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Radio-Diagnosis

A RARE CASE OF ISOLATED NEUROSARCOIDOSIS OF SPINE WITH TRIDENT SIGN.

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ABSTRACT Sarcoidosis is an inflammatory multisystem disorder of unknown cause. Most commonly, it affects the lung. Involvement of the nervous system is 5-15% of patients. Neurosarcoidosis is a serious and commonly devastating complication of sarcoidosis.Spinal cord involvement in neurosarcoidosis is extremely rare. The disease with nondiagnostic imaging and laboratory features, clinical presentation and absence of systemic involvement makes it challenging and delay in diagnosis of spinal cord involvement of neurosarcoidosis.Sequence of spinal cord involvement is Cervical>Thoracic>Lumbar.Here we describe a case ofneurosarcoidosis of spine with trident signpresenting with symptoms of myelopathy.

KEYWORDS : Sarcoidosis, neurosarcoidosis, spine, myelopathy, magnetic resonance image, trident sign.

Introduction

Sarcoidosis is a multisystem noncaseous granulomatous disease commonly affecting lungs, lymph nodes, skin and eyes. Neurosarcoidosis is a rare complication of sarcoidosis. Isolated spinal cord neurosarcoidosis in the absence of systemic disease or intracranial involvement is exceptionally rare (<0.5% cases).According to Zagilek's criteria, neurosarcoid can be diagnosed as definite, probable and possible based on clinical symptoms, MRI features, laboratory findings, presence or absence of systemic sarcoidosis, exclusion of other diagnosis, and positive nervous system histology. Often non contrast scans are done as clinical features are similar to that of myelopathy and the findings too are consistent with cervical spondylitic myelopathy.This may be further under evaluated as patient may have associated cervical stenosis and patients are often subjected to decompression surgery without any improvement. Discordance between clinical symptoms and neurological findings and rapid detoriation in neurological symptoms should lead to performance of contrast MRI scan to unravel other pathologies likeneurosarcoidosis or malignancy.

Case Presentation

68years old female presented with, neck pain since 1 month, numbness and weakness in bilateral upper limbs since 20 days, imbalance while walking since 15 days, swaying while standing since 10 days. There was no h/o fever/Koch's/trauma/bowel-bladder/HTN/DM involvement.

Routine lab investigations were ordered as per the hospital protocol. Electrocardiographic findings were within normal limits. Hematological cell counts were adequate.

Routine chest radiograph revealed no significant abnormality. Radiograph of cervical spine revealed degenerative changes in the vertebral body in form of reduction of intervertebral disc spaces, loss of cervical lordosis, marginal osteophytes. It was decided togo ahead with magnetic resonance imaging of spine as clinical suspicion of cervical stenosis and patients are often subjected to decompression surgery without any improvement.

MRI whole spine screening with dedicated cervical spine scan was performed. The scan revealed, long segment altered signal intensity within the spinal cord extension at C2-T2 vertebral body level which appears hyperintense on T2 SAG images , longitudinal expansile hyperintense lesion on STIR COR images and predominantly posterior subpial hyperintensity on T2 AXIAL images.After gadolinium administration T1 images showed posterior subpial central canal enhancement resembling a 'trident sign' on axial images and longitudinal long segment enhancement on sagittal images.

Patient was retrospectively evaluated for the systemic sarcoidosis, high-resolution CT thorax and contrast enhanced CT abdomen was performed. HRCT thorax was unremarkable, noevidence of any mediastinal or hilar lymphadenopathy and any significant changes in the parenchyma. Again CECT abdomen scan performed and it revealed incidentally detection of cholelithiasis and adrenal adenoma. Although there have been few cases of involvement of adrenal gland in sarcoidosis which may present as adrenal adenoma. It is exceptionally rare (<0.3%). There were incidental detection of adrenal adenoma with no clinical symptoms seen with respect to adrenal involvement in the case of sarcoid.

Further Positron emission tomography CT was performed and it showed linear FDG uptake in posterior part of spinal cord C3-C7, however no evidence of any demonstrable lesion.

On the basis of the imaging diagnosis of neurosarcoidosis, patient was immediately started on steroids and showed good response on follow up imaging.

Discussion

Sarcoidosis is an inflammatory multisystem granulomatous syndrome of unknown causewith a vast range of clinical manifestations involving lungs, lymph nodes, skin and eyes. The first description of sarcoidosis is given since 1869. Neurosarcoidosis is a rare complication of sarcoidosis. Isolated spinal cord neurosarcoidosis is exceptionally rare.[1] Lung is the most commonly involved organ in sarcoidosis up to 90% of patients. Nervous system involvement is 5-15% of patients.Presentation of various neurological symptomsare suggestive of the clinical diagnosis of neurosarcoidosis in multisystem sarcoidosis. Without biopsy evidence, it is very difficult to diagnose the solitary nervous-system involvement in sarcoidosis as the disease can present in many different ways. [2]

The most commonlyneurosarcoidosis is presented with clinical manifestation of involvements of singular or multiple cranial nerves palsy especially the facial nerve.However spinal neurosarcoid is relatively uncommon. It can manifest in different ways like intramedullary lesion, intradural extramedullary or extradural lesion, cauda equina syndrome and archnoiditis. In addition, spinal intramedullary neurosarcoidosis can also present similar to transverse myelopathy. The spinal intramedullary neurosarcoidosis is rare and may mimic malignancy or an inflammatory demyelinating disease. Thus it is very necessary todifferentiate intramedullary neurosarcoidosis of the spinal cord from the spinal cord tumor,

inflammatory myelitis such as neuro Behcet's, SLE, Sjogren's disease, mixed connective tissue disease, antiphospholipid syndrome, Lyme's disease, lymphoma or tuberculosis, multiple sclerosis and fungal infections. [3]

Intramedullary spinal sarcoidosis frequently affects cervical cord (56%), followed by thoracic cord (37%) and lumbar and sacral cord (7%). Classical MRI findings in intramedullary neurosarcoidosis reveal fusiform enlargement of the spinal cord with high signal on T2-weighted image and low signal on T1- weighted image, and patchy/nodular contrast enhancement. Another findings suggesting the diagnosis of intramedullary spinal sarcoidosis s even only with abnormal enhancement of spinal cord with normal appearance on T1- and T2-weighted image. [4]. Crescent shaped layering of posterior subpial enhancement accompanied by central canal enhancement giving the classical trident headappearance.

Junger et al. proposed MRI classification of intraspinal sarcoidosis in four stages, correlation with possible histological stages of the disease. Phase1, initial stage of inflammation shows the linear leptomeningeal enhancement along the spinal surfaceafter contrast administration; phase 2, leptomeningeal inflammatory process with secondary parenchymal involvementspread of through the Virchow-Robin spaces.Diffuse swelling and faint enhancement is seen within the parenchyma; phase3, mild reduction in swelling and possible normal size spinal cord, associated with focal or multiple enhancement; and phase 4 ,inflammatory process resolves in the form of normal size or atrophy of the spinal cord and no enhancement. Few other findings that can be seen as calcification, cyst formation and extradural involvement. Preoperative imaging diagnosis of intraspinal sarcoidosis may alert to look carefully for granulomas and giant cells for pathologist. It is because sarcoidosis can mimic neoplasm and lead to frozen section misinterpretation.[5]

The diagnosis and management of neurosarcoidosis can be challenging. Corticosteroids are the drug of first choice. Several cytotoxic drugs, including methotrexate, in addition have been used to treat sarcoidosis. [2] The significant therapeutic advance of novel CNS penetrant drugs depend on particularly effectivity at inhibiting granuloma formation. Future progress will be informed by a deeper understanding of the pathways underlying the granulomatous inflammation characteristic of sarcoidosis and by an increased appreciation of how sarcoid lesions evolve in the CNS microenvironment. [6]

With corticosteroid therapy, dramatic improvement was seen on MRI, including disappearance and marked reduction of swelling and enhancement. Recurrence can be seen on MRI and plasma ACE level without dramatic change in the clinical manifestations. [7]

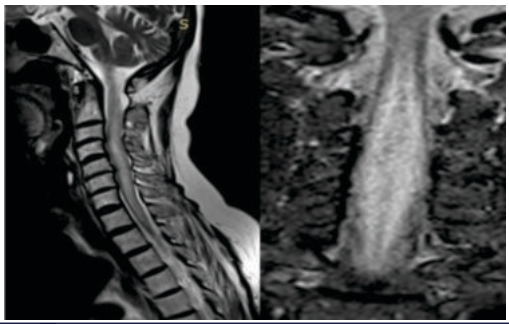
Conclusion

68 years old female presented with progressive upper limb numbness and weakness associated with neck pain and swaying on walking with sensory ataxia was evaluated. Lab investigations were normal. MR Imaging of spine demonstrated unique and characteristic sign “the trident sign”. PET CT demonstrated Linear FDG uptake in posterior part of spinal cord C3-C7 with no demonstrable lesion. Patient was diagnosed as possible neurosarcoidosis and was started on steroids and immunosuppressant. Patient has responded well to treatment.

Figure Legends

Figure 1:

Right: Magnetic resonance imaging T2 SAG image cervical spine scan reveals, long segment hyperintensity within the spinal cord at C2-T2 vertebral body level.



Left: Magnetic resonance imaging STIR COR image shows longitudinal expansile hyperintense lesion within spinal cord.

Figure 2: Magnetic resonance imaging T2 AXIAL image shows predominantly posterior subpial hyperintensity.

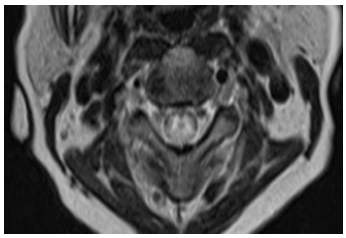
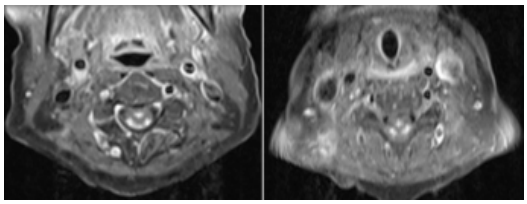


Figure 3:

Right: Magnetic resonance imaging T1 post contrast axial image shows posterior subpial cord enhancement resembling a 'trident sign'.



Left: Magnetic resonance imaging T1 post contrast axial image shows posterior subpial cord enhancement resembling a 'trident sign'.

Figure 4: Magnetic resonance imaging T1 post contrast sagittal image shows longitudinal long segment enhancement in cervical cord.

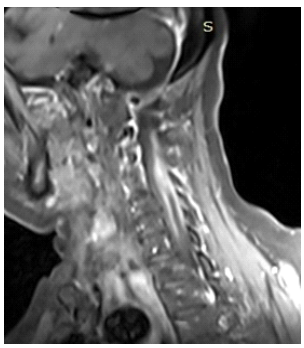
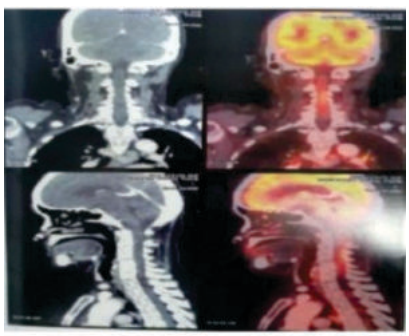
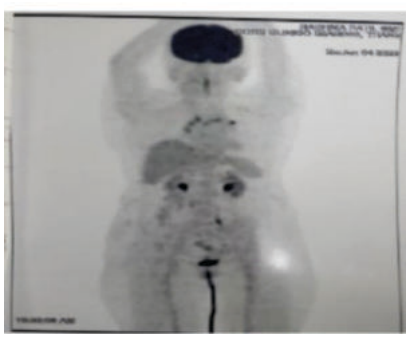


Figure 5:PET CT Linear FDG uptake shows in posterior part of spinal cord C3-C7 with no demonstrable lesion.





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