SYMPTOMATIC SARCOID MYOPATHY IN A CASE OF PULMONARY SARCOIDOSIS

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ABSTRACT

Sarcoidosis is a granulomatous disorder with a propensity of involving nearly all the systems of the body, however it predominantly affects the lungs. Muscle involvement in sarcoidosis is a highly uncommon occurrence. Here we present a rare case of a middle aged woman with Computed Tomography (CT) features suggestive of pulmonary sarcoidosis, who complained of weakness in the limb muscles,was suspected to have sarcoid myopathy as per Magnetic Resonance Imaging (MRI) findings. Histopathology confirmed the diagnosis. These findings prompted us to report this rare case of sarcoid myopathy.

Keywords: Sarcoidosis, myopathy, CT, MRI

Case Report

A 40 year old woman presented with a prolonged history of shortness of breath and was thereafter diagnosed to have pulmonary sarcoidosis as per her HRCT (High Resolution Computed Tomography) findings. CT showed extensive pleural nodules. She also complained of progressive weakness in both lower and upper limbs for the last few months. However, there was no complaint of any associated pain. On examination, she was found to have proximal muscle weakness with diminished power in the leg muscles. On MRI examination, she was found to have altered signal intensity in the muscles and intermuscular septae in T2 weighted images,which later showed enhancement in post contrast T1 FS images, suggesting muscle involvement. Histopathology revealed typical non-caseating granuloma as expected. Thereafter she was diagnosed to have sarcoid myopathy on the basis of imaging and histological findings in a prior setting of pulmonary sarcoidosis. On consequent treatment with steroids she has been gradually improving.

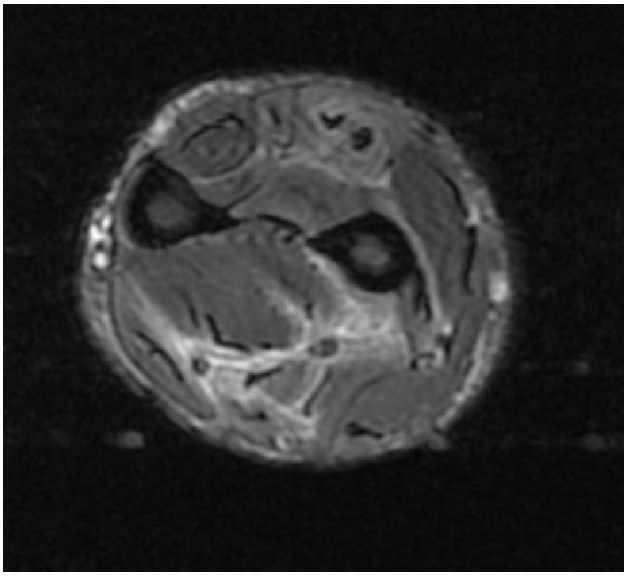

Figure 1: Axial T2 weighted image showing altered signal intensity in muscles and intermuscular septae.

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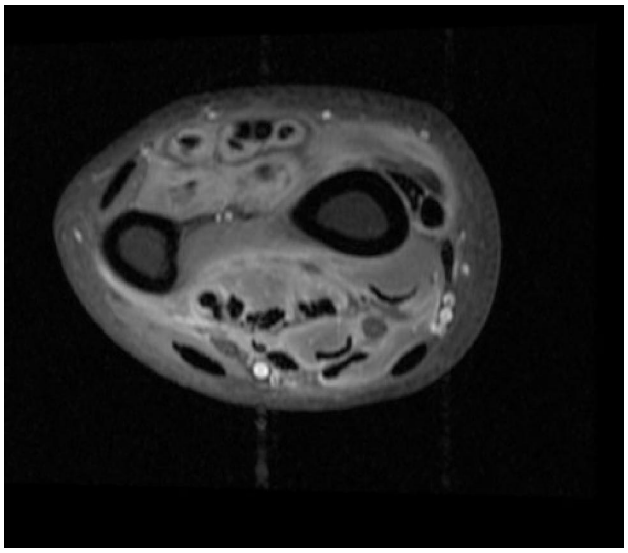

Figure 2: Axial T1 FS post contrast image showing enhancement in muscles and intermuscular septa.

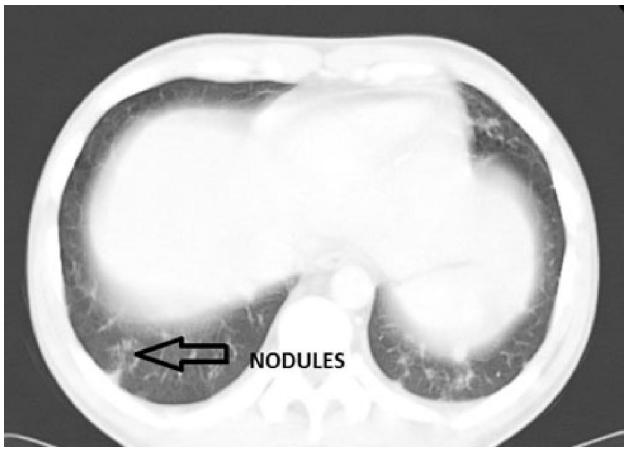

Figure 3: HRCT showing lung nodules along diaphragmatic pleura

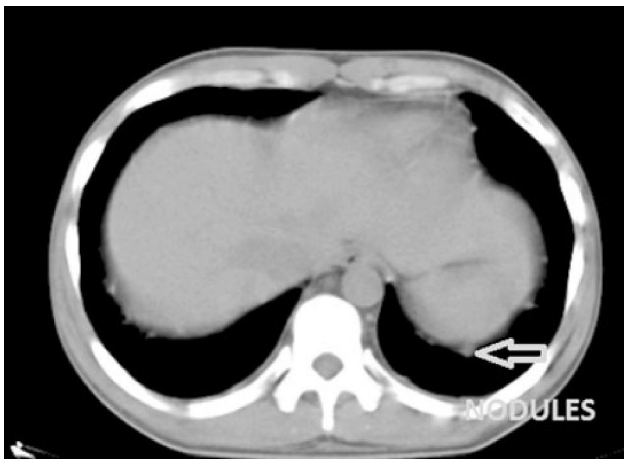

Figure 4: CT showing nodules inbronchovascular structures

Discussion

Sarcoid myopathy was initially described in 1908 by Licharew[1] in a 17 year old girl. Asymptomatic myopathy associated with sarcoidosis is seen in about 20-75% cases, however symptomatic disease occurs only in 0.5% cases.[2,3,4] Sarcoid myopathy is broadly divided into three clinical types:[2,5]

1. Acute myositis-patient presents with myalgia. This variant usually occurs early in the course of the disease. No definite imaging findings have been described. Patients are mostly diagnosed on clinical grounds.
2. Chronic myopathy: Here the patient presents with progressive weakness of muscles. Magnetic Resonance Imaging (MRI) reveals features of muscle atrophy and consequent fatty infiltration. Examination of our case revealed altered signal intensity in the muscles and intermuscular septae in T2 weighted images, which later showed enhancement in post contrast T1 FS images,suggestive of the myositis. Muscle odema may be seen (in T2 or inversion recovery sequences) as well. Also, pseudohypertrophy and contracture may be uncommonly found. Diagnosis is primarily based on imaging, blood muscle enzymes and histology.
3. Nodular variety: This is the rarest variant and patient presents with multiple palpable nodules, which may or may not be painful. MRI may reveal the following four types:[6,7] a)nodules oriented along the long axis of muscles; b) a 'star' shaped lesion in the center of the nodule, which has low signal intensity on all axial pulse sequences; c) central area shows lack of enhancement after contrast administration whereas peripheral area shows rim enhancement; d) 'Three stripes' appearance in coronal and saggital imaging (inner stripe of decreased signal intensity and outer stripes with comparatively higher signal intensity in contrast enhanced T1 weighted images).

Conclusion

Symptomatic muscle involvement in sarcoidosis is a rare entity with few cases reported in literature. Imaging in the present scenario has significantly contributed in diagnosis of sarcoid myopathy especially in those cases where histology is equivocal.

Ours is such a case where the diagnosis was based on imaging findings and on consequent treatment with steroids she has showed significant improvement. Thus in this case imaging solely has played the diagnostic role. This along with the fact that sarcoid myopathy is itself a rare entity prompted us to report such a unique case.

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