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Case Report

Rehabilitation Approach in Spastic Hemiparetic Cerebral Palsy Superimposed with Primary Central Nervous System (CNS)Vasculitis: Case Report

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ABSTRACT

Introduction: Rehabilitation of cerebral palsy (CP) varies depending on the gross motor, fine motor and communication abilities. The motor disorders seen in CP are frequently accompanied by disturbances of sensation, cognition, communication, perception, and/or behavior disorders; thus, therapy approaches are arranged to meet the individual patient's needs. Having primary Central Nervous System (CNS) vasculitis on top of cerebral palsy adds more complexity to the medical condition as more medical complications needs to be taken care of.

We report a case of a 20 year-old-female with spastic hemiparetic cerebral palsy with gross motor functional classification system I (GMFCS I) presented with seizures, fever and altered mental status. Upon further evaluation she was found to have anemia that was managed with iron supplements. Her blood workup was non-conclusive. Brain imaging showed multiple infarcts in the bilateral partial and occipital lobes. Brain biopsy showed inflammatory infiltrate with perivascular lymphocytic extending into the vessel wall that is consistent with primary Central Nervous System (CNS) vasculitis.

Patient was discharged to in-patient rehabilitation where she received her rehabilitation with interdisciplinary team. Her neurological recovery, sleep-wake cycle, seizures, spasticity, neurogenic bowel and bladder issues were addressed. She was discharged mobilizing on a wheelchair and her continuity of care was achieved with outpatient followups.

Significance: This report illustrates the significance of physiatry involvement in complex neurorehabilitation patients and the role of interdisciplinary care in optimizing quality care along with their caregivers throughout their rehabilitation journey.

Keywords: Central Nervous System (CNS); Cerebral Palsy (CP); Gross Motor Functional Classification System (GMFCS)

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INTRODUCTION

Cerebral Palsy (CP) is a group of disorders of the development of movement and posture, causing activity limitations that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain.[1] It is the most common motor disorder among the pediatric population accounts for approximately 2 in 1000 live births, and about 98% of those children aged 4 to14 years survive to age 20 years.[2] The most common cause of CP is prematurity. Rehabilitation of CP starts in the childhood and continues as these kids get older. Rehabilitation goals changes as the care priorities vary across their lifespan and include a comprehensive evaluation of their cognitive, physical, mental skills and a smooth transition to adulthood. Having a heterogenous disorder that affects the central and peripheral nervous systems like primary CNS vasculitis on top of CP adds more complexity to the medical management and glorify the role of physiatrists in managing the associated complications.

CASE PRESENTATION

A 20 year-old-female with spastic hemiplegic cerebral palsy with gross motor functional classification system I (GMFCS I) studying a double major in university presented with 1 week history of seizures, fever and altered mental status. She had an episode of syncope that required hospital admission and further workup. Initially she was found to have anemia that required iron supplement. Her blood workup was non-conclusive. Brain imaging showed multiple infarcts in the bilateral partial and occipital lobes. Her cerebrospinal fluid analysis showed lymphocytic pleocytosis and mild to moderate elevation in protein levels. Despite being on methyl-prednisone she continued to be confused. She had a brain biopsy which showed inflammatory infiltrate with perivascular lymphocytic extending into the vessel wall that is consistent with primary CNS vasculitis. She was started on Cyclophosphamide and a rehabilitation evaluation was initiated.

She required in-patient rehabilitation that focused on improving her mental status and sleep-wake cycle. During her stay physiatrists were heavily involved and her spasticity was optimized. Timed voiding was implemented to manage her upper motor neuron bladder. Physical therapy worked on optimizing her muscles' range of motion along with occupational therapy who worked on her upper extremity and fine motor control. She required adjustments of her diet and kept on puree as per speech language pathologist's recommendations. She was discharged mobilizing on wheelchair and further follow up was recommended to continue her spasticity care with chemo-denervation as outpatient. At the time of discharge given her cerebral palsy background on top of her primary CNS vasculitis the rehabilitation team recommended that she continues physical therapy as outpatient to prevent contractures.(Figure 1)

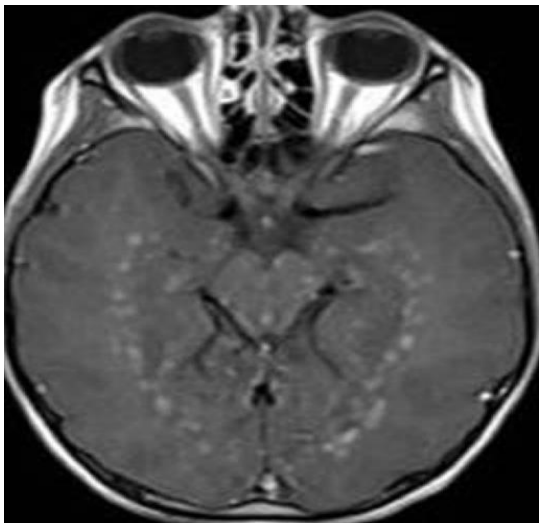

Figure 1: MRI T1W image shows multiple punctate lesions mainly around basal ganglia similar to our patient's findings.(Adopted from RadioGraphics[3])

DISCUSSION

A multidisciplinary, integrative approach to rehabilitation in patients with cerebral palsy starts early in well-equipped and staffed centers. This approach can improve appropriate treatment of patients with cerebral palsy and ensure lower risks of re-hospitalizations and improved long-term outcomes. It also allows patients to integrate and engage in the community within their capabilities and boost their quality of lives. Having physiatrists on board guide this interdisciplinary care is vital. Physiatrists manage to optimize the patient's function through medical management of spasticity, neurogenic bowel and bladder, hip and back surveillances, sialorrhea management and making sure that other allied health care professionals apply the appropriate therapies.

Having a primary CNS vasculitis on top of cerebral palsy in very uncommon. Most patients with primary CNS vasculitis first present with a prolonged and progressive prodromal period consisting of headache and cognitive changes that can last weeks to months. The most common symptoms are headache and cognitive impairment, while other manifestations include a wide variety of neurologic symptoms, such as stroke, TIA, aphasia, visual field defects, blurred vision, double vision, seizures, ataxia, focal deficits, papilledema, intracranial bleeding, and amnesia. Most patients have a combination of symptoms, and the clinical presentation may initially resemble multiple sclerosis.

In 1988, Calabrese and Mallek proposed three diagnostic criteria for primary CNS vasculitis: history and clinical findings of acquired neurological deficit that cannot be explained after a thorough initial evaluation, classic angiographic or histopathological findings of angiitis within the central nervous system, no evidence of other systemic vasculitis or any other conditions that the angiographic or histopathologic findings can be attributed to.[4,5,6]

The diagnostic criteria clearly indicates that primary CNS vasculitis is a diagnosis of exclusion. Hence laboratory studies should be performed to rule out other causes of vasculitis like systemic vasculitis, infection or neoplasm.

In 80-90% of patients, CSF is abnormal showing mild lymphocytic pleocytosis and mild to moderate elevation in protein levels. If a patient presents with ischemic manifestations regardless of etiology, a full stroke workup should be performed with EKG, echocardiogram, lipid levels and diabetes mellitus risk stratifications.[4]

Most common MRI findings are multifocal infarcts, tumor like appearance, leptomeningeal enhancement and parenchymal enhancement.[7] Conventional angiography is frequently used when clinical suspicion is high for primary CNS vasculitis even if the results are nonspecific. The key angiographic finding is alternating areas of vessel dilatation and narrowing or stenosis that are multifocal and bilateral in the medium sized cerebral arteries. Till date, the gold standard to diagnose primary CNS vasculitis is a brain biopsy.[8] This biopsy should include samples from the dura, cortex, white matter and the leptomeninges. However, brain biopsies have a false negative result in at least 25% of the patients and it has low sensitivity for diagnosis.[9]

Treatment for primary CNS vasculitis remains an area of study, however the general recommendation is to start oral prednisone upon diagnosis. Cyclophosphamide can be added if the patient is unresponsive to glucocorticoids alone. In cases of CNS vasculitis resistant to both glucocorticoids and cyclophosphamide, TNF-alpha inhibitors, azathioprine, mycophenolate mofetil, and rituximab have been suggested as adjunctive treatment to be used along with standard therapy, though efficacy has yet to be adequately assessed.[7,8] The optimal duration of therapy is still uncertain, however some advice tapering of glucocorticoids and maintenance therapy with immunosuppressants such as azathioprine in patients who achieved remission after initial treatment. Specifically, one study found that patients who received maintenance therapy have a lower relapse rate than those who did not receive maintenance therapy. Duration of maintenance therapy should be individualized and based on repeat clinical examinations and CSF and neuro radiological monitoring.[9]

Rehabilitation in this condition requires addressing issues that comes along with cerebral palsy as well as complications associated with primary CNS vasculitis. Mental status evaluation and implementation of cognitive remediation and compensatory strategies are important in getting these patients back to a routine, addressing their cognitive deficits by working with speech language pathologists.[10] It is not uncommon that cerebral palsy is associated with strabismus and retinopathy of prematurity. Hence, frequent ophthalmological evaluation is mandated. Swallowing evaluation with modified barium swallow study and a consideration of G-tube insertion to optimize nutritional demands and prevent malnutrition. Both cerebral palsy and primary CNS vasculitis have upper motor neuron bowel and bladder. Therefore, use timed voiding and proper laxatives would help with achieving social continence and prevent urinary infections and bowel obstruction. Spasticity management is the cornerstone in optimizing their functional status. Physiatrists use a comprehensive approach including serial casting, medications, chemo-denervation and sometimes intra-thecal baclofen. Mobility outcomes depending on primary CNS vasculitis response to remission therapy and exploring ambulatory aids with or without orthosis can be considered. Physiatry evaluation of medication related side effects is part of the interdisciplinary approach and a clear communication with

the neurologist is important to provide better quality of care. Glucocorticoids like prednisone when used for long term management can have a wide variety of side effects. Some of its side effects are can include: cushingoid features, skin thinning, cataracts, increased ocular pressure, osteoporosis, mood disorders and hyperglycemia.[11,12] Another therapy used in conjunction with glucocorticoid is cyclophosphamide. This therapy in particular can cause infection, transitional cell carcinoma of the bladder, infertility and hearing problems.

In conclusion, this report illustrates the significance of physiatry involvement in complex neurorehabilitation patients and the role of interdisciplinary care in optimizing quality care along with their caregivers throughout their rehabilitation journey.

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