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Effect of Vagal Nerve Stimulation in Dyke–Davidoff–Masson Syndrome with Refractory Generalized Seizures – Case Report

Dyke–Davidoff–Masson Sendromuna Bağlı Refrakter Generalize Epilepside Vagal Sinir Stimülasyonunun Etkisi

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ABSTRACT

We report a case of Dyke–Davidoff–Masson syndrome (DDMS) in whom left vagal nerve stimulation (VNS) resulted in worthwhile seizure reduction (Engel's Classification Class III). A 20-year-old woman with DDMS whose seizures were medically intractable was successfully treated using left VNS. She was born at term by unsuccessful forceps-assisted vaginal delivery. Her seizures started at the age of 4. There was no detectable mental retardation. Her seizures were intractable although she had been receiving three medications for sixteen years. She underwent left vagal nerve stimulator placement. Pre-stimulation seizure frequency was three seizures per month. This case shows that VNS is an alternative treatment procedure for medically intractable seizures in DDMS. To our knowledge, this is the first case in the world literature reporting worthwhile seizure reduction in DDMS after VNS.

KEYWORDS: Dyke–Davidoff–Masson syndrome, Refractory generalized seizures, Vagal nerve stimulation

ÖZ

Bu yazımızda Dyke-Davidoff-Masson sendromu (DDMS) olup sol vagal sinir stimulatörü ile belirgin şekilde nöbetleri azalan (Engel grade III) 20 yaşında kadın hastayı sunduk. Özgeçmişinden forseps yardımı ile zor doğum öyküsü alındı. DDMS tanısı olan hastanın 4 yaşında başlayan ilaca dirençli epilepsi öyküsü mevcuttu. Mental retardasyon tespit edilmedi. Altı yıldır 3'lü antiepileptik ilaç tedavisi almasına rağmen nöbetleri devam etmekteydi. Hastaya soldan vagal sinir stimülatörü takıldı. Postoperatif dönemde hastanın nöbet frekansı ve şiddetinde anlamlı düzelme kaydedildi. Bu vaka ile, DDMS'na bağlı ilaca dirençli epilepsilerde vagal sinir stimülasyonunun, geniş lezyonektomilere önemli bir alternatif olabileceği vurgulanmak istenmiştir. İncelenebildiği kadarı ile bu vaka, medikal literatürde vagal sinir stimülatörü kullanılarak nöbetleri anlamlı ölçüde kontrol edilebilen ilk DDMS vakasıdır.

ANAHTAR SÖZCÜKLER: Dyke–Davidoff–Masson sendromu, Refrakter jeneralize epilepsi, Vagal sinir stimülasyonu

INTRODUCTION

Dyke–Davidoff–Masson syndrome (DDMS), or cerebral hemiatrophy, is a rare neonatal, early infantile or congenital malformation that has first been reported in 1933 by Dyke et al. (5). This syndrome consists of cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses – resulting in facial asymmetry, and elevation of the sphenoid wing and petrous ridge in association with contralateral hemiplegia, seizures, mental retardation, difficulty and impairment of speech or language development. Mental retardation is not always present (9,11,12), and seizures may appear months or years after the onset of hemiparesis (12). The etiology is yet unknown.

Vagal Nerve Stimulation (VNS) is an alternative, non-destructive surgical treatment for medically intractable epilepsy. Postoperatively, 30%–50% of patients with partial seizures may experience a ≥50% seizure reduction (2,6). Patients with generalized seizures may also benefit from VNS (7,8).

We present a successful VNS procedure in DDMS manifested by tonic–clonic seizures with right hemiparesis in the late childhood.

Refractory generalized convulsions were reduced by VNS significantly.

CASE REPORT

A 20-year-old woman with intractable partial seizures initiated 16 years earlier is presented. The patient was born at term by unsuccessful forceps-assisted vaginal delivery. She had right hemiparesis since birth and her seizures started at the age of 4. Physical examination revealed facial asymmetry and right hemiatrophy. Motor examination revealed right upper extremity strength graded at 3/5, and right lower extremity at 3/5. Right plantar response was extensor. She had no sensory deficits and no detectable mental retardation. The initiation location of the seizures was the right leg, marching up into the right arm, and then to the left limbs over 10–20 second intervals. She had postictal amnesia whenever the complex motor signs appeared. Despite Carbamazapine 400 mg day-1, Primidone 750 mg day-1 and Vigabatrin 2500 mg day-1, the seizures were poorly controlled. There was no history of febrile seizures, encephalitis, metabolic disease or head trauma; however, a history of prenatal asphyxia was present. Brain magnetic resonance imaging (MRI) showed atrophy of the left cerebrum and midbrain, and dilatation of the lateral ventricle on T1-weighted images, as well as a high signal intensity area from the parietal to the occipital lobe on T2-weighted images (Figure 1). Intracarotid sodium-Amytal (Wada) testing was used to determine the side of cerebral dominance for language and memory function and the right hemisphere was found to be dominant for language. Right hippocampus and limbic functions were

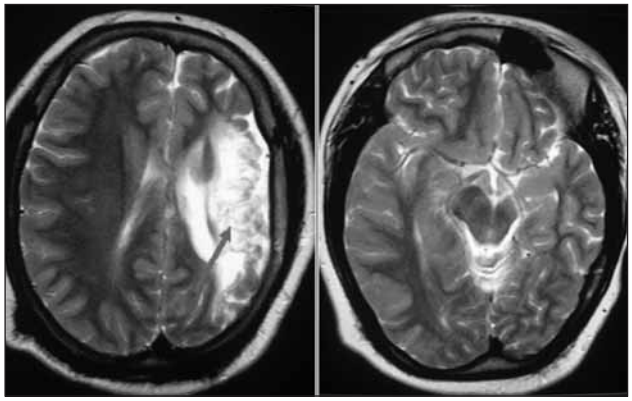

Figure 1: *Axial T2 – weighted brain MR images showing left cerebral hemiatrophy (red arrow).*

normal. Ictal EEG demonstrated the left centrofrontal origin of the seizures. She could not undergo left-sided hemispherectomy as her fine finger movements were preserved on affected side. The vagal nerve stimulator (Neurocybernetics prosthesis system, Cyberonics®, Webster, TX) was implanted in a standard fashion in the left anterior chest wall with an additional incision in the left anterior cervical region for electrode placement on the vagal nerve. The stimulator was activated in the operating room. Pulse generator testing was performed for 30 seconds, initially at 0.25 mA output current, 30 Hz frequency, and 500 μs pulse width. Postoperative output current was increased to 0.25 mA for every 4 weeks with 30 Hz frequency, 500 μs pulse width, 30-second on-time and 5-minute off-time. The patient experienced a worthwhile reduction in seizures (Engel's Classification Class III) during the second postoperative year.

DISCUSSION

DDMS is a malformation first reported in 1933 by Dyke et al. (5) Its major signs are facial asymmetry, seizures, unilateral cerebral atrophy, contralateral hemiplegia or hemiparesis, and learning difficulties. The etiology of cerebral hemiatrophy may be

classified into two groups: congenital and acquired. In the congenital type, these findings are due to cerebral injury that may occur early in life or in the uterus. The causes in the prenatal period are congenital malformation, infection and vascular insult. In the acquired form, cerebral insults occur during the perinatal period or later. The main etiologic factors involved are birth trauma, anoxia, hypoxia, infection, vascular abnormalities of the cerebral circulation, ischemic and hemorrhagic states, and, in premature infants, subependymal germinal matrix and intraventricular haemorrhage (11,12). Postnatal causes are trauma, tumor, infection and prolonged febrile seizures (1). The pathological features of the hemiatrophic brain are encephalomalacia, gliosis, porencephaly, loss of white and gray matter substance, hypoplastic cerebral peduncle, thalamus and internal capsule, ventricular enlargement and midline shift toward the atrophic side (11,12)

VNS is known to be an effective palliative therapeutic alternative in the treatment of refractory epilepsy. Many studies have confirmed long-lasting effectiveness of VNS, increasing in years, with a 23% to 50% seizure reduction 3 months after stimulation onset. (4) Patwardhan et. al., who studied children following VNS, found a 66% reduction in seizures at a 12-month median follow up, with a greater than 50% reduction in 68% of the patients (10) A ≥ 50% reduction in seizure frequency was observed in five (38.4%) out of 13 patients in the study of Buoni et al., which included series of children and young adults with epilepsy(3).

We presented a successful VNS procedure in a patient with DDMS, which resulted in a worthwhile seizure reduction and preservation of right finger movements. Multiple anticonvulsants had not induced seizure freedom in our case. Sharp waves were initially observed on the EEG. In addition, her right fine finger movements were preserved. We think that the VNS procedure has been more effective than hemispherectomy in this patient.

In conclusion, this case illustrates the role of left vagal stimulation in the treatment of DDMS. We suggest that VNS can be used as an adjunctive treatment in DDMS with medically refractory seizures not amenable to resective surgery. To our knowledge, this is the first case in the world literature reporting worthwhile seizure reduction after VNS in an adult.

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