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Median, Ulnar and Femoral Neuropathy in a Patient with Crimean-Congo Hemorrhagic Fever

Kırım-Kongo Kanamalı Ateşi Tanılı Bir Hastada Gelişen Median, Ulnar ve Femoral Nöropatisi

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Summary

Crimean-Congo hemorrhagic fever (CCHF) is an acute, tick-borne viral disease with severe hemorrhagic manifestations, high mortality rate and potential of man-to-man transmission and affects almost only humans. We report a 17-year-old female patient with right median and ulnar neuropathy and left femoral neuropathy, who had been treated for CCHF six months before. In the literature, we found only one case, in which median and ulnar neuropathy due to compartment syndrome during the course of CCHF has been reported. We are of the opinion that it should be kept in mind that median and ulnar neuropathy due to compartment syndrome and femoral neuropathy due to compression from iliopsoas hemorrhage or due to compartment syndrome resulting from hemorrhage within the iliacus fascia may develop in the course of CCHF. *Turk J Phys Med Rehab 2010;56:196-200.*

Key Words: Crimean-Congo hemorrhagic fever, neuropathy, compartment syndrome

Özet

Kırım Kongo kanamalı ateşi (KKKA), kenelerle bulaşan, genelde sadece insanları etkileyen ve insandan insana geçiş potansiyeli olan, ciddi hemorajik belirtiler ve yüksek mortalite oranına sahip, akut viral bir hastalıktır. Biz bu yazımızda, altı ay öncesinde Kırım-Kongo kanamalı ateşi tanısıyla tedavi edilen, sağ üst ekstremitesinde median ve ulnar nöropati ve sol alt ekstremitede femoral nöropati gelişen 17 yaşında bir bayan hastayı sunduk. Yapılan literatür taramasında, KKKA hastalığının seyri sırasında oluşan kompartman sendromuna bağlı gelişen median ve ulnar nöropatinin olduğu sadece bir vakaya rastlanmıştır. Kırım-Kongo kanamalı ateşi hastalığının seyri sırasında kompartman sendromuna bağlı median ve ulnar nöropatinin ve iliopsoas hemorajisinin basısına veya iliak fasya içine kanama sonucu ortaya çıkan kompartman sendromuna bağlı femoral nöropatinin gelişebileceğinin akılda tutulması gerektiği kanaatindeyiz. *Türk Fiz Tıp Rehab Derg 2010;56:196-200.*

Anahtar Kelimeler: Kırım-Kongo kanamalı ateşi, nöropati, kompartman sendromu

Case

A 17-year-old rural Caucasian female patient, who is a member of a family that deal with animal breeding, applied to our outpatient clinic with symptoms of weakness and deformity in her right hand, muscle wasting in her right forearm and left thigh. The past history revealed that she had been hospitalized at the infectious diseases clinic of a hospital, where she had been investigated and treated due to symptoms of nose bleed, hematuria, melena and prolonged menstrual bleeding. The patient had been diagnosed as having Crimean-Congo hemorrhagic fever (CCHF). During the treatment period in the hospital, diffuse ecchymoses had appeared on the left thigh and on the right hand, forearm and the distal half of the arm and, those sites had been tight and swollen.

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The musculoskeletal examination of the patient, who realized weakness in her right hand and left thigh just after discharge from the infectious diseases clinic demonstrated hyperextension of the metacarpophalangeal (MCP) joints slight flexion of the proximal interphalangeal (PIP) and distal interphalangeal (DIP) joints (Picture 1). Atrophies were detected in the muscles of the volar side of the right forearm, in the interosseous, thenar and hypothenar muscles of the right hand, and in the anterior muscle group of the left thigh. There were 4 cm of difference in the perimeter of the forearm and 5 cm difference in the perimeter of the thigh. At sensory examination, hypoesthesia was detected in the superficial and pain sensation examinations in areas corresponding to the innervation sites of the right median, right ulnar and left femoral nerves. At motor examination according to the Oxford scale, the muscle strength of the right wrist flexor muscles was 4/5 and the muscle strength of the left knee extensor muscles was 2/5.

Electroneuromyographic (ENMG) examination revealed findings compatible with partial axonal degeneration of the median and ulnar nerves proximal to the elbow showing axonal regeneration (the distal muscles were more severely affected) (Table 1). The ENMG study of the right radial nerve was normal. The presence of severe partial axonal degeneration and rare regeneration was determined in the left femoral nerve just after the innervation of iliopsoas muscle (localization is on the inguinal ligament). The ENMG studies of the left upper extremity and the right lower extremity were normal. Hyperintense signal changes on magnetic resonance imaging (MRI) with fat suppression were detected in the median and ulnar nerves on the supracondylar region of the right humerus. At this level, some hypointense irregularly bordered soft-tissue signal changes that might be compatible with fibrosis or granulation tissue around the median nerve were present on T1- and T2-weighted MRI with fat suppression (Picture 2). Hyperintense signal changes that were compatible with nerve atrophy were observed on T2- and T1-weighted MRI of the left inguinal region. With the help of the detailed history, neurological examination, ENMG studies and MRI findings, we concluded that the patient had median and ulnar nerve neuropathy due to compartment syndrome in the upper extremity and femoral neuropathy due to iliopsoas hemorrhage in the lower extremity during the course of CCHF.

During hospitalization period, range of motion exercises, isometric exercises and progressive resistive exercises were performed, hand and wrist splint was given, and the patient was discharged with home exercises programme.

Discussion

CCHF is a fatal viral disease that is seen generally in Africa, Asia, Eastern Europe and the Middle East. Although it has not been studied well yet and shows some variations among the regions, the clinical features characterized by hemorrhage, myalgia and fever frequently show a dramatic progression (1). While the cases of CCHF have been reported as an epidemic in the neighboring countries since 1970, in Turkey it has been first reported in 2002. The clinical course of CCHF infection has four phases: incubation, prehemorrhagic, hemorrhagic and convalescence (1,2). The hemorrhagic period is short. It generally begins on the 3rd-6th days of the disease and prolongs 2-3 days (2). The hemorrhagic manifestations vary from petechiae seen on the mucous membranes and skin to large hematomas (1).

Only one case was found in the wide scanning of the literature related with the occurrence of compartment syndrome in the course of CCHF. In a case reported by Moghtaderi et al. (3), median and ulnar neuropathy was observed at the 3rd month of treatment for CCHF, and it was noted that these clinical signs were developed due to compartment syndrome. Although there is no other study of neuropathy occurring in the course of CCHF, there are several studies of neuropathy in the course of haemophilia. In haemophilia, median neuropathy is most commonly seen and the ulnar and radial nerves are less commonly involved. These neuropaties occur most commonly due to intramuscular hemorrhage and less commonly due to nerve compression as a consequence of hemorrhage near the nerve. It has also been reported that hemorrhage within the nerve sheath has to be considered (4,5).

Acute limb compartment syndrome is a condition that is characterized by highly raised pressure within a fascial space that causes a decrease in the capillary perfusion below the level necessary for tissue viability (6). Acute compartment syndrome may develop due to: 1) decreased intracompartmental volume (constructive dressings and casts, closure of fascial defects, thermal injuries and frostbite); 2) increased intracompartmental content due to accumulation of edema, hemorrhage or both edema and hemorrhage (7). Regardless of the etiology-traumatic or hemorrhagic, the initial injury causes swelling within the compartment. This results in increased intracompartmental pressure due to compressive closure of thin-walled venules as a result of hypertension in the venous side of this capillary bed. The increase in the hydrostatic pressure causes an additional increment in the intracompartmental pressure, and a vicious circle develops (8). Finally, arteriolar compression leading to muscle and

Table 1. ENMG findings of the patient.

MOTOR NERVE		Lat (ms)	SD	Amp (mV)	SD	CV (m/s)	F (ms)
Right Medianus							27.5
	Wrist-APB	4.0		2.2			
	Elbow-Wrist	9.8		0.4		-83	
Right Ulnaris							-
	Wrist-ADM	2.5		0.8			
	Elbow-Wrist	6.7		0.9		24	

SENSORY NERVE		Lat (ms)	Amp (uV)	CV (m/s)	SD
Right Medianus					
	Palm-Wrist	-	-		
	Dig I-Wrist	-	-		
	Dig III-Wrist	-	-		
Right Ulnaris	Palm-Wrist	-	-		
Right Radialis					
	IOD I-forearm	1.85	28		
Right Cut antebr Lat					
	Stim 1- Rec 1	1.94	9.8	56.7	-1.6
Right Cut antebr Med					
	Stim 1- Rec 1	3.6	3.1	30.6	-6.9

Muscle (Innervation)		Fib	PSW	Amp	Dur	Poly	Stabil	IP
Right Ext indicis (radialis C7-8)	Normal	0	0/10	Normal	Normal	Normal	Normal	Normal
Right Ext dig communis (radialis C7-8)	Normal	0	0/10	Normal	Normal	Normal	Normal	Normal
Right Flex dig superfic (medianus C7 C8 T1)	Slightly inactive, neurogenic	+++	2/10	+	+	+		-
Right Abd dig min (man) (uln ramus prof. C8 T1)	Slightly inactive, neurogenic	++++	3/10	+	+	++		-
Right Flex carpi ulnaris (ulnaris C8 T1)	Slightly inactive, neurogenic	0	0/10	+	+	+		-
Right Abd pollicis brev (medianus, C8 T1)	Slightly inactive, neurogenic	+++	3/10	+	+	+		-
Muscle (Innervation)		**Fib**	**PSW**	**Amp**	**Dur**	**Poly**	**Stabil**	**IP**
Left Vastus lateralis (femoralis L_2, L_3, L_4)	Moderate subacute neurogenic	++++	3/10	+	+	+		-
Left Psoas minor (genitofemoralis L_1, L_2)	Normal	0	0/10	Normal	Normal	Normal	Normal	Normal
Left Vastus intermedius (femoralis L_2, L_3, L_4)	Moderate subacute neurogenic	++++	4/10	+	+	+		-
Left Add magnus (obturatorius L_2, L_3, L_4)	Normal	0	0/10	Normal	Normal	Normal	Normal	N o r m a l

Sensory Nerve	Lat (ms)	SD	Amp (uV)	CV (m/s)
Left Medianus (palm-wrist)	2.4	3.1	58	43.8
Left ulnaris (palm-wrist)	2.6	4.3	24	46.2
Right Paroneus superfi (foreleg-ankle)	2.2		28	40.9
Right Suralis ankle-foreleg	2.8		13	41.1

nerve ischemia appears. If an appropriate treatment is not performed immediately, muscle infarct and nerve injury occur (6). The etiologies of acute compartment syndrome may be divided into two groups: 1) related to fracture; 2) unrelated to fracture. The group that is unrelated to fracture may be divided into two subgroups: subgroup with a history of injury and subgroup with swelling unrelated to any traumatic event (9).

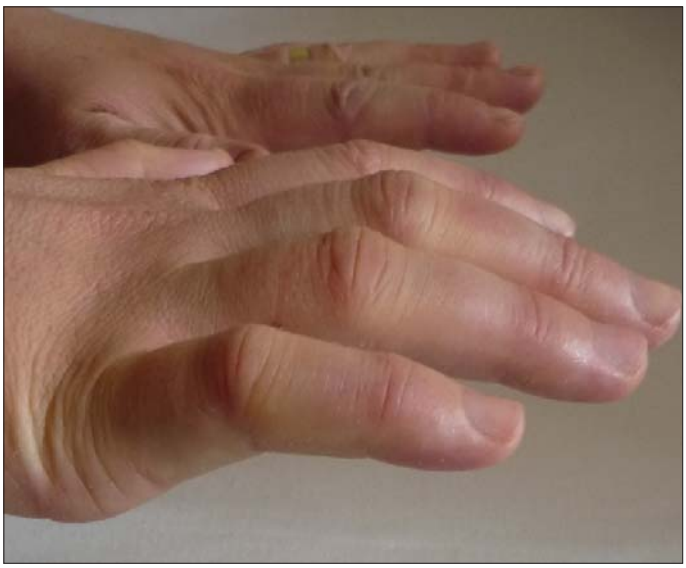
Hope MJ et al. (9) compared series of patients with acute compartment syndrome related or unrelated to fracture and found that the diagnosis was usually delayed in the group that

was unrelated to fracture. However, this delay was in terms of hours. Similarly, Prasan LM et al. (10) reported that the average time interval between the diagnosis and the fasciotomy ranged from 2 hours to 24 hours (average: 9.4 hours) in series of children with acute upper extremity compartment syndrome due to causes unrelated to fracture. The time intervals between the diagnosis of the first disease and the diagnosis of the neuropathy in the patient presented by Ali Moghtaderi (3) and in our patient are 3 months and 6 months, respectively.

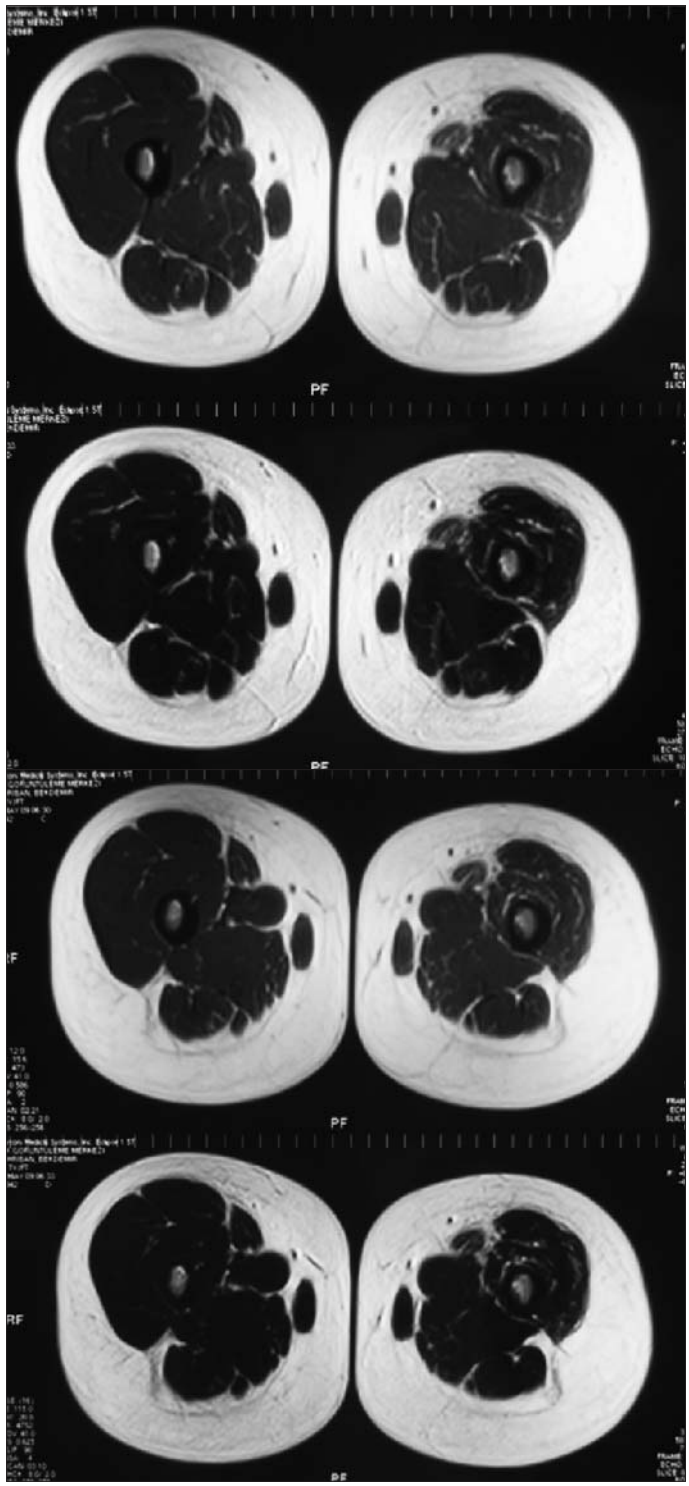
The diagnosis of compartment syndrome can be made according to clinical symptoms and sings. Largerstrom CF et al. (8) did not feel the need to measure the intracompartmental pressure in order to diagnose or confirm the diagnosis of compartment syndrome in patients with evident clinical findings. Mubarek SJ et al. (7) noticed that they could diagnose the compartment syndrome with clinical findings in most of the patients. However, they also underlined the importance of measurement of intracompartmental pressure in determining the cause of neuropathy (trauma or compartment syndrome). Therefore, they also point out the complexity of establishing the diagnosis of compartment syndrome clinically in uncooperated patients.

The symptoms of compartment syndrome are pain, pain at passive stretching and hyperesthesia in the territory of the nerve that moves inside the compartment. The physical signs of compartment syndrome are loss of motor function, taut extremity, weak capillary refilling and decreased distal pulsation (8). The acute compartment syndrome has to be kept in mind in patients with taut, swollen and ecchymotic extremity. The early diagnosis and immediate treatment of the syndrome may correct the problem before the occurrence of permanent damage (11). Our patient had noticed ecchymoses on the extremities (right upper and left lower), which had been painful, swollen and taut. These symptoms and signs of our patient were accepted in favour of hemorrhage and compartment syndrome.

Peripheral neuropathy that occurs secondary to hemorrhage proximally in the lower extremities may develop due to haemophilia, anticoagulation treatment, trauma, hip surgery and hemorrhage from arteriosclerotic aneurysm of the aorta or iliac vessels. Femoral neuropathy is the most common form of these neuropathies (12,13). In a study, Balkan C et al. (14) evaluated 146 hemophilic patients and detected femoral



Picture 1. The MCP joints were at hyperextension, and PIP and DIP joints were slightly at flexion.



Picture 2. Hypointense, irregularly bordered soft-tissue signal changes that might be compatible with fibrosis or granulation tissue around the median nerve were present on T1- and T2-weighted MR imaging with fat suppression.

neuropathy in 8 patients with iliopsoas hemorrhage confirmed by ultrasonography. The authors suggested that this neuropathy occurred as a consequence of compression of hemorrhage causing expanding of muscle tissue. They concluded that the physicians should be aware of probability of iliopsoas hematoma in patients with pain around the hip joint, femoral neuropathy and hip flexor contracture.

Goodfellow et al. (15) reported that the psoas and iliacus muscles move in separate compartments, except for the thigh region, the psoas fascia is thin and easily stretchable and able to conserve large volumes of fluid with low pressures, and it was shown that the stretched psoas fascia do not affect the femoral nerve. On the other hand, the iliacus fascia is less stretchable, and minimal swelling at the distal thick and transverse section of the fascia causes femoral neuropathy due to compartment syndrome. In our patient, we thought that femoral neuropathy developed due to compression of iliopsoas hemorrhage or due to compartment syndrome resulting from hemorrhage within the iliacus fascia. Neuropathy that develops due to intraneural hemorrhage has been reported in the literature, even though it is rare (16). This matter should also be considered in etiologic evaluations.

Finally, it may be useful to keep in mind that median, ulnar and femoral neuropathies may develop in the course of CCHF. After discharge from hospital, detailed neurological examination of patients has to be performed on routine follow-up visits, and if needed, ENMG evaluation may help to detect neuropathy.

References

1. Ergönül Ö. Crimean Congo haemorrhagic fever. Lancet Infect Dis 2006;6:203-14. [Abstract] / [Full Text] / [PDF]
2. Vorou R, Pierroutsakos IN, Maltezoo HC. Crimean-Congo hemorrhagic fever. Curr Opin Infect Dis 2007;20:495-500. [Abstract]
3. Moghtaderi A, Alavi-naini R, Azimi H. Compartment syndrome: an unusual course for a rare disease. Am J Trop Med Hyg 2005;73:450-2. [Abstract] / [PDF]
4. Lancourt JE, Gilbert MS, Posner MA. Management of bleeding and associated complications of hemophlia in the hand and forearm. J Bone Joint Surg Am 1977;59:451-60. [Abstract]
5. Kaymak B, Özçakar L, Çetin A, Erol K, Özçakar B. Concomitant compression of median and ulnar nerves in a hemophiliac patient: a case report. J Bone Spine 2002;89:397-412. [Abstract] / [Full Text] / [PDF]
6. Tiwari A, Haq AI, Myint F, Hamilton G. Acute compartment syndromes. Br J Surg 2002;89:397-412. [Abstract] / [PDF]
7. Mubarak SJ, Hargens AR. Acute compartmant syndromes. Surg Clin North Am 1983;63:539-65.
8. Lagerstrom CF, Reed RL 2nd Rowlands BJ, Fisher RP. Early fasciotomy for acute clinically evident posttraumatic compartment syndrome. Am J Surg 1989;158:36-9. [Abstract] / [PDF]
9. Hope MJ, McQueen MM. Acute compartment syndrome in the absence of fracture. J Ortop Trauma 2004;18:220-4. [Abstract]
10. Prasarn ML, Quellette EA, Livingstone A, Giuffrida Y. Acute pediatric upper extremity compartment syndrome in the absence of fracture. J Pediatr Ortop 2009;29:263-8. [Abstract]
11. Grattkau BE, Epps HR, Di Scala C. Compartment syndrome in children and adolescents. J Pediatr Surg 2005;40:678-82. [Abstract] / [Full Text] / [PDF]
12. Dingeman RD, Mutz SB. Hemorrhagic neuropathy of the sciatic, femoral and obturator nerves. Clin Orthop Relat Res 1997;127:133-6. [Abstract]
13. Katz SG, Nelson IW, Atkins RM, Duthie RB. Peripheral nerve lesions in hemophilia. J Bone Joint Surg Am 1991;73:1016-9. [Abstract] / [PDF]
14. Balkan C, Kavaklı K, Karapınar D. Iliopsoas haemorrhage in patients with haemophilia: Results from one centre. Haemophilia 2005;11:463-7. [Abstract] / [Full Text] / [PDF]
15. Goodfellow J, Fearn CB, Matthews JM. Iliacus haematoma. A common complication of haemophlia. J Bone Joint Surg Br 1967;49:748-56. [Abstract] / [PDF]
16. Mandal A, Colville J, Jones M, Erdmann MW. An unusual case of haemorrhagic median neuropathy. Ann R Coll Surg Engl 2004;86:258-9. [Abstract] / [PDF]