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Case Report

Neurosarcidosis Presentation with Psychotic Features- A Case Report

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Abstract

Sarcoidosis is an inflammatory, granulomatous condition of unknown etiology, involving multiple organs. In nearly 5% of the patients, central nervous system involvement can occur, manifesting as neurosarcoidosis. Neurosarcoidosis presents in many different forms such as cranial mononeuropathy with facial nerve most commonly involved either unilateral or bilateral, hypopituarism, seizures, communicating or non-communicating hydrocephalus and meningeal involvement. In our case, the patient presented with features of delusion and agitation which is a very rare presentation of neurosarcoidosis. In less than 20% cases of neurosarcoidosis, we have patients with some psychotic features. Our case is a 46-year-old Asian male, who was brought to the emergency department by his partner with psychotic features who was later diagnosed by the help of radiological findings and history of pulmonary involvement. Conclusion is to raise awareness for keeping neurosarcoidosis in mind when a patient presents with these kind of symptoms.

Keywords: *Neurosarcidosis; Psychotic Features; Sarcoidosis*

Introduction

Sarcoidosis is a multi-organ granulomatous disorder of most probably an autoimmune etiology that affects individuals worldwide. It is characterized pathologically by the presence of non-caseating granulomas in involved organs. The most common organ involved are the lungs and the lymph nodes [1]. The incidence of Sarcoidosis is not uniform throughout the world. According to an estimate average incidence of 16.5 per 100,000 in men and 19 per 100,000 in women. The disease is less common in most of the Asian countries and on the other hand, its incidence is high in some of the developed European countries like Sweden and Iceland where approximately incidence rate is more than 60 per 100,000 [2]. In the United States, the disease is more common in people who belong to the group of African Descent where the reported incidence is 35 cases per 100,000, on the other hand, Caucasians have an incidence rate of 10.9 per 100,000 [3]. The disease is not only limited to the pulmonary or endothelial organs but it can affect almost every organ of the boy including dermal, cardiac, musculoskeletal and even nervous system. Neurologic complications occur in approximately 5 to 10 percent of patients with sarcoidosis [4]. Neurosarcoidosis presents in many different forms such as cranial mononeuropathy with facial nerve most commonly involved either unilateral or bilateral, hypopituitarism, seizures, communicating or non-communicating hydrocephalus and meningeal involvement [5-8]. We here are reporting a case in which the patient presented with features of delusion and agitation which is a very rare presentation of neurosarcoidosis. Reportedly only 1% patients of sarcoidosis present with these psychotic symptoms [9].

Case Report

A 46-year-old Asian male, a diagnosed case of sarcoidosis from last 16 years was brought to the emergency department by his partner with presenting complains of agitation for the past few weeks. Patients had some delusional thoughts and his wife told us that he considers himself a famous tv news reporter. According to her, there has been a gradual increase in his sleep as well. He used to sleep 6 - 7 hours daily but for the last one and half month, the daily sleep requirements have increased to more than 13 hours. There have been multiple absentees from his job because of his excessive somnolence. His partner told us that the most worrying episode occurred today when he woke up with a disoriented speech. His thoughts appeared disorganized and he became aggressive when he was confronted.

The patient had a past history of consumption of multiple illegal substances like marijuana and few episodes of cocaine consumption but that was 8 - 9 years back. He has no other personal or family history of any psychiatric illness. Surgical history includes a laparoscopic cholecystectomy which was done five years ago. He had been under regular follow up care with his pulmonologist for the sarcoidosis management. There was no dermal, optic or joint involvement noted in any of his previous visits. Excessive fatigue, mood disturbance and a decrease in libido were the other symptoms mentioned at the time of patient evaluation.

During the interview with a psychiatrist in the emergency department, slight psychomotor retardation was noted. His mood was normal, constricted affect which was otherwise appropriate. Patient had no suicidal or homicidal tendency. His judgment, insight and impulse control were normal. On mental status examination, he was oriented to time, place and person. Vital signs were normal with pulse 68 beats per minute; blood pressure 124/86 mm Hg; afebrile. Pulmonary, cardiac, joint, skin, neuro exam including cranial nerves and peripheral nerve examination was unremarkable. A limited ophthalmologic examination of visual fields along with visual acuity, reflexes, and ocular muscle movements were normal. There were no signs of meningeal irritation. Chest radiographs showed hilar adenopathy and some parenchymal changes which are typically present in the case of sarcoidosis. The urine toxicology screen was also negative. As a part of routine examination, 12 lead electrocardiographs was ordered which showed non-specific repolarization changes in the chest leads. In the next step Gadolinium enhanced brain magnetic resonance imaging (MRI) was ordered to see any visible abnormality. It showed some mild enlargement of the ventricles, dense enhancement of the basal cisterns and parafalcine leptomeninges with adjacent bi-frontal edema that were consistent with neurosarcoidosis. After the MRI, we conducted an electroencephalogram which showed some non-specific pattern of generalized slowing, and sporadic bi-temporal spikes but without any propagation of the activity.

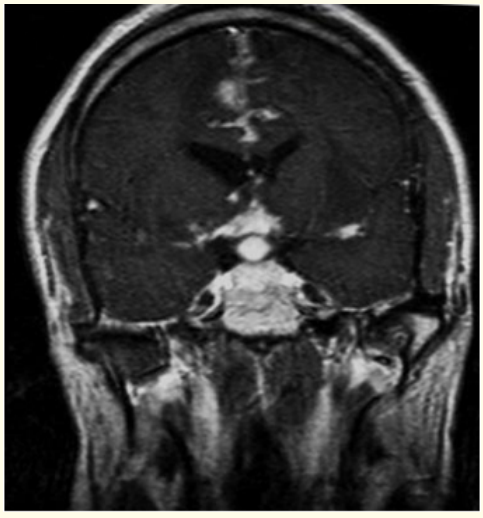

Figure: *Showing MRI image showing supra-sellar enhancement suggestive of sarcoidosis.*

Laboratory evaluation showed some strange findings which somehow revealed the possible cause of his low energy, increased sleep, and loss of libido. These laboratory findings were consistent with the state of pan hypopituaritism.

Test	Result	Normal Values
Serum TSH	0.045 microIU/mL	0.35 - 4.8 microIU/mL
Free thyroxin	0.68 ng/dL	0.89 - 1.90 ng/dL
Serum FSH	0.7 U/ml	1.0 - 14.0 U/ml
Luteinizing hormone	< 0.06 U/L	1.5 - 9.3 U/L

TSH: Thyroid Stimulating Hormone; FSH: Follicular Stimulating Hormone.

The patient was then started with a high dose of corticosteroids for the neurosarcoidosis treatment the dose of corticosteroids was adjusted to 60 mg daily. The patient was hospitalized for 08 days during which he had few episodes of mood disturbance and headache but there were no episodes of agitation and delusions. On 9th Day he was discharged from the hospital on oral corticosteroids with a follow up from medicine department for the management of hypopituitarism.

Discussion

This case exemplifies a complex clinical presentation encountered with neurosarcoidosis. Our case, a diagnosed patient of sarcoidosis with pulmonary involvement, presented with psychiatric symptoms highly suggestive of thought disorder. The patient was also reported to be agitated, aggressive and increasingly somnolent. The previous history of illicit drug abuse, sarcoidosis, and current delusions raised concerns for multiple suggestive etiologies. Further evaluation and neuroimaging supported the presumptive diagnosis of neurosarcoidosis.

Sarcoidosis is an inflammatory, granulomatous condition of unknown etiology, involving multiple organs. In nearly 5% of the patients, central nervous system involvement can occur, manifesting as neurosarcoidosis [10]. Cranial nerve involvement being the most frequent [5] while meningitis, hydrocephalus, peripheral neuropathy, encephalopathy, and seizures can be the clinical presentations of neurosarcoidosis [6,10]. In addition to neurological symptoms, neurosarcoidosis can present with psychosis and disorders of mood, memory, and thought. Some cases of delirium, dementia, hallucinations, delusions, euphoria, agitation and cognitive dysfunctions have also been reported [10]. There should be a higher level of suspicion for neurosarcoidosis in patients with previous sarcoidosis and neurological and psychiatric symptoms. The complex and subclinical nature of symptoms seen in CNS sarcoidosis make it challenging to correctly diagnose and manage the condition [11].

When the diagnosis of neurosarcoidosis is in question extra neural site involvement should be sought. Chest X-Ray or CT scan should be performed as an initial diagnostic tool in suspicious cases. Around 24 - 68% of the patients presenting with neurosarcoidosis have radiological findings on chest radiology [6].

Histopathology is the gold standard in diagnosing but difficult to perform but the inaccessibility of affected site may make it difficult to perform at times [7]. Serum angiotensin converting enzyme levels are helpful but are non-specific and are not necessarily elevated in isolated CNS sarcoidosis [13]. Ophthalmic, sinus or endoscopic examination can also be performed to locate the extraneural site of sarcoidosis. At times magnetic resonance, gallium and fluorodeoxyglucose positron emission tomography scans are ordered to locate the site and it might reveal an accessible site to perform histopathology.

Though magnetic resonance imaging provides indirect information of neurosarcoidosis, this still is the most sensitive tool to evaluate neurosarcoidosis [13].

CSF analysis has been frequently used in the evaluation of neurosarcoidosis [13].

Corticosteroids have been the mainstay treatment of neurosarcoidosis and early intervention with corticosteroids is still advocated for neurosarcoidosis. But newer agents are also emerging as effective treatment options. Immunosuppressive therapy with methotrexate, cyclophosphamide, and azathioprine are being frequently considered. Role of antitumor necrosis factor agents like infliximab and mycophenolate has also been found useful to address the neurological involvement of sarcoidosis [12].

Following flowchart will help us understand the management plan of neurosarcoidosis

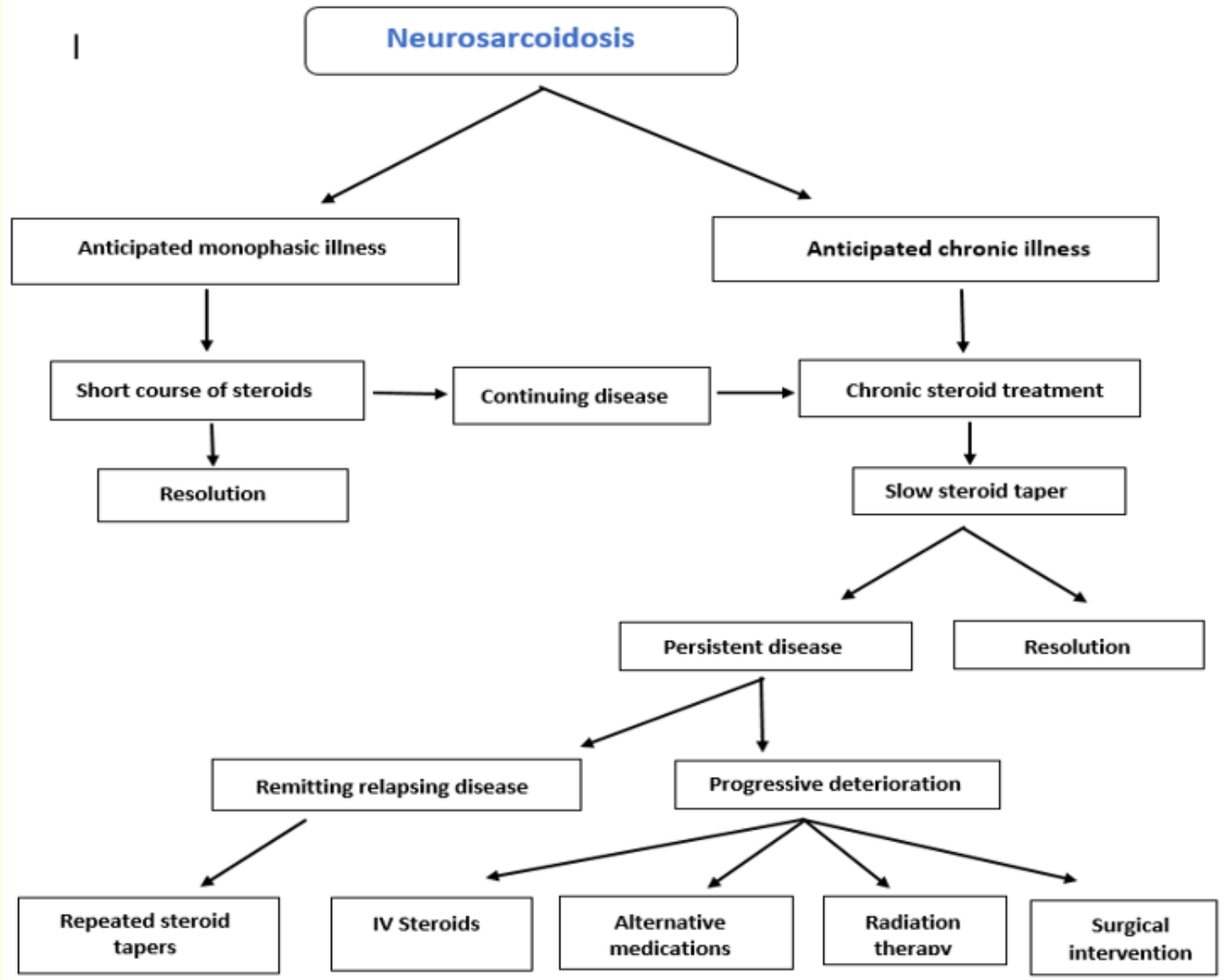

Figure 1

Conclusion

Conclusion is to raise awareness for keeping neurosarcoidosis in mind when a patient presents with these kind of symptoms.

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