

Chronic Inflammatory Demyelinating
Polyneuropathy in Older AdultsBegum R^{1,2*} and Suh T¹⁻³¹Division of Geriatric & Palliative Medicine, Department of Internal Medicine, University of Michigan, USA²VA GRECC, Ann Arbor VA Hospital, USA³Huron Valley PACE, Ypsilanti, USA

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Abstract

Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is a rare immune mediated disorder of the peripheral nervous system which can occur at any age but is more common in older males. Clinical features can include weakness, sensory loss, and gait impairment which may lead to significant functional loss and difficulty managing ADLs and IADLs independently in the elderly population. This case report will highlight why primary care providers need to be aware of CIDP.

Introduction

CIDP is a rare neurological disorder consisting of inflammation of peripheral nerves and destruction of myelin sheath. The prevalence is estimated from 1-7.7 per 100,000 and appears to be more common in older males [1]. Prevalence is higher in males than females with the mean age of onset being 47.6 years [2]. Usual symptoms are loss of strength and sensation, difficulty climbing stairs, raising arms, pain, and imbalance¹. Older patients usually have slow progressive distal pattern unlike younger patients where proximal weakness is more common [3]. Usually a sensory or combination of sensory and motor involvement are present in older patients versus predominantly motor symptoms in younger patients [4]. Treatment consists of IVIG, plasmapheresis, and/or corticosteroids.

Case

A 70-year-old female with history of two episodes of AIDP who continued to have an inflammatory neuropathy consistent with CIDP. In 1993 she had an episode of sensory loss following a flu-like illness and established care with the neuromuscular clinic. A nerve biopsy revealed vasculitis. EMG in 2002 showed sensory greater than motor axonal neuropathy. Multiple hospitalizations for treatment of CIDP consisted of IVIG and Solumedrol. Her neuropathy limits her mobility and produces a disabling tremor. IVIG infusion is currently being provided to her at home through PACE, but there have been several instances where it was difficult to get home infusions.

Discussion

The pathogenesis of CIDP is not fully understood and it is often under diagnosed due to several factors including its rarity, many variants, and presentation of vague symptoms. CIDP accounts for about 20% of initially undiagnosed neuropathies and represents about 10% of all patients referred to neuromuscular clinics [3]. A thorough history, physical, and neurological exam should be performed to help determine the cause of neuropathy. Symptoms can often present as weakness, numbness, pain, difficulty climbing stairs, getting out of chairs, and difficulty walking which can lead to falls. Therefore, CIDP can affect quality of life owing to these functional impairments and may possibly impact mental and emotional state of patients.

Early recognition and treatment of CIDP is important to avoid significant disability. In most cases the diagnosis of CIDP can be confirmed by clinical and Electromyography (EMG) criteria and nerve biopsy may also be helpful although it is not often required [3]. Furthermore, it is important for primary care providers to understand and be able to distinguish the differences in characteristics of CIDP between younger and older adults (Table 1). Elderly patients usually have a slow progressive distal pattern unlike younger patients where proximal weakness is more common. Relapsing course is seen less frequently in the elderly population and usually a sensory or combination of sensory and motor involvement is present versus predominantly motor symptoms in younger patients [4]. In addition to the progressive course more common in older adults, this population also has more chronic health problems which may contribute to the worse prognosis.

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Table 1: Differences in characteristics of CIDP between younger and older adults.

| CHARACTERISTICS OF CIDP | YOUNGER ADULTS | OLDER ADULTS |
|-------------------------|---------------------|-------------------------|
| LOCATION OF WEAKNESS | PROXIMAL | DISTAL |
| TYPE OF COURSE | RELAPSING-REMITTING | SLOWLY PROGRESSIVE |
| SENSORY VS. MOTOR | PREDOMINANTLY MOTOR | SENSORY OR SENSORIMOTOR |
| PREFERRED TREATMENT | PREDNISONE | PLASMA EXCHANGE OR IVIG |
| PROGNOSIS | BETTER | WORSE |

Goals of treatment for CIDP should include reduction of symptoms and improvement of functional status. Treatment consists of IVIG, plasmapheresis, and/or corticosteroids. Alternative immunosuppressive regimens can also be considered if patients have not improved with conventional therapies, or has improved but has frequent relapses, or if the patient has developed adverse reactions with the conventional treatment; however, the effectiveness of these alternative treatments for CIDP remains uncertain [3]. Physical and occupational therapy as well as maintaining an exercise regimen may help with gait, balance, and endurance. Counseling can also be helpful to address symptoms of depression and anxiety related to loss of functional capacity.

Conclusion

This case highlights the importance of primary care providers to be aware of CIDP as it can cause gait abnormalities, motor, and sensory deficits which can severely limit patient's independence in performing ADLs and IADLs. Treatment options are also challenging given that patients may not be able to present to the clinic for IVIG infusions and may need home infusions. Challenges in providing home IVIG infusion include ensuring safe administration, cost of therapy, home care agencies administering infusions, and coordination among patients, healthcare providers, and health plans

[5]. Plasma exchange can also be difficult because it is invasive, can be time consuming, and requires need for special equipment and well-trained staff in facilities familiar with the procedure.³ In addition, many older adults have issues with transportation and mobility that may limit access to plasma exchange. These challenges can delay treatment; thus, primary care providers need to be aware of them.

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