Neurosarcoidosis

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Abstract

We report a case of Neurosarcoidosis in absence of pulmonary features. It is estimated that less than 1% of patients have an isolated CNS involvement, without systemic evidence of disease. This case also had an unusual clinical presentation. A 28 years old female, presented with headache for 6 months. Her MRI Brain showed multiple ring enhancing lesions in the left cerebellum and vermis. Patient underwent posterior fossa craniotomy with biopsy of left cerebellar lesion which revealed non-caseating chronic granulomatous inflammation, consistent with sarcoidosis.

Keywords: Neurosarcoidosis, Cerebellar lesions, Magnetic Resonance Imaging.

Introduction

Sarcoidosis is a systemic granulomatous disease of unknown origin, characterized by the presence of non-caseating granulomas in affected organs. Lesions are commonly seen in the lungs, lymphatic system, eyes, skin, liver, spleen, salivary glands, heart, nervous system, muscles and bones.[1] 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. The exact cause of sarcoidosis is unknown. Genetic factors confer increased susceptibility. Neurosarcoidosis has been described in 5% of patients with sarcoidosis. It is estimated that less than 1% of patients have isolated central nervous system involvement, without systemic evidence of disease.[2]

Case Report

A 28 years old female patient was admitted for evaluation of persistent headache for 6 months. Clinical examination showed her Glasgow Coma Scale to be 15/15, pulse rate 62, blood pressure 110/80 and was afebrile. Neurological examination revealed unremarkable motor and sensory examination. Laboratory investigations showed white cell count of 8.8, erythrocyte sedimentation rate 35. Kveim test, Anogiotensin Convesting Engyme level and Cerebrospinal Fluid were not done. Her chest x-ray showed no evidence of hilar adenopathy or prominent pulmonary markings. Computed Tomography Scan of the head showed multiple hypodense areas in the cerebellum, inferior herniation of cerebellar tonsils with obstructive hydrocephalus. Basal cisterns were effaced. She underwent magnetic resonance imaging of Brain with Gadolinium which revealed multiple small abnormal signal intensity areas in left cerebellum and vermis. These were associated with surrounding oedema causing effacement of adjacent cerebellar folia and pressure on brainstem and fourth ventricle. This shows predominantly hypointense signals on T1-Weighted, iso to hyperintense signals on T2-Weighted and fluid attenuated inversion recovery (FLAIR) images. Post contrast images showed peripheral enhancement. Based on these findings the most probable diagnosis of tuberculomas were suggested. To confirm the diagnosis, the patient was subjected to biopsy of the lesion. She underwent posterior fossa craniotomy of left cerebellar lesion. Pathology of specimen revealed non-caseating chronic granulomatous inflammation, consistent with neurosarcoidosis. The patient had improvement of her symptoms after treatment with Dexamethasone. She was discharged from hospital without further complications and advised to continue regular follow-up.

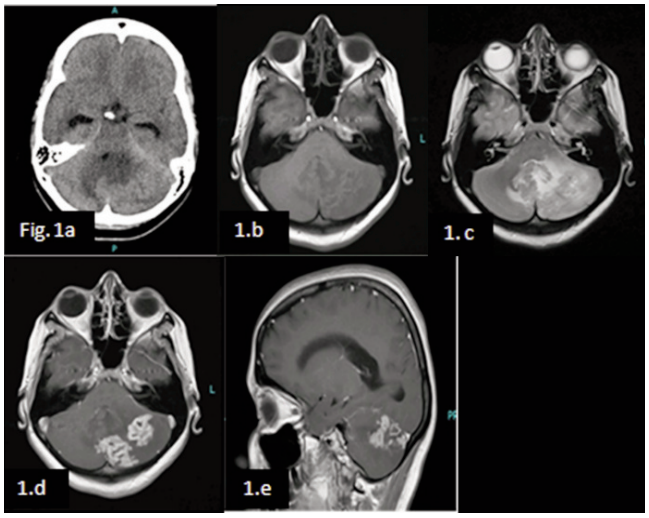

Figure: (a) Non-contrast CT Scan showed hypodense areas in the cerebellum and prominent temporal horns of lateral ventricles. (b) MRI, T1-Weighted axial image: shows hypointense areas in left cerebellum and vermis. (c) T2-Weighted axial image: iso to hyperintense lesions in left cerebellum and vermis with marked surrounding oedema. (d) Contrast enhanced axial T1-Weighted image: shows heterogenous enhancement of the lesions. (e) Contrast enhanced T1-Weighted sagittal image showing multiple enhancing lesions in left cerebellar hemisphere.

Discussion

Neurologic affliction in sarcoidosis has been described in 5% of patients with sarcoidosis.[3] Imaging findings include dural thickening or mass, leptomeningeal involvement, enhancing and non-enhancing parenchymal lesions, cranial nerve involvement and spinal or nerve root enhancement. Clinical symptoms of neurosarcoidosis depend on the site of granuloma involvement and are non-specific. Facial nerve paralysis and vision loss are common symptoms, as are headaches, seizures, and signs of meningeal irritation.

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 is 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.[5]

Magnetic resonance imaging presentations of neurosarcoidosis are quite variable. In an MR imaging study by Zajicek et al,[5] the most common neurologic presentation was optic nerve disease followed by involvement of other cranial nerves, spinal cord lesion, brain stem or cerebellar lesion, meningeal lesion, cognitive decline, hydrocephalus and pituitary or hypothalamic involvement, respectively.[5]

Lexa and Grossman reported patients most commonly exhibiting white matter periventricular and periaqueductal hyperintensity on long TR/TE sequences.[6] Leptomeningeal enhancement and a parenchymal mass with enhancement were also seen.

Christoforidis et al[4] found that MR imaging findings usually consisted of hypointensity or mixed intensity on T2-Weighted sequences and enhancement with Gadolinium.[4]

Seltzer et al,[7] demonstrated hypothalamic and pituitary thickening with enhancement as well as a case of sagittal sinus thrombosis.

The diagnosis of definite neurosarcoidosis is confirmed by biopsy results showing non-caseating granuloma, with an absence of organisms or other causes.[2] Corticosteroids remain the mainstay of treatment and patient may improve rapidly.[2]

Conclusion

Neurologic involvement is a significant cause of mortality and morbidity in patients with sarcoidosis and isolated neurological symptoms are the sole presenting abnormalities in the absence of pulmonary or other systemic findings. However, with timely diagnosis, these can be successfully managed with corticosteroid therapy.

References

1. Shah R, Roberson GH, Cure JK. Correlation of MR imaging findings and clinical manifestations in Neurosarcoidosis. AJNR Am J Neuroradiol 2009; 30: 953-61.
2. Smith JK, Matheus MG, Castillo M. Imaging manifestations of Neurosarcoidosis. AJR Am J Roentgenol 2004; 182: 289-95.
3. Hodge MH, Williams RL, Fukui MB. Neurosarcoidosis presenting as acute infarction on diffusion weighted MR imaging: summary of radiologic findings. AJNR Am J Neuroradiol 2007; 28: 84-6.
4. Christoforidis GA, Spickler EM, Recio MV, Mehta BM. MR of CNS sarcoidosis:correlation of imaging features to clinical symptoms and response to treatment. AJNR Am J Neuroradiol 1999; 20: 655-69.
5. Zajicek JP, Scolding NJ, Foster O, Rovaris M, Evanson JF, Saadding JW, et al. Central nervous system sarcoidosis-diagnosis and management. Q J M 1999; 92: 103-17.
6. Lexa FJ, Grossman RI. MR of sarcoidosis in the head and spine: spectrum of manifestations and radiographic response to steroid therapy. AJNR Am J Neuroradiol 1994; 15: 973-82.
7. Seltzer S, Mark AS, Atlas SW. CNS sarcoidosis: evaluation with contrast enhanced MR imaging. AJNR Am J Neuroradiol 1991; 12: 1227-33.