SKELETAL SARCOIDOSIS: AN UNCOMMON IMAGING FINDING IN MULTISYSTEM SARCOIDOSIS

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Introduction

Sarcoidosis is a chronic inflammatory disorder of an unknown etiology which can involve multiple systems, by formation of non-caseating granulomas. The lungs are the commonly affected organs and skeletal system sarcoidosis are uncommonly reported (1-13% of reported sarcoidosis patients[2,5]). In about 50% patients sarcoidosis is an incidental finding[3].
Our report highlights a histologically confirmed case of sarcoidosis with axial skeletal involvement which is an uncommon finding[2,5].

Case presentation

A 45 year old male, on follow-up for diabetes mellitus for 12 years, presented with dry cough and postural dizziness for 2 months with significant loss of weight. There was no history of evening pyrexia or loss of appetite. On examination, he was found to be normotensive, with hepatosplenomegaly, cervical and inguinal lymphadenopathy which were multiple discrete nodes which were non-tender and bilateral. Neurological examination revealed that proprioception was affected in bilateral lower limbs.

On investigation his haemoglobin was 10.8mg/dL with normochromic normocytic cells which was likely due to a chronic illness. Creatinine and liver function tests were within normal range. ESR was elevated at 88mm/hr and the LDH level was normal at 148 U/l. His serum uric acid levels were increased at 9.5mg/dL and both total calcium (3.89mg/dL) and ionized calcium (2.6mg/dL) were elevated. Thyroid profile and iron profile were both normal. ACE level was also normal.
EMG revealed abnormalities confined to lower limbs and suggest chronic, bilateral, multiple lumbo-sacral root, plexus, proximal nerve involvement with a differential of diabetic lumbo-sacral plexopathy or sarcoidosis associated non compressive plexopathy
Echocardiogram was normal with ejection fraction of >60%

The chest x-ray revealed bilateral, symmetrical hilar shadowing with reticular shadows in lower zones.

The abdominal ultrasound reveled an enlarged liver with coarse echotexture without focal lesions. Para-aortic lymphadenopathy was identified with bilateral inguinal lymphadenopathy. No splenomegaly was identified.

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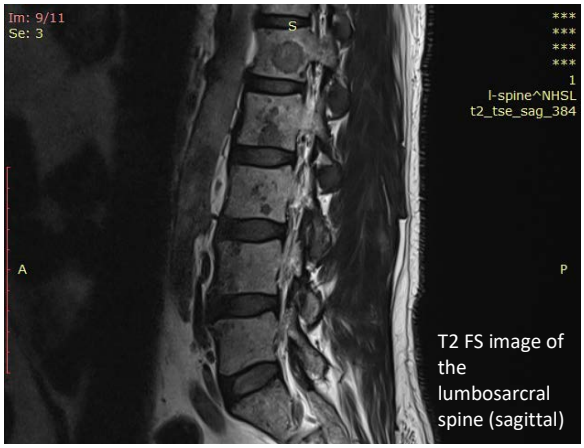
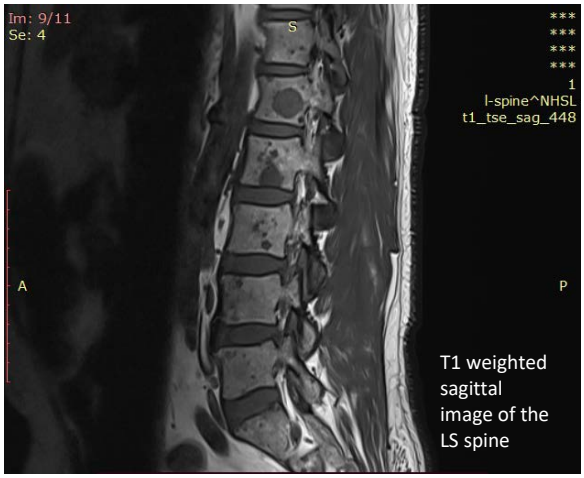
There was diffuse enlargement of bilateral parotid glands on ultrasound of the parotid region. Ultrasound of the neck revealed multiple thyroid nodules.

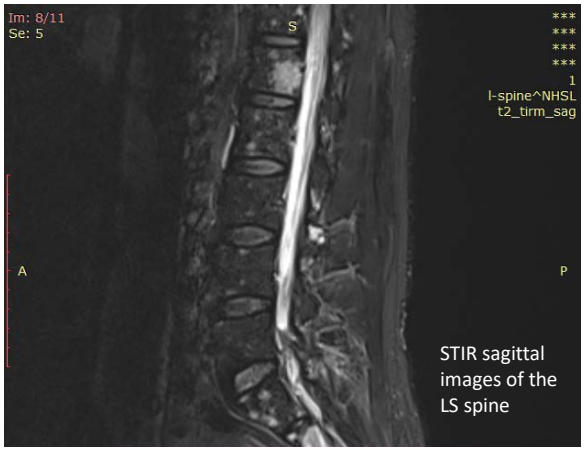
HRCT chest revealed multiple enlarged lymph nodes in pretracheal, paratracheal, prevascular, precarinal, paraaortic, subcarinal and bilateral hilar groups with lung nodules distributed in a peri-fissural, peri-lymphatic distribution predominantly in the right upper and mid zones. Pleural thickening was noted in the right lung base.
The CECT chest, abdomen and pelvis showed no enhancing/ enlarged lymph nodes in the neck, chest findings similar to the HRCT, mild enlargement of the liver with multiple non enhancing, sub-centimeter nodules scattered throughout the liver, prominent spleen with multiple on-enhancing sub-centimeter areas throughout the spleen. There were multiple enlarged, mildly enhancing lymph nodes in coeliac axis, hepatoduodenal, pancreatoduodenal, retrocaval, aortocaval, paraaortic, iliac, obturator, and superficial inguinal lymph nodes. Imaged bones appeared normal except for degenerative changes.

Diagnosis was made of a systemic granulomatous process more in favour of sarcoidosis with a differential of lymphoma.
MRI lumbosacral spine was performed which showed multiple lesions of varying sizes in the lumbo-sacral spine including vertebral bodies as well as posterior elements, with T1 and T2 hypointensity which were hyperintense on STIR imaging. Para aortic lymphadenopathy was also noted. After reviewing the CECT chest, abdomen and pelvis, the diagnosis was made in favour of lymphoma with suggestion of biopsy from lymph nodes.

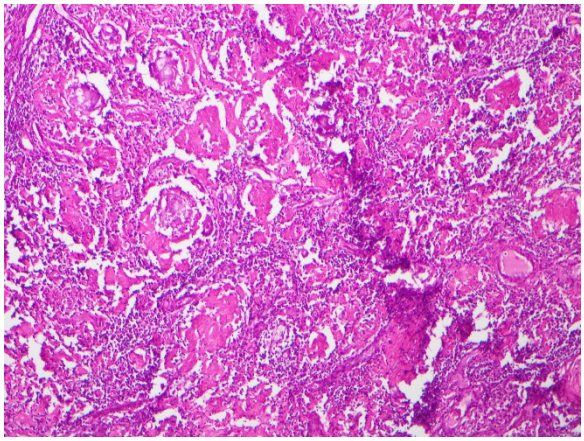


T2 FS image of the lumbosarcral spine (sagittal)



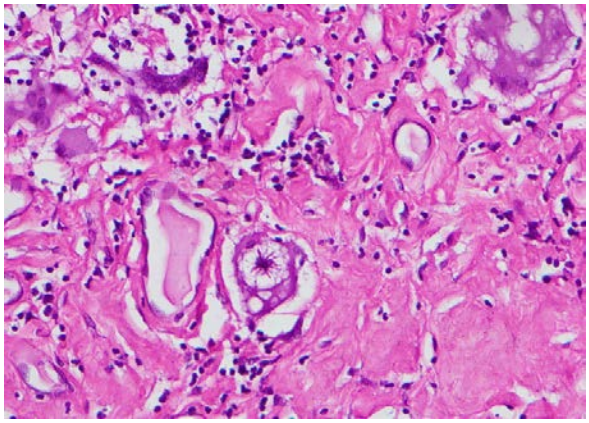


Left inguinal lymph node biopsy was performed which showed granulomatous inflammation compatible with sarcoidosis. Gene xpert for tuberculosis and culture for tuberculosis were negative in the sample.

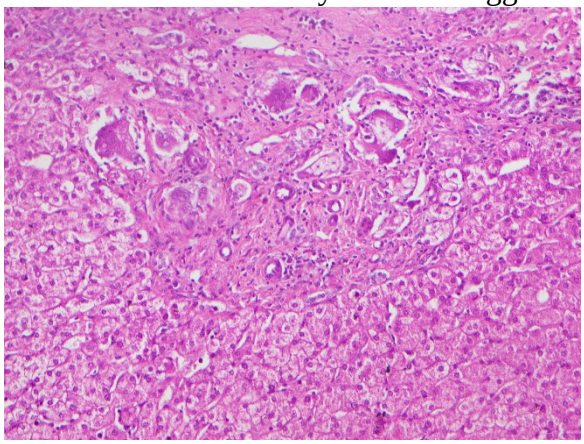


Granuloma in the lymph node biopsy specimen

A para-aortic lymph nodal and liver biopsy were also performed, both of which revealed non caseating granulomatous inflammation most compatible with sarcoidosis.



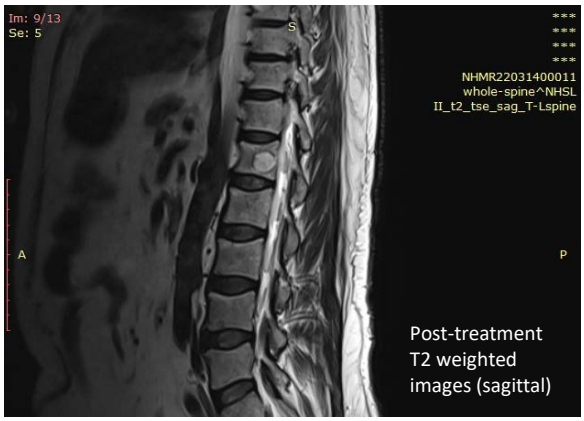
Presence of asteroid body which is suggestive of sarcoidosis

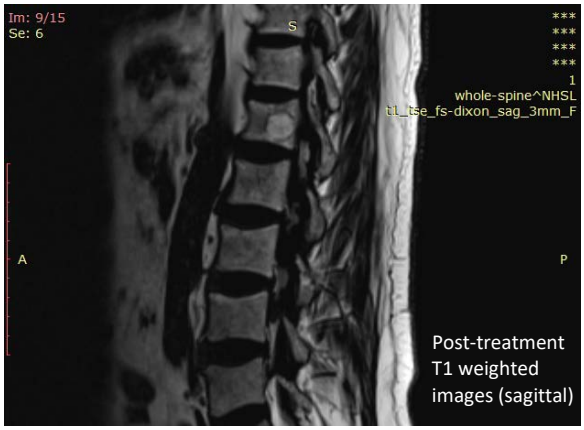


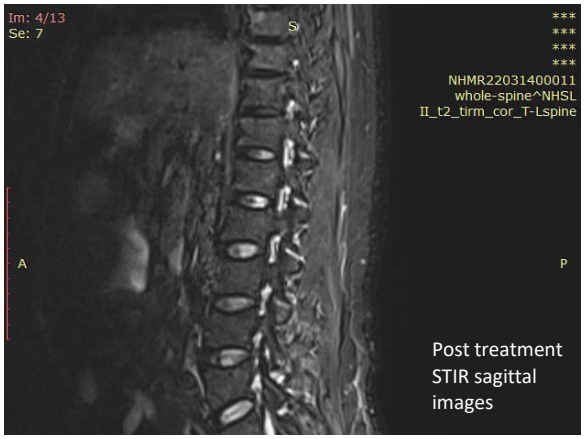
Non caseating granuloma in the liver

The patient was started on steroids and immunosuppressant and MRI spine was repeated after about a year.

The MRI revealed the presence of previously noted lesions in the vertebral bodies and posterior elements but with reduction in the number and some with changes in signal intensity from T1 and T2 hypointensity to T2 hyperintensity. Para-aortic lymphadenopathy was identified, albeit to a lesser extent.







Discussion

Diagnosis of sarcoidosis is made using clinic-radiological data supported by histology with biopsies from sited like lymph nodes and liver. As the histology shows non-caseating granulomas, tuberculosis and other granulomatous disorders need to be excluded. Other supportive evidence include ACE level and presence of hypercalcemia.

The axial skeleton appearance of sarcoidosis can be quite similar to metastatic deposits and can be misdiagnosed. The large bone lesions of skeletal sarcoidosis could be lytic or sclerotic, indistinct or well marginated and of varying sizes[2].

In this case, the patient was being investigated for weight loss and cough and was found to have mediastinal lymphadenopathy with pulmonary changes (stage II). Para-aortic lymphadenopathy, liver and splenic involvement was also identified. Due to the back pain, when the spine was imaged multiple bone lesions with T1 and T2 hypointensity with STIR hyperintensity were noted. Due to supra and infra diaphragmatic involvement, involvement of liver, spleen, lungs as well as bone, the main diagnosis was lymphoma. Hence the biopsies were performed, which revealed features in favour of sarcoidosis. The serum ionized calcium and total calcium levels both were elevated. ESR was elevated as well. Tuberculosis was excluded by gene expert testing.

The post treatment MRI revealed varying T1 and T2 intensity lesions which have reduced in size compared to previous MRI suggesting good response to treatment.

Acknowledgement

Department of Pathology, National Hospital of Sri Lanka for the use of histological images

Consent was obtained from the patient after explanation of the contents of the article.

References:

1. Pulmonary Sarcoidosis: typical and atypical manifestations at high resolution CT with pathologic correlation, Eva Carido et al., Radiographics 2010; 30: 1567-1586, DOI 10.1148/rg.306105512
2. Musculoskeletal sarcoidosis: Spectrum of Appearances on MR Imaging, Sandra L Moore et al., Radiographics 2003; 23: 1389-1399, DOI 10.1148/rg236025172
3. Radiological manifestations of sarcoidosis in various organs. Takayashi Koyama et al. Radiographics 2004, 24:87-104, DOI 10.1148/rg.241035076
4. Clinical and radiological features of extrapulmonary sarcoidosis: a pictorial essay, Stefano palmucci et al, Insights imaging (2016) 7:571-587. DOI 10.1007/s13244-016-0495-4
5. Sarcoidosis from Head to Toe; What the Radiologist Needs to Know. Dakshinamoorthy Ganeshan et al. RadioGraphics 2018; 38:1180–1200 DOI 10.1148/rg.2018170157