Mirror movements in progressive hemifacial atrophy

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Abstract

Mirror movements are simultaneous, involuntary, identical movements occurring during contralateral voluntary movements. These movements are considered as soft neurologic signs seen uncommonly in clinical practice. The mirror movements are described in various neurological disorders which include parkinsonism, cranio veretebral junction anamolies, and hemiplegic cerebral palsy. These movements are intriguing and can pose significant disability. However, no such observation regarding mirror movements in progressive hemifacial atrophy have been reported previously. We are reporting a teenage girl suffering from progressive hemifacial atrophy and epilepsy with demonstrable mirror movements in hand.

Key Words

Hemifacial atrophy, mirror movements, parry romberg syndrome

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Introduction

The mirror movements (MMs) are simultaneously occurring unintended movements in homologous muscles during contralateral voluntary movements. It was first described by Erlenmeyer in 1857. It was truly defined by Cohen *et al*. in 1991.[1] The MMs are rarely observed in clinical practice. These are tricky, unintentional movements, sometimes causing injury to the patient. These MMs are exhibited in cerebral palsy, craniovertebral junction anamoly, parkinsonism, post stroke phase, and myelomeningocoele.[2] In this report, we intended to present an unsual association of MMs in a young girl suffering from progressive facial atrophy along with cerebral hemiatrophy. The literature is silent about presence of MMs in progressive facial atrophy.

Case Report

A 17-year-old girl has generalized tonic clonic seizures followed by loss of consciousness since last 12 years. Initially, these movements are well-controlled with drugs with 4-5 episodes per year but since last 3 years, these movements increases in frequency; and now since last 2 months, patient have daily one to two episodes of these movements. There was no history of headache, vomiting, rash, arthralgia, delayed menarche, and peripheral limb weakness. There is no short neck, low hair line, or decreased neck movement.

On examination, she has atrophy of her left face with slight deviation of mouth and nose towards left side [Figure 1]. During nervous system examination, we found that when she held the examiner finger by her left hand she perform the similar kind of movement (MM) in her right hand [Video 1]. There was no movement noted proximally in upper limb and lower limb. Rest of her neurological examination is normal.

Routine hematological and biochemical parameters of the patient were within normal limits. Electroencephalography (EEG) shows generalized slowing. Magnetic resonance imaging (MRI) brain shows left cerebral atrophy [Figure 2]. The patient was treated with antiepileptic drugs with leviteracetam 500 mg and oxcarbazepine 300 mg twice daily. Her seizures are well-controlled at follow-up after 5 months.

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Discussion

The MMs can be normally observed till 10 years at the point of complete myelination of corpus callosum.[3] The persistence of MMs beyond this age is usually pathological[4] and represent abnormality of central motor drive to relaxed limb. The MMs have been described in cerebral palsy, cranio-vertebral junction anomaly, Klippel feil anamoly, Kallaman syndrome,

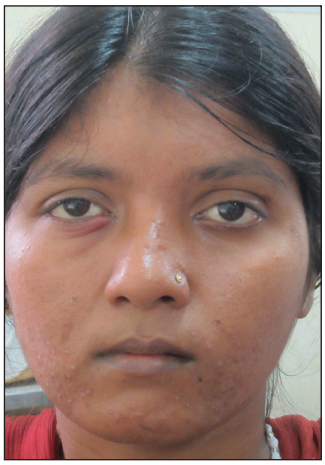

Figure 1: Photograph of the patient showing atrophy of her left face with slight deviation of mouth and nose toward left side

Parkinson's disease, corticobasal degeneration, post stroke, essential tremors, focal hand dystonia, Creutzfeldt-jakob's disease, and Huntigton's disease. However, rarely, it is seen in normal individuals and mostly while performing complex unimanual tasks.[5,6] The MMs can often impede the bimanual tasks and at times can be very disabling. As found in most cases, in our patient, MMs were limited to distal upper limb (wrists and finger movements)[6] and our patient did not have any disability due to MMs.

MM either seems to arise due to activation of both hemispheres when only one of the hemispheres is intended to fire.[7] Another possible mechanism is presence of fast conducting ipsilateral pyramidal tract that does not decussate instead projects on the ipsilateral spinal motor neurons. Both mechanisms can occur in tandem with one another and contribute to clinical symptomatology. However, it has been observed that over-activation of ipsilateral pyramidal tract plays a major role in congenital disorders associated MM.[8]

MM also occur in several acquired and degenerative neurological disorders but the mechanism of mirroring in such conditions remains elusive. The theory of physiological cross talk was proposed because of formation of false synapses. Other mechanisms attributed to MMs were deficient corpus callosum, incomplete pyramidal decussation, loss of inhibition, and over-activation of ipsilateral pyramidal tract.[9]

Our patient was diagnosed as progressive facial atrophy, repetitive focal seizures, and cerebral hemiatrophy with demonstrable MMs. Progressive facial atrophy (Parry Romberg syndrome) is characterized by unilateral facial atrophy which involves muscles, subcutaneous tissues, and even underlying bones.[10] The neurological manifestations include migraine headache, trigeminal neuralgia, focal seizures, and sometimes associated with cerebral hemiatrophy.[11] En coupe De Sabir (localized Morphea) and Parry Romberg syndrome can also have similar appearance and almost 17% of such patients have shown to present with MRI brain abnormalities. However, the authors concluded that whether there is a causal relationship or true association cannot be ascertained.[12]

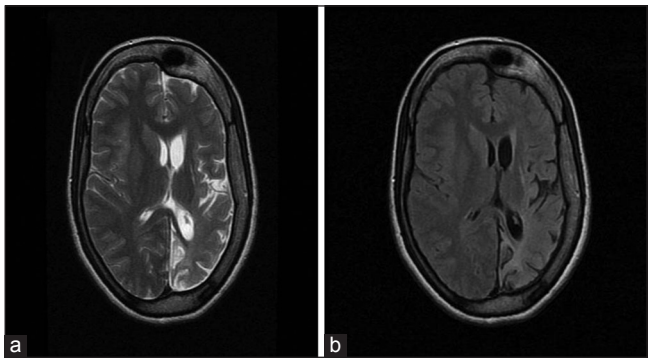

Figure 2: Magnetic resonance imaging of the cranium (a) T2-weighted image (b) T2 fluid attenuated inversion recovery image depicting left sided cerebral hemiatrophy

The MMs in our patient could be related to cerebral hemiatrophy. The plausible explanation for this association may be deficient transcallosal connections, increased activation of non-diseased cerebral cortex, or formation of aberrant corticopyramidal connections are postulated mechanisms. Our hypothesis is supported by previous observations made in a case of acquired MMs in a case of refractory epilepsy after focal cortical resection.[13] A similar case reported by Takajo *et al*. of MMs along with focal onset seizures was in a patient with open lip schizencephaly. The authors postulated neuronal reorganization and reallocation of motor control on the non-diseased cerebral hemisphere. The loss of transcallosal inhibitory control from affected hemisphere may lead to overflow of the stimulatory signals from the non-diseased hemisphere and thereby cause simultaneous homologous movement.[14]

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