PM R XXX (2016) 1-5

www.pmrjournal.org

Case Presentation

Generalized Weakness in a Transplant Patient: A Case Presentation

Cora Brown, Ernesto Cruz, MD

Abstract

Generalized weakness in transplant patients is a major complaint in tertiary rehabilitation hospitals. The diagnosis and management of generalized weakness in this population pose challenges for physicians. We present the case of a transplant patient with generalized weakness who was eventually diagnosed with calciphylaxis using a multidisciplinary diagnostic approach of electrodiagnostics, vascular study, and skin biopsy. Calciphylaxis is a rare cutaneous disorder that mimics other collagen vascular diseases in its presentation and fulminant course. Physiatrists should be cognizant of calciphylaxis, as it signals a poor prognosis if not correctly diagnosed and treated in a timely manner, with high incidence of sepsis, wound pain, and disability.

Introduction

Calciphylaxis is an uncommon and poorly understood cutaneous syndrome seen in patients with end-stage renal disease and transplant patients on immunosuppressive therapies [1-3]. The cardinal features consist of cutaneous purpuric lesions and necrosis, along with histopathologic characteristics of calcification of cutaneous arterioles [1-3]. Because of its unpredictable and unique clinical presentation mimicking other collagen vascular diseases, it poses a diagnostic challenge for physicians [1-3]. Management of the disease involves a multidisciplinary approach ranging from conservative therapies such as sodium thiosulfate, bisphosphonates, and cinacalcet to surgical parathyroidectomy, with variable success [4,5]. Rehabilitation of patients with calciphylaxis involves facilitated range of motion, strengthening, and endurance exercises as well as wound care and pain management.

Case Presentation

A 47-year-old woman with a medical history of polymyositis diagnosed at age 14 years, idiopathic pulmonary fibrosis status after a left lung transplant at age 30 while on chronic immunosuppressive medications, hypertension, and end-stage renal disease on hemodialysis at age 40, presented with a 2-week course of progressive generalized weakness and a painful lesion of the right lower leg. At baseline, the patient stated that she was able to drive herself to the dialysis center, to perform her activities of daily living (ADLs) independently, and to ambulate around the house and outdoors without use of an assistive device. She reported that, 2 weeks before admission, she was feeling too weak to get out of the bed, to rise from the chair, or to drive the car. She noted that the weakness involved the hands and feet in addition to the shoulder and hip muscles. The patient reported that the lesion started out like a small pimple that became nonhealing, painful, and ulcerated after sustaining a minor trauma to the leg.

Examination of the upper and lower limbs revealed hypotonia of all 4 extremities. There was no fasciculation observed. The digits were warm and without cyanosis bilaterally. The peripheral pulses were 2+ symmetrically. An ulcerated lesion of the right lower leg was tender to palpation and measured 10 × 5 × 0.5 cm in diameter. Manual muscle testing revealed grade 4/5 strength of proximal arm muscles, 0/5 strength of wrist extension, 2/5 strength of proximal leg muscles, and 4/5 strength of ankle dorsiflexion and ankle plantar flexion bilaterally. Muscle stretch reflexes were absent in the upper and lower limbs, even with Jendrassik maneuvers. Hoffman's reflexes were absent and Babinski response revealed down-going toes. Sensation to light touch was intact in all dermatomal and peripheral nerve

1934-1482/$ - see front matter
http://dx.doi.org/10.1016/j.pmrj.2016.04.001

Table 1
Electrodiagnostic studies: Anti-sensory summary

Site	NR	Onset (ms)	Norm Onset (ms)	P-T Amp (μV)	Norm P-T Amp	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Right median anti-sensory (second digit)											
Palm		2.0	<1.3	5.9	>20	Palm	Second digit	2.0	7.0	35	>50
Wrist		3.5	<3.4	14.9		Wrist	Palm	1.5	7.0	47	>50
Elbow		8.3		2.6		Elbow	Wrist	4.8	25.4	53	>50
Right radial anti-sensory (first web space)											
Wrist 10 cm		2.3	<2.2	28.9	>15	Wrist 10 cm	First web space	2.3	10.0	43	>50
		0.9		29.0							
Right sup peroneal anti-sensory (ant lat mall) Tech difficult wound											
14 cm	NR				5	14 cm	Ant lat mall		14.0		
Right sural anti-sensory (lat mall)											
Calf	NR				>15	Calf	Lat mall		14.0		
Right ulnar anti-sensory (fifth digit)											
Wrist		3.7	<3.6	6.1	>15	Wrist	Fifth digit	3.7	14.0	38	>50
BE	NR					BE	Wrist		17.5		

distributions bilaterally. Tinel sign was negative over the median and ulnar nerves at the wrists. Sitting straight leg test results were negative bilaterally. Functional assessment on admission to the acute medical service revealed that the patient was able to perform sit-to-stand transfer and had bed mobility and ambulation of about 150 feet with moderate assistance of one-person assistance.

The rheumatologic work-up showed an elevated erythrocyte sedimentation rate and creatinine phosphokinase levels. Antinuclear antibody, double-stranded DNA and rheumatoid factor results were negative. The patient was diagnosed with exacerbation of her polymyositis and treated with high-dose prednisone (80 mg daily) for 1 week and 2 sessions of intravenous immunoglobulin. At 2 weeks after the hospitalization, she reported no improvement of the weakness and showed functional decline. At this point, the patient required maximal assistance with one-person assistance for sit-to-stand transfer and bed mobility. Her ambulation was decreased to 5 feet with maximal assistance of one-person assistance. Further diagnostic studies were performed to aid in diagnosing the patient's condition. She underwent magnetic resonance imaging of the bilateral femurs, which showed focal patchy areas of muscle signal possibly related to myositis or other muscle diseases. A left quadriceps muscle biopsy was performed by a general surgeon, which revealed atrophy of muscle secondary to disuse without evidence of metabolic myopathy or myositis. The patient was later transferred to an acute inpatient rehabilitation service for rehabilitative management of the generalized weakness. The unique challenge of treating this patient was to make a correct diagnosis. Because of her different pattern of the weakness in the upper extremities that affected distal muscles greater than proximal muscles but the reverse in the lower extremities, further diagnostic studies were performed. The patient underwent electromyographic and nerve conduction velocity studies, which showed axonal peripheral polyneuropathy. A vascular workup was performed, which showed an ankle brachial index of 0.53 of the bilateral lower limbs, with normal pulse volume recording waveforms consistent with mild to moderate

Table 2
Motor summary

Site	NR	Onset (ms)	Norm Onset (ms)	O-P Amp (mV)	Norm O-P Amp	L-R Lat (ms)	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)	Norm Vel (m/s)
Right median motor (abd poll brev)												
Wrist		4.4	<4.2	2.5	>5		Elbow	Wrist	5.8	25.2	43	>50
Elbow		10.2		1.4								
Right peroneal motor (ext dig brev)												
Ankle		5.2	<6.0	0.5	>3		B fib	Ankle	8.6	28.2	33	>40
B fib		13.8		0.3								
Right tibial motor (abd hall brev)												
Ankle		6.5	<6.0	2.6	>5		Knee	Ankle	9.0	32.5	36	>40
Knee		15.5		2.2								
Right ulnar motor (abd dig minimi)												
Wrist		4.1	<3.6	2.6	>5		B elbow	Wrist	4.2	20.0	48	>50
B elbow		8.3		2.1			A elbow	B elbow	2.4	9.5	40	>50
A elbow		10.7		1.8								