P-155

Microscopic Polyangiitis with Initial Neurological Manifestations Mimicking Focal Neuropathies

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Introduction

Microscopic polyangiitis (MPA) is a small-vessel vasculitis typically affecting the kidneys, lungs, and other capillary beds. When neurologic manifestation predominates, it is often misdiagnosed as focal neuropathy or spinal disease. We report a case of MPA presenting with acute foot drop and hand paresthesia, initially suggested by electrodiagnostic studies and confirmed by serological testing.

Case description

A 58-year-old man was admitted to the neurosurgery with a 1-month history of right foot drop, ankle swelling, and bilateral hand numbness following flu-like symptoms. Laboratory studies showed systemic inflammation. He had previously received empirical antibiotics for presumed cellulitis without clinical improvement.

As depicted in Figure 1, neurologic examination revealed a distal motor deficit in the right lower extremity. Sensory testing demonstrated hypoesthesia and paresthesia in right feet and both hands. Despite spinal MRI showing diffuse disc bulging and stenosis, the distribution of deficits was atypical; therefore, he was referred to the Department of Rehabilitation Medicine for electrodiagnostic evaluation of suspected combined focal distal neuropathies. Electrodiagnostic testing demonstrated absent sensory nerve action potentials (SNAP) in the right median, bilateral sural and superficial peroneal nerves, with reduced SNAP amplitude in the left median nerve. Compound muscle action potentials (CMAP) were not elicited in the right peroneal and tibial nerves and amplitudes were reduced in both median and the left tibial nerve. (Table 1). Needle electromyography showed active denervation in multiple non-contiguous muscles. (Table 2). These findings indicated an asymmetric sensorimotor axonal polyneuropathy, suggestive of mononeuritis multiplex.

On hospital day 6, the patient developed right-sided hemiparesis due to a 1-cc ICH in the left basal ganglia. (Figure 2) Given the diagnosis of mononeuritis multiplex, these combined clinical features suggested an underlying systemic vasculitis. Subsequent serologic testing revealed a markedly elevated anti–myeloperoxidase antibody titer (>700 U/mL), confirming MPA. The patient underwent induction therapy with corticosteroids, mycophenolate mofetil, and rituximab. Management of the right foot drop included an ankle–foot orthosis (AFO), neuromuscular electrical stimulation, and gait training. After 2 months of treatment, he achieved independent indoor ambulation with a right AFO.

Figure 2. Brain CT on hospital day 6

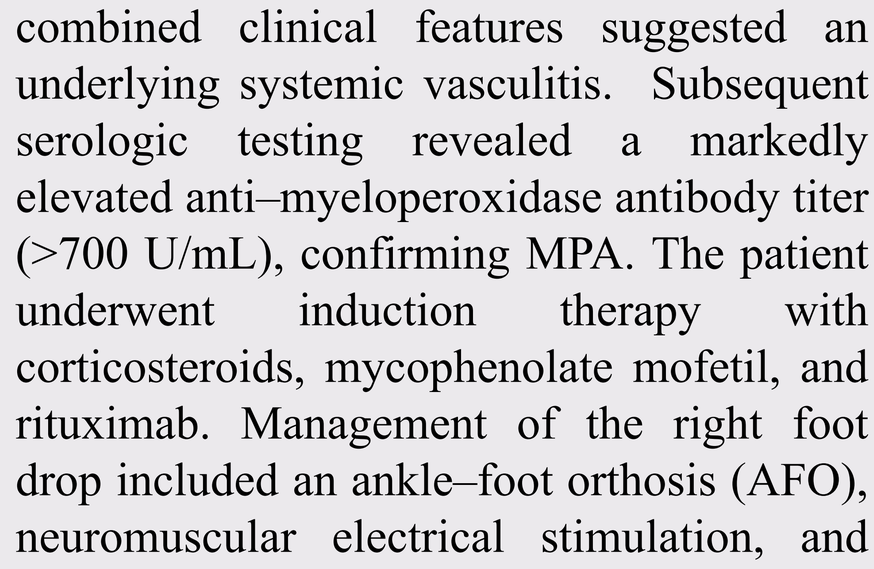

Figure 1. Initial neurologic examination of the patient

Motor power by Medical Research Council grade		
	Right	**Left**
Shoulder flexion	5	5
Elbow flexion	5	5
Elbow extension	5	5
Wrist flexion	5	5
Wrist extension	5	5
Finger flexion	4	5
Finger extension	5	5
Hip flexion	5	5
Knee flexion	3	4
Knee extension	5	5
Ankle dorsiflexion	0	3
Ankle plantaflexion	0	4
1st toe extension	0	0

Sensory
Both palm-finger paresthesia (finger 1-4th) Right leg near anesthesia (10%) Paresthesia at right lateral-posterior calf, foot dorsum, and sole Intact sensation above knee
Deep tendon reflex and Pathologic reflex
Biceps jerk (1+/1+) Knee jerk (1+/1+) Hoffman's sign (-/-) Babinski sign (-/-) Ankle clonus (-/

Table 1. Result of nerve conduction study

Nerve/Site	Latency (ms)		Amplitude*		CV (m/s)	
	Right	**Left**	**Right**	**Left**	**Right**	**Left**
Sensory nerve						
Median – Digit III (Wrist)	**NE**	3.1	**NE**	**5.2**	-	57
Median – Digit III (Palm)	**NE**	1.8	**NE**	**6.0**	-	57
Ulnar – Digit V (Wrist)	3.0	3.4	14.8	12.5	63	54
Dorsal ulnar cutaneous (Wrist)	2.9	2.3	10.4	15.9	51	53
Radial – Snuff box (Forearm)	1.9	2.0	25.3	25.5	80	69
Sural (Ankle)	**NE**	**NE**	**NE**	**NE**	-	-
Superficial peroneal (Foot)	**NE**	**NE**	**NE**	**NE**	-	-
Saphenous (Lower leg)	3.4	2.9	**6.6**	**7.3**	**38**	44
Motor nerve						
Median – APB (Wrist)	**4.8**	3.4	**0.3**	**0.8**	**45**	**44**
Median – APB (Elbow)	**10.0**	8.7	**0.3**	**0.6**		
Ulnar – ADM (Wrist)	2.9	2.7	**4.5**	6.5	61	61
Ulnar – ADM (Below elbow)	6.3	6.1	**3.8**	6.2		
Ulnar – ADM (Above elbow)	7.7	7.5	**3.8**	6.1	71	71
Tibial – AHB (Ankle)	**NR**	5.3	**NR**	**1.0**	-	44
Tibial – AHB (Popliteal fossa)	**NR**	13.1	**NR**	**0.5**		
Peroneal – EDB (Ankle)	**NR**	**NR**	**NR**	**NR**	-	-
Peroneal – EDB (Fibula head)	**NR**	**NR**	**NR**	**NR**		
Peroneal – TA (Fibula head)	**NR**	4.2	**NR**	**3.9**	-	56
Peroneal – TA (Popliteal fossa)	**NR**	5.8	**NR**	**3.3**		

* Microvolt in sensory study and millivolt in motor study
CV: conduction velocity; NE: not evoked; NR: no response; APB : abductor pollicis brevis; ADM : abductor digiti minimi; AHB : abductor hallucis brevis; EDB : extensor digitorum brevis; TA : tibialis anterior

Table 2. Result of needle electromyography

Muscle	Spontaneous activity			Motor unit action potential			
	IA	**Fib**	**PSW**	**Amp**	**Dur**	**Poly**	**Interference pattern**
L C7-T1 paraspinal	**Increased**	None	None	Normal	-	-	-
L L3-L5 paraspinal	**Increased**	None	None	Normal	-	-	-
R Abductor pollicis brevis	Normal	**2+**	**2+**	Normal	Normal	None	Normal
L Abductor pollicis brevis	Normal	None	**1+**	Normal	Normal	None	Normal
R Tibialis anterior	Normal	None	**3+**	Normal	-	-	-
R Peroneus longus	Normal	None	**1+**	Normal	-	-	-
R Tibialis posterior	Normal	None	**1+**	Normal	-	-	-
R Gastrocnemius	Normal	None	**1+**	Normal	-	-	-
R Biceps femoris	Normal	None	**1+**	Normal	Normal	None	**Reduced**
L Tibialis anterior	Normal	**1+**	**1+**	Normal	Normal	**Poly**	Normal
L Peroneus longus	Normal	**2+**	**1+**	Normal	Normal	**Poly**	**Discrete**
L Tibialis posterior	Normal	None	**2+**	Normal	Normal	**Poly**	**Discrete**
L Gastrocnemius	Normal	None	**2+**	Normal	Normal	**Poly**	**Reduced**
L Biceps femoris	Normal	None	**2+**	Normal	Normal	None	Normal

IA : insertional activity; Fib : fibrillation; PSW : positive sharp wave; Amp: amplitude; Dur: duration; Poly: polyphasic

Conclusion

Acute multifocal neurological deficits can be the primary manifestation of systemic vasculitis. Electrodiagnostic evidence of multiple axonal involvement, especially if inflammatory markers are elevated, should prompt evaluation for systemic vasculitis to prevent serious complications including ICH.