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Rare presentation of Sarcoidosis

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ABSTRACT

Sarcoidosis is an idiopathic, noncaseating granulomatous disorder with wide systemic involvement. Lungs, eyes, and skin are the organs most commonly affected. Bone involvement, which is very rare, was reported as present in 3 to 13% of affected cases. Here we present a case of young female with ribs, vertebrae and pulmonary involvement, lesion mimicking as metastasis or tuberculosis thus emphasizing the importance of differential diagnosis.

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Introduction

Sarcoidosis is a multisystemic inflammatory disorder of unknown etiology, characterized by T-lymphocyte infiltration, granuloma formation, and impairment of the normal micro-architecture (1). The pulmonary involvement has been found to be the most common presentation, however as a multisystem disorder it can involve any organ including skin, as well as the osteoarticular system. This disorder has been found to have a higher prevalence in young individuals with female predominance (2) and bimodal distribution pattern with peaks in the 3rd and 5th decades.In 1904, Kreibich was the first to describe the involvement of bone in patients with sarcoidosis. He detected multiple radiolucent areas in distal phalanges of second finger in four patients with sarcoidosis(3). The involvement of bone in sarcoidosis has been found to be uncommon with the frequency ranging from 3 to 13% (4,5). The lesions seen may be sclerotic, lytic or mixed type (6). Most commonly the phalanges are involved in the skeletal system causing dactylitis. The other bones can be involved like skull, maxilla, facial bones, vertebrae, ribs and pelvic bones (7).

Skeletal with pulmonary sarcoidosis has been rarely reported. We intend to describe the clinical presentation, radiological

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features and pathological description of the lesions in the patient with skeletal and pulmonary sarcoidosis. This is to direct the importance of sarcoidosis as it can mimic tuberculosis as well as malignancy which require high index of suspicion with multidisciplinary approach to reach the final diagnosis.

Case Report:

A 27 year married female, non-diabetic, non-hypertensive, with no other comorbidity, presented in the pulmonary emergency department with the chief complaints of pain on the right side of the chest since 1 month which was localised to the right anterior side of the chest and was non referring, non-radiating , increased on deep breathing, there was no postural variation. She also complained of fever since last 20 days, undocumented usually low grade, not associated with chills or rigors, with no diurnal variation. It was associated with loss of appetite and weight loss. There was no history of any chronic illness, tuberculosis, trauma in past.

At time of admission she was afebrile with all vitals parameters stable. On respiratory system examination there was mild tenderness all over the right side of the chest wall below mammary region, no lump and swelling or fistula was visible on the chest wall. On palpation, percussion and auscultation there was no other abnormality observed. Cardiovascular, neurological and abdominal system examination were normal.

Patient was admitted and was started on symptomatic and supportive care with empirical antibiotics and analgesics. Routine blood investigations showed mild anaemia (Haemoglobin 10.3 g/dL) with normal leucocyte counts and platelet count. Liver function tests and kidney function tests,serum calcium level were within normal limit. Viral markers were non-reactive.

Chest X-ray showed bilateral hilar prominence with bilateral reticular opacities with involvement of right side more than left side, lytic opacities in the 8th and 9th ribs on the right side (Figure1). We performed contrast CT thorax which showed multiple paralymphatic nodules with mediastinal and bilateral hilar lymphadenopathy, suggestive of sarcoidosis. The CT also showed multiple sclerotic lytic vertebral (D8, D9, D10, and D 12) and lytic cystic ribs(1st, 2nd, 8th, 9th and 12th on right side and 8th rib on left side) lesions suggestive of skeletal sarcoidosis (Figure 2,3).

Serum ACE levels were slightly elevated than the normal limit (76 U/L). Mantoux test was negative. We performed bronchoscopy which showed multiple endobronchial small nodules in the right upper and middle lobe bronchi (Figure 4). The biopsy from the endobronchial nodule showed epitheloid cell granulomas with few Langerhan's giant cells and at places Schaumann like bodies seen with incoscpicuous necrosis, all features were suggestive of sarcoidosis(Figure 5).

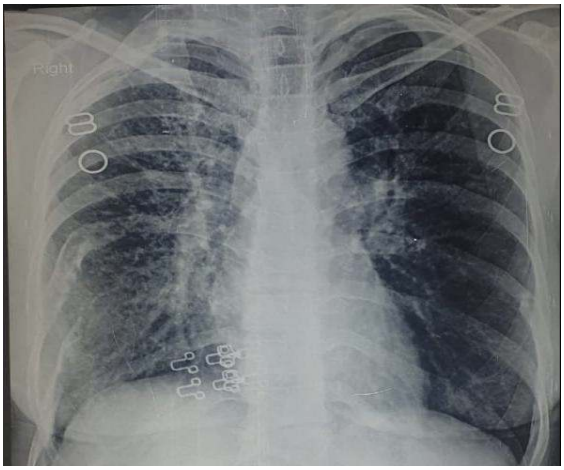

Figure 1: Chest x-ray: reticular opacity in right upper lobe and left hilar prominence

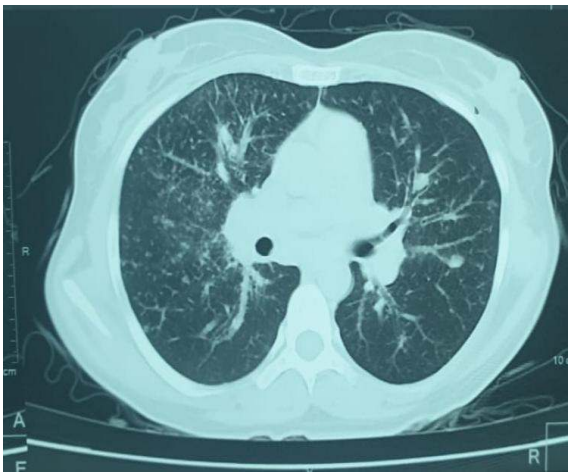

Figure 2: CECT Thorax: paralymphatic nodules

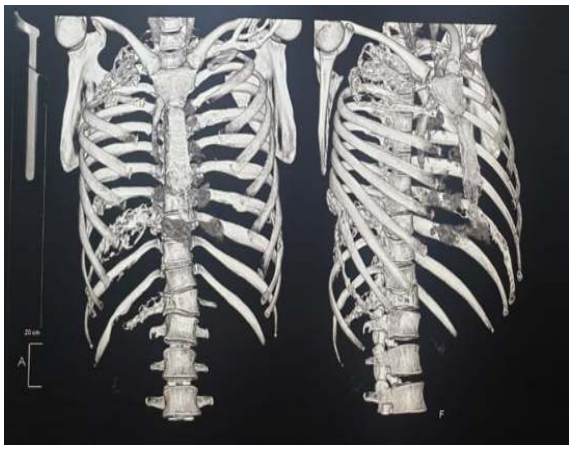

Figure 3:CT Thorax:multiple sclerotic lytic vertebrae(D8,D9,D10 and D12) and lytic cystic ribs(1,2,8,9,12 on right side and 8th rib on left side)

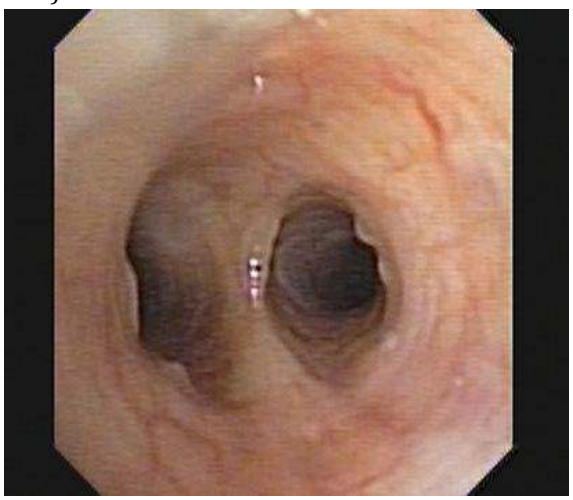

Figure 4: Bronchoscopy image: Endobronchial nodules in right middle lobe

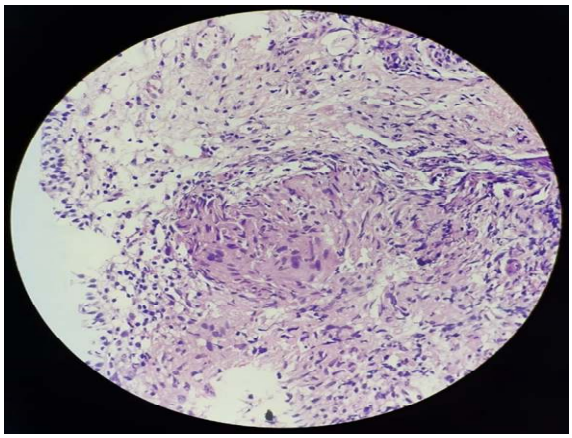

Figure 5: HPE: epitheloid cell granulomas with few Langerhan's giant cells and at places Schaumann like bodies seen with incoscpicuous necrosis, suggestive of sarcoidosis

CT guided biopsy was performed form the ribs which was inconclusive. Patient was advised for the open biopsy from the vertebral lesions which was refused by the patient.

Considering all the evidence patient was diagnosed as the case of skeletal and pulmonary sarcoidosis and was started on oral prednisolone 1mg/kg/day along with symptomatic treatment. Patient's symptoms improved and was discharged. Patient has been doing well and is on regular follow up.

Discussion:

Sarcoidosis, also previously known as Besnier-Boeck-Schaumann's disease, is a systemic disorder. The exact etiology and pathogenesis has yet not fully understood but there is cellular immune system activation with inflammatory response involving almost all the systems. This disorder has been associated with various genetic and environmental factors. It is found more in the younger age groups with female predominance(4). Most common involved organ is the lung followed by the lymph nodes (7). Most common cause of death in sarcoidosis is due to cardiac involvement. Other common cause of mortality reported is due to pulmonary fibrosis leading to respiratory insufficiency (8-10).

The bone sarcoidosis shows lesions that vary from lytic, cystic, sclerotic and mixed type. The lesions can also be invasive causing trabecular irregularity, there may also be periosteal reaction and disseminated lesions. Such varied presentation leads to delay in reaching the diagnosis and management. Sarcoidosis is an important differential for metastatic bone lesions as well as myeloma. High prevalence of tuberculosis in India also adds on to the diagnostic dilemma as bone tuberculosis also has such varied presentation as bone sarcoidosis. The commonly involved bones are the skull, vertebrae, nasal bones and the phalanges which when involved present as dactylitis. Ribs are rarely involved in sarcoidosis (11-14) and in our patient there was involvement of ribs and vertebrae along with pulmonary involvement. The radiological features include cystic lesions, may also present as cortical defects, reticularizations of cortical bones.

Sometimes sclerotic and destructive lesions may also be seen. There may be pathological fractures due to destructive lesions. In MRI T2 sequence shows increased nonspecific signal intensity whereas T1 sequences show decreased nonspecific intensities .Differential diagnosis of bone sarcoidosis also include various other granulomatous diseases of bone such as Langerhans cell histiocytosis, fungal infections , viral infections like Epstein-Barr virus and cytomegalovirus and Ewing sarcoma (15). Pulmonary involvement (80-90%) is commonly seen with osseous sarcoidosis as was present in our case and are more prone to having cutaneous involvement, especially lupus pernio (48-70%), which was not found in our case. After confirming the diagnosis of the bone lesions, it is important to investigate other systems for the involvement. Corticosteroids are mainstay of treatment. There are different alternative drugs also available which are effective in treatment which include chloroquine, methotrexate, azathioprine, leflunomide, cyclosporine, minocycline, cyclophosphamide, anti-TNF and rituximab. Confirmation of the diagnosis is always made by the histopathological examination which shows granuloma in the medullary cavity and destruction in the surrounding bone tissue. Non necrotizing histiocytic granulomas are characteristic for diagnosis. The early and definitive diagnosis is important for adequate treatment of this disease.

Conflicts of interest

The authors have no competing interest.

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