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CASE REPORT

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A rare form of sarcoidosis: a case of bone sarcoidosis

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Abstract

Sarcoidosis is an inflammatory disease which is characterized by chronic, systemic noncaseating granuloma formation. This disease generally affects the respiratory system. Bone lesions in sarcoidosis are developed in the cases with 3-13 percentage that generally asymptomatic. In this study we presented a patient of sarcoidosis that had been pulmoner symptoms a long time, and then diagnosed with excisional biopsy from her rib bone.

Keywords: Sarcoidosis, bone involvement, granuloma, rib

Introduction

Sarcoidosis is an inflammatory disorder characterized by noncaseating granulomas within tissues. Although the etiology of sarcoidosis remains unknown, infectious, occupational agents, and factors associated with the seasons of the year are most likely involved (1-4). It is a multisystem disease that most frequently involves lungs, lymph nodes, skin, and eyes (5). The presenting features are usually relatively non-specific general signs of weight loss, asthenia, fever, deterioration in general health and evidence of mediastinal lymphadenopathy in suggestive sites, often associated with parenchymal disease. Note that 50% of patients suffering from thoracic sarcoidosis are asymptomatic.

The musculoskeletal system is involved in 1% to 13% (average, 5%) of cases (6). Rheumatic manifestations of sarcoidosis, although rare, include inflammatory arthritis, periarticular soft tissue swelling, tenosynovitis, dactylitis, bone involvement, sarcoid myopathy, and bone loss. (7)

Arthralgia, along with synovitis and periarthritis, dominates the clinical picture of sarcoidal arthropathy. Acute arthritis presents as symmetric, migratory, or additive polyarthritis that often involves the knees, ankles, wrists, elbows, and the proximal interphalangeal joints and may be associated with Lofgren's syndrome.

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Bone lesions develop in 1% to 13% of patients with sarcoidosis with an estimated average of 5 %.(8,9,10). Classic bone lesions in sarcoidosis are described in the small bones of hands and feet and are typically lytic lesions, which may lead to fracture, bone collapse, and deformity (1,8,9). The important diagnostic clues are often cysts in the phalanxes, which are seen in 14% of patients. The specific pattern of involvement is most evident on radiographic images, especially the images from middle and distal phalanxes. Soft tissue swelling may present with or without skin lesions on the proximal interphalangeal joints and middle phalanxes.

Any bone, including the skull, ribs, nasal bone, and long bones, can be affected in Positron emission tomography/computed tomography (PET/CT) imaging can be useful in the assessment of bone involvement in sarcoidosis patients. (11)

Case

A 52 year old female patient who has been suffering from cough, exhaustion and backache for seven years has been examined by the chest diseases clinics and pre-diagnosed with lung tuberculosis, sarcoidosis. The patient had a history of tuberculosis at the age of ten. The patient who couldn't be diagnosed despite all the medical examinations started to have increasing pains in her right hemithorax. In the Torax CT (computed tomography) in 2014 a lesion causing parenchymal protrusion on the side of the mediasten containing calcifications of 2.5x2 cm on the 6th rib in addition to the multiple lymph nodes in mediastinum and in both lungs was detected. Then rib resection was done to the patient in March, 2015 in our hospital. After the resection the patient was diagnosed with bone

sarcoidosis due to the non-caseating granulomas found out in the pathology and she was directed to our clinic for the follow-ups. The pain was evaluated as VAS:8. Besides the rutin blood tests, graphs (X-Rays) of the hands and the feet and pelvis AP were taken. In the graphs there weren't any cystic, lytic, sclerotic etc. lesion. She had a control Thorax CT in which it has been observed that the mass generating protrusion to the lung in the 6th rib was regressed. After the whole body bone scintigraphy it has been interpreted that the findings of the anterolateral of the right 5th and 6th costas can depend on the postop change and that the diffuse heterogeneous involvement can depend on the degenerative process besides the sarcoidosis involvement. After all the examinations the bone sarcoidosis diagnosis of the patient was confirmed and treatment of prednisolone 7.5 mg per day and methotrexate (MTX) 7.5 mg per week was started. In the follow up visit 15 days after the discharge the patient mentioned that she had less pain and the pain was evaluated as VAS: 4 with action. Followed in MTX dose of 15 mg a week was removed. MTX was discontinued in patients because it's bone pain continued.30 mg corticosteroid and 100 mg azothioprin was started. Then 5 mg of corticosteroid was tapered per week.The patient's bone pain was significantlydecreased in control of sixth months. Not additional problems in the patient's follow-up.

Discussion

Sarcoidosis is a multisystem disorder characterized by noncaseating granulomatous infiltration. Sarcoidosis can hardly diagnosed since the findings of the bone lesions cannot be found out easily.

Our case had pulmonary symptoms such as cough and exhaustion. She had a lymph node biopsy form the mediastinum but she couldn't be diagnosed. After the biopsy taken from the mass in the right 6th rib, the patient was diagnosed with bone sarcoidosis due to the noncaseating granuloma. This involvement is rare.

The histological findings of sarcoidosis in the bone are precisely the same as they are in other tissues of the body. The lesion of sarcoidosis is a focal well-defined granuloma formed by the accumulation of epithelial cells, multinucleated giant cells, lymphocytes, macrophages and fibroblasts.

The reported incidence for osseous sarcoidosis varies widely between 3 to 13% on conventional imaging by plain radiographs and 39 % using the combination of bone scintigraphy and plain radiographs.(12,13) However, the prevalence is unknown. In the era of frequent use of MRI, CT and technetium bone scans osseous sarcoidosis lesions are found more often and may be revealed as incidental findings in a subject imaged for other indications. Uncommonly, sarcoidosis bone lesions resembling bone metastases may be the initial presentation of undiagnosed disease.(14,15) As in other forms of sarcoidosis, a biopsy is required to establish the diagnosis of osseous lesions. It is not possible to distinguish granulomatous bone involvement from other causes by MRI or other imaging studies, making biopsy necessary for diagnosis.(16)

With torax CT we had found out the granuloma generating protrusion to the lung in the 6th costa of our patient who had pain in her right hemithorax and after the resection we realized that the mass was lost in the control Torax CT.(Fig. 1-2)

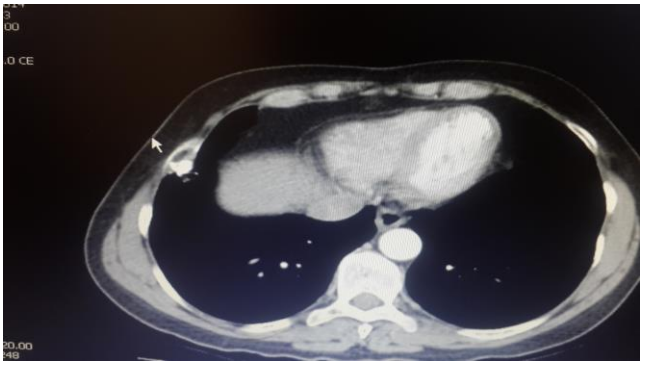

Figure 1

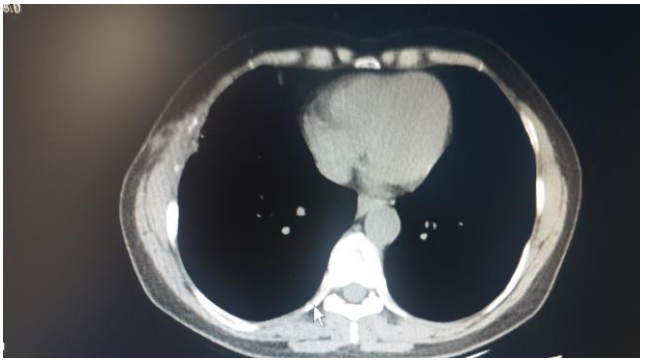

Figure 2

Conventional radiography usually reveals the location of sarcoid bone lesions in the small bones of the hands and feet. The bony lesions are usually lytic (also called bone cysts, but the term is the misnomer) as the result of osteolysis. The following radiographic features of 136 cases of osseous sarcoidosis involving hands and feet described by Yaghmai: diffuse marrow infiltrate (identified better by bone scintigraphy), punched-out lesions, permeative lesions (reticular pattern), destructive lesions (dactylitis) and sclerotic lesions (rare, usually seen in the late stage of disease after long-term treatment). (17)We didn't encounter any bone involvement such as lytic, sclerotic, diffuse etc. in the hand and feet graphs of our patient.

ESR(erythrocyte sedimentation rate) , alkaline phosphatase and serum calcium are frequently normal, but may be elevated. ACE(angiotensing converting enzyme) level has

been found to be elevated in up to 75% of sarcoid patients, but has limited specificity in osseous sarcoidosis. The ESR, alkaline phosphatase ,serum calcium and ACE levels were among the normal range for our patient.

There are two major controversies in the treatment of skeletal sarcoidosis. The first is whether asymptomatic lesions that have been revealed by sensitive imaging modalities but are not evident on radiographs or CT should be aggressively treated with corticosteroids. The course of the disease is quite variable, and spontaneous improvement has sometimes been reported. (16,18) Furthermore, there are no data indicating that sustained asymptomatic lesions can induce osteolysis in the long term. Therefore, to date, therapy is indicated only in cases of painful and destructive skeletal sarcoidosis. The second controversy is over the optimal test or sequence of tests that should be used to assess the response to the treatment of skeletal sarcoidosis. There are several reports that elevated ESR and ACE levels normalize after successful therapy. (16,19) Additionally, a follow-up technetium-99 m methylene diphosphonate (Tc-99 m MDP), Gallium-67 (Ga-67) scan and Fluorine-18 (F-18) fluoro-2-deoxyglucose (FDG) positron emission tomography (PET) may reveal reduced uptake post-treatment.(19,20)

Conclusion

In conclusion, long bone and vertebral especially costas involvement on sarcoidosis is a not so rare. We report that a patient with rib involvoment of sarcoidosis.

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