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Case Report / Olgu Sunumu

Brainstem Hypoplasia Presenting with Mirror Movement

Ayna Hayali Hareket ile Prezente Olan Beyin Sapı Hipoplazisi

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Summary

A 20 year old female patient, who was operated for congenital syndactyly of her left hand at age of 5, admitted to neurology policlinic with involuntary movements of her hands. We saw mirror movement (MM) of her right hand when she was writing and catching something with her left hand. Also we saw MM of her left hand when she was doing something with her right hand with a lower amplitude. Cervical magnetic resonance imaging (MRI) revealed no abnormality. Brain MRI revealed hypoplasia of right middle, inferior cerebellary peduncle, olive and pyramis. MM shows homolog muscle activity which imitates contralateral movement, during a spesific task. This movement usually affects upper extremity especially hand. Corticospinal tract dysfunction is often considered in the pathogenesis. MM may present as part of cervicomedullary junction abnormality, cerebral palsy, cerebrovascular disease, Parkinson's disease. We wanted to discuss the pathogenesis of MM in our patient with syndactyly and MRI abnormality.

Keywords: Mirror movement, brainstem hypoplasia, syndactyly

Öz

Doğuştan sol elinde sindaktili olan ve 5 yaşında opere olan 20 yaşında kadın hasta polikliniğe ellerinde istemsiz hareketler şikayeti ile başvurdu. Hastanın sol eli ile yazı yazma, kavrama gibi hareketler yapması sırasında sağ elinde ayna hayali hareket görüldü. Aynı hareket daha düşük amplitüdlü olarak hasta sağ eli ile bir şeyler yaptığında sol elinde de görülüyordu. Çekilen servikal manyetik rezonans görüntülemesinde (MRG) patoloji gözlenmedi. Beyin MRG'sinde sağ orta, alt serebellar pedinkül, olive ve piramid hipoplazisi görüldü. Ayna hayali hareket belirli bir görevin yerine getirilmesi sırasında karşı taraftaki hareketi taklit eden homolog kas aktivitesini gösterir. Sıklıkla üst ekstremitede; özellikle elde görülür. Patogenezinde sıklıkla kortikospinal yol disfonksiyonu düşünülmüştür. Ayna hareketleri servikomedüller bileşke anomalisi, serebral palsi, serebrovasküler hastalık, Parkinson hastalığı gibi birçok nörolojik hastalık ile birlikte tarif edilmiştir. Hastamızdaki ayna hayali hareketin sindaktili, sağ bulber, olive ve pedinkül hipoplazisi ile beraberliğini paylaşmak ve patogenezi tartışmak istedik.

Anahtar kelimeler: Ayna hayali hareket, beyin sapı hipoplazisi, sindaktili

Introduction

The term mirror movement was first used by Erlenmeyer in 1879. Cohen et al. (1) described mirror movement (MM) is an involuntary and synkinetic/synchronized movement of a part of body that appears during voluntary movement of the opposite side of body. MM shows homologous muscle activity, which imitates contralateral movement during a specific task (2). MM frequently effects the upper extremities, especially the hands. It can be seen in infants and tends to disappear by age 10 after myelination of the corpus callosum. It can be seen in normal adults after excessive exercise or due to exhaustion (3). MM may present as a part of some

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neurologic and physciatric diseases including cervicomedullary junction abnormalities, cerebral palsy, cerebrovascular diseases, Parkinson's disease, symptomatic epilepsies, Friedreich's ataxia, phenylketonuria, amyotrophic lateral sclerosis, Kallman syndrome, and obsessive compulsive disorder (2). In hereditary cases, this condition is inherited in an autosomal dominant or recessive pattern.

Hypoplasia of the brain stem, olivary nucleus, and peduncle are reported in many genetic syndromes and frequently cause severe neurologic sequelae. Syndactyly is a malformation that can accompany genetic syndromes and other malformations.

Herein, a patient with MM and hypoplasia of the brain stem and olivary nucleus is reported and the pathogenesis of MM is discussed.

Case Report

A woman aged 20 years with left hand dominance presented to our polyclinic with involuntary movements of her right hand. It was learned that her symptoms started after she learned to write. Involuntary movements of right hand were observed while she was doing something with her left hand, especially during finer movements of the fingers. Also during voluntary movements of her right hand, involuntary movements of the left hand were observed, but with lower amplitude. When she was aged five years, she had had syndactyly that effected the first two fingers of the left hand and underwent plastic surgery. She had no family history of MM. In the physical examination, she had sequela that effected the first two fingers of the left hand and also the fiingers on her left hand were smaller than on the right. The neurologic examination was normal except for the involuntary movements ofthe right hand while she was catching something or writing using the left hand (Figure 1, 2, 3, 4, 5, 6, 7 are taken from a video recording of the patient while she was handling a reflex hammer). Routine laboratory investigations were normal. Cervical magnetic resonance imaging (MRI) showed no cervicomedullary junction abnormality. T2 sequences of cranial MRI showed hypoplasia of the right middle and inferior cerebellary peduncle, right side of bulbus, olivary nucleus, and pyramis (Figure 8, 9, 10).

Discussion

MM is an involuntary and synkinetic movement of a part of body that appears during voluntary movement of the opposite side, and it can accompany various diseases. The pathophysiology of MM is not wellunderstood. Pyramidal dysfunction is thought to be the reason in congenital cases (4). However, MM is also one of the early signs of Parkinson's disease and this suggests circuits of basal ganglion can also be involved in the pathogenesis (5). Two mechanisms have been suggested to explain the formation of MM (2). First, MM is caused by fast conducting fibers that originat from the hand area in the primary motor cortex (M1) and terminate in the ipsilateral spinal cord without crossing over. This mechanism is suggested for congenital cases. Second, MM is caused by synchronized abnormal activation of both hemispheres. This mechanism is suggested for acquired cases. MM in cervicomedullary junction abnormalities is thought to be caused by involvement of spinal motor neurons and crossed-over pyramidal fibers in the brain stem, which innervate distal muscles (6). Green J. B. (7) suggested that involvement of pyramidal fibers that cross over can result with inhibitory effects on secondary motor neurons and cause MM. An electrophysiologic study on a patient with cervical meningomyelosel showed that reorganization of pyramidal fibers that had crossed over caused MM (8). Presence of MM in a patient with malformation of high cervical spinal cord also supported this theory (9). Our patient hadhypoplasia of the right middle and inferior cerebellary peduncle, right side of bulbus, olivary nucleus, and pyramis; therefore, we think that reorganization of pyramidal fibers that cross over can cause MM, as shown in the literature. Hypoplasia of the brain stem frequently causes clinically severe syndromes. An autopsy study of sudden fetal and infant death revealed hypoplasia of the olivary nucleus

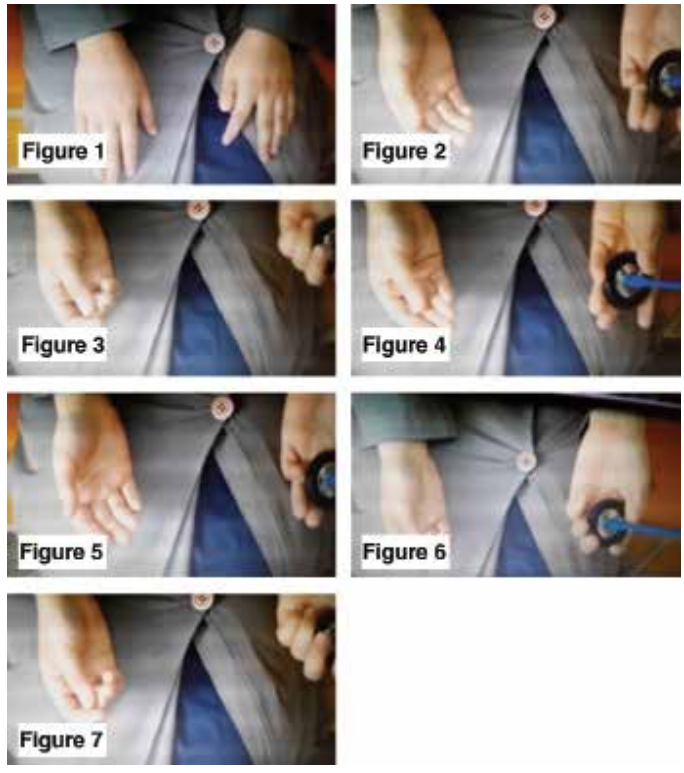

Figure 1, 2, 3, 4, 5, 6, 7. Pictures taken from a video recording of the patient while she was handling a reflex hammer

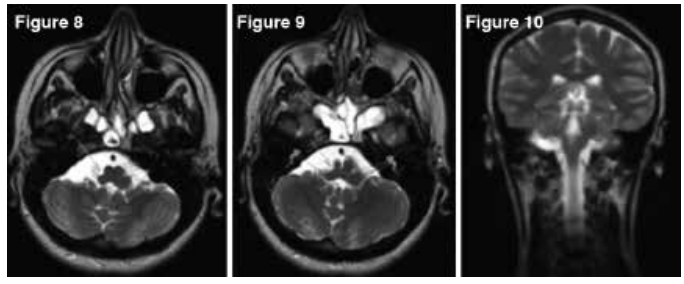

Figure 8, 9, 10. T2 sequences of cranial magnetic resonance imaging showed hypoplasia of the right middle and inferior cerebellary peduncle, right side of bulbus, olivary nucleus, and pyramis

in many cases. Death was thought to be caused by disruption of respiratory rhythm (10). Conversely, our patient had a normal neurologic examination except syndactyly and MM. To our knowledge, this is the first patient with MM and hypoplasia of the brain stem. MRI is important for detection of congenital/acquired neurologic malformations of the brain stem in patients with MM. As a result, we think that dysfunction of the pyramidal tractus and reorganization of pyramidal fibers, which crossed over, were the pathogenetic factors that caused MM in our patient. For detection of rare disorders such as MM, a careful examination is required in patients with involuntary movements and once noticed, cranial MRI is needed to investigate the etiology.

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