CLINICOLABORATORY PROFILE OF SARCOIDOSIS - OUR EXPERIENCE

Aruna Talatam¹, Phani Kumar Reddy²

¹Assistant Professor, Department of Pulmonary Medicine, NRI Medical College, Guntur, Andhra Pradesh.
²Assistant Professor, Department of General Medicine, NRI Medical College, Guntur, Andhra Pradesh.

ABSTRACT

BACKGROUND
Sarcoidosis is a multisystem disorder of unknown aetiology affecting middle age group. Most commonly affected organs are lymph nodes, lungs, skin and eyes. It is characterized by noncaseating granulomatous inflammation of sites of disease. Diagnosis is based on clinical, radiological and histological examination.

AIM
The purpose of our study is to present clinicolaboratory profile of established sarcoidosis cases which is grossly underdiagnosed in our country and to create awareness in the minds of clinicians regarding diagnostic workup and discuss the treatment modalities which are usually tailor made.

MATERIAL AND METHODS
12 patients diagnosed to have sarcoidosis who came to our outpatient department were included in this study. A detailed clinical evaluation, laboratory investigations were carried out and treated accordingly.

RESULTS
In this study, we present 12 cases of sarcoidosis with varied presentations. Most frequent presentation was stage II sarcoidosis (50%). Most commonly affected organs were lymph nodes (90%), lungs (83%), eyes (41%) followed by skin, cranial nerves, parotid, liver, heart, etc. Elevated serum ACE levels were noted in 9 cases (80%), all the cases were with extrapulmonary involvement. Hypercalcaemia and hypercalciuria was seen in 2 cases. There was remission with glucocorticoids in 50% of cases and two cases were treated with glucocorticoids and azathioprine.

DISCUSSION
Sarcoidosis is a multisystem disorder of unknown cause. It commonly affects young and middle-aged adults and frequently affects lung, lymph nodes, eye and skin. The diagnosis is established when clinicoradiographic findings are supported by noncaseating epithelioid cell granulomas in two separate organ systems. The mainstay of treatment is corticosteroids. The duration of therapy is 3-6 months in tapering doses. If there is no clinical response, cytotoxic agents are added.

CONCLUSIONS
Even today sarcoidosis, a multisystem disorder remains a challenging diagnosis. The first step of success is to have a high index of suspicion in clinician’s mind particularly in our country where tuberculosis is rampant.

KEYWORDS
Sarcoidosis, Lymph nodes, Lungs, Glucocorticoids.

INTRODUCTION: Sarcoidosis is a multisystem disorder of unknown origin characterized by noncaseating granulomatous inflammation of sites of disease. The disease can affect any organ in the body, the most frequent presentations being bilateral hilar adenopathy, pulmonary infiltration and skin or eye lesions. The disease was first identified in 1869 by Hutchinson. It has been more than 100 years since the disease is known,(¹) but the cause is yet to be identified. The disease can have devastating effect on the overall quality of life of patients who suffer from it. The fascination with sarcoidosis lies in its variable presentation, and its unpredictable clinical course. Even today it remains a challenging diagnosis. The treatment has to be tailored to the individual needs of the patient. In spite of the advances in diagnostic modalities and development of newer drugs, the clinical outcome remains a “mystery”.

AIM: The purpose of our study is to present clinicolaboratory profile of established sarcoidosis cases and to discuss the treatment modalities which are usually tailor made.
MATERIALS AND METHODS: Patients diagnosed to have sarcoidosis were included in this study. A detailed history, clinical evaluation and series of tests were carried out. Chest x-ray, ultrasound of abdomen and CT chest was done in all patients. Specific biochemical tests like serum calcium, 24 hours urinary calcium, serum ACE levels. Bronchoscopic procedures like bronchial washings, transbronchial needle aspiration, endobronchial lymph node biopsies were done. In few cases, skin biopsy, conjunctival biopsy, nasal mucosal biopsy and liver biopsy were done for histological confirmation. Mantoux test was negative in all the cases. Asymptomatic cases were followed under close observation. All patients with symptoms were treated with glucocorticoids for six months with slow tapering. All those patients with no evidence of disease progression for at least one year of followup period were considered cured. Few cases required immunosuppressants in addition to glucocorticoids.

RESULTS: We present a total of 12 cases of sarcoidosis. Out of which 3 cases with bilateral hilar lymphadenopathy, 6 cases with bilateral hilar lymphadenopathy and pulmonary infiltrates and 3 cases with fibrocystic changes (Fig. 1, 2, 4). All the cases were diagnosed according to American Thoracic Society guidelines. In all the cases, the clinicoradiographic features were highly suggestive of sarcoidosis supported with the histologic evidence of noncaseating epithelioid cell granulomas. Granulomatous inflammation was present in >= two organs in majority of cases.

An interesting case of stage II sarcoidosis was a 48-year-old lady who initially developed subcutaneous nodule on wrist for which excision was planned. Her surgical profile revealed radiologic changes suspicious of sarcoidosis. Her mother was an established case of sarcoidosis with multiorgan involvement. The patient also had associated ulcerative colitis and lichen planus. Her blood parameters, serum ACE levels, serum calcium levels were normal. Meanwhile she developed skin rash (Fig 3). She being a medical professional was reluctant to use steroids and there was spontaneous regression of subcutaneous nodule. She was put on hydroxychloroquine 200 mg once daily for skin rash for which she is responding well.

DISCUSSION: Sarcoidosis is a multisystem disorder of unknown cause. It commonly affects young and middle-aged adults and frequently affects lung, lymph nodes, eye and skin. The liver, spleen, salivary glands, heart, nervous system, muscles, bones and other organs also may be involved. The diagnosis is established when clinicoradiographic findings are supported by noncaseating epithelioid cell granulomas in two separate organ systems. Granulomas of unknown origin and local sarcoid reactions must be excluded. Frequently observed immunologic features are depression of cutaneous delayed type hypersensitivity and a heightened Th-1 immune response at sites of disease. Circulating immune complexes along with signs of B-cell hyperactivity also may be found.\(^{(2)}\)

Sarcoidosis has been reported in India but its similarity to tuberculosis has led to underdiagnosis. Our study includes 12 cases of sarcoidosis out of which lymph node involvement was commonest (91.6%), followed by lung (83%), eyes in 41% of cases. Skin and cranial nerve involvement in 16.6% of cases. Parotid, liver and upper respiratory tract involvement was seen in one case each. Erythema nodosum was reported in one case. Type II sarcoidosis was found to be commonest (50%) in our study, followed by type I and IV 25% each. Our data was compared with other Indian studies as suggested in table 1. Elevated serum ACE levels were noted in 9 cases (80%), all the cases with extrapulmonary involvement compared to raised ACE levels in 75% of cases in a study by Gupta et al. Hypercalcaemia and hypercalciuria was seen in 2 cases.

The mainstay of treatment is corticosteroids.\(^{(3,4)}\) The duration of therapy is 3-6 months in tapering doses. If there is no clinical response, cytotoxic agents are added.\(^{(5)}\) Commonly used cytotoxic agents are methotrexate, azathioprine, mycophenolate, cyclophosphamide, leflunomide.\(^{(6)}\) The antimalarial agents have been reported as useful in chronic cutaneous disease and nephrolithiasis.

There was spontaneous resolution in 1 case. Three cases are being followed up with close observation and they are not on steroids. Six patients used steroids and their symptoms have resolved.

Two patients are on steroids and azathioprine. All the patients are leading a fairly good quality of life.

The disease has heterogenous outcome. In some, there may be spontaneous regression or progression of extensive fibrotic lesions as a postgranulomatous fibrosis. A statement on sarcoidosis reported in 1999 described the course and prognosis of disease as self-limiting in acute cases whereas progressive fibrosis in those with insidious onset.\(^{(7)}\)

CONCLUSIONS: Sarcoidosis, an enigmatic multisystem disorder, has many different faces. Even today, it remains a challenging diagnosis. The first step of success is to have a high index of suspicion in clinician’s mind particularly in our country where tuberculosis is rampant. As far as treatment is concerned protocol based treatment does not hold good. In our experience, we noted the treatment has to be individualised according to the clinical presentation and all the patients are maintaining a good quality of life even in those who were diagnosed 2 decades ago.

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<table>
<thead>
<tr>
<th>Present study (In %) (n=12)</th>
<th>Kumar et al(^{(8)})</th>
<th>Joshi et al(^{(9)})</th>
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<tr>
<td>Age &gt; 40 years</td>
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<tr>
<td>Stage I</td>
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<tr>
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<td>IV</td>
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Table 1: Comparison of our study with other Indian studies

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<th>Other Study 2</th>
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<tr>
<td>Skin</td>
<td>16.6</td>
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<tr>
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<td>Serum ACE levels</td>
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REFERENCES: