Rheumatic disease in a Nigerian lady with sarcoidosis: Case report

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Abstract

Sarcoidosis is a rare disease. It is a systemic granulomatous disease that primarily affects the lungs and lymphatic systems of the body. It has not been reported in rheumatology practice in Nigeria. The essential factors for diagnosis include compatible clinic-radiologic features, histologic proof of non-caseating epithelioid granulomas, and exclusion of similar diseases. The patient was a 63 year old lady who had a skin lesion overlying the upper lip. She developed cough which was associated with occasional chest pain. She had multiple joint pains, fever, oral ulcers and weight loss with associated anorexia. She developed significant hair loss, fatigue, redness of the eyes; with poor vision, and hearing impairment of about 6 years duration at the time of presentation. Essential findings were those of hearing impairment and a few crepitations in the lung bases. Her chest radiograph revealed bilateral hilar lymphadenopathy. The antinuclear antibody and anti double stranded DNA were both negative while serum angiotensin converting enzyme (sACE) was elevated (58.5IU). A punch biopsy of the skin overlying her upper lip revealed granulomatous dermatitis. Her electrolytes, urea and creatinine, liver function tests and full blood count were essentially normal.

Key words: Sarcoidosis, Granulomatosis, Joint pains, Serum angiotensin converting enzyme

Introduction

Sarcoidosis is a systemic granulomatous disease that primarily affects the lungs and lymphatic systems of the body[1]. The cause of sarcoidosis remains unknown, however genetic and environmental factors have being incriminated. The possible agents involved in its aetiology are micro organisms, organic and inorganic dusts such as aluminium, zircomium and talc[1]. Sarcoidosis occurs globally and has its highest geographic prevalence in northern European countries (5-40 cases per 100,000 people)[2]. The disease appears to be slightly more frequent in women than men[3]. Sarcoidosis may present at any age but prevalence peaks between 20 and 40 years of age, with a second peak in women over 50 years[3]. Sarcoidosis appears to be three to four times more common in blacks than whites[4]. Immunogenetic differences between patients may determine their clinical manifestations of sarcoidosis, and could underlie the heterogeneity of the disease[5,6]. Blacks tend to present with more acute and severe disease than whites, who tend to present with asymptomatic and chronic sarcoidosis[7].

Being a multisystemic disease, sarcoidosis also does affect the musculoskeletal system. To the best of our knowledge, it has not been reported in rheumatology practice in Nigeria. We present a case of a middle aged Nigerian lady with sarcoidosis attending a rheumatology clinic.

Case report

The patient was a 63 year old lady who was apparently well until about 10 years prior to presentation when she developed a lesion on the skin overlying the upper lip. She developed cough which was productive of sputum but no associated hemoptysis. There was only occasional chest pain but no difficulty with breathing. She had multiple joint pains involving the shoulders, knees, ankles and wrists. She had experienced fever, oral ulcers, and weight loss with associated anorexia. She also developed significant hair loss, fatigue, redness of the eyes; with poor vision and hearing impairment of about 6 years duration at the time of presentation.

She had a skin nodule on the left foot with associated numbness of the digits. There was a history of neck pain radiating down to the limbs. She only occasionally takes alcohol but does not smoke. She is neither hypertensive nor diabetic. She has never worked in an industry. Essential findings were those of hearing impairment and a few crepitations in the lung bases. Her chest radiograph revealed bilateral hilar lymphadenopathy while

radiography of the cervical spine showed spondylosis. The antinuclear antibody and anti double stranded DNA were both negative while serum angiotensin converting enzyme (sACE) was elevated (58.5IU). A punch biopsy of the skin overlying her upper lip revealed granulomatous dermatitis. This consisted of focal thinning and mild acantholysis of the epidermis. The papillary dermis exhibited band-like zone of inflammation characterized by foamy histiocytes, macrophages with melanin incontinence, lymphocytes and a few poorly formed giant cells. Thyroid function test showed minimally elevated T3 and T4 with a normal thyroid stimulating hormone. Her electrolytes, urea and creatinine, liver function tests and full blood count were essentially normal. Her erythrocyte sedimentation rate was 79mm/Hr. Her serum calcium was within normal limits (9.2mg/dl). She is presently on methotrexate, folic acid, prednisolone and omeprazole.

Discussion

Due to its nonspecific presentation, the diagnosis of sarcoidosis can be challenging. It has been shown recently that the diagnosis is often delayed[8]. The essential factors for diagnosis include compatible clinico-radiologic features, histologic proof of non-caseating epithelioid granulomas, and exclusion of similar diseases[1,9].

The reported case has a myriad of symptoms and laboratory features in keeping with sarcoidosis. The age of the patients fits in to the second peak of patients presenting with sarcoidosis[4]. Though 63 years old, her symptoms started 10 years prior to presentation hence in keeping with an earlier report by Giovinale *et al*[10] who described a 53 year old with systemic sarcoidosis. Being female is in tandem with the greater frequency of sarcoidosis in the female gender[1]. Cough is a common presentation in sarcoidosis as it primarily affects the lungs in more than 90% of patients[11]. There are four stages of pulmonary sarcoidosis. Patients with stage I or II disease may have no symptoms, whereas stages III and IV can be characterized by progressive dyspnea, loss of lung function, and fibrosis[9]. Twenty-five percent of patients have skin involvement[11]. Lesions can range from nonspecific maculopapular eruptions, such as plaques and nodules, to erythema nodosum and lupus pernio[11]. The reported case had a lesion on the skin overlying the upper limb whose biopsy revealed granulomatous dermatitis. Lupus pernio consists of indurated plaques and discoloration of the nose, cheeks, lips, and ears and usually indicates a chronic disease course that is unlikely to result in spontaneous remission[11] as has been in the reported case. The presence of granulomas alone is not diagnostic as the same lesions may also be observed in chronic berylliosis, tuberculosis, histoplasmosis, coccidioidomycosis, lymphoma, Hodgkin's disease, bronchogenic carcinoma, foreign body granuloma, schistosomiasis, syphilis, and leprosy[12]. The reported case had articular involvement. Osteoarticular involvement in sarcoidosis is uncommon[9]. The incidence is around 3–13% and is clinically relevant only in 2–5% of cases, often presenting an asymptomatic course[13].

Patients with bone involvement often also have lung disease[14] (up to 90% of cases) and skin involvement (in a proportion that rises to 25%) mainly in the form of lupus pernio as is the case in the reported patient. The presence of acute polyarthritis associated with lymphadenopathy and erythema nodosum is called Löfgren syndrome[15]. In the American Thoracic Society/European Respiratory Society/World Association of Sarcoidosis and other granulomatous disorders statement on sarcoidosis, constitutional symptoms, such as fever, weight loss, fatigue and malaise, were mentioned as being present in approximately one-third of patients[16]. Neurosarcoidosis is a rare manifestation which may present as cranial nerve palsies, meningitis, seizures, and neuropsychiatric symptoms[17,18]. The most common neurologic manifestation of sarcoidosis is cranial neuropathy secondary to nerve granulomas, raised intracranial pressure, or granulomatous meningitis[19]. The facial nerve is the most frequently affected cranial nerve while the vestibulocochlear nerve is involved in 1–7% of cases of neurosarcoidosis[19] as in this case. Bilateral hilar lymphadenopathy is characteristic as was seen in this case and usually accompanied by paratracheal lymphadenopathy[20]. Serum angiotensin converting enzyme is increased in 30 to 80% of patients with sarcoidosis as was in this case[21]. Serum Angiotensin Converting Enzyme (ACE) has a poor predictive value in sarcoidosis and can also be elevated in several disorders such as tuberculosis, multiple sclerosis, and Guillain-Barre Syndrome[21].

Conclusion

Sarcoidosis is a rare multisystemic disorder with protean clinical, laboratory and radiologic manifestations. The musculoskeletal system is not excluded from its presentation. The presence of arthritis, hilar adenopathy and skin manifestation should raise the suspicion about Löfgren syndrome.

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