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Extrathoracic sarcoid: A case report from Bangladesh

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Abstract

Sarcoidosis is a multi-system granulomatous disease characterized by the presence of non-caseating granuloma. Though the respiratory symptoms are the commonest presentation, a wide range of extrathoracic involvement frequently occurs which often leads to misdiagnosis. In a tuberculosis endemic area like Bangladesh, sarcoidosis is very difficult to diagnose when it presents with a nonspecific presentation along with peripheral lymphadenopathy. Here we reported a 60 years lady who presented with fatigability for 6 months associated with significant weight loss. She has non-tender cervical and axillary lymphadenopathy. Her chest X-ray and HRCT chest were insignificant. Subsequent excision biopsy of lymph node demonstrates non-caseating granuloma, suggestive of sarcoidosis after exclusion of all other possible diagnosis The aim of reporting this case to aware physicians regarding the atypical extrathoracic clinical presentation of sarcoidosis and how it can mimic tuberculosis.

Keywords

Sarcoidosis; granulomatous disease; respiratory symptoms; X-ray

Introduction

Sarcoidosis is an inflammatory disease characterized by non-caseating granuloma of unknown eitiology [1]. The incidence is higher among 20-40 years of age group with slightly female preponderance. Only 5% of sarcoidosis patients have a positive family history [2].

Historically sarcoidosis was thought to be more prevalent in developed countries. However recent data showed that it is being more prevalent in Tuberculosis endemic countries as well, eg, indian subcontinent [3]. One of the probabilities of this increase is that the diseases was overshadowed by the presence of tuberculosis. As recently tuberculosis has been controlled and can be excluded confidently utilizing investigations so that the incidence of sarcoidosis has inclined [4].

A diagnosis of sarcoidosis is established based on the compatible clinical history and radiologic findings which is confirmed by histologic evidence of non-caseating epithelioid-cell granulomas in one or more organs. The disease predominantly affects lung, but can also any organ including skin, eye and abdominal organs [5].

Because sarcoidosis can involve any organ, the clinical presentation is often variable. 50% of cases are asymptomatic which are diagnosed incidentally. Patient commonly presents with non-specific symptoms, such as fatigue, weight loss, fever and night sweats [6]. After the nonspecific symptoms, the commonest mode of presentation is respiratory and musculoskeletal symptoms [7].

Though Chest Xray is the key investigation, high-resolution CT (HRCT) chest may be needed if chest Xray is inconclusive in suspected cases, as parenchymal change can be best delineated on HRCT. Bilateral hilar lymphadenopathy is the earliest and one of the most common manifestations of sarcoidosis, peripheral lymphadenopathy is less likely to present first [8].

Since sarcoidosis is a multisystem disorder, evidence of granulomatous inflammation in at least 2 organs is required for diagnosis. However, biopsy confirmation from one organ is sufficient when background is compatible and alternative diagnoses have been excluded [9,10].

Bronchoalveolar lavage fluid analysis, transbronchial biopsy are helpful to exclude the differential diagnosis such as tuberculosis, fungal infections, and malignancy. Additional investigations would be high serum calcium and high Serum ACE level, lung volume estimation and diffusing capacity of carbon monoxide. Other tests includes analysis of 24-hour urinary calcium and IFN-γ release assays [11].

Fortunately, many patients do not need treatment as they get spontaneous remission. Corticosteroid remains the mainstay of primary treatment. Among the steroid-sparing agents, Methotrexate, Chloroquine, Hydroxychloroquine, Cyclophosphamide, Azathioprine, Chlorumbucil, Cyclosporin, Infliximab, Thalidomide are variably used [12].

The overall prognosis is excellent. only 5% to 8% may have fatal outcome [13].

Case Story

A 60 years lady presented with increasing fatigability for the last 6 months. She had no myalgia, joint pain, and no respiratory symptoms. But she had a weight loss of about 15kg during this period.

She had been diabetic for the last 8 years, on oral hypoglycaemic agent and it was well controlled. She did not have any tuberculosis contact. There were multiple nontender cervical lymph nodes of variable size. The largest one was 8×7 cm. Her right axillary lymph nodes were also palpable. They were of variable size and shape, the largest one was about 9×7 cm. All the lymphnodes were firm, non-tender, not fixed with the underlying structure and overlying skin was free and there is no discharging sinus.

At this point, our possible differential diagnosis was Tuberculosis, Sarcoidosis, atypical fungal di-

sease. Blood counts were within the normal range. The serum calcium was 13 mg/dl. The Mantoux test was 3 mm. Her diabetic status was controlled. Renal and liver function were within normal limits. Ultrasonography (USG) of the abdomen showed mild hepatosplenomegaly. HRCTof the chest showed ground glass soft tissue densities in both lungs.

First, we performed Fine needle aspiration cytology (FNAC) of the lymph node which showed chronic granulomatous inflammation. As it did not give us the definite diagnosis, later The lymph node was excised and the histopathologic study revealed non-caseating multiple discrete granulomas composed of a tight cluster of epithelioid cells, lymphocytes, Langerhans cells, and foreign body type giant cells (Figure 1).

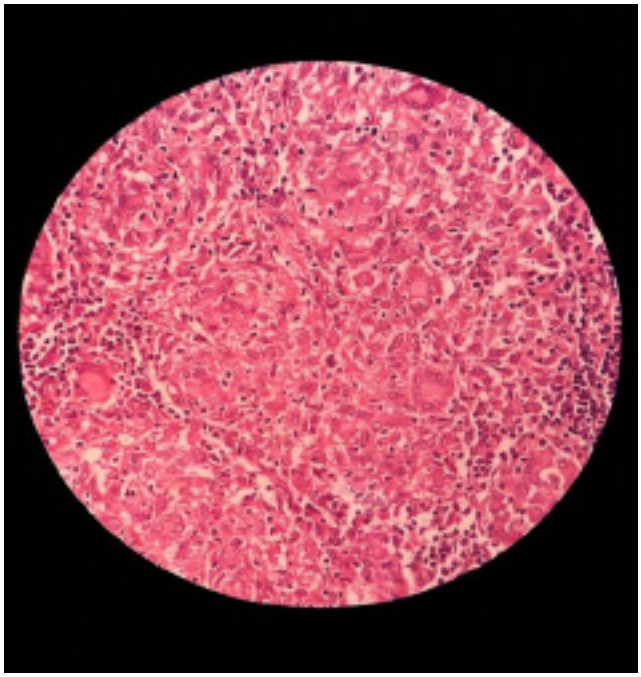

Figure 1: Lymph node Biopsy suggesting of Non-caseating multiple discrete granulomas composed of a cluster of epithelioid cells, lymphocytes, Langerhans cells, and foreign body type giant cells.

Discussion

Sarcoidosis is a granulomatous disease involving multiple organs. Babu K described in his article that , unlike earlier years, when sarcoidosis is thought to be prevalent in develpowd countries, now it is being common in developing countries [14]. Specially Sharma mentioned the increase in incidence in india. It has been estimated that sarcoidosis constituted 10 to 12 cases/1,000 new registrations annually at a Respiratory Unit at Kolkata and 61.2/100,000 new cases seen at the Delhi [15].

Despite this sarcoidosis is one of the under-reported diseases in our country on account of its similarity to tuberculosis, which is further compounded by the lack of awareness among physicians and pathologists.

The presentation of sarcoidosis is variable. It may vary from asymptomatic to thoracic or extrathoracic presentation.

Rao et al described in their literature that Isolated extrapulmonary manifestations of sarcoidosis

occur in only 10% of cases [16].

In our case, though the patient had severe non specific symptoms which can be presentation of tuberculosis, fungal disease and even malignancy. The patients Mantoux test was 3 mm that was suggestive of presence of anergy which is also found in most of the sarcoidosis [17]. The ground glass opacity in chest HRCT and high serum calcium were also suggestive of sarcoidosis.

As there was no respiratory symptom, despite ground-glass opacities in the HRCT chest, we did not perform Bronchoscopy. Serum ACE level is not available in our center. So we could not order it Due to unavailability, we could not perform any immunological tests.

To exclude another differential diagnosis of deep fungal infection, we performed fungal stain and culture of the specimen and it showed no growth.

As the histological diagnosis matched with clinical presentation and serum calcium, so we confirmed it and treated with oral steroid. The patient responded well in oral prednisolone for several months. She reported improvement in fatigability and gained 6 kg weight in four months.

Conclusion

The incidence of sarcoidosis is incrasing in the developing countries like Bangladesh. More over,peripheral lymphadenopathy as the sole and initial presentation of sarcoidosis is rare reported cases even in southeast Asia. The high prevalence of tuberculosis can potentially compound the problem of overlapping clinical presentations of TB and sarcoidosis in Bangladesh.

This case reminds clinicians that sarcoidosis should be considered as an uncommon cause of peripheral lymphadenopathy along with extrathoraic nonspecific symptoms.

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