Original Research Article

Sarcoidosis with atypical clinical presentation: A case report

Manoj Kumar[1*], Dhara Mahendra[2]

[1] Consultant Radiologist, Department of Radiology, Manipal Hospital, Jaipur, Rajasthan, INDIA.
[2]Associate Professor, Department of Ophthalmology, SMS Medical College, Jaipur, Rajasthan, INDIA.
Email: drdharamanoj@gmail.com

Abstract

Background: Sarcoidosis is a rare multisystemic inflammatory disease of unknown origin characterized by growth and organization of inflammatory cells and the formation of noncaseating granulomas in any organ of human body. The most common sites are lungs, lymph nodes, eyes and skin. Central nervous system (CNS) represents a rare site of sarcoidosis. This condition called as Neurosarcoidosis. **Case presentation**: In this paper, we describe a case of a 39-year-old woman with neurosarcoidosis who presented with initial atypical symptoms such as facial paresthesia, confusion, auditory dysfunction and blurring of vision without respiratory or other systemic illness. Our patient fits the picture of probable neurosarcoidosis as per the Zajicek *et al.* criteria, with the imaging feature of neurosarcoidosis, evidence of inflammation in the CNS and systemic sarcoidosis and exclusion of other diseases. There was subsequent improvement in symptoms with steroids. **Conclusion:** The diagnosis and management of neurosarcoidosis remain difficult because of limitations of available diagnostic tests.

Keywords: diagnosis, neurosarcoidosis, treatment.

[*]**Address for Correspondence:**
Dr Manoj Kumar, Consultant Radiologist in Manipal Hospital, Jaipur, Rajasthan, INDIA.
Email: drdharamanoj@gmail.com

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INTRODUCTION

Sarcoidosis is a rare multisystemic granulomatous disease of unknown etiology which typically affects the lungs and mediastinal lymph nodes.[1] Sarcoidosis can involve other organ like central nervous systems, bones, eyes, skin, spleen, and liver. Neurosarcoidosis (NS) affects various sites of the central nervous system, including the cranial nerve, meninges, brain parenchyma, hypothalamus, and pituitary gland. Sarcoidosis is a very rare disease affecting 0.01%-0.02% of the global population and the infrequent event of sarcoidosis affecting solely the central nervous system (CNS) is defined as an isolated neurosarcoidosis (INS).[2,3,4]

Sarcoidosis usually diagnosed between 20 and 40 years of age.[5] Neurosarcoidosis occurs in the central or peripheral nervous system and is usually associated with other sarcoidosis organ involvement. Neurosarcoidosis can mimic more common disease processes, such as meningioma, glioma, or metastases. Involvement of the nervous system is rare but can result in serious complications.[6] Neurosarcoidosis is often suspected in patients with systemic sarcoidosis who develop neurological disorders.[7] However, when sarcoidosis develops exclusively in the nervous system (isolated neurosarcoidosis), diagnosing neurosarcoidosis may be difficult because of the nonspecific clinical features[8]. Furthermore, definite diagnosis of neurosarcoidosis requires histological confirmation, which is often not performed in patients with isolated central nervous system localization of sarcoidosis. INS usually progresses slowly and exhibits a variety of clinical manifestations, making timely, and transparent diagnosis difficult in most cases.[9,10,11]

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CASE

The aim of the paper is to report a confusing atypical initial clinical presentation of sarcoidosis. In this paper, we present the case of a 39-year-old woman who presented with facial paresthesia, confusion, auditory dysfunction and blurring of vision. No evidence of systemic manifestation or respiratory symptoms. She had no significant clinical history and was not on any medication. Initial laboratory results were unremarkable. Physical examination results and blood pressure were also normal. The results showed no abnormalities in complete blood count, erythrocyte sedimentation rate, serum electrolytes, and ultrasonography of the pancreas, spleen, kidney, liver and parotid gland. Chest radiograph and EEG (electroencephalogram) were also normal. Normal opening pressure was shown in CSF (cerebrospinal fluid) sampling, which includes 7/mL white blood cells, 0.81 g/L protein, and 3.04mmol/L glucose. MRI brain with contrast study revealed diffuse enlargement of pituitary gland and stalk, enhnacment of intracanalicular and intracranial portion of right optic and bilateral facial nerves along with pachymeningeal enhancement, thickening andenhancement along the bilateral meckels cave and prepontine cistern. Considering the MRI brain finding, probable diagnosis of neurosarcoidosis was kept. Further workup was done with evaluation of serum ACE levels and CECT chest and abdomen CT. CECT chest showed mediastinal nonnecrotic lymphadenpathy. Abdomen CT showed multiple splenic foci-suggestive of granulomatous lesion. Serum-ACE (angiotensin-converting enzyme) slightly increased (48 IU/L). Our patient fits on the picture of probable sarcoidosis as per the Zajicek *et al.* criteria, with the MRI brain imaging feature of neurosarcoidosis, with evidence of inflammation in the CNS and systemic sarcoidosis and exclusion of other diseases. She was started on steroids resulting in subsequent improvement in symptoms.

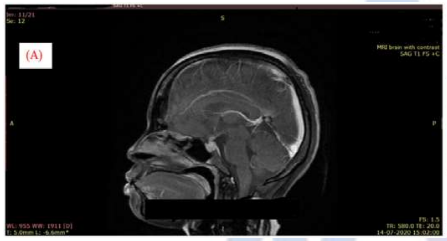
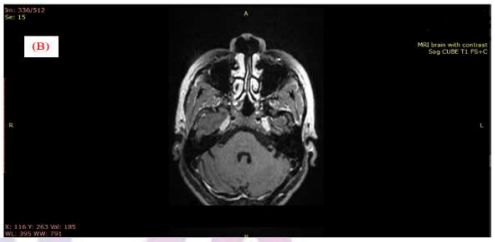


Figure 1



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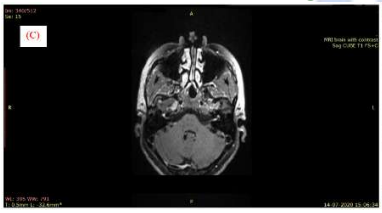

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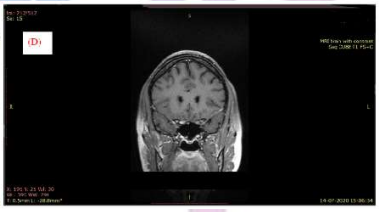

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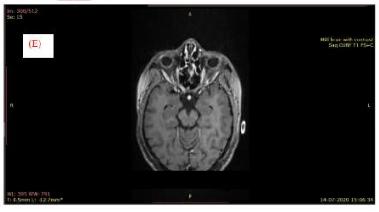
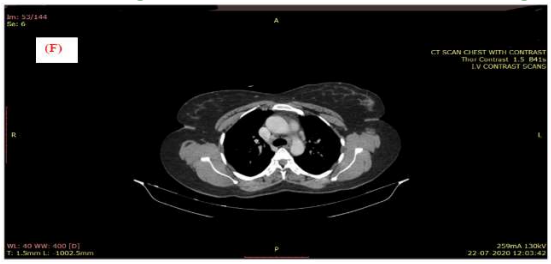


Figure 5



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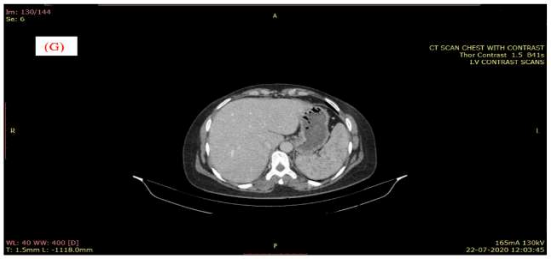

Figure 7

Figure 1: (A) MRI T1 sag post gadolinium image shows diffuse enlargement of pituitary gland with thickened stalk and enhancement; **Figure 2:** (B) MRI T1 axial post gadolinium-Diffuse thickening and pachymeningeal enhancement surrounding the meckels cave; **Figure 3:** (C) MRI T1 axial post gadolinium image-Enhancement of bilateral facial nerves at tympanic segments; **Figure 4 and 5:** (D and E) MRI T1 axial and cor post gadolinium images show enhancement of right optic nerves; **Figure 6:** (F) Contrast CT chest axial section shows enlarged non necrotic mediastinal nodes; **Figure 7:** (G) Contrast CT abdomen axial section shows multiple innumerable hypo dense splenic lesions suggestive of granulomas.

DISCUSSION

Neurological symptoms of NS vary widely depending on the site of focal lesion, such as the cranial nerve, dura mater, cerebral cortex, brainstem, and diencephalo-hypophysial area. More than half of the patients with NS primarily show neurological symptoms, making prompt diagnosis difficult. Microvascular changes are identified in the majority of patients with NS according to postmortem studies.[12] The pathogenesis of sarcoidosis remains elusive, although it is believed that type IV allergic reactions occur due to a specific, yet unidentified antigen, and that granulomas are formed in various systemic organs.[13] Cerebral MRI imaging presentations of NS show various abnormalities. Pawate *et al.* reported T2 hyperintense white matter lesions (52%) and leptomeningeal enhancement (19%) to be typical images of NS.[14] Vincent *et al.* also reported patients with strokes due to NS via MR imaging, in which T2 hyperintense lesion was the most common (69%), followed by leptomeningeal thickening or enhancement (44%).[15] Multiple white matter lesions followed by meningeal enhancement is always found in the MRI of the neurosarcoidosis patient. Gallium scintigraphy is reported by Marangoni *et al.*[16] for the use of diagnosis of probable sarcoidosis owing to high sensitivity of the test. A recent study suggests that fluorodeoxyglucose-positron emission computed tomography could be applied as a valid alternative imaging test, for it leads to better uptake in central nervous system sites.[17] The usefulness of CSF, ACE levels for the diagnosis of neurosarcoidosis is controversial. CSF abnormalities in neurosarcoidosis are usually nonspecific and can also be found in multiple sclerosis, Guillain–Barre syndrome, Behcet disease, brain tumors, and neurodegenerative diseases.[18] When no extraneural organ is identified as possibly being involved with sarcoidosis, biopsy of the lesion in CNS is needed for the last diagnosis.

CONCLUSION

The diagnosis and management of neurosarcoidosis remain difficult because of limitations of available diagnostic tests and atypical clinical manifestation. So, it is important to keep neurosarcoidosis in mind when diffuse enlargement of pituitary gland and involvement of cranial nerve is seen. In our case this leads to the appropriate treatment of patient. There is a need in the future for a multicenter prospective study in order to develop diagnostic and therapeutic standards of neurosarcoidosis.

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