Supernumerary phantom limbs in ICU patients with acute inflammatory demyelinating polyneuropathy

Alexander N. Melnyshyn,
MD
Teneille E. Gofton, MD,
FRCPC
Valerie Schulz, MD,
FRCPC

Correspondence to
Dr. Gofton:
teneille.gofton@lhsc.on.ca

ABSTRACT

Objective: To explore the previously undescribed phenomenon of phantom limb generation in patients with severe acute inflammatory demyelinating polyradiculoneuropathy (AIDP).

Methods: Between April 2011 and January 2014, we encountered 3 patients with AIDP in our intensive care unit who experienced features of self-limited supernumerary phantom limbs (SPLs) during their course.

Results: The following case series describes the phenomenon of SPLs in AIDP.

Conclusions: This report aims to raise awareness of the possibility of SPLs in the course of AIDP. The pathophysiology and management strategies for this clinical phenomenon are unknown.

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GLOSSARY

AIDP = acute inflammatory demyelinating polyradiculoneuropathy; **SPL** = supernumerary phantom limb.

Phantom limbs—the sensations of body parts not physically there—are associated with a variety of disease states including stroke, brain tumors, cranial and spinal trauma, multiple sclerosis, and epilepsy.¹ The most recognized diathesis is limb loss. Reports suggest an incidence of 90%–98% in traumatic amputations.² Two thirds of phantom limb patients develop concurrent neuropathic pain.³ Supernumerary phantom limbs (SPLs) occur when patients experience feedback from an appendage lacking an anatomical correlate, even when the body remains intact. Phantoms extend from the body, often moving appropriately relative to other parts. Mechanisms underlying these sensations are incompletely understood and an active area of study. We describe 3 patients (table) with acute inflammatory demyelinating polyradiculoneuropathy (AIDP) who experienced supernumerary phantom limb sensations (April 2011–January 2014). To our knowledge, current literature does not reference SPLs in the context of AIDP.

Case reports. Patient AB described having 4 arms and 4 legs, with 2 of the latter crossed atop her real ones. She maintained insight into the illusory nature of the phantoms and had delayed notifying staff because she “knew them to be wrong.” During this time, she developed waxing and waning neuropathic pain, exacerbated by passive range of motion. Neuropathic pain agents including gabapentin provided minimal benefit. Interestingly, onset of pain preceded the return of local power and sensation. As she improved in sensation and strength, her phantoms and pains diminished, leaving her with burning neuropathic pain in her ankles and soles by 6 months. This improved somewhat with escalating doses of gabapentin and hydromorphone in follow-up.

Patient CD described burning dysesthesias in her legs and arms, well-controlled with hydromorphone and gabapentin. An entry from the nursing notes on day 72 is illuminating in retrospect: CD “was complaining of burning pain in her legs last evening and overnight. Initially, she would request for her legs to be repositioned, but upon seeing (with a mirror) that her legs were straight, she would settle...she continued teary-eyed secondary to pain and frustrated because it is far more difficult for her to communicate her...pain.” On day 77, CD asked why she sensed 4 hands. By day 99, she complained of 4 legs, consciously controllable. The duplicate phantom appendages gradually telescoped to hands and feet. CD experienced pain only in her SPLs. Sensations were absent some days, returning the next, always greater in her feet than hands. As her

From the Departments of Clinical Neurological Sciences (A.N.M., T.E.G.) and Anesthesia and Perioperative Medicine (V.S.), Western University, London, Canada.

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Table Patient characteristics, clinical presentation, paraclinical investigations, primary treatment, latency of phantoms from symptom onset, and 6-month follow-up

Patient	Age, y/sex	Prodrome	Symptoms	LP ^a	EMG-NCS	TX	Δ (SX – phantom) onset, d	6-mo follow-up
AB	31/F	Sore throat day of onset	Ascending numbness, bilateral LE weakness, rapid progression to quadriplegia, dysphagia, respiratory distress, diplopia, ptosis, dysautonomia: Constipation, tachycardia, labile BP	ACD	Consistent with demyelination, AIDP, axonal loss, motor fiber predominance	5 d course IVlg	54 ^b	Phantoms resolved
CD	57/F	Diarrheal illness 1 week prior	Bilateral hand paresthesias, LE weakness, nonambulatory, rapid progression to quadriplegia, respiratory distress, dysautonomia: Constipation, tachycardia, labile BP	ACD	Consistent with severe demyelination, AIDP, severe axonal loss	5 d course IVlg	72	Persistent symmetric LE paresthesias, altered padding sensation and tightness, symmetric fingertip paresthesias
EF	68/F	None discernible	Shoulder and thoracolumbar pain, paresthesias in all 4 extremities, LE weakness, nonambulatory, rapid progression to quadriplegia, facial and bulbar weakness, respiratory distress, dysautonomia: Constipation, labile BP	ACD	Consistent with demyelination, AIDP	5 d course IVlg	46	Phantoms resolved, residual neuropathic pain in hands and feet

Abbreviations: ACD = albuminocytologic dissociation; AIDP = acute inflammatory demyelinating polyneuropathy; BP = blood pressure; EMG-NCS = electromyography and nerve conduction studies; IVlg = IV immunoglobulin; LE = lower extremity; LP = lumbar puncture; SX = symptoms; TX = treatment.

^aFor the purposes of this article, ACD is defined as a raised CSF protein with normal cell count.

^bApproximate onset by documentation. Patient experienced phantoms earlier without reporting.

sensorimotor function improved, the pain resolved entirely, leaving nonpainful phantom feet. She maintained residual paresthesias in her palms and soles with hyperalgesia and filmy sensation at 6 months. Her pain was adequately controlled with pregabalin, hydromorphone, and fentanyl.

Patient EF developed burning, lancinating dysesthesia in her tongue and low back with constant dull aching and extreme fatigue in her 4 limbs. She sensed 8 limbs in total, the surplus appendages layered atop her physical ones. The phantoms were sporadically painful and seemed to move at will. She occasionally sensed a “swirl” or pirouette (she had been a professional dancer for years). Other times her phantoms contorted painfully, as if “tied in a knot.” Sensations were nighttime predominant, affecting her sleep. By day 78, her upper extremity SPLs began telescoping inwardly. By morning, power was returning to her finger extensors. By day 81, the telescoping continued; phantom hands extended from her elbows bilaterally, coinciding with local return of power. Her SPL pain waxed and waned; hydromorphone and pregabalin were somewhat helpful. By day 110, her phantoms came and went, reactivating with exertion. When her arms and phantoms were made to overlap, she noted significant pain reduction. On more than one occasion, passive repositioning facilitated overlap and pain reduction. Since this was our third patient with this experience, we openly discussed this phenomenon with the patient and nurses, to remove the stigma of it being inappropriate, and asked the patient to teach us how to reduce the pain experience. The patient discovered SPL pain relief by carefully, passively moving her real leg over the position of her SPL. Later, the patient could consciously move her phantoms with concentration.

Patient EF improved slowly, describing tight, wire-like pain surrounding her phantoms. Telescoping persisted, with phantom fingers now extending from metacarpophalangeal joints; the full arm could be reactivated randomly with exertion. EF’s lower extremity phantoms weighed “heavy like stones,” impeding physiotherapy.

DISCUSSION In AIDP, effective deafferentation occurs on an acute to subacute basis,⁴ disrupting normal patterns interpreted centrally. This may be similar to the abrupt change occurring with amputation, despite limbs and nerve roots remaining intact. When patients progress to tetraplegia, the effect occurs in all limbs.

Some salient observations extend from our patients’ experience: (1) latency from symptom onset to SPL recognition is approximately 50 days; (2) development of SPLs appears to follow emergence of neuropathic pain; (3) discomfort and phantom

sensations improved significantly with return of local motor function; and (4) the SPL sensations resolved almost entirely over time. Canavero⁴ has suggested a mechanism of reverberating corticothalamic circuits that retain sensory memories of our internal representation. These representations may be substituted in times of deafferentation, resulting in phantoms. Staub et al.⁵ found increased blood oxygen level-dependent signal during SPL movement, suggesting abnormal thalamocortical closed-loop activity may underlie phantom phenomena. They suggest that intact premotor processes translating intention into action can result in phantoms in a setting of deafferentation, which impairs correction of mismatch between expected and actual movements. Perhaps returning sensorimotor feedback supersedes these circuits to which the brain had defaulted following the nadir phase of AIDP, explaining resolution.⁵ Patients CD and EF indicated the severity of neuropathic pain experienced correlated with divergence of phantoms from the visually confirmed position of their anatomical correlates. Flor et al.⁶ suggest that pain may result from incongruence between motor intention and sensory feedback, a theory supported by mirror studies producing painful and non-painful paresthesias using mirrors to disjugate actual and seen movements. The search for a unifying theory of phantom limbs continues, with current inquiry examining all levels of the neuraxis.^{3,7}

Our case series highlighted SPL experiences in patients with AIDP that resulted in pain, psychosocial distress, and functional compromise. Some common features of their SPLs included latency from AIDP symptom onset, appearance after the onset of neuropathic pain, improvement with return of motor function, and resolution over time. This report aims to raise awareness of the possibility of SPLs in the course of AIDP. The pathophysiology and management

strategies for this clinical phenomenon remain elusive.

AUTHOR CONTRIBUTIONS

Alexander N. Melnyshyn: clinician, primary author, drafting and editing of manuscript. Teneille E. Gofton: clinician, secondary author, corresponding author, development of manuscript outline, editing of manuscript. Valerie M. Schulz: clinician, secondary author, development of manuscript outline, editing of manuscript.

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