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Case Report

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An Interesting Case with Extrapulmonary Manifestations of Sarcoidosis

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Abstract

Sarcoidosis is characterized by a various courses and clinical presentation and in this era, some patients may have some symptoms difficult to establish the sarcoidosis diagnosis. Here we report a new case of sarcoidosis with some learning points for the clinicians. We report a 39-year-old male patient attending to the clinic due to productive cough, dyspnea, and skin lesions as papule as well as erythematous plaques in extensor regions of elbows and knees. Regarding the biopsy results of skin lesions, hillar, mediastinal lymphadenopathy, and serum ACE level the sarcoidosis was proposed. Fatigue, spinal pain, and paraplegia plus paraparesis of lower limb, tachycardia, multiple lesions at the lumbar spine, skin lesions, and posterior fossa toxoplasmosis were the findings at later visits, representing a confusing course. Finally, according to the presentation of the case reported in this paper, we concluded that the patients with sarcoidosis may present with multiple extrapulmonary manifestations that would result in some confusing points for diagnosis. Prompt clinical and imaging assessment would result in faster diagnosis and better prognosis to decrease the burden of disease.

***Keywords*:** Extrapulmonary Manifestation, Sarcoidosis

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Introduction

Sarcoidosis is a multi-system disease of unknown cause with fluctuations in its course and severity, characterized by formation of some granulomas in a variety of organs, essentially in the lungs and lymphatic system (1-3). Specific genetic polymorphisms may be associated with increased risk of sarcoidosis or may affect the disease presentation (4). Mortality rate, mainly due to pulmonary fibrosis, is higher in patients with sarcoidosis than normal people (1, 2).

Diagnosis of sarcoidosis is based on the clinical-radiological representation as well as histopathological evidence of noncaseating granulomas. Other diseases mimicking the sarcoidosis, mostly infections and other granulomatoses, should be excluded before the final diagnosis (5, 6). Sarcoidosis is characterized by various courses and clinical presentation (1-3), and in this era,

some patients may have some symptoms difficult to establish diagnosis of the sarcoidosis. Here we report a new case of sarcoidosis with some learning points for the clinicians.

Case Presentation

We report a 39-year-old male patient attending to the clinic due to productive cough, dyspnea, skin lesions as papule, and erythematous plaques in extensor regions of elbows and knees. Regarding the biopsy results of skin lesions, hillar, mediastinal lymphadenopathy, and serum ACE level, the sarcoidosis was proposed. Also the blood urea, CK-MB, LDH, and CRP levels along with leukocyte and platelet counts were high but the serum calcium level was low.

At the next visit, we observed that he had fatigue, spinal pain, and paraplegia plus paraparesis of lower limb and without reflexes. The diagnosis was Guillain-Barre syndrome, and plasmapheresis were performed five times leading to significant improvement. During hospital stay, the patients developed tachycardia. For this matter, echocardiography was performed that revealed right atrial mass probably clot or mass. Further assessment with cardiac MRI (Figure 1) ruled out the clot and showed mobile pedunculated mass close to IVC accompanied by reduced LV systolic function (EF=35%) and severely reduced RV systolic function (EF=19%). The mass size was reduced with steroid therapy, according to serial cardiac MRIs.

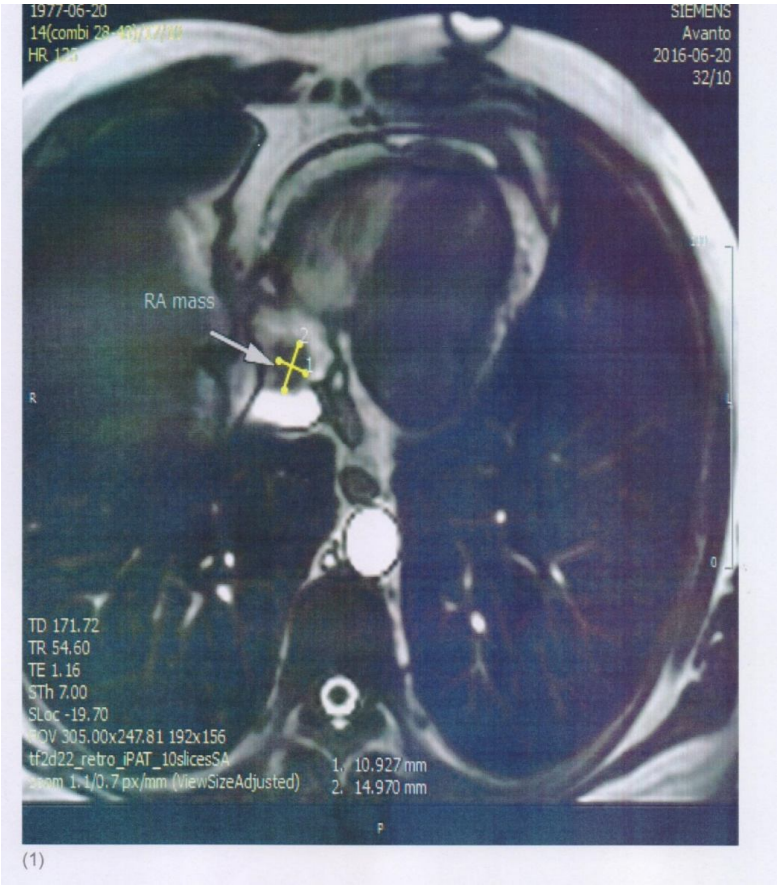

Fig. 1. Cardiac MRI showing mobile pedanculated mass close to IVC

Whole body bone scan and MRI revealed multiple lesions at lumbar spine, showing sarcoidosis or metastasis (Figure 2). The lesions were relieved in next MRI after steroid therapy. Also the ophthalmic scan was performed and showed no abnormal finding.

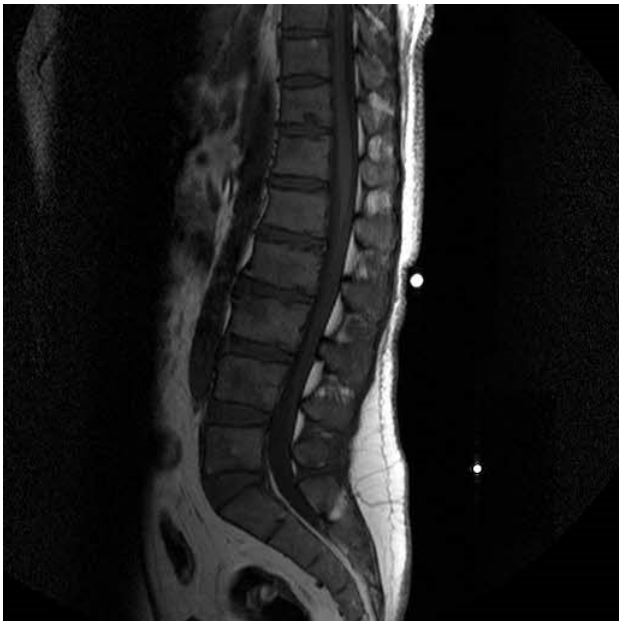

Fig. 2. Multiple lesions at lumbar spine showing sarcoidosis

The CT scan results also hypothesized presence of metastasis and after craniotomy, the diagnosis was toxoplasmosis. At last admission for headache, imbalance, and multiple skin masses, the diagnosis was posterior fossa toxoplasmosis (Figure 3).

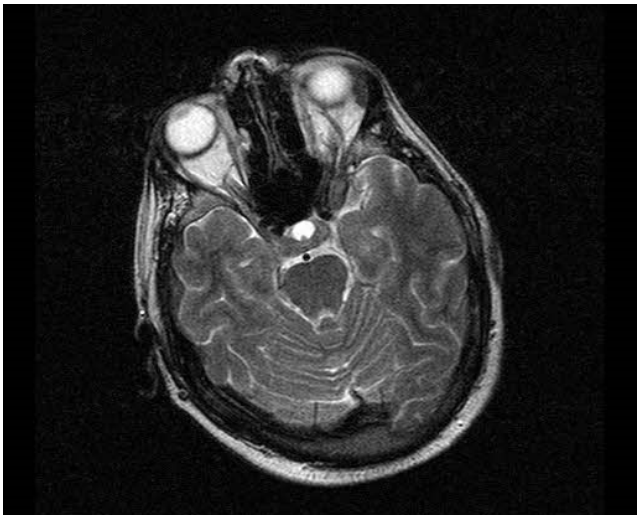

Fig. 3. Posterior fossa toxoplasmosis

The patients underwent craniectomy. There were abscess-form lesions in the abdominal and chest walls and thigh with pus drainage and staphylococcal positive culture. Also the skin lesions were due to staphylococcal infection, that was treated with antibiotic therapy and drainage and the patient was discharged with good general condition. At the last follow-up visit, the patient was asymptomatic and the steroid dose was reached to 5 grams daily.

Discussion

Though the diagnostic criteria have not changed, sarcoidosis remains yet a diagnosis of the exclusion process, best supported by biopsy specimens from the tissue, which demonstrates noncaseating granulomas in the patients with compatible radiological and clinical features of the disease (7, 8). Sarcoidosis is typically accompanied with pulmonary findings in more than 90% of the patients (9). However, our reported patient had some respiratory complaints such as cough and dyspnea; the pulmonary findings were not clearly obvious unless the hillar and mediastinal lymphadenopathy.

Extrapulmonary involvement is common in sarcoidosis, and all organs may be involved (especially lymph nodes, joints, eyes, and central nervous system), although isolated extrapulmonary disease is rare (9). Skin manifestations such as papules and plaques were the major findings in our patient. However the skin lesions would mimic various common dermatological disorders, resulting in confusion about the diagnosis and also some problems for the management (10). The presence of skin granuloma may be useful clinical finding (11).

The diagnosis of sarcoidosis is difficult and the typical laboratory findings of sarcoidosis such as ACE are not diagnostically present (9). Cardiac involvement may be found in approximately five percent of the patients with sarcoidosis (12). The course of the disease is variable, ranging from benign arrhythmias to complete heart block or sudden death, and the diagnosis is difficult (12). However, the tachycardia in our patient led to assessment with echocardiography and finding the disease.

The neurological manifestations of disease are also confusing. Posterior fossa toxoplasmosis is an extremely rare finding in the patients with sarcoidosis. However, Kaur et al. (13) reported in their case report study a patient with neurosarcoidosis mimicking lymphoma. Also, the Guillain-Barre syndrome secondary to sarcoidosis was previously reported by Fahoum et al. (14). It is usually a subacute or chronic sensorimotor axonal polyneuropathy (14), and the intravenous immune globulin and prednisolone would lead to progression discontinuation as also been seen in our reported patient (15). Also, some authors have reported patients with sarcoidosis who had cardiac masses either in the ventricles or atria at left or right sides (16-18).

Finally, according to the presentation of reported case in this paper, it may be concluded that the patients with sarcoidosis may present with multiple extrapulmonary manifestations that would result in some confusing points for diagnosis. Prompt clinical and imaging assessment would result in faster diagnosis and better prognosis to decrease the burden of the disease.

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This work has no contributors to recognize.

Conflict of interest

No bias in the study.

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