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Case Report

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Rehabilitation Results of a Hemiparatic Subject with Sturge-Weber Syndrome and Intractable Epilepsy

Nezire KÖSE [a], Mehmet Gürhan KARAKAYA, Saadet OTMAN

Hacettepe Üniversitesi Fizik Tedavi ve Rehabilitasyon Yüksekokulu, Nöroşirurji Ünitesi, ANKARA

ABSTRACT

This study presents the results of a rehabilitation program in a spastic hemiparetic subject who had undergone hemispherectomy because of Sturge-Weber syndrome and intractable epilepsy. Sturge-Weber syndrome includes unilateral cerebral cortical angiomatosis, which often leads to progressive cerebral dysfunction and epileptic seizures that are medically difficult to control. A three-year-old girl who was followed-up for 2.5 years after a diagnosis of Sturge-Weber syndrome, intractable epilepsy and right spastic hemiparesis was hospitalized at the Department of Neurosurgery for epilepsy surgery since the frequency of her seizures had increased. The functional status and quality of life of our subject, whose seizures disappeared after hemispherectomy, improved after the rehabilitation program. *©2004, Fırat Üniversitesi, Tıp Fakültesi*

Key words: *Sturge-Weber, Intractable Epilepsy, Rehabilitation*

ÖZET

Sturge Weber Sendromu ve Dirençli Epilepsi Nedeniyle Hemipleji Olmuş Bir Vakanın Rehabilitasyon Sonuçları

Bu çalışmada Sturge Weber ve dirençli epilepsi nedeni ile hemisferektomi yapılan spastik hemiparetik bir olgunun, rehabilitasyon sonuçları sunulmaktadır. Sturge Weber sendromu, tıbbi olarak kontrolü zor olan epilepsi nöbetleri ve ilerleyici serebral fonksiyon bozukluğu ile karakterize tek taraflı kortikal serebral angiomdur. Sturge Weber Sendromu, dirençli epilepsi ve sağ spastik hemiparezi tanısı ile 2,5 yıldır takip edilen 3 yaşındaki bir kız olgu, epilepsi nöbetlerinin artması nedeniyle epilepsi cerrahisi için nöroşirurji departmanına yatırılmıştır. Çalışma sonunda; hemisferektomi sonrası nöbetleri tamamen kaybolan olgunun, rehabilitasyon programı sonrasında fonksiyonel durumunda ve yaşam kalitesinde önemli bir iyileşme görülmüştür. *©2004, Fırat Üniversitesi, Tıp Fakültesi*

Anahtar kelimeler: *Sturge-Weber Sendromu, Dirençli Epilepsi, Rehabilitasyon*

Sturge-Weber syndrome is a rare neurocutaneous disorder, characterized by a facial nevus flammeus and extensive angiomatous changes involving the leptomeninges, the dura, and the vessels of the gray and white matter. It may come to the physician's attention in infancy or childhood because of mental impairment and epilepsy that is often medically intractable. Surgery greatly improves the prognosis of this disease with the performance of a hemispheric resection on appropriately selected patients. This procedure has been shown to reduce or alleviate seizures, and may improve functional and intellectual capacity [1,2]. This study presents the results of a rehabilitation program in a spastic hemiparetic subject who had undergone hemispherectomy because of Sturge-Weber syndrome and intractable epilepsy.

CASE REPORT

A three-year-old girl who was followed-up for 2.5 years after a diagnosis of Sturge-Weber syndrome, intractable epilepsy and right spastic hemiparesis was hospitalized at the Department of Neurosurgery for epilepsy surgery since the frequency of her seizures had increased. Physical examination findings were; spread hemangioma in the left side of the face, hand, thorax, leg and foot, and the neurological examination findings were; exophthalmoses in the left eye, inability to move eyeball inward in right eye, left spastic hemiparesis, Babinski sign and increased deep tendon reflexes in the right upper and lower extremities. She was unable to sit, stand or walk. Cerebellar tests were performed (finger to nose, rapid alternating movements, etc.) and, all tests in the left side of subject were normal. As for right side, no serebellar test was performed in the right side since she was right hemiparesis. Magnetic resonance imaging findings were; left lateral ventricule and choroid plexus expansion, an atrophic corpus callosum, cortical calcification in the left hemisphere, pial and leptomeningeal angiomatosis, and left diffuse cortical atrophy (Figure 1).

The subject was operated on with a diagnosis of congenital anomaly, angiomatosis, Sturge-Weber syndrome and intractable epilepsy. A left cortical hemispherectomy was performed (Figure 2-3). Following surgery the subject was hypoactive and spastic hemiparetic, and the intensity of hemiparesis was similar to the pre-operative level.

On the postoperative 10th day, the subject was referred to us by the neurosurgeons to have physical therapy and a rehabilitation program in the same department. The following evaluations were performed before and after the rehabilitation program;

[a] Yazışma Adresi: Dr. Nezire Köse, Hacettepe Üniversitesi Fizik Tedavi ve Rehabilitasyon Yüksekokulu, Nöroşirurji Ünitesi ANKARA
Tel: 0 312 324 38 47 Fax: 0 312 311 11 31 e-mail: nezire67@yahoo.com

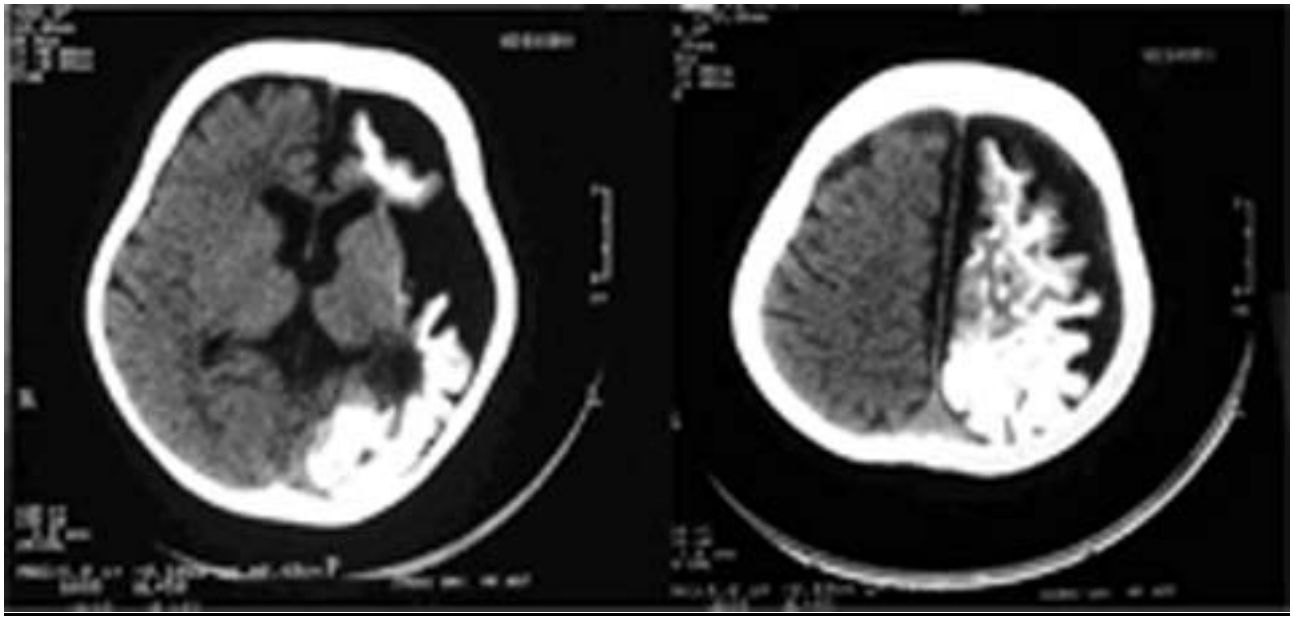

Figure 1. Pre-operative computerized tomography images.

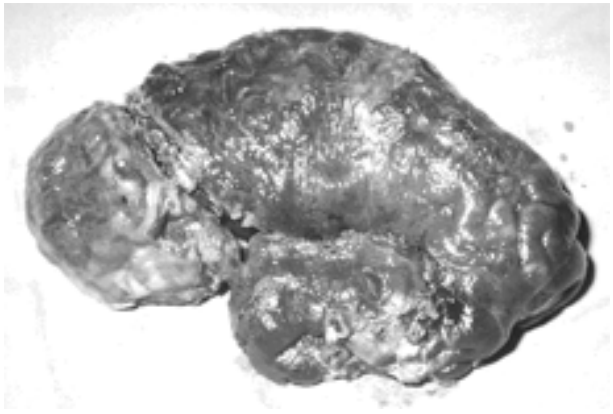

Figure 2. The making resection left hemisphere

* Motor function was scored according to the motor item scale of the NIH stroke scale for arm or leg (0 = no drift; 1 = limb drift; 2 = some effort against gravity; 3 = no effect against gravity; 4 = no movement) [3,4]
* Muscle tone was assessed by the Modified Ashworth Scale (1-2-3-4-5) [3]
* Gross motor function was evaluated according to the Gross Motor Function Classification System (GMFCS) (Levels 1-2-3-4-5) [5,6]
* Balance impairment was graded as; 0: normal, 1: minimum impairment, 2: mild impairment, 3: significant impairment) [7]
* Level of functional impairment was assessed by the Functional Independence Measure for Children (Wee FIM) [8].

The rehabilitation program was applied to the subject for five sessions per week, over three months. The GMFCS level of her was five before rehabilitation program. She has limited voluntary control of movement. She was unable to hold up their head or trunk against gravity and require adult assistance to roll. In addition, she was unable sit, stand or walk. The other pre- and post-rehabilitation evaluation results are presented in Table 1.

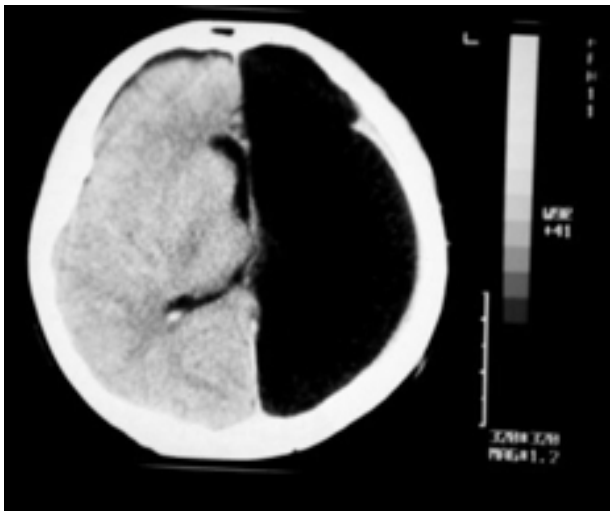

Figure 3. Post-operative computerized tomography image

Rehabilitation Program: Cold application and stretching in the anti-spastic position were used for spasticity inhibition in the muscles involved. In addition, the neurodevelopmental treatment technique for motor learning was used [6,9]. Parallel to the post operative clinical improvement, the rehabilitation program progressed and balance and weight transfer was initiated in the fourth week of rehabilitation and gait training was initiated in the fifth week.

At the end of third month, the subject was re-evaluated and discharged under a home-program. The epilepsy seizures completely disappeared after the epilepsy surgery. After rehabilitation, gross motor function improved from level 5 to level 3 according to the GMFCS, and functional dependency decreased from 79% to 43% according to Wee FIM, and intensity of spasticity and balance impairment was reduced.

Table 1. The results of evaluation before and after the rehabilitation program.

			Before Rehabilitation	After Rehabilitation
Motor Function (NIH stroke scale) *	Right Upper Extremity	Proximal	2	1
		Distal	3	2
	Right Lower Extremity	Proximal	2	0
		Distal	3	1
Muscle Tone †	Right Upper Extremity	Shoulder extensors	1	0
		Shoulder internal rotators	3	1
		Shoulder adductors	1	0
		Elbow flexors	2	1
		Elbow extensors	2	0
		Pronators	2	0
		Wrist flexors	1	1
		Finger flexors	1	1
	Right Lower Extremity	Hip flexors	2	1
		Hip adductors	2	1
		Hip external rotators	2	1
		Knee flexors	1	0
		Plantar flexors	4	2
Balance Impairment		Sitting	1	0
		Crawling	3	1
		On the knees	3	1
		Standing	3	3
Gross Motor Function Classification System			Level 5	Level 3
Wee FIM ‡			26/126	71/126

Abbreviations and using symbols. *According to NIH (National Institutes of Health) stroke scale; †: According to the Modified Ashworth Scale; ‡: Functional Independence Measure for Children (Total independency score is 126)

DISCUSSION

Sturge-Weber syndrome, classified among the neurocutaneous syndromes, is often recognized by its cutaneous manifestations but becomes clinically important because of its cerebral vascular pathology. An abnormal development of the embryonic vasculature is probably responsible for the vascular malformation in this disease. Angiomatous formation occurs in the pial and dural veins and capillaries overlying the cerebral cortex, but spares the cerebral arteries. This hypervascularity is evident both grossly and microscopically. The vascular malformation most commonly affects the parieto-occipital region but may also affect the frontal or temporal lobes. Unihemispheric involvement is most frequent, but bihemispheric lesions are occasionally observed. Much of the interest in this syndrome stems from the radiographic findings of intracranial calcifications. These calcifications are located in the second and third layers of the cortex but also in the vessel wall, in the perivascular space, in the white matter, and, rarely, within the neuron. Many authors have proposed that the deposition of calcium is a secondary phenomenon related to cerebral anoxic injuries. However, others suggest that altered vascular permeability or a primary vascular factor is responsible. In addition to the calcifications, extensive gliosis, atrophy, and loss of neurons may be found in the involved regions. The disease may be progressive and frequently clinically correlated with worsening development and poor seizure control [1,2].

Parallel to this information, the subject presented in this paper had isolated left hemisphere involvement, an atrophic corpus callosum, cortical calcification in the left hemisphere, pial and leptomeningeal angiomatosis and left diffuse cortical atrophy.

In addition, although they are less common, variants have been described in which the leptomeningeal angiomatosis is present in the absence of facial nevus [1]. In our subject, spreaded hemangioma was present in the left side of the face and thorax, left hand, leg and foot.

Focal or generalized seizures occur in 75 to 89% of cases, and are more likely and begin earlier in patients with bilateral involvement. The onset of seizure activity usually begins in infancy, though sometimes not until childhood. An earlier onset of seizures appears to be associated with a worse prognosis for mental development [1,2]. Generalized seizures in our case initiated in the sixth month but the subject did not have mental retardation. Mental retardation can be severe, but it is variable feature of this syndrome. Mental retardation is seen in more than half of patients with unilateral involvement[1].

The treatment goals in this syndrome are to minimize or eliminate seizures and to maximize intellectual potential. Therefore, patients with medically intractable seizures and children at risk of mental deterioration should be selected for early surgical intervention. Surgical options for this disease include anatomical and functional hemispherectomy, localized cortical resection, and corpus callosotomy. Surgical decisions concerning patients with an onset of seizures in late infancy or childhood or with forme fruste disease are more difficult; some of these patients may benefit from surgery. For example, anatomical or functional hemispherectomy offers the best results for children with hemiplegia and epilepsy that are refractory to medication [1,2,10,11]. Left cortical hemispherectomy was applied to our subject, who had hemiparesia and intractable epilepsy.

The best results in the management of Sturge-Weber syndrome have been obtained with hemispherectomy procedures. Treatment of children with later onset and less severe courses should be directed at treating the seizures with

local cortical resection, if possible, and hemispherectomy should be reserved for patients with extensive disease and hemiplegia [1].

In these types of patients, whether or not they undergo surgery, neurological loss and problems such as resistant epilepsy influence the quality of life to a wide extent. Therefore, the aim of treatment should be to improve the quality of life and to reduce mortality and morbidity. One of the ways of improving quality of life in these patients is managing or decreasing the effects of neurological deficits with the appropriate rehabilitation program. The rehabilitation program increases the life quality, prevents complications (contracture, etc.), decreases the present inefficiency in variable activities in daily living, increases patient's independence, and obtains the best psychosocial support for the patient and her relatives. The functional status and quality of life of the three-year-old subject whose seizures disappeared after hemispherectomy improved after the rehabilitation program.

We think that brain plasticity, neural shooting, functional reorganisation and replacement may have played a role in the improvement of our subject. Recently developed functional neuroimaging tools now make it possible to study non-invasively several aspects of human brain functional reorganization in response to injury. Clinical models that are suitable for the study of developmental brain plasticity include patients who have undergone cortical resections for the improvement of intractable epilepsy, patients who have sustained unilateral cerebrovascular insults at various periods of development, and patients with chronic progressive unilateral brain injury such as in Sturge-Weber syndrome [12].

Parallel to our study, Rebolledo et al. presented a case with epilepsy partialis continua follow up for seven years. They obtained good results from their study, and they explained their results in terms of brain plasticity and neuronal restoration in the left cerebral hemisphere after removal of the abnormal cerebral epilepsy focus. According to Rebolledo et al., this procedure may permit the restoration of the inadequate neuronal environment and normalization of neural physiological stability. The postulated mechanisms of reorganization of function are unmasking, nonsynaptic diffusion neurotransmission and receptor plasticity, trophic factors, synapsins and neurotransmitters. The neuropsychological mechanism to preserve the functions would involve a cortical reorganization with axonal and dendritic development beside sprouting and synaptogenesis [13].

In addition, Bach-y-Rita revealed human models of recovery of function which include hemispherectomy patients who have regained bilateral function, facial paralysis patients who recover function (with appropriate rehabilitation) after VII-XII cranial nerve anastomosis, and patients with muscle transpositions to re-establish lost motor functions. The role of early and late rehabilitation, with attention to psychosocial and environmental factors, appears to be critical for recovery [14].

In conclusion, the importance of early rehabilitation in addition to medical and surgical procedures in patients with Sturge-Weber syndrome and intractable epilepsy in reducing morbidity and increasing quality of life appears evident.

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