Diffusion Tensor Imaging Of Peripheral Nerves In Churg-Strauss Syndrome

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Running title: Diffusion tensor imaging of peripheral nerves in CSS

Abstract

We here describe a case of a 41-year-old woman with asthma, eosinophilia, and multiple mononeuropathies due to Churg-Strauss syndrome (CSS). In this case, multiple mononeuropathies caused allodynia and nerve-stretch pain in the lower extremities. A nerve conduction study and diffusion tensor imaging (DTI) indicated that the major site of lesions was at the right popliteus. Peripheral nerve DTI was found to be a feasible method to help evaluate peripheral nerve function.

Dear sir

Churg-Strauss syndrome (CSS), also called allergic granulomatous angiitis (AGA), is a rare autoimmune disease that was first described in 1951 by two pathologists, J. Churg and L. Strauss.[1] Since then, the presence of asthma, eosinophilia, and small vessel vasculitis with granuloma have been regarded as the key features of this disease. In 1990, the American College of Rheumatology proposed six classification criteria for CSS.[2] CSS belongs to the class of antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis, which also includes Wegener's granulomatosis (WG) and microscopic polyangiitis (MPA), and is clearly distinct from classical polyarteritis nodosa (PAN), which involves medium-sized arteries and is usually not associated with ANCA. Eosinophilic infiltration and/or ischemic damage due to vasculitis is often detected in the peripheral nerves. Clinically, multiple mononeuropathies are typically seen in such cases. We here describe a case of a woman with asthma, eosinophilia, and multiple mononeuropathies due to CSS. In this case, both a nerve conduction study and diffusion tensor imaging (DTI) helped in evaluating the peripheral nerve lesions.

A 41-year-old woman developed numbness and pain in the lower extremities. She had a history of bronchial asthma from childhood and allergic rhinitis more recently. Her numbness and pain continued and four months later, she developed gait disturbance that brought her to our hospital. On admission to our hospital, she was alert and cooperative. Her cranial nerves were normal. She could walk with assistance; however, she had mild weakness in the extremities (Medical Research Council classification, 4/5) that was especially prominent in foot dorsiflexion (1/5). Her deep tendon reflexes were normal. She had a mild tingling sensation in the plantar area bilaterally. She could not stand on her feet because of severe allodynia (pushing in the plantar area and dorsum of the feet caused burning pain), particularly on the right side. She kept bending her knee joint because of nerve-stretch pain along the tibial and peroneal nerves, where nerve-pushing also evoked severe pain. Her knee joints themselves were not painful. The pain sensation was markedly decreased in the distal extremities, while the touch sensation was preserved. The position sensation was abolished in the right foot, while the Romberg sign was negative. Laboratory tests revealed an increased white blood cell count of 21200/μL with eosinophilia (eosinophil 62%), and an IgE of 7290 IU/ml, perinuclear (P)-ANCA of 69.0 U/ml, soluble interleukin-2 receptors of 2000 U/ml, and gamma-globulin of 34.6%. Repeated chest X-rays and CT showed transitory infiltrative shadows bilaterally. The cerebrospinal fluid examination was normal. The nerve conduction study revealed multiple mononeuropathies in the lower extremities, particularly at the right popliteus area **(Table 1)**. The clinical and laboratory findings fulfilled the clinical diagnosis of CSS.[2]

Administration of one course of methylprednisolone pulse (1 g/day * 5 days),[3] followed by 60 mg/day oral prednisolone, lessened her pain markedly, and she became able to stand on her feet and to extend her knee joints. Her muscle weakness began to ameliorate gradually. Four months after steroid pulse therapy, a second nerve conduction study revealed that the right tibial nerve function had slightly improved, whereas the right peroneal nerve function had worsened. Three months after steroid pulse therapy, we performed DTI tractography of peripheral nerves by 3.0T MRI system.[4,5] As a result, at the right mid-thigh and popliteus area, the common peroneal nerve was not visualized and the tibial nerve was poorly visualized **(Figure 1)**. These findings corroborated the nerve conduction study results.

Our patient showed typical clinical features of CSS. Among these, it is noteworthy that she exhibited right-side dominant allodynia that hindered her from standing on her feet, and nerve-stretch pain and focal tenderness along the tibial and peroneal nerves that kept her from bending her knee joint. These clinical features and the nerve conduction study results indicated that, among multiple mononeuropathies, the main lesion might be located at the right popliteus. Corresponding to this, on DTI tractography of the peripheral nerves, the common peroneal nerve was not visualized and the tibial nerve was poorly visualized at the right mid-thigh and popliteus area. Although we did not measure fraction anisotropy or diffusivity in our patient, these findings most probably reflect nerve injury[4,5] due to vasculitis. Recently, DTI has been applied to the study of peripheral nerves, e.g., median (carpal tunnel syndrome)[6,] ulnar[6], radial[7], and tibial nerves[8]. These studies, including ours, demonstrate the feasibility of performing DTI of peripheral nerves to help evaluate peripheral nerve lesions. We here describe a case of a woman with asthma, eosinophilia, and multiple mononeuropathies due to CSS. In this case, multiple mononeuropathies caused allodynia and nerve-stretch pain in the lower extremities. A nerve conduction study and DTI indicated that the major site of the lesion was at the right popliteus. Peripheral nerve DTI was found to be a feasible method to help evaluate peripheral nerve function.

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	Nerve		Patient		Normal limit
			before	4 months after steroid pulse	
			R	R	
Motor					
	Median	Distal latency (ms)	3.2	3.4	4.5
		Conduction velocity (m/s)	63	57	48
		CMAP amplitude (mV)	13.3	15.1	4.4
	Ulnar	Distal latency (ms)	2.2	2.6	3.5
		Conduction velocity (m/s)	53	69	48
		CMAP amplitude (mV)	9.7	7.5	4.6
	Tibial	Distal latency (ms)	5.6	3.8	5.6
		Conduction velocity (m/s)	34.0	41	38
		CMAP amplitude (mV)	0.47	0.70	5.8
	Peroneal	Distal latency (ms)	5.0	NE	6.2
		Conduction velocity (m/s)	36	NE	39
		CMAP amplitude (mV)	1.2	NE	1.2
Sensory					
	Median	Conduction velocity (m/s)	57	58	44
		SNAP (ÊV)	58	37	11.1
	Ulnar	Conduction velocity (m/s)	59	49	42
		SNAP (ÊV)	24	43	7.8
	Sural	Conduction velocity (m/s)	44	50	40
		SNAP (ÊV)	5.2	6.3	3

Table 1 Nerve conduction study in the patient.

Four months after steroid pulse therapy, the patient still showed severe multiple mononeuropathy, e.g., CMAP of the peroneal nerve was not evoked, and CMAP of the tibial nerve was decreased in the right foot.

CMAP: compound muscle action potential

SNAP: sensory nerve action potential

NE: not evoked

Normal limit is determined by 100 patients in Chiba University Hospital.

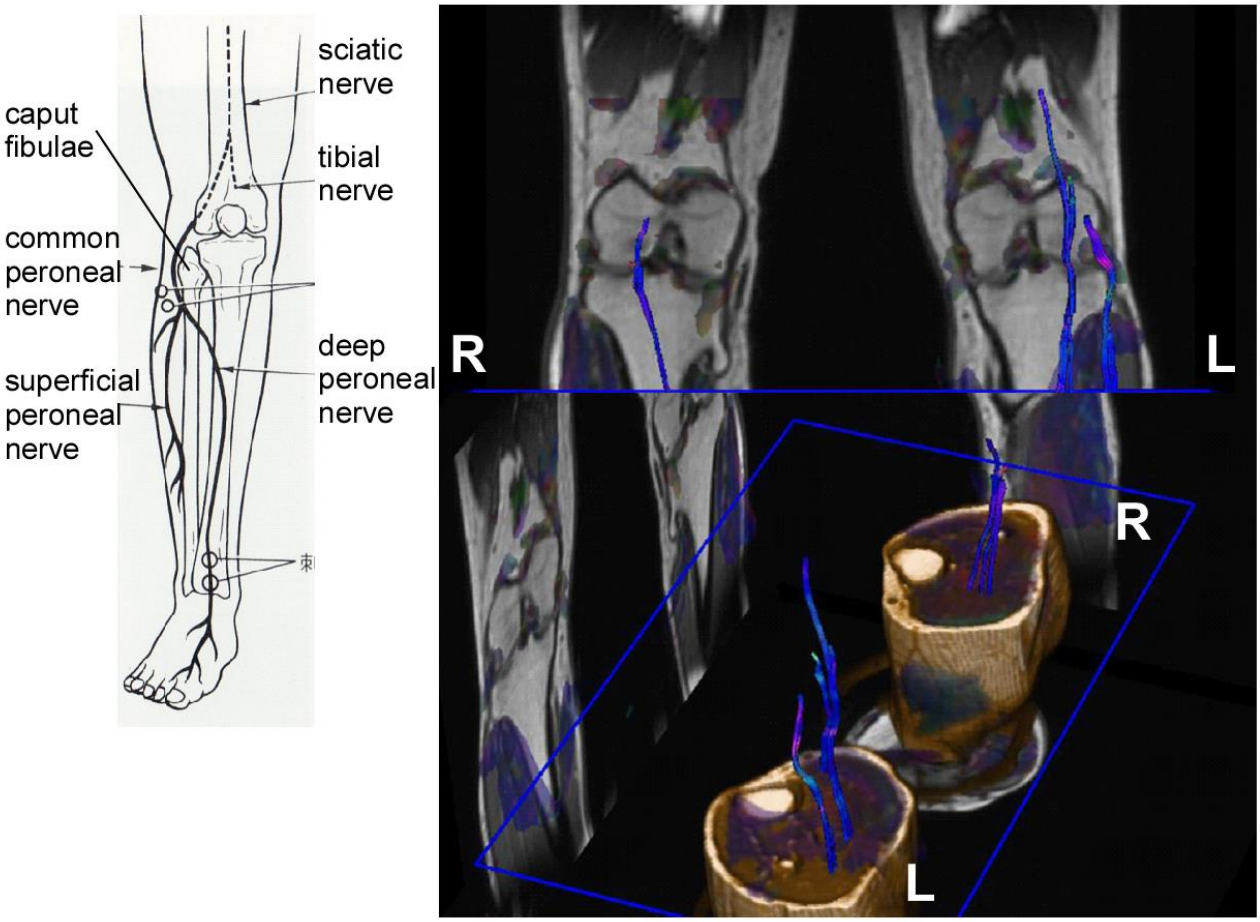

Figure 1 MRI neurography at the right popliteus area of the patient 3 months after steroid pulse therapy.
Note that at the right mid-thigh and popliteus area, the common peroneal nerve is not visualized and the tibial nerve is poorly visualized. These findings correspond well with the nerve conduction study results.