Dermatomyositis

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Introduction

Sarcoidosis, a multisystem noncaseating granulomatous disease of unknown cause, has occasionally been associated with connective tissue diseases. African Americans and women have higher rates of sarcoidosis [1,2]. Its association with dermatomyositis is uncommon and most of cases are described in Japanese patients [3-13]. We report a patient with sarcoidosis who developed dermatomyositis.

Case Report

In September of 2013 a 38 year old woman presented to us with hands erythema, periorbital edema, (Figures 1,2) some subcutaneous masses in the right forearm and several years' history of muscle weakness. She had personal history of post radiotherapy hypothyroidism (1992) treated with levothyroid and sarcoidosis diagnosed in 1998 (probably Lofgren syndrome: bilateral hilar lymphadenopathy, erythema nodosum, fever) and treated with prednisolone. Laboratory examination: aldolase, LDH and creatin kinase (1084 U/L) levels were elevated; Histological examination of previously biopsied erythematous skin lesion on her knees, showed lymphocytic inflammatory infiltrate around upper dermis blood vessels and mucin deposition (Figure 3); a muscle biopsy and electromyogram confirmed dermatomyositis. Histological examination from a forearm nodule revealed noncaseating epithelioid granulomas in dermis and subcutaneous tissue (Figure 4).

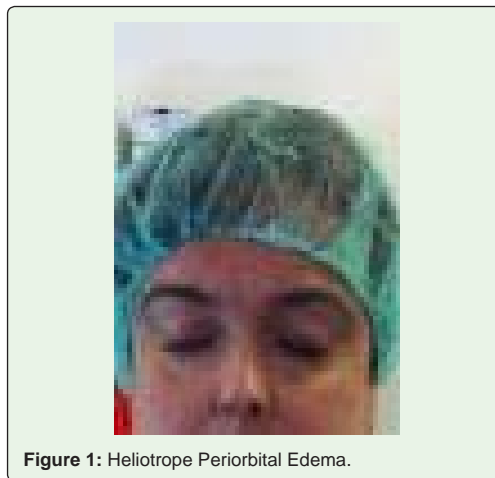


Figure 1: Heliotrope Periorbital Edema.



Figure 2: Gottron 'S Papules.

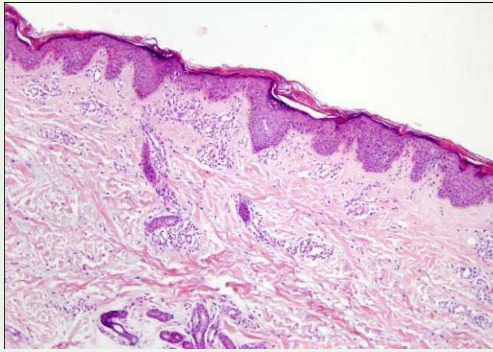


Figure 3: Lymphocytic Inflammatory Infiltrate around Upper Dermis Blood Vessels and Mucin Deposition (Skin Biopsy).

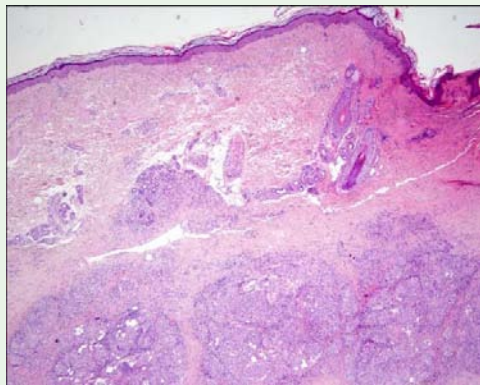


Figure 4: Non-Caseating Epithelioid Granulomas in Dermis and Subcutaneous Tissue (Forearm Biopsy).

A diagnosis of dermatomyositis coexisting with subcutaneous sarcoidosis was made. Conventional cancer screening was performed and no malignancy was found. Skin lesions showed improvement when prednisolone dose was increased to 8 mg /day.

Discussion

Sarcoidosis can affect any organ. The current hypothesis is that occurs in genetically susceptible people exposed to unknown environmental agents. Dermatomyositis (DM) is one of the idiopathic inflammatory myopathies with characteristic cutaneous findings as heliotrope rash (violaceous rash involving periorbital skin) and Gottron's papules (eruption on the dorsal hands, particularly over the knuckles) [14,15].

Sarcoidosis is rarely associated with dermatomyositis. Patients with sarcoidosis have been shown to have reduced number of regulatory T cells called CD1d restricted natural killer T cells [2] Loss of immunoregulation could explain the amplified T cell activity that characterises sarcoidosis and other autoimmune diseases [2].

Most patients with sarcoidosis do not need systemic treatment. Absolute indications for oral steroids include hypercalcaemia, neurological involvement, cardiac or ocular involvement. Corticosteroids are the accepted first-line treatment but other treatments as antimalarials, methotrexate, mycophenolate mofetil, leflunomide, tacrolimus, intravenous immunoglobulins and rituximab may be beneficial in recalcitrant cutaneous DM [16].

A relationship between dermatomyositis or sarcoidosis and malignancies is documented [3,5].

Dermatologists play an important role in the diagnosis, evaluation of extracutaneous sarcoidosis and associated dermatomyositis. Diagnosis of DM is difficult. It is important to examine each rash. Laboratory and histological examinations from different cutaneous lesions are useful for diagnosis of cutaneous lesions in patients with sarcoidosis. An associated malignancy must be ruled out. Age and sex specific malignancy screening should be considered in all adult patients with DM. A recent comparative study highlighted the potential value of PET/CT, a single and minimally invasive investigation, comparable to conventional cancer screening [15].

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