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A Case of Systemic Sarcoidosis presenting with UMN Paraparesis and Neuropsychiatric Manifestations

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ABSTRACT

Introduction: Sarcoidosis is a multiorgan granulomatous illness with uncertain cause. Neurological sarcoidosis is a rare manifestation of this disease and occurs in approximately 5-10 percent of patients.[1] Neuroendocrine dysfunction, focal or multifocal encephalopathy, peripheral neuropathy, myopathy, aseptic meningitis, and cranial mononeuropathy are common neurological disorders.

Case-summary: We describe a case report of a 43-year-old woman who presented with chief complaints of bilateral lower limb weakness for two months and impaired sensorium for one day. She also gave a history of lesions on her nose and cheeks and edema of upper and lower limb digits for the last six months. During the hospital stay, the, the patient developed a sudden onset of blurring of vision. On neurological examination, the patient had reduced power in lower limbs, 2/5 at all movements bilaterally, along with increased tone, atrophy of lower limbs, and exaggerated reflexes. Plantar reflex was extensor bilaterally. There were no sensory or autonomic features. Contrast MRI done of the brain and spine showed multifocal non-enhancing T1 hypointense and T2/FLAIR hyperintense lesions involving the cortex and subcortical white matter with thickened and enhancing optic chiasma, including the pre- and retro-chiasmatic region up to the right lateral geniculate body, and leptomeningeal enhancement in the brain along with multiple ill-defined areas of enhancement along the length of the spinal cord in the cervicodorsal region with associated patchy areas of leptomeningeal enhancement. CECT Thorax revealed multiple subpleural, perifissural, and peribronchovascular nodules coalescing around the central bronchovascular structures; smooth interlobular septal thickening in bilateral lung fields; and multiple discrete and confluent bilateral hilar and mediastinal lymph nodes, with some of them showing calcifications within and a few showing heterogenous enhancement. Biopsy of skin lesion showed granulomas. A diagnosis of systemic sarcoidosis with neurological involvement was made and treated with IV methylprednisolone pulse 1 gram OD for 3 days, followed by oral steroids 60 mg OD. The patient showed significant clinical improvement in power, sensorium, skin lesions and neuropsychiatric symptoms.

Conclusion: This instance emphasizes the necessity of considering sarcoidosis as a differential diagnosis and initiating early therapy in individuals with myelopathies and other multisystem involvement.

Keywords: Biopsy, Neuroendocrine, Sarcoidosis.

INTRODUCTION

Sarcoidosis is a multisystem condition that mainly affects young adults under 40, with peak between the ages of 20 and 29, with slight female preponderance. A suitable clinical and/or radiological image, histological evidence of noncaseating granulomas, and the elimination of other conditions that could provide a comparable histological or clinical picture are the basis for the diagnosis of sarcoidosis.

Neurologic issues are the first symptom in 50–70% of cases of sarcoidosis, and they affect 5–10% of individuals initially. Research indicates that rates of asymptomatic involvement may be higher because in one study, neurosarcoidosis was found at postmortem in up to 15% of people with systemic illness who had never received a diagnosis.[1] The three primary criteria used to diagnosis sarcoidosis are a suitable clinical presentation, the presence of non-necrotizing granulomatous inflammation in one or more tissue samples (though this is not always required), and the exclusion of other granulomatous disorders.[3]

CASE SUMMARY

A 43-year-old female presented with bilateral lower limb weakness for two months and altered sensorium for one day. Weakness of lower limbs started from the right lower limb, initially noticed as slippage of

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slippers from the right foot, which later progressed to the left lower limb. The weakness progressed over time. Initially she could walk with a stick; however, gradually she became bedbound. She complained of a band-like sensation around the umbilicus and a tingling sensation in the lower limbs. Around 3.30 AM, on the day of the presentation, family members noted tightening of lower limbs followed by clenching of teeth, loss of speech for around 1 hour followed by a period of slurred speech. She regained normal speech after a few hours. This was associated with a single episode of non-projectile, non-bilious vomiting. There was no history of tongue bite, drooling of saliva, up-rolling of eyes, or incontinence during this time.

On further evaluation, the patient provided a history of lesions on her nose and cheek and multiple swellings on her hand for six months, along with a history of abnormal behaviour and irrelevant talks intermittently for the last 2-3 months and complaints of a weak stream, difficulty in voiding, and a feeling of incomplete evacuation of her urinary bladder for around one year. She had been admitted for the same six months back and was diagnosed with bilateral hydroureteronephrosis and had a Foley urinary catheter inserted, which has been in situ since then. There was no history of fever, headache, shortness of breath, or cough.

On examination, she was confused initially with GCS E3V4M5, but by the next day, she was well oriented. However, she had intermittent episodes of altered behaviour over the next few days. She had ill-defined erythematous indurated plaques with perilesional hyperpigmentation over the tip of her nose and left cheek. She also had fusiform swelling of the right middle finger and left second toe. She had no thickened nerves. Neurological examination revealed atrophy, increased tone, and motor weakness of bilateral lower limbs. Knee and ankle reflexes were brisk bilaterally. There was an extensor plantar reflex on both sides. Sensory examination was normal. The right pupillary reflex was sluggish. Other system examination was unremarkable.

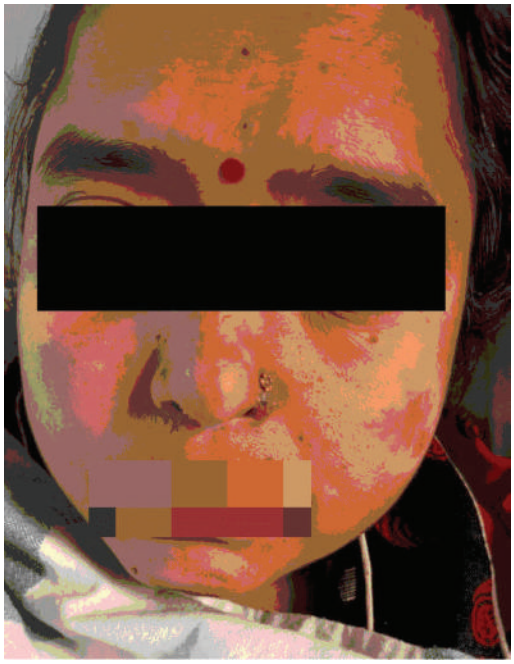

Figure 1: Erythematous indurated plaques with perilesional hyperpigmentation over the tip of her nose and left cheek.

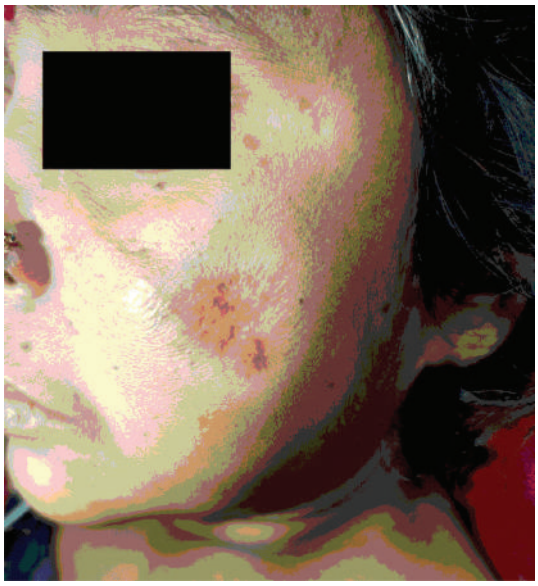

Figure 2: Erythematous indurated plaques with perilesional hyperpigmentation over left cheek.

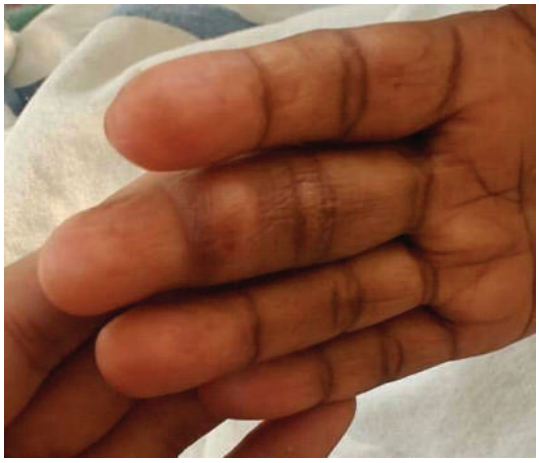

Figure 3: Fusiform swelling of the right middle finger.

Table 1: CSF analysis.

Investigation	Result	Normal range
Hemoglobin	9.3 mg/dL	12-15 mg/dL
WBC	8890 µL	4000-10000/uL
Platelet	371000/uL	150-450k/uL
Urea	26 mg/dL	13-43 mg/dL
Creatinine	0.52 mg/dL	0.7-1.3 mg/dL
Calcium	8.25 mg/dL	8.6-10 mg/dL
CRP	5.8 mg/dL	0-5 mg/dL
Vitamin B12	>1000	More than 190
HIV	Non-reactive	
Blood culture	sterile	
IGRA for TB	Non-reactive	

Contrast MRI of the brain and whole spine showed multifocal non-enhancing T1 hypointense and T2/FLAIR hyperintense lesions involving the cortex and subcortical white matter with thickened and enhancing optic chiasma, including the pre- and retrochiasmatic region up to the right lateral geniculate body, and leptomeningeal enhancement in multiple brain areas along with multiple ill-defined areas of enhancement along the length of the spinal cord in the cervicodorsal region with associated patchy areas of leptomeningeal enhancement. CECT of the thorax revealed multiple subpleural, perifissural, and peribronchovascular nodules, with nodules coalescing around the central bronchovascular structures, smooth interlobular septal thickening in bilateral lung fields, and multiple discrete and confluent bilateral hilar and mediastinal lymph nodes. Some of them had calcifications within, and a few had heterogeneous enhancement.

Table 2: CSF analysis.

Investigation	Result	Normal range
Cell count and microscopy	10 cells, 100% mononuclear	Less than 5 cells
Protein	487 mg/dL	15-40 mg/dL
Glucose	32 mg/dL	40-70 mg/dL
Gram stain	Negative	
ZN Stain	Negative	
CBNAAT	Non-reactive	

Histopathology of the skin lesion showed granulomatous pathology with ZN stain for leprosy and TB being negative. She complained of decreased vision during her hospital stay. An ophthalmology consultation was taken. They noted bilateral pale discs, tessellated fundi, and peripapillary atrophy.

A diagnosis of systemic sarcoidosis involving skin, lung, and the nervous system (myelopathy, optic nerve, and laboratory evidence of aseptic meningitis) was made. Three doses of intravenous methylprednisolone pulse 1000 mg followed by oral prednisolone 1.5 mg/kg/day were administered to the patient. She showed remarkable improvement in skin lesions, which almost disappeared during the hospital stay. There was partial recovery of the motor power in the lower limbs.

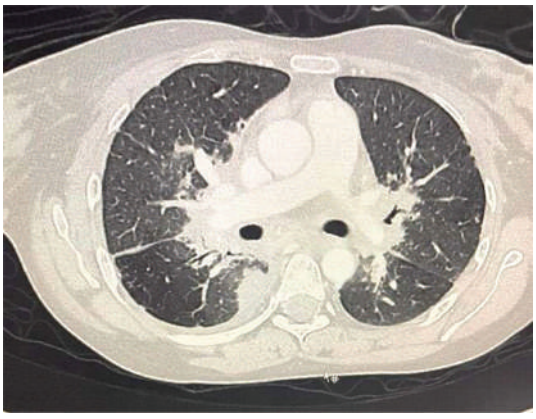

Figure 4: HRCT window showing Perifissural and peribronchovascular nodules, with nodules coalescing around the central bronchovascular structures.

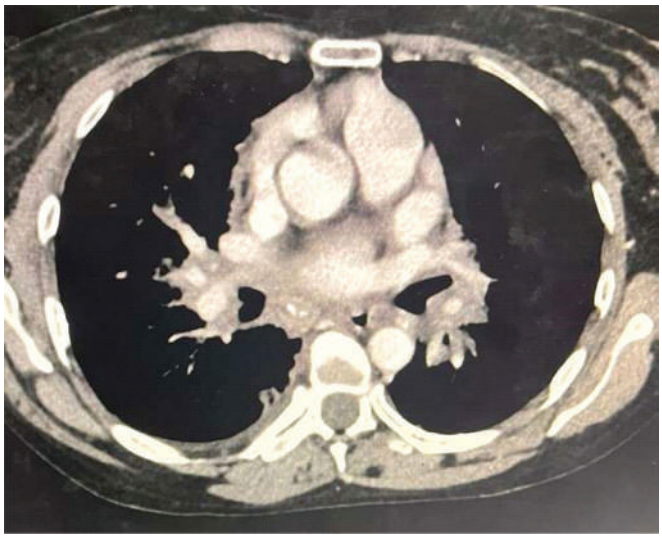

Figure 5: Bilateral hilar and mediastinal lymph nodes.

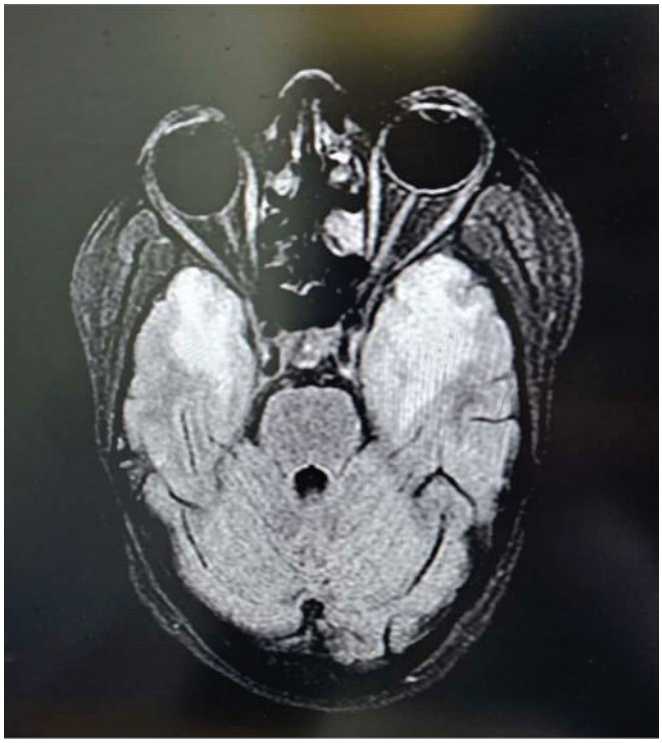

Figure 6: CEMRI Brain showing Multiple hyperintense areas in cortex and subcortical white matter.

DISCUSSION

Sarcoidosis is considered to result from an exaggerated cellular immune response in genetically predisposed individuals exposed to environmental triggers. Reported prevalence varies globally, with estimates ranging from fewer than 1 to over 40 cases per 100,000. While the lungs and lymphatic system are most frequently involved, the nervous system may also be affected, sometimes as the initial or only manifestation.[2] Neurologic complications occur in approximately 5 to 10 percent of patients with sarcoidosis.

Because of its non-specific clinical presentation and neuroradiological imaging characteristics, intracranial neurosarcoidosis remains a very difficult diagnosis, particularly in the absence of systemic signs of the disease. Any area of the brain may be affected by intracranial neurosarcoidosis, which typically affects the cranial nerves and has a preference for the basal leptomeninges. This can lead to a variety of clinical symptoms. The commonest clinical manifestation is involvement of the cranial nerves. Extra- or intra-axial parenchymal lesions and nodular or widespread leptomeningeal thickening are features of intracranial sarcoid. On neuroradiological imaging, intracranial sarcoid may resemble several types of meningitis as well as intracranial mass lesions like meningioma, lymphoma, and glioma. Magnetic resonance imaging is a very sensitive diagnostic method for detecting cerebral abnormalities brought on by neurosarcoidosis, and angiotensin-converting enzyme levels in serum and cerebrospinal fluid can be increased, decreased, or normal. Although the results are not specific, lumbar puncture is useful in ruling out other neurological illnesses, especially infectious ones.[4]

Treatment consists of Glucocorticoids[5] and other immunosuppressive drugs, including mycophenolate mofetil, azathioprine, methotrexate, and cyclophosphamide. Infliximab may be effective in refractory pulmonary as well as extrapulmonary manifestations.[6] About two-thirds of patients with neurosarcoidosis may experience monophasic, relapsing-remitting, or chronic progressive disease pattern. High-risk patients may have hydrocephalus or parenchymal illness of the central nervous system (CNS). Because of sarcoidosis or the immunosuppressive medications other than glucocorticoids, these patients are susceptible to malignancies, especially lymphoma.[7]

CONCLUSION

The pathogenesis of sarcoidosis, a multisystem granulomatous disease, remains uncertain. Neurological involvement in aseptic meningitis, myelopathy, mononeuropathies, polyneuropathies, myopathy, and cranial nerve involvement is present in about 5% of cases. Establishing the diagnosis requires a comprehensive approach incorporating clinical suspicion, imaging, and histologic evaluation. When there are no respiratory symptoms, as in this instance, the diagnosis can be difficult and calls for a high index of suspicion. In refractory cases, immunosuppressive therapy and steroids are the main forms of treatment. Prompt initiation of therapy can result in meaningful clinical recovery and reduce the risk of long-term sequelae. The disease can have a variable clinical course.

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