

## PURPOSE

The purpose of this case report is to heighten awareness of Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsiveness to Steroids (CLIPPERS) and describe rehabilitation services that supplemented medical management of this rare condition in a recreational athlete.

## BACKGROUND/ SIGNIFICANCE

CLIPPERS is chronic inflammatory disease of the central nervous system that was first described by Pittock in 2010, and approximately 100 cases have been reported world-wide to date.<sup>1,2</sup> The average age of onset is 50 years-old, but cases have been reported in patients aged 13- 86 years.<sup>3</sup> The male to female ratio is 3:1.<sup>3</sup> The disease primarily affects the perivascular spaces of the pons, mid-brain, and cerebellum.<sup>1,2</sup> Diagnostic criteria include (1) curvilinear lesions with “pepper-like speckles” on gadolinium-enhanced magnetic resonance images usually in the pons and cerebellum but lesions have been found in the cerebrum and spinal cord, (2) corresponding clinical impairments, (3) rapid clinical and radiological improvement with steroid administration, and (4) absence of alternative diagnoses.<sup>1,2,4</sup> Medical management of the initial attack includes high doses of intravenous (IV) steroids until a reduction in clinical signs occurs, then oral steroids are administered.<sup>5</sup> During the relapse free period of the disease, oral steroids are gradually tapered to reduce the negative effects of prolonged steroid use.<sup>5</sup> Prognosis depends on how quickly CLIPPERS is recognized and steroid therapy begins, but even when treated quickly, axonal damage can occur and cause atrophy of involved structures.<sup>6</sup> Recovery is influenced by the amount of atrophy that remains.<sup>6</sup> There is a gap in the literature regarding the role of rehabilitation in management of CLIPPERS.

## CASE DESCRIPTION

A 34-year-old male warehouse worker and recreational basketball player, with no prior medical history, was admitted to a hospital with acute onset of altered mental status, impaired vision and ataxia. He received high doses of IV steroids and was discharged to an acute rehabilitation hospital with oral steroids, then referred to an outpatient therapy clinic one-month post-onset of CLIPPERS with ongoing use of oral steroids. His chief complaints were unsteadiness, difficulty walking, double vision, weakness tingling in the face, inability to play basketball. His goals

## CASE DESCRIPTION (Continued)

were to return to his prior level of function and reduce double vision. Physical and occupational therapy examination findings follow.

**Observation:** The patient presented with a left eye patch, resting hand tremors, and ataxic gait with rolling walker.

### Functional Outcomes Measures:

--Dynamic Gait Index (DGI) 15/24

-BERG: 51/56

### Cranial Nerve (CN) Impairments:

CN 2: impaired vision while wearing left eye patch

CN 3: difficulty moving eyes up/down, slight B eyelid ptosis

CN 4 and 6: Difficulty moving eyes laterally and inferiorly

CN 5: Impaired facial sensation (peri-oral area)

CN 7: Slightly asymmetrical smile

Upper Extremity Strength: 3/5 to 5/5

Lower Extremity Strength: 4/5 to 5/5

### Balance Testing:

Dynamic Standing Balance: Needed assistance to recover

Single Leg Stance: Right: 0 seconds, Left: 0 seconds

Unable to tandem walk

Able to braid t(slowly) to the right side but not to the left

### Motor Control Assessment:

(+) Dysmetria (finger to nose and heel to shin tests)

(+) Ocular dysmetria, and saccades

Decreased fine motor control

## METHODS

The patient participated in 8 physical and 6 occupational therapy sessions over 5 weeks to address his impairments. The interventions focused on oculo-motor gait, balance, coordination training, and return to sport activities. Oculomotor training included ocular calisthenics targeting the recti muscles by smooth pursuit tracking. Ocular coordination training included activities to improve integration of visual, vestibular and proprioceptive systems. A Marsden ball, was used to challenge both visual accuracy and vergence reaction time due to the semi-predictable nature of this moving pendulum target. The patient was instructed to track the ball, tap the letter, while simultaneously performing standing balance on compliant and non-compliant surfaces. Vestibular ocular reflex (VOR) training was utilized to recalibrate and improve VOR gain. As his condition improved, fine motor activities, return to running and return to basketball activities were incorporated.

## OUTCOMES

All physical and occupational therapy problems resolved except Intermittent binocular diplopia and oculomotor fatigue which were quickly corrected by closing one eye briefly. The DGI and BERG scores improved to 22/24 and 56/56 respectively. The patient returned to running and playing recreational basketball without limitations. Due to residual oculomotor deficits, the patient was referred to a neuro-optometrist for further evaluation. One year post -discharge the patient was contacted by phone for follow up. The patient reported that approximately 3 months after his last therapy session his double vision had fully resolved. Shortly afterwards he abruptly discontinued his oral steroids without medical guidance or approval. He remained off steroids for several months but sought medical attention when his double vision recurred. Once immunosuppressive therapy was re-prescribed the patient's diplopia promptly resolved

## DISCUSSION

Steroid therapy has been established as a standard treatment for patients with CLIPPERS. However, there is currently a void in the literature regarding the role of rehabilitation for patients diagnosed with this rare disease. This case report described a targeted rehabilitation program that supplemented pharmaceutical management of CLIPPERS.

## CONCLUSION

To our knowledge, this is the first report to describe a rehabilitation program for a patient with CLIPPERS. This report supports targeted interventions to address impairments, but more research is needed to determine if rehabilitation is an effective adjunct to pharmaceutical management of CLIPPERS.

## REFERENCES

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