Journal of Experimental and Clinical Medicine
https://dergipark.org.tr/omujecm

Case Report

J Exp Clin Med
2022; 39(3): 910-912
doi: 10.52142/omujecm.39.3.60

Adamkiewitz syndrome in Marfan syndrome and rehabilitation outcomes-3 cases and review of the literature

Zeynep KIRAÇ ÜNAL[1,*], Gülnur ÇELİK[1], Sadik Yiğit YILDIZ[1], Damla CANKURTARAN[1] Ece ÜNLÜ AKYÜZ[1], Ilker İNCE[2]

[1]Department of Physical Medicine and Rehabilitation, University of Health Sciences Diskapi Yildirim Beyazit Education and Research Hospital, Ankara, Türkiye
2 Department of Cardiovascular Surgery Clinic, University of Health Sciences Diskapi Yildirim Beyazit Education and Research Hospital, Ankara, Türkiye

Received: 26.05.2022 • **Accepted/Published Online:** 04.07.2022 • **Final Version:** 30.08.2022

Abstract

Acute Spinal Cord Ischemia Syndrome (ASCIS) is rare but clinically significant. Aortic dissection (AD) is one reason for ischemia of the spinal cord. The artery of Adamkiewicz provides the major blood supply to the anterior thoracolumbar spinal cord, and injury to this artery can cause consequential neurologic damage.Three male patients applied to our rehabilitation clinic with paraplegia. All three had AD due to Marfan Syndrome and clinical findings due to Adamkiewitz artery ischemia. Although we later learned that one of the patients later died from a new ruptured aortic aneurysm, we achieved good motor gains in these rare cases with rehabilitation. Although paraplegia is rare as the initial symptom in AD, vascular etiology should not be forgotten in acute paraplegia. The prognosis of ASCIS is known to be poor, but a good and individualized rehabilitation program is required.

Keywords: Acute Spinal Cord Ischemia Syndrome, Marfan syndrome, paraplegia, rehabilitation

1. Introduction

The spinal vasculature has unique anatomical connections, but there are regions of the spinal cord that can easily become ischemic (1). The artery of Adamkiewicz (AKA), which supplies approximately one-quarter of the spinal cord, is the largest vessel that reaches the spinal cord. AKA mostly originates between T8 and L1 (89%) and has been reported in some people in T4-8 (2). The involvement of the AKA causes ischemia in the anterior regions of the spinal cord, while the posterior cord fed from the posterior spinal artery is less affected. Lesions in T4-6, in particular, are common because this is a 'watershed' area (1).

Acute Spinal Cord Ischemia Syndrome (ASCIS) is rare. However, it is a disease that can cause permanent sequelae. In a study on ASCIS, 16% of cases were associated with aortic pathology (3). Aortic Dissection (AD), whose mortality is 80%, may cause malperfusion or occlusion of segmental arteries feeding the spinal cord, and paraplegia may develop in 2-8% of patients (4,5). Marfan Syndrome (MS) is a genetic disease associated with reduced life expectancy due to the risk of AD and rupture (6). Due to its rarity, 3 MS cases with AD-related AKA Syndrome and their rehabilitation results will be presented here.

2. Case Presentations

2.1. Case 1

A 25-year-old male patient was admitted to our clinic for rehabilitation with complaints of inability to walk. The patient, diagnosed with MS at the age of 14, had a history of surgery due to aortic valve replacement (AVR) and mitral valve replacement (MVR). The patient was admitted to the emergency department due to tearing back pain, an aortic aneurysm rupture was detected, and he was operated. We learned that cerebrospinal fluid (CSF) drainage was inserted in the patient, who developed motor and sensory deficits in the bilateral lower extremity on the postoperative first day, to provide reperfusion in the Medulla Spinalis.

There were a pectus excavatum and an operation scar on the anterior chest wall in the physical examination. The patient had sitting balance. While bilateral lower extremity distal muscle strength was 2/5 globally, right lower extremity proximal muscle strength was 1/5 and left lower extremity proximal muscle strength was 2/5. There was no apparent sensory deficit. We evaluated the patient on the ASIA disorder scale (ABS) D. There was grade 1 spasticity in the lower extremities globally according to the Modified Ashworth Scale. Bilateral patella and Achilles reflexes were hyperactive; pathological reflexes were positive. Routine biochemistry tests were normal.

*Correspondence: zeynepkirac88@gmail.com

We diagnosed the patient with AKA Syndrome in light of the present findings. The patient underwent bilateral lower extremity electrical stimulation, verticalization, cycling exercises, balance and coordination exercises, gait training, breathing exercises, and occupational therapy in our clinic and was discharged to be ambulated with a pair of walking sticks under surveillance. We found from the patient records that the patient died after 1.5 years due to the aortic aneurysm rupture again.

2.2. Case 2

A 25-year-old male patient diagnosed with MS applied to the emergency department due to stabbing pain in his back. The patient was urgently operated for aortic aneurysm rupture and applied rupture repair and aortic valve replacement. External lumbar drainage was applied to provide spinal cord reperfusion. The patient, whose medical stabilization was achieved, was referred to our clinic due to weakness and loss of sensation in the lower extremities.

The general condition of the patient, who was transferred to our service for ASCIS rehabilitation one month after the event, was good, and his systemic examination was normal. The bilateral lower extremity muscle strength was 3/5 in the proximal and 2/5 in the distal. We evaluated the patient's neurological injury level as L2 ABS C. We could not detect the patient's Patella and Achilles tendon reflexes, who had sitting balance and could be ambulated with a wheelchair. or any pathological reflex. There was no incontinence. The patient's Functional Independence Scale (FIM) score was 74 points. We started a rehabilitation program considering the patient's spinal cord damage due to AKA ischemia.

In the rehabilitation program, we gave lower extremity electrical stimulations to the patient and cycling exercises, ambulation training, balance and proprioception exercises, and occupational therapy.

On the 30th day of hospitalization, DTRs were taken as hyperactive, and Babinski's reflex became positive. When he was discharged from the rehabilitation clinic, the patient had ABS C, neurological level L2, and his FIM score reached 84 points. The patient could ambulate up to 40 meters with a pair of soft knee orthoses and a walker.

After two months, the patient was discharged with appropriate anticoagulant treatment and a home exercise program.

2.3. Case 3

A 23-year-old MS patient applied to the emergency department with the complaint of chest pain. Rupture repair with AD diagnosis and external lumbar drainage were performed. On the fifth post-op day, a cardiac pacemaker was implanted in the patient with deep bradycardia. After medical stabilization, the patient was referred to our clinic because of weakness and loss of sensation in the lower extremities.

Three weeks after the event, the patient was transferred to our service with the diagnosis of ASCIS. We performed ABS C and detected neurological level T12. The general condition was good, the systemic examination was roughly normal, the proximal muscle strength of the patient was 3/5, and the distal muscle strength was 2/5. The patient, who was ambulatory at the wheelchair level, had sitting balance. The FIM score was 82 points. The patient did not have pressure sores, had no incontinence and spasticity, could not obtain DTR, and did not have pathological reflexes.

Considering that the patient had ASCIS due to ischemia of the AKA, we initiated a rehabilitation program. We could not give electrical stimulation because he had a cardiac pacemaker. We gave lower extremity strengthening exercises, verticalization with a tilt table, ambulation training, cycling exercises, balance and proprioception exercises, and occupational therapy.

We started pregabalin for neuropathic pain and increased it to 225 mg/day. After a while, the patient's pain subsided; thus, we stopped pregabalin. We evaluated the patient as neurological level L2 and ABS C at the discharge examination. The patient could be ambulated in a short distance with one soft knee orthosis and a walker. The FIM score improved by 13 points.

We advised the patient to continue with the home exercise program and warned him about the risk of falling. He was discharged after three months to be examined in the follow-up.

3. Discussion

The midthoracic part of the spinal cord, which is usually fed by a single anterior radicular artery, is the weakest part of the blood supply; the lower thoracic and lumbar parts are supplied by the anterior radicular arteries, the largest of which is named "AKA" (2).

Spinal cord ischemia usually develops on the basis of atherosclerosis (7). Depending on the infarct site, various spinal cord syndromes may occur. These are Anterior Spinal Artery (ASA) Syndrome, Posterior Spinal Artery Syndrome, Spinal Sulcal Artery Syndrome, "Man-in-the-barrel" syndrome and Syndrome of the AKA (8,9).

Syndrome of the AKA appears as complete transverse spinal cord syndrome. It means that flaccid paresis at the level and spastic paraparesis below the level of infarction, positive pathological reflexes, loss of sensation, and bladder and bowel dysfunction can be seen (8,9). These are similar to our cases' clinical appearance, and except for our third case, we obtained pathological reflexes in all of them.

When we examined their histories, it was remarkable that all of our cases were diagnosed with MS. One of the most critical Ghent criteria for the diagnosis of MS is AD, and the life expectancy is reduced in those with large aortic arch and arch diameters (10). Spinal cord ischemia in dissecting aneurysms is observed at a high rate of 38% (11).

The prognosis of ASCIS is generally not good. In one study, 115 patients with spinal cord ischemia were followed for three years, and approximately 23% of the patients died, 42% of the survivors used a wheelchair, 26% were mobilized with support, and only 32% could walk without support (12). Although we later heard about the death of our first case, all our patients were discharged with support at the end of the rehabilitation program. Ambulatory functional and clinical outcomes, which were likely due to multiple factors, including differences in mean age, etiology of injury, length of follow-up, and most notably, our higher proportion of ASIA A patients, indicating the significant poor prognostic predictor of a severe initial cord injury, differ between 20% and 70% (13).

In conclusion, although paraplegia is rare as the first symptom in AD, vascular etiology should be considered in acute paraplegia. There is no specific treatment other than prevention of complications, treatment of the underlying cause and rehabilitation. Relatively poor prognosis should be recognized, and these cases should be managed in rehabilitation centers.

Conflict of interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, or publication of this article.

Funding

The author(s) received no financial support for this article's research, authorship, and/or publication.

Acknowledgments

None.

Authors' contributions

Concept: Z.K.Ü., E.Ü.A., Design: Z.K.Ü., D.C., E.Ü.A., Data Collection or Processing: G.Ç., S.Y.Y., İ.İ., E.Ü.A., Analysis or Interpretation: Z.K.Ü., G.Ç., S.Y.Y., D.C., İ.İ., E.Ü.A., Literature Search: Z.K.Ü., E.Ü.A., Writing: Z.K.U., D.C., E.Ü.A.

References

1. Weidauer S, Nichtweiß M, Hattingen E, Berkefeld J. Spinal cord ischemia: aetiology, clinical syndromes and imaging features. Neuroradiology 2015; 57(3): 241-257.
2. Takagi H, Ota H, Natsuaki Y, Komori Y, Ito K, Saiki Y, et al. Identifying the Adamkiewicz artery using 3-T time-resolved magnetic resonance angiography: its role in addition to multidetector computed tomography angiography. Jpn J Radiol 2015; 33: 749–756.
3. Nedeltchev K, Loher TJ, Stepper F, Arnold M, Schroth G, Mattle HP, et al. Long-term outcome of acute spinal cord ischemia syndrome. Stroke 2004; 35(2): 560-565.
4. Khan IA. Clinical manifestations of aortic dissection. J Clin Basic Cardiol 2001; 4: 265.
5. Prakash, B, Pai RK., Chaitra V, Ramasamy P, Shuba N. A Case of Acute Paraplegia Due to Aortic Dissection in Marfan Syndrome.Journal of Neurosciences in Rural Practice 2017; 8(2): 316-319.
6. Pyeritz RE. Marfan syndrome: 30 years of research equals 30 years of additional life expectancy. Heart 2009; 95: 173–175.
7. Alblas CL, Bouvy WH, Lycklama À Nijeholt GJ, Boiten J. Acute spinal-cord ischemia: evolution of MRI findings. J Clin Neurol 2012; 8: 218-223.
8. Novy J, Carruzzo A, Maeder P, Bogousslavsky J. Spinal cord ischemia. Clinical and imaging patterns, pathogenesis and outcomes in 27 patients. Arch Neurol 2006; 63: 1113–1120.
9. Mull M. Acute spinal cord ischemia: diagnosis without therapeutic options? Clin Neuroradiol 2005; 15: 79–88.
10. Loeys BL. Revised Ghent criteria for the diagnosis of Marfan syndrome (MFS) and related conditions. J Med Genet 2010; 47: 476-485.
11. Elmacı T. Kardiovasküler cerrahi girişimlerde spinal iskemi. Journal of Istanbul Faculty of Medicine 2011; 63(4).
12. Robertson CE, Brown RD Jr, Wijdicks EF, Rabinstein AA. Recovery after spinal cord infarcts: long term outcome in 115 patients. Neurology 2012; 78:114.
13. Ge L, Arul K, Stoner M, Mesfin A. Etiology and Outcomes of Spinal Cord Infarct: A Case Series From a Level 1 Trauma Center. Global Spine J 2020; 10(6): 735-740.