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흉부 혈관내 대동맥박리 재건술 이후 발생한 허혈성 단일사지 신경병증: 케이스 보고

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Ischemic Monomelic Neuropathy after Thoracic Endovascular Aortic Repair (TEVAR) : Case Report

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Ischemic monomelic neuropathy (IMN) is a rare type of peripheral neuropathy presenting with sensory and motor impairment after vascular surgery. It is particularly rare for one to experience IMN following thoracic endovascular aortic repair (TEVAR); however, we have recently experienced such a case. A 56-year-old male with traumatic aortic dissection underwent TEVAR. One day after, he suddenly complained of numbness of the left hand with reduced grip strength and wrist movement. Upon physical examination, weak left radial arterial pulse and significantly low blood pressure in the left arm (85/60 mmHg) compared to right arm (130/85 mmHg) were observed. Electrophysiologic findings were compatible with left radial, median and ulnar neuropathy. On follow-up 1 year later, his motor weakness was mildly improved, but constant neuropathic pain and ulnar claw deformity were observed.

Key Words: ischemia, mononeuropathy multiplex, endovascular procedure

Introduction

Ischemic monomelic neuropathy (IMN) is a rare type of peripheral neuropathy resulting from shunting of blood flow or acute non-compressive occlusion of the major proximal limb artery after vascular surgery.[1] The exact pathophysiology of IMN is poorly understood. However, it has been reported that IMN mainly occurs in patients with a pre-existing microvascular disease. Diabetic mellitus is a particularly well-established risk factor of IMN.[2]

The diagnosis of IMN can be made clinically. And immediate treatment for IMN is very important because its prognosis depends on the prompt recognition, diagnosis, and correction of the vascular problem.

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Thus, accurate history taking and physical examination are necessary to identify the cause of neurologic deficits following vascular surgery.[1,2]

There are a few reported cases of ischemic complications of the upper extremity after vascular access for hemodialysis. However, cases of IMN after TEVAR have rarely been reported and the electrodiagnostic findings of the IMN after TEVAR have never been precisely described. In this report, we present a case of IMN following TEVAR confirmed by electrodiagnostic study in a patient with traumatic aortic dissection.

Case Report

A 56-year-old male with hypertension and gout was admitted to another hospital after a traffic accident. He had multiple rib fractures, a right scapular fracture, and lung contusion. A chest CT scan revealed aortic rupture in the proximal portion of the descending thoracic aorta (Fig. 1A). He was then transferred to our hospital for intensive care including thoracic endovascular aortic repair (TEVAR). He underwent TEVAR under general anesthesia in an angiography room. After deploying a stent graft (Zenith TX2, Cook Japan, Tokyo) in the descending aorta with coverage of the left subclavian artery (LSA), post-chest CT showed no endoleak and well-maintained patency in LSA (Fig. 1B, C).

One day after TEVAR, he suddenly complained of numbness of the left hand with reduced grip strength and wrist movement. However, because of his pain in multiple joints and posterior neck resulting from the trauma, we were not able to precisely measure his neurologic deficits. Three days later, his left-hand grip and wrist flexion improved to grade II and sensory tests were normal.

Nerve conduction studies (NCSs) were performed at 1 week after the onset of symptoms. The findings of motor NCSs were markedly reduced amplitude of compound motor action potentials (CMAPs) at the left median and radial nerve with minimal drop in CMAPs at ulnar nerve; sensory NCSs were normal (Table 1). The needle electromyography demonstrated no motor unit action potentials (MUAPs) in the left hand intrinsic muscles, flexor carpi radialis, flexor carpi ulnaris and extensor digitorum without denervation (Table 2). These electrodiagnostic results were suggestive of mononeuropathy multiplex. Pallor, coolness, trophic skin or hair changes, tenderness and swelling of affected limb were absent. Additional clinical symptoms were weak left radial artery pulse and significantly low blood pressure in the left arm (85/60 mmHg) compared to the right arm (130/85 mmHg). Laboratory tests including complete blood cell count, serum electrolyte,

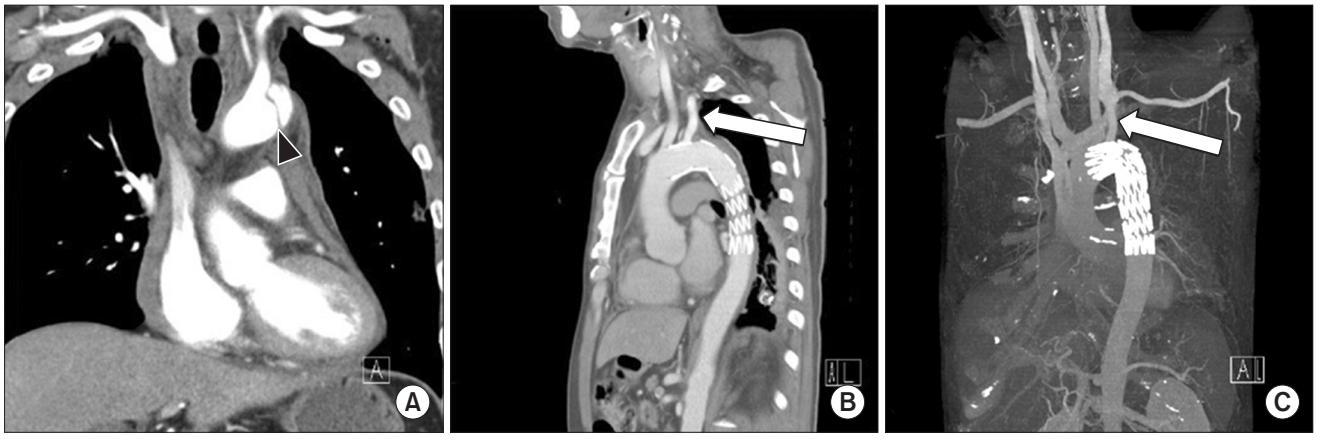

Fig. 1. (A) Pre-TEVAR CT scan showed aortic rupture in the proximal portion of the descending thoracic aorta (black arrowhead). (B) Post-TEVAR CT scan demonstrated proper apposition of the graft with no evidence of endoleak and maintenance of patency in a left subclavian artery (white arrow). (C) Post-TEVAR CT MIP images also showed maintenance of patency in a left subclavian artery (white arrow).

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Table 1. Nerve Conduction Study

	Study	Nerve	Latency (*R/L, ms)	Amplitude (*R/L, mV/uV)	Velocity (*R/L, m/s)
1 Week after	CMAP	Median	3.75/4.05	5.7/3.4	50.0/56.3
Onset of symptoms		Ulnar	2.45/2.80	10.8/7.5	57.1/56.4
		Radial	1.60/1.85	9.7/4.0	
	SNAP	Median	2.65/2.65	26.9/25.0/	
		Ulnar	2.20/2.35	36.3/33.4	
		Radial	1.95/1.70	38.0/37.0	
3 Months after	CMAP	Median	4.10/4.85	8.7/1.0	54.8/54.1
Onset of symptoms		Ulnar	2.65/4.10	9.9/0.9	57.5/46.5
		Radial	1.55/NR	8.6/NR	
	SNAP	Median	2.60/2.70	27.1/22.9	
		Ulnar	2.25/2.95	24.5/27.5	
		Radial	1.90/1.65	32.7/31.6	
1 Year after	CMAP	Median	4.15/5.00	10.8/2.2	55.6/52.8
Onset of symptoms		Ulnar	2.85/5.50	13.8/1.4	60.6/59.7
		Radial	2.10/3.20	7.7/1.7	
	SNAP	Median	2.80/3.45	27.6/24.9	
		Ulnar	2.50/4.15	31.5/9.4	
		Radial	1.20/1.65	35.6/36.3	

*R/L: right/left, CMAP: Compound muscle action potential, SNAP: Sensory nerve action potential, NR: No response

Table 2. Needle Electromyography

	Muscle	Spontaneous activities	MUAP	Recruitment
1 Week after	Lt. Deltoid	None	Normal	Normal
Onset of symptoms	Lt. Biceps brachii	None	Normal	Normal
	Lt. Triceps brachii	None	Normal	Normal
	Lt. Flexor carpi radialis	None	No MUAP	–
	Lt. Flexor carpi ulnaris	None	No MUAP	–
	Lt. Extensor digitorum	None	No MUAP	–
	Lt. First dorsal interosseus	None	No MUAP	–
	Lt. Abductor digiti minimi	None	No MUAP	–
	Lt. Abductor pollicis brevis	None	No MUAP	–
3 Months after	Lt. Deltoid	None	Normal	Normal
Onset of symptoms	Lt. Biceps brachii	None	Normal	Normal
	Lt. Triceps brachii	None	Normal	Normal
	Lt. Flexor carpi radialis	PSW 2+	Normal	Reduced
	Lt. Flexor carpi ulnaris	PSW 1+	Normal	2 MUAP
	Lt. Extensor digitorum	PSW 3+	Normal	Reduced
	Lt. First dorsal interosseus	PSW 4+	Normal	2 MUAP
	Lt. Abductor digiti minimi	PSW 2+	Polyphasic	2 MUAP
	Lt. Abductor pollicis brevis	PSW 2+	Polyphasic	2 MUAP
1 Year after	Lt. Deltoid	None	Normal	Normal
Onset of symptoms	Lt. Biceps brachii	None	Normal	Normal
	Lt. Triceps brachii	None	Normal	Normal
	Lt. Flexor carpi radialis	PSW 3+	Normal	Reduced
	Lt. Flexor carpi ulnaris	PSW 3+	Normal	Reduced
	Lt. Extensor digitorum	PSW 2+	Normal	Reduced
	Lt. First dorsal interosseus	PSW 3+	Normal	2 MUAP
	Lt. Abductor digiti minimi	PSW 2+	Polyphasic	2 MUAP
	Lt. Abductor pollicis brevis	PSW 2+	Polyphasic	2 MUAP

MUAP: Motor unit action potential, PSW: Positive sharp wave

blood urea nitrogen and serum creatinine were within normal range. Unfortunately serum creatine kinase (CK) level was not tested. For these reasons, we finally diagnosed his neurologic deficits as ischemic monomelic neuropathy (IMN).

At three months after onset of symptoms, follow-up

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electrodiagnostic studies were performed. On motor NCSs, the amplitude of CMAPs in the median, radial, and ulnar nerve was lower than that of the initial test. Sensory NCSs demonstrated no interval change (Table 1). The needle electromyography showed denervation potentials in the muscles above (Table 2).

The last electrodiagnostic studies were conducted approximately 1 year later. Reduced amplitude of sensory nerve action potential (SNAP) in ulnar nerve was newly developed (Table 1). Except for that, the results were relatively similar to the previous studies (Table 1, 2). His motor weakness improved mildly, but constant neuropathic pain and ulnar claw deformity were observed. And low blood pressure in the left arm (80/55 mmHg) compared to the right arm (130/80 mmHg) was still checked.

Discussion

The term, "ischemic monomelic neuropathy (IMN)" was first used in 1983 by Wilbourn et al.[1] to describe a peripheral neuropathy developed following the shunting of blood flow or due to acute non-compressive occlusion of the major proximal limb artery after vascular surgery. The pathophysiology of IMN is unclear but has been considered as relative ischemia resulting from sudden diversion or transient occlusion of the blood supply to the nerves of the forearm and hand.[3,4] The diagnosis of IMN can be essentially based on clinical symptoms. In general, neurologic deficits appear immediately after surgery and may present with wrist drop, reduced thumb opposition, or difficulty with finger flexion. Paresthesia, numbness, and diminished sensation in a distal upper limb may be combined with motor deficits. Unlike vascular steal syndrome, the hand is warm, and the radial pulse may be present.[5] Nevertheless, there are no pathognomonic signs for diagnosing IMN. Additionally, many patients present with severe neuropathic pain and allodynia overshadowing these functional losses. Many clinicians have difficulty diagnosing IMN.[1,2] Electrodiagnostic studies are helpful to differentiate IMN from other conditions. NCSs usually show an axonal loss and reduced motor and sensory nerve conduction velocities of median, radial, and ulnar nerves. The needle electromyography reveals primarily acute denervation and reduced motor unit recruitment of distal upper limbs.[1-3,5]

In our case, it was difficult to diagnose IMN because of several ambiguous findings. First, chest CT after TEVAR revealed well-maintained patency in LSA. Second, initial electrodiagnostic study showed only reduced CMAPs at the left median and radial nerve with minimal drop in CMAPs at ulnar nerve and normal sensory SNAP amplitude. We were concerned about the possible causes of his neurologic deficits such as spinal cord infarction, vascular steal syndrome, compartment syndrome, blood pressure cuff compression injury and etc. Upon physical examinations, his neurologic deficits were compatible with lower motor neuron lesion and his lower extremities were intact until one year after the onset. Signs of ongoing vascular insufficiency such as pallor, coolness, trophic skin or hair changes were absent. There were no signs of muscle infarction like tenderness and swelling of affected limb. Therefore, we excluded the possibility of spinal cord infarction, vascular steal syndrome and compartment syndrome. There was also no chance of blood pressure cuff compression injury because an arterial line on the right arm was used to monitor blood pressure. Additional clinical symptom was significantly low blood pressure in the left arm (85/60 mmHg) compared to the right arm (130/85 mmHg). Hence, we thought that his neurologic deficits resulted from IMN after TEVAR rather than others.

In fact, it is not clearly explained why neurologic deficits occurred despite well-maintained patency in LSA. IMN after dialysis access surgery has been known as a complication which is different from vascular steal syndrome and its pathophysiology is explained by the presence of pre-existing microvascular disease combined with a loss of perfusion pressure and high shunt takeoff from the brachial artery.[2] In our case, it is

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assumed that high shunt takeoff from LSA produces a loss of perfusion pressure of left distal upper limb and eventually results in IMN.

In addition, it is unclear why median, ulnar and radial SNAP amplitude were preserved until at least three months after the onset of symptoms. Acute ischemic neuropathy after creation of an arterio-venous fistula often gives rise to axon-loss mononeuropathy multiplex.[5] In previous electrophysiologic studies of patients with lesser degrees of ischemia, NCSs revealed correspondingly fewer and less severe abnormalities.[6] And one case report demonstrated that sural SNAP amplitude was preserved in ischemic tibial neuropathy as a complication of popliteal artery embolization.[7] We thought that the preservation of sensory SNAP amplitude might be related to less degree of ischemia with well-maintained patency in LSA. Further cases are encouraged to be reported to figure out electrophysiologic features in IMN.

TEVAR have some complications such as stroke, spinal cord ischemia, upper extremity ischemia, vocal cord paralysis, and mortality. It is thought that these are mainly associated with the coverage of the left subclavian artery (LSA) during TEVAR.[8,9] Zamor et al.[9] reported on the importance of LSA revascularization during TEVAR based on evidence of higher rates of stroke and upper extremity ischemia on unrevascularized group compared with revascularized one. Nevertheless, few studies have reported low incidences of upper extremity ischemia following TEVAR. Klocker et al.[8] showed 0.8% incidence of upper extremity ischemia and concluded that it has no impact on quality of life.

We questioned how to best manage our patient because of his well-maintained patency in LSA. In general, the treatment of upper extremity ischemia after the TEVAR is immediate LSA revascularization. In 2010, the Society for Vascular Surgery suggested guidelines for management of the LSA with TEVAR. Following these guidelines, LSA revascularization should be performed depending on urgency before or after the TEVAR.[10] However, in cases of well-maintained patency in LSA, there is no consensus about the treatment of upper extremity ischemia after TEVAR. We performed conservative management including a range of motion exercises and pain control. His motor weakness was mildly improved, but the constant neuropathic pain and ulnar claw deformity were still observed 1 year later.

In conclusion, we report a rare case of IMN after TEVAR confirmed by an electrodiagnostic study. IMN could be developed by several vascular accesses including TEVAR. Improved awareness of disease entity and clinical suspicion of neurologic deficit in the immediate post-operative period provides an early diagnosis with optimal outcome.

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