Clinical and Neuroimaging Features of Sarcoid Associated Myelopathy

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Introduction

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology that affects individuals worldwide. It is characterized pathologically by the presence of non-caseating granulomas in involved organs. In sarcoidosis, granulomas in the disease involve mostly lungs and lymph nodes but other organs may be affected. Neurosarcoidosis (NSC), the clinical involvement of the nervous system occurs approximately in 5-10% of patients with sarcoidosis, and granulomatous inflammation may affect the meninges, hypothalamus, pituitary gland, and cranial nerves. Although NSC is often suspected in patients with systemic sarcoidosis who develop neurological symptoms, approximately half of patients with neurological involvement present as new onset sarcoidosis, which makes it difficult to diagnose as NSC. Spinal sarcoidosis is a rare manifestation of the disease appearing as inflammatory, intramedullary, or extradural lesions as well as cauda equina syndrome. We present the clinical, neuroimaging and natural history features of a large series of patients with spinal cord NSC.

Material and Methods

This is a descriptive retrospective analysis of patients with neurological involvement of NSC diagnosed and followed at the Johns Hopkins Translational Myelitis Center, from 2003 to August 2012. The diagnosis of neurosarcoidosis was based on the JAPS neurosarcoid diagnostic criteria. Patients with defined, probable and possible diagnosis NSC and myelopathic forms were included. The clinical and neurological profile, diagnostic imaging, CSF and laboratory findings were evaluated. Particular attention was given to the temporal evolution of symptoms and presentation (e.g., hyperactive <5 hours; acute 6-48 hours; Subacute 48 hours-21 days; chronic >21 days), clinical manifestation (e.g., motor, sensory or sphincter involvement) and overlap with other neurological involvement (e.g., cerebral, meningeal, and retinal). The disability outcome was determined by the ASIA score in the outpatient clinic as a minimum six months after the first manifestation of myelopathy, the pattern of relapses and cause of relapses in those with at least 10 months of follow up. The drug response and level of spinal cord involvement were based on examination of the MRI available and analyzed the cytological characteristics as well as oligoclonal band and IgG index when possible.

Table 2. Clinical Profile

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Number (Percentage)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>19 (38.0)</td>
</tr>
<tr>
<td>Female</td>
<td>30 (62.0)</td>
</tr>
<tr>
<td>Age (median)</td>
<td>48.3 (range 33-67)</td>
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<tr>
<td>Age &gt;40</td>
<td>39 (79.6)</td>
</tr>
<tr>
<td>Gender&lt;40</td>
<td>4 (8.2)</td>
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<tr>
<td>Gender&gt;40</td>
<td>25 (51)</td>
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<tr>
<td>Testicles</td>
<td></td>
</tr>
<tr>
<td>Skin</td>
<td></td>
</tr>
<tr>
<td>Meningeal</td>
<td></td>
</tr>
<tr>
<td>Sensory</td>
<td></td>
</tr>
<tr>
<td>Other associated neurosarcoidosis syndromes</td>
<td>5 (10.2)</td>
</tr>
</tbody>
</table>

Results

We are grateful with our patients and families for their participation in this trial. johns hopkins project restore.

If a, b, c, d, and e are described by the medical records. We extracted the characteristic of the CSF when available and analyzed the cytological characteristics as well as oligoclonal band and IgG index when possible.

Clinical neurological syndrome supported by findings in MRI and/or CSF and plus:

1-Definite: Clinical neurological syndrome supported by histologic documentation in meninges of inflammatory changes consistent with granulomatous inflammatory disease and
Exclusion of other pathologies associated to neoplastic, rheumatologic or neurological infectious diseases by CT scan or FDG-PET scan and/or serological studies.

2-Probable: Clinical neurological syndrome supported by findings in MRI and/or CSF and plus:
- Histologic evidence of sarcoidosis in other organs
- Exclusion of other pathologies such as rheumatologic or neurological infectious diseases by CT scan or FDG-PET scan and/or serological studies.

3-Possible: Clinical neurological syndrome supported by findings in MRI and/or CSF and plus:
- Clinical systems involvement suggestive of sarcoidosis without histologic confirmation
- Exclusion of other pathologies such as rheumatologic or neurological infectious diseases by CT scan or FDG-PET scan and/or serological studies.

4-Suspected: Clinical neurological syndrome supported by findings in MRI and/or CSF and plus:
- A clear evidence of systemic involvement suggestive of sarcoidosis
- Exclusion of other pathologies such as rheumatologic or neurological infectious diseases by CT scan or FDG-PET scan and/or serological studies.

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Figure 2. Patients followed up more than 18 months

Figure 3. Suspected cause of the relapsing disease

Figure 4. A sagittal T2w image, B sagittal contrast enhanced STIR image, C axial T2w image, D axial contrast enhanced T1w image, E axial T2w image, F axial T2w image showing a subacute spinal cord lesion.

Figure 5. A sagittal T2w image, B sagittal contrast enhanced T1w image, C axial T2w image, D axial contrast enhanced T1w image, E axial T2w image showing a subacute spinal cord lesion.

Conclusions

The clinical profile of spinal cord sarcoidosis is predominantly a chronic, progressive myelopathy and principally manifests with sensitive symptoms.

- Usually one of two MRI patterns are identified, a tumefactive central cord lesion or patchy multilevel lesions, almost all of them with contrast enhancement during the symptomatic phase and some with nodular meningeal enhancement.

In this series of patients neurosarcoidosis was diagnosed as a first manifestation of sarcoidosis in 75% of the cases and in 85.7%, myelopathy was the first manifestation of the disease.

Most patients with neurosarcoidosis showed abnormalities in standard CSF analysis. Specific pattern were not found, but in most of them we observed a lymphocytic predominant pleocytosis with increased proteinorachia.

- In most of the cases the relapses were presented during the steroids tapering or were due to lack of adherence to the immunosuppressive treatment.