Neurosarcoidosis presenting as psychiatric Symptoms: a diagnostic challenge

Farheen Niazi  
PAEC General hospital, Islamabad, Pakistan, farheenniazi@gmail.com

Tariq Hussain  
PAEC General hospital, Islamabad

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Recommended Citation
Available at: http://ecommons.aku.edu/pjns/vol12/iss2/12
INTRODUCTION

Sarcoidosis is a rare inflammatory disorder of unknown etiology, and is characterized by noncaseating granulomatous lesions. Sarcoidosis may affect many systems of the body, most commonly the lungs (87%) and thoracic lymph glands (28%) are involved. Eyes, kidneys, nervous system, heart, bones, and joints may also be affected. Most patients with sarcoidosis have no symptoms at all; the disease is often detected on routine chest radiographs with bilateral perihilar lymph nodes enlargement. Symptoms, if present may include cough, shortness of breath, and arthritis. Nervous system involvement (neurosarcoidosis) is reported in 5-15% of sarcoidosis patients.

Neurosarcoidosis is an uncommon but severe, sometimes life-threatening manifestation of sarcoidosis. Isolated neurosarcoidosis without involvement of othersystems is a rare occurrence. Intracranial neurosarcoidosis is a diagnostic challenge especially in the absence of systemic signs and symptoms of the disease elsewhere and also due to its non-specific clinical presentation and neuroradiological findings.

CASE REPORT

A 26 years old male, scientific assistant by profession had history of bowel irregularities for last one year for which he was diagnosed as having Irritable bowel syndrome. Afterwards, he had numerous somatic complaints which were diagnosed as psychogenic and depression related. For the past several months he was having multiple complaints regarding his health including headaches, generalised body aches, vertigo etc. Such patients are difficult to diagnose as sarcoidosis is rare in our part of the world and symptoms are often nonspecific. A high index of suspicion should be kept to identify these cases. We emphasize the importance of recognizing sarcoidosis in patients presenting with variable neuropsychiatric manifestations.

ABSTRACT

Among patients having neurosarcoidosis, psychiatric manifestations can be seen in 20% of the patients. Sarcoidosis is extremely rare in Asia, being almost unknown in some countries like China and Southeast Asia. Neurosarcoidosis occurs in approximately 5% of patients with sarcoidosis. Neurosarcoidosis presenting as psychiatric manifestations has rarely been reported. We report a case of a young male who presented with psychiatric symptoms, was diagnosed with neurosarcoidosis and responded well to oral steroids. His symptoms were diffuse headaches, and feelings of movement of fluid on top of head and certain other vague somatic complaints like body aches, vertigo etc. Such patients are difficult to diagnose as sarcoidosis is rare in our part of the world and symptoms are often nonspecific. A high index of suspicion should be kept to identify these cases. We emphasize the importance of recognizing sarcoidosis in patients presenting with variable neuropsychiatric manifestations.

KEYWORDS: Neurosarcoidosis, Sarcoidosis, headache

Corresponding to: Dr. Farheen Niazi, PAEC General hospital, Islamabad, Pakistan

Email: farheenniazi@gmail.com

Date of submission: January 11, 2017 Date of revision: February 26, 2017 Date of acceptance: March 17, 2017
His ANA was negative. ESR was found to be normal. His ENA profile was negative. Serum complement levels showed low C4 but normal C3. Serum ACE levels were found to be high i.e 131U/l (range 8-53U/l). HRCT chest showed right hilar lymphadenopathy.

On the basis of high ACE levels and neuropsychiatric symptoms diagnosis of neurosarcoidosis was made and patient was given steroids, i.e. prednisolone 40mg per day which was tapered gradually to a maintenance dose of 10mg once daily on alternate day over several months.

He showed good initially recovery of his symptoms. Follow up MRI brain after 3 months showed resolution of lesions.

Hilar lymph node biopsy was offered to the patient but was refused. Later on he also developed diabetes insipidus. His urine osmolarity was found to be low i.e 272 mosm/kg (range 300-900mosm/kg). Endocrinologist was consulted and was again thought to be due to hypothalamic involvement in neurosarcoidosis. He was treated conservatively for diabetes insipidus and showed good recovery.

DISCUSSION

Sarcoidosis is a multisystem granulomatous disease of unknown etiology. Sarcoidosis can be seen in patients of all ages, but most commonly affects young adults.\(^1\)

Neurosarcoidosis develops in 5-15% of sarcoid patients, and among neurosarcoidosis patients, 20% may manifest psychiatric symptoms.\(^2,4,5\)

Symptoms may range from mental status changes associated with delirium or dementia, to a range of psychiatric symptoms like delusions, hallucinations, euphoria, depressive personality changes, aggressiveness, apathy, and cognitive deficits.\(^2,4,5\)

The diagnosis of neurosarcoïdosis is difficult given the unusual clinical presentation of nonspecific psychiatric symptoms and headaches. A high index of suspicion is required on behalf of doctors especially general physicians/internists and psychiatrists for recognition of such atypical presentation of neurosarcoïdosis.

Our case highlights that neurosarcoïdosis can present with neuro-psychiatric manifestations, in addition to hypothalamic disturbances which is manifested in form of diabetes insipidus.

Neurosarcoidosis is one of the many medical illnesses that may present with psychiatric symptoms requiring a careful search for organic etiologies when medical evaluation raises clinical suspicion. In our patient no response to antidepressants, history of uveitis, low complement levels prompted us to do neuroimaging. Further finding of high ACE levels, high signal lesions on MRI brain and presence of hilar lymphadenopathy confirmed our diagnosis.

The prompt and definitive improvement of neuro-psychiatric symptoms noted in our patient with high-dose steroid therapy further supports the diagnosis of neurosarcoïdosis. Such response to steroid therapy has been reported in other cases.\(^6\)

CONCLUSION

The diagnosis of isolated neurosarcoïdosis should be kept in mind in young patients with nonspecific somatic complaints and depressive symptoms especially if there is no cause/stressors for such symptoms. Keeping a high index of suspicion is crucial in reaching to the diagnosis of neurosarcoïdosis.

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Conflict of interest: Author declares no conflict of interest.
Funding disclosure: Nil

Author's contribution:
Farheen Niazi; data collection, data analysis, manuscript writing, manuscript review
Tariq Hussain; data collection, data analysis, manuscript writing, manuscript review