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A Rare Case of Pulmonary and Extra-Pulmonary Sarcoidosis.

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ABSTRACT

Sarcoidosis, considered to be an immunological disorder by many experts, can present either as a pulmonary subtype or as an extra-pulmonary subtype, with the former being more common in clinical practice. Common extra-pulmonary sites include lymph nodes, liver, spleen, heart and skin, in decreasing order of frequency. Presence of rare features in a patient with sarcoidosis can delay the diagnosis, and hence, the treatment as well. This case report highlights a rare presentation of a patient with pulmonary and extra-pulmonary (involving lymph nodes, spleen and musculoskeletal system) sarcoidosis.

Keywords: Splenomegaly, Musculoskeletal, Granuloma, Non-caseating, Schaumann

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INTRODUCTION

Sarcoidosis is an inflammatory disorder characterized pathologically by the presence of non-caseating granulomas in the affected tissues [1]. The most common age group of presentation is 10 to 40 years, with the female sex being affected more commonly (2:1 ratio). Aetiology is unknown [2]. Studies have revealed clues of the aetiology to begenetic, infectious, environmental or nutritional. Current theories suggest that the disease develops in genetically susceptible hosts who are exposed to certain antigens that trigger an overwhelming inflammatory immune response leading to granulomatous reactions, making it an immunological disorder [3].

The most commonly affected sites include lymph nodes, lungs, liver, spleen, heart and skin. It has been reported that almost 20 to 40% of sarcoidosis patients do not have any symptoms, and are incidentally diagnosed on a routine chest X-ray. Others may present with cough, dyspnoea and other respiratory symptoms, as lungs are common sites of sarcoidosis [4].

Here, the authors present a rare case of sarcoidosis affecting the lung as well as a few extrapulmonary sites.

CASE REPORT

A 55-year-old female presented to us with history of non-productive cough, pain at the ankle joint and weight loss (of around 14 kg) over the past 6 months. The patient gave a past history of nephrolithiasis was present. On general examination, she was found to be pale, and also showed axillary and cervical lymphadenopathy. Her per-abdominal examination revealed splenomegaly. She had bilateral ankle joint swelling associated with tenderness.

Laboratory blood tests showed a haemoglobin level of 10 g/dL, an ESR of 51 mm/hour, serum creatinine of 2 mg/dL and serum calcium of 14.9. Also, her parathormone (PTH) level was 9.3, with a phosphorus value of 3.3 and vitamin D of 18. Urine microscopy revealed hypercalciuria and calcium oxalate stones. The patient's calcium level remained high despite anti-hypercalcaemic measures.PTH, phosphorous and vitamin D were low. Hence a bone scan was done,which was normal. Chest X-ray showed bilateral hilar prominences (as shown in Figure 1).

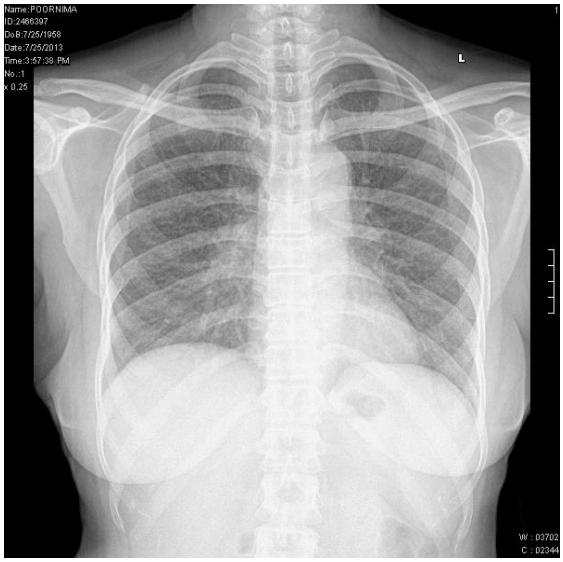

Figure 1: Chest X-ray showing bilateral hilar prominences

A HRCT of thorax was done, which showed multiple smooth and irregular nodular densities in both upper lobes and right middle lobe (as seen in Figure 2), and multiple enlarged pre-tracheal,paratracheal and pre-vascular lymphnodes (as seen in Figure 3).

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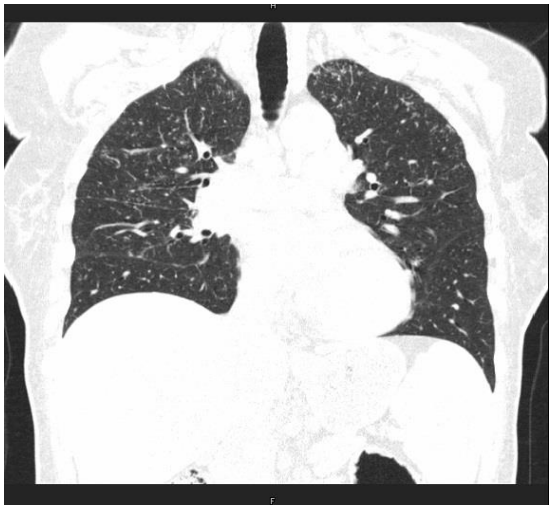

Figure 2: HRCT of thorax showing multiple nodules in both upper lobes and the right middle lobe

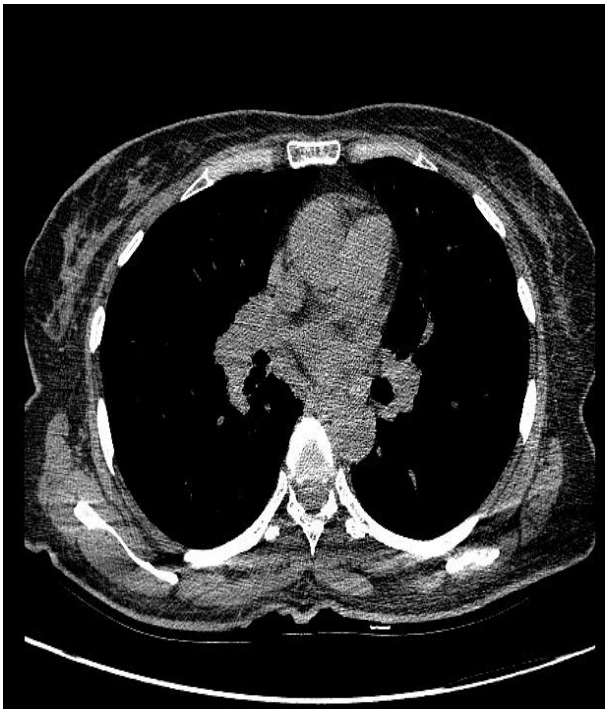

Figure 3: HRCT showing enlarged lymph nodes

The most likely differential diagnoses considered at this time were tuberculosis, sarcoidosis and lymphoma. Axillary lymph node biopsy was done. Histopathology of this specimen revealed non-caseating granulomas with asteroid and Schaumann bodies, consistent with the diagnosis of sarcoidosis.Serum ACE levels were measured, which turned out to be high (102) and the diagnosis of sarcoidosis was confirmed.

Following this, the patient was started on corticosteroids.She improved clinically, and was discharged on oral steroids.In the subsequent follow-up visits, the dose of steroids was tapered and continued. During the last follow-up, the patient was asymptomatic, and repeat laboratory tests were within normal limits.

DISCUSSION

Although the most common age for presentation of sarcoidosis is below 40 years, our patient was 55 years old at the time of diagnosis.Lungs are predominantly affected in sarcoidosis, but extra-pulmonary

involvement can occur in association with, or in the absence of pulmonary involvement. Also, isolated extra-pulmonary involvement can occur only in 10%of cases [5].

Hepato-splenic and musculoskeletal involvement are rare presentations of sarcoidosis, as seen in the present case [6]. Splenomegaly is the most common presentation, if there is splenic involvement in sarcoidosis. Spleen is usually homogenous, but rarely, multiple nodular lesions can be noted. These nodules can be mistaken for infections, lymphoma or metastasis of carcinomas [7].

Other granulomatous disorders to be excluded before confirming the diagnosis of sarcoidosis include lymphomas, Wegener's granulomatosis, berylliosis and pneumoconiosis [8]. Also, other causes of hypercalcaemia like hyperparathyroidism, malignancy and thyrotoxicosis are to be ruled out. A combination of bilateral hilar lymphadenopathy, histopathological detection of non-caseating granulomas and exclusion of other diseases with similar presentation suggested the diagnosis of sarcoidosis in our patient.The unusual feature of this case was splenomegaly and musculoskeletal involvement along with pulmonary involvement, which made it a rare presentation of sarcoidosis.

CONCLUSION

To conclude, this case highlights the importance of considering sarcoidosis as one of the several differential diagnoses when an elderly patient presents with hypercalcaemia and bone pain, even though the probability of malignancy is higher.

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