Neurosarcoidosis - A Rare Cause of Rapidly Progressive Dementia

Suman Kushwaha1*, Preeti Singla1, Neha Agarwal1, Sujata Chaturvedi1 and Deepak Jha1

1Institute of Human Behavior & Allied Sciences - Delhi, India

Abstract

An elderly presenting with rapidly progressive cognitively decline should be investigated for unusual causes of dementia. Neurosarcoidosis as a cause of dementia is a rare presentation. The clinicians should recognize this rare cause of dementia as optimal therapeutic intervention will help in reversing the cognitive decline.

Key Word: Neurosarcoid; Dementia; Rapidly Progressive

Abbreviation: RPD- Rapidly Progressive Dementia; CSF- Cerebrospinal fluid; CECT- Contrast enhanced computed tomography; PCR- Polymerase Chain reaction; ZN Stain- Zeal Nelson Stain

The Case

65-year-old female presented with 5 months’ history of progressive decline in interaction with family members and loss of interest in household activities. Simultaneously she developed forgetfulness for recent events and was unable to recognize family members. She also started neglecting her personal hygiene and later developed urinary and fecal incontinence. She became dependent for activities of daily living and was bed bound in three months into the illness. On examination, she was conscious but disoriented to time, place and person. Meningeal signs were positive. Cranial nerve and motor system examination was normal. All reflexes were 2+ and bilateral planters were flexor. Gross sensory examination was normal. Higher mental function evaluation shows she was apathetic, comprehensible for simple verbal commands and slow in performing all tests. Repetition and Naming was impaired, verbal fluency was decreased. Attention and immediate memory were markedly impaired. She had dressing apraxia. The clinical diagnosis of chronic meningitis was kept and investigated.

Complete blood count and routine biochemical tests were normal. Contrast enhanced MRI brain showed thick irregular leptomeningeal enhancement and altered signal intensity in the region of dura along the right petrous temporal bone extending anteriorly to involve the right cavernous sinus and posteriorly to involve the right tentorium, right cerebellum, right temporoparietal dura and posterior falx. CSF analysis showed clear, acellular, sugar 53mg%, protein 75mg/dl, and chloride was 124meq/L. India ink, gram stain, and acid fast bacilli stains were negative. No Growth on CSF cultures for tuberculosis was negative. CECT chest showed emphysematous lung fields with subcentricmetal mediastinal lymphadenopathy. CECT abdomen was normal. Serum ACE levels were 75 ug/L. Right posterior temporal dural biopsy was done.

Intraoperative findings showed a thickened pale dura. Biopsy showed several distinct well circumscribed, coalescent granulomas comprising of lymphocytes, plasma cells, epithelioid cells and giant cells consistent with no necrotizing chronic granulomatous lesions (Figure 1). Necrosis was not seen. ZN stain for acid fast bacilli and PAS and silver methenamine stain for fungal elements was noncontributory.

Figure 1: Dural Biopsy showing distinct well circumscribed, coalescent granulomas comprising of lymphocytes, plasma cells, epithelioid cells and giant cells consistent with non-necrotizing chronic granulomatous lesions without necrosis.

*Corresponding author: Suman Kushwaha, Associate Prof Neurology, Room No 112 – Academic Block, Institute of Human Behavior & Allied Sciences, Delhi, India, E-mail: sumankushwaha@gmail.com


Citation: Suman Kushwaha, Preeti Singla, Neha Agarwal, Sujata Chaturvedi and Deepak Jha (2017) Neurosarcoidosis - A Rare Cause of Rapidly Progressive Dementia. BAOJ Neurol 3: 037.

Copyright: © 2017 Suman Kushwaha, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.
The compatible clinical profile, supportive lab and imaging findings along with histologic findings are consistent with neurosarcoidosis in our case. She was started on high dose of oral prednisone 60 mg day. The improvement in her clinical condition started within a week. The cognitive functions gradually improved over a period of 6 weeks and she became functionally independent in 8 weeks. After 6 months of treatment, patient is improved and stable on tapering dose of steroids.

Neurosarcoidosis has protean manifestations and can masquerade as many other diseases [1]. Rapidly progressive dementia (RPD) is a rare presentation. Parenchymal inflammation, hypercalcemia, or increased intracranial pressure due to space occupying granulomas can lead to abnormal mental status in neurosarcoidosis [2,3]. MRI is very sensitive in detecting abnormalities in neurosarcoidosis, but it is nonspecific [4]. The definitive diagnosis of neurosarcoidosis requires the exclusion of other causes and the identification of non caseating sarcoid granulomas by histologic analysis are hallmarks of sarcoidosis and reveal the inflammatory character of the disease [5]. The granuloma formation begins with exposure to an antigen and is followed by T-cell and macrophage activation [6].

Closest differential diagnosis of neurosarcoid is CNS tuberculosis differentiated by presence of caseating granulomas. Corticosteroids are the first line of treatment precise mechanisms of action of corticosteroids in treating neurosarcoidosis are not known, but presumably due to the known anti-inflammatory and immunomodulating effects. Benefits vary from substantial improvement about half to no benefit [7]. Neurosarcoidosis is a rare disease with many presentations and a commonly devastating complication of sarcoidosis. It should be investigated in patients presenting with rapidly progressive dementia as early recognition and Immunomodulation will help in reversal of cognitive decline.

References