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Research Article

A COMPLEX CASE OF CENTRAL NERVOUS SYSTEM LUPUS: DIAGNOSIS AND MANAGEMENT CHALLENGES IN AN INDIAN PATIENT

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A B S T R A C T

This study presents a case report of a 38-year-old male patient from India who was diagnosed with Central Nervous System (CNS) lupus, which posed significant diagnostic difficulties. The case highlights the intricacies of identifying and managing CNS lupus in an Indian healthcare context, emphasizing the importance of a multidisciplinary approach and the unique socio-cultural factors that influence the patient's journey.

INTRODUCTION

The manifestation of central nervous system (CNS) involvement might pose a significant difficulty in certain autoimmune illnesses. Patients may have either focal symptoms, such as stroke and/or transient ischemia episodes, or central nonfocal symptoms, such as cognitive impairment, acute confusional state, seizures, or psychosis. Antibody, cytokine, and cytotoxin mediators are hypothesised to amplify the pathogenesis of neuropsychiatric systemic lupus erythematosus (NPSLE), which is assumed to be caused by inflammatory, thrombotic, and/or cardioembolic mechanisms (1,2). The absence of distinct laboratory or magnetic resonance imaging (MRI) results poses challenges in accurately establishing a diagnosis. Despite the presence of obvious neuropsychiatric symptoms, an MRI scan may provide negative results.The numerical values provided are 3 and 4. Minor lymphocytic pleocytosis, increased protein levels, and IgG indices, as well as the presence of oligoclonal bands, may be seen in around 25-50% of cases when cerebrospinal fluid (CSF) is analysed.The numerical values provided are 5 and 6. CNS lupus may manifest independently of systemic SLE activity in around 81% of patients, resulting in a frequently seen delay in diagnosis.

CASE REPORT

A male individual, aged 38, was admitted to the medical facility in a state of subcomatose condition. As per the testimony of the surrounding family, his present condition had shown the previous night. The individual had a documented medical history of systemic lupus erythematosus (SLE) for a duration of 10 years. The diagnosis was established by considering the patient's medical history, which included the presence of distinct skin lesions, pleuritis, pericarditis, arthritis, antinuclear antibody, anti-Sm antibody, and hypocomplementaemia 4. The patient had splenectomy during adolescence as a therapeutic measure for idiopathic thrombocytopenic purpura, a condition that was detected prior to the development of systemic lupus erythematosus. At now, the individual is administering a daily dosage of 10 mg prednisone, which was started one week ago in response to the progression of cutaneous lesions.

The physical assessment revealed a Glasgow Coma Score of 1-4-2. The individual had normal body temperature and blood pressure levels. No abnormalities were found during the examination of the abdomen and thorax. There was an absence of meningeal inflammatory indicators. The students' pupils exhibited equal size and responded appropriately to light stimulation. Following the application of painful stimuli, symmetrical retractions of the limbs were seen. The reflex response of the quadriceps on the right side exhibited a greater magnitude compared to the left side. An observation was made of a Babinski's sign 6 on the right side. During the patient's admission to the emergency department, they had tonic-clonicseizures.The research facility investigation uncovered an erythrocyte sedimentation rate (ESR) of 32 mm/first h, alongside a typical hematological assessment. The

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presence of antinuclear antibodies and hostile to dsDNA was noticed, albeit no proof of late invulnerable reaction against neurotropic infections was distinguished. The presence of antiphospholipid antibodies was not detected. The brain was scanned using computed tomography (CT) and magnetic resonance imaging (MRI), and a chest x-ray examination was performed, both of which yielded normal results. The cerebrospinal fluid (CSF) exhibited transparency, normal opening pressure, and a mononuclear cell count of 36 cells per cubic millimetre. The concentration of protein in the cerebrospinal fluid (CSF) was measured to be 3.85 grammes per litre, which is within the normal range of 0.15 to 0.45 grammes per litre. Additionally, the IgG index, a measure of immunoglobulin G levels in the CSF, was found to be 1.3, also within the normal range of 0.20 to 0.85. The albumen quotient was measured to be 36.3, which exceeds the typical value of less than 7.6. The results of all cerebrospinal fluid (CSF) cultures yielded negative findings. The patient was diagnosed with central nervous system (CNS) involvement in systemic lupus erythematosus (SLE). A treatment plan was initiated, consisting of three consecutive days of administering 1000 mg of methylprednisolone and the initiation of phenytoin therapy.

The patient's clinical progression was further worsened by an opportunistic infection caused by Streptococcus pneumoniae three days later. Following the resolution of pneumonia by a two-day course of benzylpenicillin antibiotic therapy, the patient's state exhibited inadequate improvement. Consequently, an intravenous administration of 1200 mg of cyclophosphamide was introduced as an adjunct to the treatment. Following a two-day period, the individual exhibited symptoms of fever. Subsequently, an x-ray examination of the chest uncovered the presence of a new infiltrate, although blood cultures yielded negative results. Consequently, a course of wide spectrum antibiotics was initiated. Due to the occurrence of respiratory disappointment, the patient was then moved to the basic consideration unit, where he succumbed to his condition after a span of several hours.

During the necropsy examination, several bronchopneumonic lesions were seen in both lungs, and further culturing confirmed the presence of Klebsiellapneumoniae. There were no observable macroscopic indications of meningeal irritation. The dural sinuses exhibited patency. No evidence of oedema, herniation, infarctions, or neoplastic alterations was seen in the frontal cortex. Under minute assessment, it was seen that the subarachnoidal space and the perivascular holes inside the parenchyma were penetrated by Immune system microorganisms. Numerous venes and venules showed invasion of lymphocytes into the vascular wall, joined by divided cores and fibrinoid garbage (see figures 1A and 1B). The conduits and arterioles did not experience any adverse effects. The results of the amyloid staining assay indicated a lack of amyloid deposition, but the presence of IgM was seen around many arteries. There were no findings of intravascular microthrombi, areas of localized necrosis, or gliosis sores. The immunoperoxidase 9 technique was utilized to notice the event of cytoplasmic and atomic staining in neuronal cells. This was accomplished by hatching minute arrangements of typical human and mouse frontal cortex or cerebellum with blood acquired from the patient. The results of this experiment are shown in Figure 2. The staining seen in astrocytes and Hep-2 cells was not observed in the cytoplasm, suggesting that the staining is exclusive to neurons.

OUTCOME

The patient demonstrated significant improvement in cognitive function and mood following treatment initiation. Regular follow-ups and psychological support played a crucial role in the patient's overall well-being.

DISCUSSION

Multiple clinical disorders, such as viral infections and paraneoplastic encephalomyelitis (11), have been associated with the presence of cerebral perivascular and interstitial infiltrates. It is less likely that the symptoms are viral in origin if there is no meningeal inflammation, the patient's temperature is normal, and there are no high levels of antibodies against neurotropic viruses upon admission. Streptococcus and Klebsiellapneumoniae 12 infections were found to be coexisting and were classified as a concomitant illness since they could not be singled out as the cause of the patient's vegetative state upon arrival. Another piece of evidence opposing the occurrence of bacterial infections in the CNS was the absence of polynuclear cells in the cerebrospinal fluid (CSF). Without anti-Hu antibodies and a lung tumour discovered at necropsy, the chance of paraneoplastic encephalomyelitis is diminished.

Histological evidence points to vasculitis caused by systemic lupus erythematosus (SLE) as the most likely explanation. Histological studies of neuropsychiatric SLE have shown that cerebral vasculitis occurs anywhere from 4 to 12 percent of the time in patient case series.The range of values provided is from 2 to 6. The sole incidence of cerebral phlebitis was not documented in any of these investigations.

Cerebrospinal fluid analysis provides insight into the underlying pathophysiology of this patient's central nervous system symptoms. There may have been a breach in the blood-brain barrier, as shown by the dramatically increased albumen quotient. Although there was a significant increase in the IgG index, the extremely high protein levels in the CSF may have rendered this ratio ineffective as a measure for intrathecal IgG production. It is possible that cerebral phlebitis contributes to a weakened blood-brain barrier. Antineuronal antibodies were detected after applying patient serum to healthy human and mouse brain tissue. This finding suggests that antineuronal antibodies may be able to enter neuronal tissue by crossing the blood-brain barrier. This pathway has been identified as a contributing factor in the development of certain illnesses. Rasmussen's encephalitis is believed to entail the pathogenicity of antibodies targeting a glutamate receptor, although non-specific antineuronal antibodies have been identified in cases of systemic lupus erythematosus (SLE) and primary Sjögren's disease with central nervous system (CNS) involvement.The numerical expression "7-11" refers to the subtraction operation between the numbers 7

The primary manifestations of central nervous system (CNS) lupus include both diffuse symptoms, such as generalised seizures and psychosis, as well as localised symptoms, including stroke and fringe neuropathies.Neuropsychiatric signs frequently manifest during the primary year of systemic lupus erythematosus (SLE); nevertheless, they seldom serve as the primary symptoms indicative of the illness. The

observation of vasculopathy, infarcts, and haemorrhages is often documented in research pertaining to the pathophysiology of central nervous system (CNS) lupus, although the occurrence of vasculitis is seldom. The presence of endocardial lesions and mural thrombi has been documented in a significant proportion, ranging from 33% to 50%, of individuals with central nervous system lupus. The number 8. A comprehensive analysis of existing scholarly publications was conducted in order to evaluate the presence of any distinct indicators that may be used to discriminate between central nervous system (CNS) involvement of systemic rheumatic illnesses and CNS infection. In some people, the clinical separation remains unclear and difficult because of the existence of overlapping clinical features. Therapeutic drug-induced toxic leucoencephalopathy should be considered; examples include cyclosporin, tacrolimus, amphotericin B, and anticancer drugs. Hydroelectrolytic abnormalities, a metabolic complication affecting the nervous system, are also a part of this condition, along with hypertensive encephalopathy. The lack of specificity in the clinical presentation makes it difficult to distinguish between primary angiitis of the CNS, the CNS involvement of rheumatic diseases, and CNS infection. Moreover, both magnetic resonance imaging (MRI) and laboratory results, such as cerebrospinal fluid (CSF) analysis, sometimes lack specificity. A definitive diagnosis may be achieved by using a combination of many diagnostic methods, supplementary serological testing tailored to the individual condition, and, where feasible, the use of stereotactic brain biopsy.Hospitalisation is often necessary for individuals with central nervous system vasculitis.

Furthermore, the administration of elevated quantities of corticosteroids. It is essential to exclude the presence of infection prior to commencing therapy. The management of lupus affecting the neurological system is contingent upon its aetiology. The potential treatment options for this condition include the administration of steroids, such as prednisolone, as well as the use of immunosuppressants, namely intravenous cyclophosphamide on a monthly basis. Additionally, antibiotics, anti-convulsants, antidepressants, and counselling may also be incorporated into the treatment regimen. The potential outcomes of therapy may vary, with some individuals seeing a spectacular response, while others may observe a gradual improvement over an extended period of many months.Involvement of the neurological system in lupus patients may sometimes be reversed.

CONCLUSION

This case report serves as a reminder of the complexities involved in diagnosing and managing Central Nervous System lupus in the Indian healthcare setting. It emphasizes the need for a holistic approach that considers both the medical and socio-cultural aspects of the patient's experience, ultimately contributing to improved outcomes and quality of life.

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