Case Report / *Olgu Sunumu*

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Primer Raynaud's Phenomenon with sensorimotor demyelinating polyneuropathy: a case report

Sensörimotor demiyelizan polinöropati ile birlikte primer Raynaud Fenomeni: bir olgu sunumu

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ABSTRACT

Raynaud's phenomenon (RP) is characterized by color changes and episodic vasospasm in the distal parts of the body. Raynaud's phenomenon associated with sensorimotor demyelinating polyneuropathy has been rarely reported in the literature. Here, we aimed to present a 36-year-old male patient who was admitted to our clinic with complaints of discoloration, numbness, and tingling in the upper and lower extremities. A mixed type of sensorimotor demyelinating polyneuropathy was detected in nerve conduction studies of the patient. A significant improvement was observed within 2 months after pregabalin, acetylsalicylic acid, and nifedipine treatment.

Keywords: Primer raynaud's phenomenon, sensorimotor demyelinating polyneuropathy, case report.

ÖZ

Raynaud fenomeni (RF), vücudun distal kısımlarında renk değişiklikleri ve epizodik vazospazm ile karakterizedir. Sensörimotor demiyelinizan polinöropati ile ilişkili RF literatürde nadiren bildirilmiştir. Biz burada üst ve alt ekstremitelerde renk değişikliği, uyuşma, karıncalanma şikayetleri ile kliniğimize başvuran 36 yaşında erkek hastayı sunmayı amaçladık. Hastanın sinir ileti çalışmalarında mikst tip sensorimotor demiyelinizan polinöropati saptandı. Hastada pregabalin, asetilsalisilik asit ve nifedipin tedavisi sonrası 2 ay içinde belirgin düzelme görüldü.

Anahtar Sözcükler: *Primer raynaud fenomeni, sensorimotor demiyelinizan polinöropati, olgu sunumu.*

INTRODUCTION

Raynaud's phenomenon (RP) is described with a triphasic color change with initial white or pallor (ischemic phase), then blue or cyanosis (deoxygenation phase), and followed by red or erythema (reperfusion phase) (1). It is a symptom complex caused by digital vascular compromise and divided into two subtypes as primary and secondary (2). The primary Raynaud's phenomenon (PRP) is the most common form and the pathophysiological causes of PRP is not yet fully known. It has been focused on vascular, intravascular, and neural mechanisms such as central impairment of autonomic function, sympathetic denervation (3). The etiology of secondary RP includes a broad range of diseases such as hematological, thrombo-embolic, autoimmune rheumatic diseases, vasculitis, carpal tunnel syndrome, acromegaly, and malignancies (1, 2). Gunatilake et al. (4) reported the first case of Guillain–Barré syndrome presenting with RP in a 21-year-old male case. The pathophysiological mechanism linking these two conditions was thought to be associated with dysautonomia with altered sympathetic activation, however, it was emphasized that it should be further evaluated by experimental studies (4).

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In a study, underlying subclinical small fiber dysfunction has been reported in a subset of patients with PRP (5). As an important outcome of the study, testing of peripheral autonomic and somatic small-diameter peripheral nerve fibers was highlighted as a potentially useful assessment method for PRP (5). Raynaud's phenomenon with sensorimotor demyelinating polyneuropathy has been reported rarely in the literature. We aimed to report a case with RP and sensorimotor demyelinating polyneuropathy.

CASE REPORT

A 36-year-old male patient was admitted to our clinic with complaints of changes in the color exacerbating by cold, numbness, and tingling of the distal parts of the upper and low extremities for the last 6 months (Figure-1). There was no medical history about smoking and any diseases for the patient. The general condition was moderate, oriented, cooperative, and the vital signs were within normal ranges. On physical examination, the patient had bilateral RP at the distal parts of his extremities. The patient had no joint tenderness, swelling, deformity, skin rash, or oral ulcers. On neurological examination, sense of vibration in both extremities was paresthetic and deep tendon reflexes were decreased. Fasting blood glucose, vitamin B12, and thyroid function tests were normal. Rheumatoid factor, anti-cyclic citrullinated peptide, antinuclear antibody, anticardiolipin immunoglobulin M, immunoglobulin G, lupus anticoagulant, complement 3, complement 4, direct Coombs test, and anti-neutrophil cytoplasmic antibodies were negative. No cryoglobulins and monoclonal proteins were detected. The patient's cerebrospinal fluid analysis was as follows: a protein count of 46,7 mg/dL, glucose 65mg/dL, and normal cell count. The cerebrospinal fluid culture was sterile. Nerve conduction studies of the patient were reported as mix-type sensorimotor demyelinating polyneuropathy (Table-1). Magnetic resonance angiography of the extremities and the capillaroscopic findings were normal. Pregabalin (600mg/day), acetylsalicylic acid (100 mg/day), and nifedipine (30 mg/day) were started to the patient. B12 replacement therapy was not given because the vitamin B12 level was within normal limits. A significant clinical improvement was observed in 2 months of the treatment. The complaints of numbness and tingling in the distal parts of the upper and lower extremities decreased, and the vibration sense and deep tendon reflexes returned to normal.

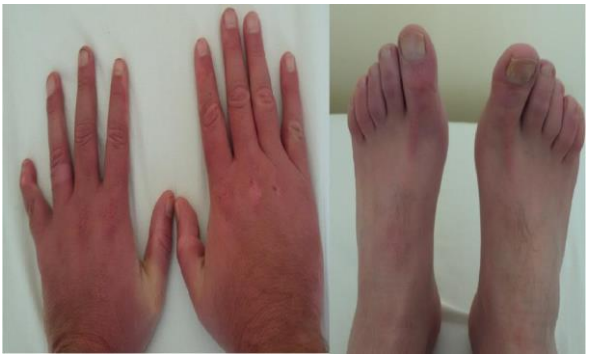

Figure-1. Discoloration of the patient's upper and lower extremities (Raynaud's phenomenon).

Table-1. Nerve conduction study of the patient.

Anatomic Site	Amplitude (mV)	Latency (ms)	Conduction Velocity (m/sec)
Right Median Motor			
Wrist	8.8	3.4	36.0
Elbow	6.1	10.2	
Right Ulnar Motor			
Wrist	9.7	2.8	35.8
Elbow	8.6	9.5	
Right Tibial Motor			
Ankle	9.6	4.9	27.7
Knee	2.1	19.0	
Right Peroneal Motor			
Ankle	15.3	4.4	37.0
Knee	5.4	14.4	
Left Median Motor			
Wrist	8.4	3.2	35.8
Elbow	6.2	9,6	
Left Ulnar Motor			
Wrist	8.7	2.7	32.6
Elbow	7.6	9.1	
Left Tibial Motor			
Ankle	9.2	4.8	28.1
Knee	2.4	17.6	
Left Peroneal Motor			
Ankle	14.8	4.2	36.5
Knee	5.1	13.2	

DISCUSSION

The prevalence of RP has been reported to be approximately 5% in the general population (6). The dysregulation of neural, endothelial, and intravascular control mechanisms was mentioned in RP (3). The assessment of RP includes careful history, examination, laboratory investigations, and imaging studies (2). The complete blood count, serum glucose, serum protein immunofixation, thyroid function tests, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibody, and nail fold capillaroscopy are useful for evaluating RP (7). Angiography is used to exclude obstructive vascular diseases. Patient education and lifestyle changes such as stopping smoking cessation are important for patients with RP (7). The calcium-channel blockers, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, alpha-blockers, phosphodiesterase type 5 inhibitors, and selective serotonin reuptake inhibitors are some of the pharmacological therapies for the treatment of RP (7). The surgical intervention is an option in patients with refractory RP to medical treatment (3). Our patient's laboratory investigation was negative for autoimmune diseases and malignancy. And, he was diagnosed as primary RP.

The clinical features of neuropathies are neuropathic sensory symptoms, and deep tendon reflexes are usually reduced or absent (8). A careful history, examination, and appropriate investigations are needed for peripheral polyneuropathies' diagnosis. In addition, erythrocyte sedimentation rate, renal and liver function tests, thyroid profile, fasting blood glucose, vitamin B12, folate, serum protein electrophoresis, antinuclear antibody, double-stranded DNA antibodies, and extractable nuclear antigens analysis are helpful in diagnosis. Nerve biopsy is rarely required, however, it may be used in the diagnosis of vasculitis (8). Non-pharmacological management includes input from the multidisciplinary team. A multidisciplinary team should include physiotherapists, occupational therapists, speech-language therapists, and psychotherapists. Orthotics are used for drop foot or upper limb palsies. Gait training, stretching and muscle-strengthening exercises were applied to our patient as rehabilitation methods. The management of peripheral neuropathies includes specific treatments and general measures. Specific treatments are vitamin B12 replacement therapies. General measures are symptomatic drug treatment for neuropathic pain including antidepressants (amitriptyline, nortriptyline, venlafaxine, duloxetine) or anti-epileptic agents (carbamazepine, gabapentin, pregabalin) (8). We used pregabalin (600mg/day), acetylsalicylic acid (100 mg/day), and nifedipine (30 mg/day) for the treatment of Raynaud's phenomenon with sensorimotor demyelinating polyneuropathy

CONCLUSION

We report a case of PRP with sensorimotor demyelinating polyneuropathy, which highlights the importance of neural mechanisms for RP. Similar pathogenetic mechanisms play role in these two diseases. The neural mechanisms in both RP and sensorimotor demyelinating polyneuropathy can explain the common pathophysiology (3, 9). Clinicians should consider the sensorimotor demyelinating polyneuropathy in PRP, and the patients should be examined in this respect.

Conflict of interest: We have no conflict of interest.

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