CASE REPORT

Sarcoidosis - A Case Report

RA Jahan[1], S Majumder[2]

[1]Lecturer, Department of Pathology, Rajshahi Medical College, Rajshahi, [2]Assistant Professor, Department of Respiratory Medicine, Rajshahi Medical College, Rajshahi.

Abstract

A 26 year old man presented for evaluation of bilateral lymphadenopathy on chest radiograph and multiple firm, non-tender, subcutaneous, mobile nodules on his left index and middle finger. Clinical and histopathologic features were consistent with pulmonary and cutaneous sarcoidosis. Typical skin findings are painless, firm, mobile nodules without overlying epidermal involvement and with a predilection for the trunk and extremities. Histopathologic alterations include epilthelioid cell tubercles in the subcutaneous fat. As this is a diagnosis of exclusion, other etiologies of granulomatous inflammation must be ruled out with tests and special stains. The mainstay of treatment is with oral glucocorticoids.

Key words : Caseating, Epithelioid, Granuloma, Glucocorticoid & Sarcoidosis7

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Introduction

Sarcoidosis is a multisystemic disease of unknown etiology, characterized by the formation of non-caseating granulomas in the affected organs.[1] The involvement of the skin occurs in 25% of the cases and it can develop in any phase of the disease although it is more common at the beginning.[2] The first description of sarcoidosis in 1800, was related to its cutaneous manifestations. The term 'sacoidosis' derives from a report from Boeck in 1899 and it is due to the clinical similarities of the lesions with benign sarcomas. At the beginning of the year 1900, sarcoidosis was described involving lungs and other internal organs.[1] Sarcoidosis usually occurs in young adults, with two peaks of incidence: between 25 & 35 and 45 & 55 years of age.[3] We report a case of systemic sarcoidosis with cutaneous involvement.

Case Report

In January, 2011 a 26 year old man presented to a respiratory medicine specialist of Rajshahi with the complaints of non-productive cough, exertional dyspnea, fever, night sweats for few weeks and weight loss. He had no clinical response to oral antibiotic. He had smoking habit for last 5 years. He denied any history of tuberculous exposure, recent travel or occupational exposure. Physical examination revealed a temperature of 100.6°F, respiratory rate 32/min, pulse rate 120/min, and blood pressure 120/80 mm Hg. He appeared ill and had mild respiratory distress. Head, eye, ear, nose, and throat examination was unremarkable. The neck was supple with no lymphadenopathy. Heart sounds were normal, with no murmurs or gallops. Examination of the abdomen, trunk, lower extremities, and neurologic function was unremarkable. His chest radiograph revealed bilateral lymphadenopathy. He subsequently developed multiple firm, non-tender, subcutaneous, mobile nodules on the left index and middle finger.

Laboratory Data :

A complete blood count, basic metabolic panel, liver function tests, urinalysis, and pulmonary function tests were normal. A chest radiograph showed bilateral hilar fullness. Hepatitis B antigen and antibody, and hepatitis C antibody tests were negative. A purified protein derivative skin test was negative. An excisional biopsy of one of the subcutaneous nodule on the left index finger was performed.

Histopathology :

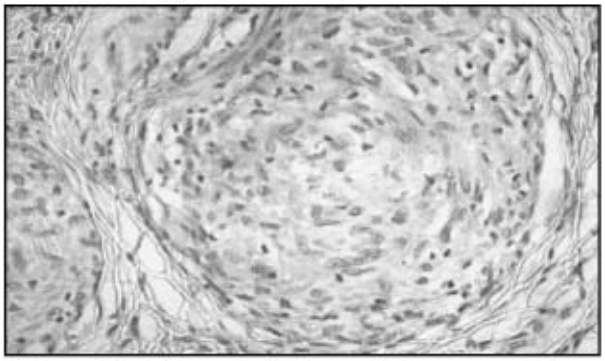

Figure 1: Characteristic sarcoid non-caseating granulomas in skin nodule with many giant cells.

Discussion

Sarcoidosis can affect any organ system, but pulmonary manifestations typically dominate.[4] Abnormalities on chest radiographs are detected in 85–95% of patients. However, 30–60% of patients with sarcoidosis are asymptomatic, with incidental findings on chest radiographs.[4] The clinical course is heterogeneous. Spontaneous remissions occur in nearly two-thirds of patients, but the course is chronic in 10–30%.[5]

The radiographic staging system for pulmonary sarcoidosis was developed more than four decades ago.[6]

This classification schema defines the following stages:

Stage 0	Normal chest radiograph
Stage I	Bilateral hilar / mediastinal lymphadenopathy
Stage II	Bilateral hilar/mediastinal lymphadenopathy plus interstitial infiltrates
Stage III	Interstitial infiltrates without evidence of hilar/mediastinal lymphadenopathy
Stage IV	Dense fibrosis, bullae formation, architectural distortion

The most characteristic finding (present in 50–85% of cases) is bilateral hilar lymphadenopathy (BHL), often with concommitant enlargement of the right paratracheal lymph nodes. Rare features (occurring in 1–3% of patients) include pleural effusions, unilateral segmental infiltrates or mass lesions, unilateral hilar lymphadenopathy, large nodular opacities simulating metastases, cavitation and diffuse ground-glass opacities.[4]

Subcutaneous sardoidosis is a rare, specific subtype of nodular cutaneous sarcoidosis that was originally described by Darier and Roussy in 1904. Although these nodules are often referred to as Darier-Roussy sarcoid, this is actually a term that is used to describe many subcutaneous inflammatory disorders and is therefore nonspecific. Hence, this eponym has been largely abandoned in exchange for the more accurate term subcutaneous sarcoidosis.[7] The diagnosis of subcutaneous involvement in sarcoidosis is made by the histopathologic identification of noninfectious sarcoidal or epithelioid granulomas with multi-nucleated giant cells and with lymphocytic inflammation involving predominantly the panniculus.[7] This diagnosis is associated with extra-cutaneous systemic disease involvement, especially bilateral hilar lymphadenopathy. Subcutaneous sarcoidosis presents as painless, firm, mobile nodules without overlying epidermal involvement.[8] These nodules have a predilection for the trunk and extremities, but also occur on the face. The range of the number of lesions is 1 to 100 and the average size of these lesions is 0.5 to 2 cm.[9]

The relation between cutaneous and systemic sarcoidosis has been evaluated.[10] In this study, the patient is a young adult and he presented with non-productive cough. He showed bilateral hilar lymphadenopathy on his chest radiograph and multiple skin nodules on his left index and middle finger. Histopathology of the nodule revealed epilthelioid cell tubercles in the subcutaneous fat. The nodules arise in the deep dermis and adipose tissue and may rarely become calcified. Appropriate tests and special stains always should be performed to rule out additional etiologies of these granulomas. Mantoux test was done and it was negative in this case. The mainstay of treatment for subcutaneous sarcoidosis is with oral glucocorticoids. Traditional dosages range from 20 to 40 mg daily, with responses noted within 4 to 8 weeks after initiation of therapy.[11] Patients also have been treated with nonsteroidal anti-inflammatory drugs, clofazamine, methotrexate, hydroxychloroquine, intralesional glucocorticoids, dapsone, allopurinol, minocycline, and potassium iodide with various responses.[11] In this case oral glucocorticoid therapy was briefly used and the patient now improved.

Conclusion

Sarcoidosis is a multi-systemic inflammatory disorder, but affects the lungs in 90% of cases. Non-productive cough, dyspnoea and chest pain are the most common features of pulmonary sarcoidosis. The diagnosis of pulmonary sarcoidosis is suggested by bilateral hilar

lymphadenopathy, with or without parenchymal changes on chest radiographs and is supported by noncaseating granulomata in tissue biopsies. Radiographic staging of pulmonary sarcoidosis as well as clinical and laboratory findings can be prognostic. Treatment of sarcoidosis typically includes corticosteroids, but other therapeutic agents may have benefit, and treatment needs to be individualised. Lung transplantation remains a viable therapeutic alternative for patients with pulmonary sarcoidosis who do not respond to pharmaceutical agents.

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