

Neurosarcoidosis- A case report

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Abstract: Neurosarcoidosis is an uncommon multisystem granulomatous disease which can affect the nervous system. The diagnosis of neurosarcoidosis is difficult in patients who lack either pulmonary or systemic manifestations of sarcoidosis. Further more the clinical features of neurosarcoidosis are variable. We are reporting one such case of systemic sarcoidosis with neurological manifestation. A 60 year old lady presented with headache and seizures for 6 months and imageology of brain revealed multiple dural based lesions in brain and dura. biopsy revealed chronic non caseating granulomatous lesion suggestive of sarcoidosis .

Key words: dura, headache, sarcoidosis

I. Introduction

Sarcoidosis is a multisystem inflammatory granulomatous disease of unknown etiology although current opinion favors an immune response to an as yet unknown antigen. Although the characteristic non-caseating granulomas can occur in any organ system, the lungs and the draining mediastinal lymph nodes are the most common sites of involvement. Neurosarcoidosis is thought to be much rarer occurring in less than 5% of patients with systemic sarcoidosis. It is estimated that less than 1% of patients have isolated central nervous system involvement, without systemic evidence of disease.

Sarcoidosis can affect patients of all ages and races but is most common in the third and fourth decades. Women are more frequently affected than men. Genetic factors confer increased susceptibility.

II. Case report

A 60 year old lady presented with persistent headache for the duration of period of six months which was holocranial and dull aching with no diurnal or postural variations and not associated with vomitings.

Head ache accompanied with two episodes of generalized tonic clonic seizures over period of one month with no post ictal weakness or confusion.

Her general physical and nervous system examination were unremarkable.

Ancillary blood investigations showed Hb – 13 gm% TLC 6900 cells/ml ESR 90mm/hr

Mantoux test was negative

Chest xray revealed mediastinal lymphadenopathy

MR brain plain with contrast revealed multiple enhancing dural based lesions in frontal, parietal, in the left temporal convexity regions and in the falx along the tentorium with edema of subcortical white matter of bilateral frontal , anterior temporal and bilateral posterior occipital lobes along the splenium suggestive ? metastasis? Tuberculomas

Screening of cervical spine also revealed extra enhancing dural multiple lesions .

Patient underwent dural biopsy in the frontal region under local anesthesia to confirm the diagnosis.

Per operatively dura was unusually vascular with firm greyish white gritty lesion seen adherent to the inner surface of the dura but was not infiltrating the brain surface.

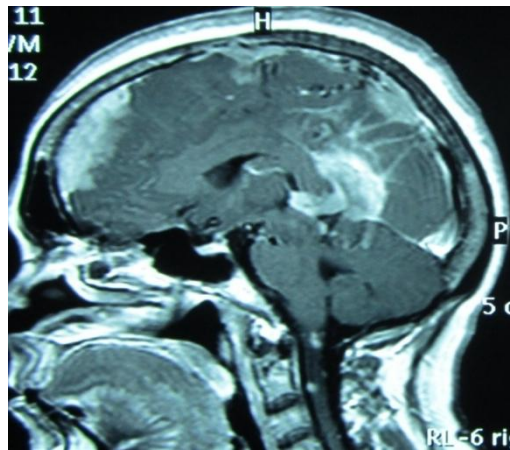
Histopathology was suggestive of chronic granulomatous lesion with no caseation suggestive of sarcoidosis

Post operatively she was started on steroids and immune suppressants with anti epileptic agents

She was seizure free at 3 / 6/ 12 months follow up and relieved of headache

At 12 months follow up , CT chest was done which showed complete resolution of lymphadenopathy

ESR levels showed decrease from 90 to 50mm/hr

Imageology**III. Discussion**

Clinical symptoms of neurosarcoidosis depends on the site of granuloma involvement and are non specific 1

Cranial neuropathies were the commonest manifestation of NS, occurring in 35 of the 54 patients (63%). Of the cranial neuropathies, optic neuropathy was the commonest, affecting 35% of patients while facial nerve involvement was seen in 11%. Myelopathy, presenting with sensory and motor symptoms in the upper or lower extremities, was seen in 19%. Seizures (17%) and headaches (17%) were other features seen in 10% or more of the patients in a study of 54 cases of neurosarcoidosis by s pawate et al 2

In our patient headache and seizures were the main manifestation with no other systemic manifestations

Seizures are estimated to occur in 5–20% of neurosarcoidosis patients. The neurologic manifestation of neurosarcoidosis that may contribute to development of seizures are protean and include parenchymal abnormalities including encephalopathy/vasculopathy (5–10%), intraparenchymal mass lesion(s) (5–10%), endocrinopathy with metabolic disturbance including hypothalamic and/or pituitary dysfunction (10–15%), meningeal disease (including aseptic meningitis and meningeal mass) (10–20%), and hydrocephalus (10%). Most reports indicate a poor prognosis with progressing or relapsing course when seizures are associated with sarcoidosis 3

Neurosarcoidosis can be diagnosed in patients with (i) a clinical presentation suggestive of neurosarcoidosis with (ii) exclusion of other possible diagnoses, as follows: Neurosarcoidosis can be diagnosed in patients with (i) a clinical presentation suggestive of neurosarcoidosis with (ii) exclusion of other possible diagnoses, as follows:

1. Definite NS: Positive central nervous system histology

2. Probable NS: [a] Laboratory evidence of CNS inflammation (elevated levels of CSF protein and/or cells, the presence of oligoclonal bands and/or MRI evidence compatible with neurosarcoidosis), and [b] Evidence for systemic sarcoidosis (either through positive histology, including Kveim test, and/or at least two indirect indicators from Gallium scan, chest imaging and serum ACE)

3. Possible NS: Where the above criteria are not met central nervous system histology 2. Probable NS: [a] Laboratory evidence of CNS inflammation (elevated levels of CSF protein and/or cells, the presence of oligoclonal bands and/or MRI evidence compatible with neurosarcoidosis), and [b] Evidence for systemic sarcoidosis (either through positive histology, including Kveim test, and/or at least two indirect indicators from Gallium scan, chest imaging and serum ACE) 3. Possible NS: Where the above criteria are not met 4

From a radiologic perspective, neurosarcoidosis has been described by a number of different modalities, including single photon emission computed tomography (SPECT), computed tomography (CT) and magnetic resonance (MR) imaging. 4 CT findings of neurosarcoidosis generally non-specific and includes hydrocephalus, periventricular hypoattenuation and contrast enhancement, calcification, meningeal contrast enhancement, white matter lesions and lesions at the optic nerve or chiasma

Tumors are isointense on T1-weighted MRI images and hyperintense on T2-weighted images, with uniform enhancement after gadolinium administration 65

Intraoperatively, these lesions are firm, fibrous masses attached to the dura. 5

Histologically, pseudomeningiomatous neurosarcoidosis is similar to sarcoidosis elsewhere in the body, with multinucleated giant cells and epithelioid histiocytes that react with antibodies to MAC387 and CD68 but not

with S-100 protein. Limited numbers of lymphocytes and plasma cells and extensive fibrosis are also present. Schaumann or asteroid bodies may or may not be seen in giant cells and are not specific for sarcoidosis [5]. The diagnosis of definite neurosarcoidosis is confirmed by biopsy results showing non-caseating granuloma, with an absence of organisms or other causes. Corticosteroids remain the mainstay of treatment along with supplementary immunosuppressants and patient may improve rapidly [1], as seen in our patient.

IV. Conclusion

Neurosarcoidosis should be kept as differential diagnosis in cases of multiple dural based lesions and

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