

Sarcoidosis in Nepal: Epidemiology, Clinical Presentation and Ethnic Differences

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ABSTRACT

Background & Objectives: Sarcoidosis is a multi-system granulomatous disorder of unknown origin. The frequency and the clinical presentation of sarcoidosis vary among the geographical regions. This study is a retrospective analysis of the sarcoidosis patients diagnosed in Nepalese population. The aim of this study was to find the prevalence of sarcoidosis in Nepalese population which has high prevalence of tuberculosis and to understand the clinical profiles and ethnic differences. **Materials & Methods:** A retrospective chart analysis was done in all the patients from January first, 2005 to December 31st, 2010 who had abnormal chest radiograph and who were diagnosed with sarcoidosis. The study was done at Nepal Medical College Teaching Hospital and Everest Nursing Home. **Results:** A Forty two patients were diagnosed to have sarcoidosis over a five year period, of which 23/42 (55.0%) were females and 19/42 (45%) were males. The mean age was 36 years with minimum age being 18 years and maximum 72 years. As far as Ethnicity is concerned, 15/42 (36.0%) were Brahmins, 08/42 (19.0%) were Marwadi, and 07/42 (17.0%) were Chhetri. The most common presenting symptom was cough 19/42 (45.0%). Out of the 42 patients, eight were being treated for pulmonary tuberculosis. Most common radiographic findings were bilateral hilar lymphadenopathy 15/42 (36.0%). **Conclusion:** Sarcoidosis is common in Nepal and should be considered in the differential diagnosis before treating the patient empirically for tuberculosis.

Key words: Epidemiology; Nepalese population; Sarcoidosis

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INTRODUCTION

Sarcoidosis is a systemic disorder of unknown cause that is characterized by its pathological hallmark, the non caseating granuloma. Its presenting features are protean, ranging from a symptomatic but abnormal findings on chest radiography in many patients to progressive multiorgan failure in an unfortunate minority. The illness can be self-limited or chronic, with episodic recrudescence and remissions. Because the lungs and thoracic lymph nodes are almost always involved, most patients report acute or insidious respiratory problems, variably accompanied by symptoms affecting the skin, eyes, or other organs.

In a study from eight countries of Asia and Africa in 1976, fewer than 30 patients were reported from India, Malaysia, Thailand, Taiwan, and United Arab Emirates, while none from Singapore and Korea.¹ However, there has been increasing reports of sarcoidosis from countries like India,²⁻⁶ Singapore,⁷ and Malaysia.⁸ There has been no study from Nepal looking into the prevalence of sarcoidosis. Remarkable clinical similarities with tuberculosis make the differential diagnosis of the two conditions difficult especially in countries like Nepal which has high burden of tuberculosis. The objective of this study was to find the prevalence of sarcoidosis in Nepalese population

which has high prevalence of tuberculosis and to understand the clinical profiles and ethnic differences.

MATERIALS AND METHODS

Setting: The study was done at Nepal Medical College Teaching Hospital which is a tertiary care teaching hospital and at Everest Nursing home which is a private nursing home. Both located in Nepal.

Study design: We did a retrospective chart analysis of all the patients from January one, 2005 to December 31, 2010 who was diagnosed with sarcoidosis. Demographics including ethnicity, clinical presentation, and diagnostic work up were noted. Diagnostic work up including chest imaging, bronchoscopy and tissue biopsy results were noted in detail. We also gathered pertinent microbiological data of these patients which included sputum gram stain, AFB and culture reports. Laboratory tests such as total white cell count, differentials including lymphocyte level, serum calcium level, and serum angiotensin converting enzyme levels (SACE) were also collected. Other pertinent lab tests including anti-nuclear antibody and rheumatoid factor were also checked in some of the patients to rule out connective tissue diseases. Presence of non caseating granulomas in the lung and the absence of TB or fungal disease were regarded as confirmation of the diagnosis of Sarcoidosis.

Subjects: All the patients who were diagnosed with Sarcoidosis during this time frame.

RESULTS

A total of 42 patients were diagnosed to have Sarcoidosis over a five year period, of which 23/42 (55.0%) were females and 19/42(45.0%) were males. The mean age was 36 years (range 18 to 72 years), with 81.0% of the patients falling in the age group of 20-45 years. As far as ethnicity is concerned, 15/42 (36.0%) were Brahmin, 08/42

(19.0%) were Marwari, and 07/42(17.0%) were Chhetri. The most common symptoms were cough and joint pains (table 1). Ocular symptoms were reported by two patients and two patients were asymptomatic. Out of the 42 patients, eight(19%) were being treated for pulmonary tuberculosis. Physical examination was unremarkable in most patients except in the two patients who were found to have erythema nodosum. A total of 32 out of 42 patients (76%) had stage I disease on chest radiograph (mediastinal and/or hilar lymphadenopathy), three patients had stage II disease (bilateral pulmonary infiltrates in addition to mediastinal/hilar lymphadenopathy) and seven patients had stage III disease (only pulmonary infiltrates) (table 2). None of the patients had stage IV disease or lung fibrosis. Transbronchial and or endobronchial lung biopsy confirmed findings suggestive of sarcoidosis in 30 (71%) patients. In 12 patients, diagnosis was made by transbronchial needle aspiration of the mediastinal and or hilar lymph nodes.

DISCUSSION

Our study shows that Sarcoidosis is common in Nepal. The study also indicates that it is more common in Brahmins and it is equally distributed in Chhetri and Marwari population. We did not find Sarcoidosis in other ethnic groups in Nepal like Newar, Gurung and Sherpas. Our findings also showed that sarcoidosis is mistaken for tuberculosis and many patients (19% in our study) are empirically treated with anti-tuberculosis treatment. Tuberculosis still remains the most prevalent infectious disease in Nepal, but the awareness regarding sarcoidosis has increased and it is diagnosed with greater frequency in recent times. Current study also shows that there are improved efforts on the part of the treating physicians to make a diagnosis. The availability of computed

Table 1: Symptoms

Symptoms	No. of patients (%)
Cough	25 (60%)
Joint pains	13 (30%)
Eye symptoms	02 (5%)
Asymptomatic	02 (5%)
Total	42

Table 2: Most common radiographic findings

Radiographic findings	Stage	No. of cases
Mediastinal and bilateral hilar lymphadenopathy	I	32
Pulmonary infiltrate with mediastinal/hilar lymphadenopathy	II	03
Pulmonary infiltrate only	III	07
Pulmonary fibrosis	IV	00
Total		42

tomography and fiberoptic bronchoscopy for transbronchial lung and lymph node biopsies has also helped in making the diagnosis; unlike in the past when chest abnormality was invariably considered to be due to tuberculosis.

The disease affects people in their most productive years in life as was seen in our study. In terms of ethnicity, the reports from eastern India suggest involvement of Marwari community more often. In our study, 19% patients were Marwari. In the west regarding ethnicity is concerned, the Puerto Ricans in America and the Scandinavians in Europe suffer from this disease more commonly, also blacks are several times more affected compared to whites.⁹⁻

¹¹The clinical presentation and natural course of sarcoidosis vary greatly.¹² Although almost any organ of the body can be affected, the lungs or intrathoracic lymph node are involved in almost all the cases (>90%). In our study, all the patients had involvement of respiratory system.

Radiologically, as in our study, stage I disease is most common presentation of sarcoidosis.¹² Determination of a general prognosis, based on the patient's initial clinical presentation, is possible. Stage I disease, which is defined as the presence of hilar adenopathy without parenchymal infiltrates on radiography, remits in 60 to 80 percent of cases without treatment; stage II (hilar adenopathy with infiltrates) remits in 50 to 60 percent; and stage III (infiltrates without adenopathy) remits in less than 30 percent.¹³ Patients with Lofgren's syndrome have the best prognosis.

The limitation of this study is the fact that this was conducted in Kathmandu valley. Involving multiple academic centers throughout Nepal would give us the true prevalence of sarcoidosis in Nepal. The study was conducted five years back but given the lack of another study about Sarcoidosis in Nepal and the fact that the prevalence of tuberculosis remains the same this study and the findings still remain relevant.

CONCLUSION

The true prevalence of sarcoidosis in Nepal is difficult to assess due to the high prevalence of tuberculosis. It is more common in Brahmins and equally distributed in Chhetri and Marwari population. Physicians should have high index of suspicion and sarcoidosis should be in the differential diagnosis of young patients with mediastinal/hilar lymphadenopathy with or without pulmonary infiltrates.

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