Trigeminal ganglion involvement in sarcoidosis – case report.

Acometimento do gânglio trigeminal na sarcoidose – relato de caso.

Jairo Silva dos Ângelos
Guilherme Brasileiro de Aguiar
Edgar Manuel Garcete Fariña
Maud Parise
Alexandre Martins Cunha
Elington Lannes Simões
Flávio Nigri
Carlos Telles

ABSTRACT
Sarcoidosis is a disease of unknown etiology presenting as noncaseating granulomatous infiltration in tissues. Central Nervous system is rarely affected, usually as dissemination from other organs, mainly lungs. We report on a case in which the patient had only central nervous system involvement, presenting with a mass in the right gasserian ganglion and another lesion in right parietal cortex, observed in magnetic resonance. Diagnosis was given by histopathological study of the lesion and the patient improved with corticosteroid treatment.

Key-words: sarcoidosis, Gasser’s ganglion, central nervous system

INTRODUÇÃO
Sarcoidosis is a systemic disease of unknown etiology, presented as noncaseating granulomas in the involved tissues. Any tissue can be affected, but in the majority of cases, pulmonary sarcoidosis precedes the appearing of the disease in other sites. Manifestation in the nervous system is a rare feature.

In the present article we report a case of neurosarcoidosis in a patient with trigeminal nerve symptom and seizures. We also review the literature with special emphasis in cranial nerves involvement.

CASE REPORT
A 63-year-old woman was admitted at our institution with a history of partial-complex seizures that began one year before. Neurological examination: hypoesthesia in left body and in right face, with facial hypoesthesia respecting the maxillary division of trigeminal nerve. CT images showed a right parietal lesion, next to midline, measuring about 2 cm in diameter, with a nodular aspect, poor evident borders, slightly hyperdense, with perilesional edema (Figure 1A) and moderate contrast enhancement (Figure 1B). Magnetic resonance image (MRI) with enhancement on T1-weighted sequence showed also adjacent paquimeningitis and dural thickening on right Meckel’s cavum (Figures 1C, 1D and 2A).
Patient was submitted to a biopsy of the parietal lesion and the histopathological analysis revealed a noncaseating granuloma. After disclosing other granulomatous diseases, a diagnosis of sarcoidosis was done, and treatment with prednisone was soon initiated. The patient had a radiological improvement (Figure 2B) and became seizure-free, nevertheless remaining with the trigeminal symptom referred.

In central NS, sarcoidosis takes the form of a granulomatous infiltration of meninges and adjacent neural structures, frequently in skull base. Granulomas might be present as tumoral masses, or as focal infiltrative lesions, in cortical or subcortical regions. Therefore, clinical signs depend directly on the site of the granulomatous lesion, including cranial nerves syndromes, aseptic meningitis, encephalopathy, vasculopathy, psychiatric manifestations, hydrocephalus, hypothalamic disorders, spinal cord and peripheral nerves signs. Also, there are asymptomatic or subclinical forms of the reported disease.

Cranial nerves damage is the commonest neurological manifestation of sarcoidosis, being the facial nerve the more frequently affected. The other cranial nerves are involved in the following order of decreasing frequency: optic nerve, vestibulocochlear, glossopharyngeal, vagus, oculomotor, trigeminal, hypoglossal, abducens, accessory, and trochlear.

Trigeminal nerve involvement is extremely rare, and only few cases have been reported on the literature. Braido et al describe a case of bilateral involvement of the Gasserian ganglion cistern, with trigeminal signs, but associated with Heerfordt syndrome (uveitis, parotiditis, facial palsy, fever) and sarcoidosis on mediastinum and parotid gland. Bonet et al report on a case of a young female patient, who had only trigeminal involvement, due to a mass involving the nerve; in that case the diagnosis of sarcoidosis was achieved only with the histopathological study of the lesion. Arias et al also report a rare case of trigeminal sarcoidosis, with the first and the second branches of fifth nerve affected by a lesion in the right gasserian ganglion.

The case reported here illustrates a patient without previous diagnosis and without clinical evidence of sarcoidosis. Initially, the complaint of hypoesthesia in the face was not recognized as a specific symptom, but this became relevant when radiologic alterations observed in the Gasser’s ganglion region disappeared after the treatment with prednisone.

Seizures represent unspecific signs, indicating chronicity and poor prognosis. Also unspecific are the findings in the cerebrospinal fluid (CSF), including pleocytosis, high protein content and the increase in the concentration of angiotensin converting enzyme (ACE), which can be high in more than 50% of neurosarcoidosis cases. This finding, however, has more utility when monitoring the disease activity and treatment response.

MRI is the best diagnostic tool for identifying sarcoid lesions in NS, but the images found are also non specific. On T1-weighted sequences, lesions might appear isointense or hypointense and hyperintense or hypointense on T2-weighted sequences. The reason for this variability is unknown, but it might depend on the amount of connective tissue stroma and fibrosis in the lesion. Enhancement of sarcoid lesions is reported as a common finding.

Due to the various clinical and radiological features, there are...
diagnostic criteria for recognizing sarcoidosis in NS (Table 1). According to these criteria, our patient had a definite diagnosis of neurosarcoidosis, due to the histopathologic evidence and the exclusion of other granulomatous diseases applicable to this case. The radiologic improvement of Gasser’s ganglion alterations assures the diagnostic conclusion.

Treatment is based on the use of corticosteroids, as first-choice drugs, for sarcoid lesions in NS and in other tissues.

This case is a rare example of sarcoidosis in NS, including the involvement of trigeminal nerve, which is another uncommon finding. So, it’s reasonable to think in sarcoidosis, even before a biopsy, when we have a detailed radiologic study of the Meckel’s cavum in a patient with trigeminal symptoms. Nevertheless, in this and other clinical situations, where pulmonary sarcoidosis is not present, the diagnostic certainty is given only by the histopathological study of the suspicious lesions.

Table 1: Criteria for the diagnosis of neurosarcoidosis

<table>
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<tr>
<th>Definite</th>
<th>Probable</th>
<th>Possible</th>
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<tr>
<td>Clinical presentation compatible with neurosarcoidosis</td>
<td>Laboratory support of central NS inflammation*</td>
<td>Clinical presentation compatible with neurosarcoidosis</td>
</tr>
<tr>
<td>Exclusion of other possible causes</td>
<td>Exclusion of other possible causes</td>
<td>Exclusion of other possible causes</td>
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<td>Positive nervous system histology</td>
<td>Evidence of systemic sarcoidosis**</td>
<td>(abstr.)</td>
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*High concentrations of CSF protein and high numbers of cells, the presence of oligoclonal bands, or MRI evidence compatible with neurosarcoidosis.

**Positive histology or at least two indirect indicators from gallium scan, chest imaging, and serum angiotensin-converting-enzyme. Adapted from Hoitsma et al.

References


