VAGAL NERVE STIMULATOR MALFUNCTION WITH CHANGE IN NECK POSITION: CASE REPORT AND LITERATURE REVIEW

Erin D’Agostino1, Vyacheslav Makler2, David F. Bauer2
1 Geisel School of Medicine, Dartmouth College, Hanover, New Hampshire
2 Section of Neurological Surgery, Dartmouth-Hitchcock Medical Center, Lebanon, New Hampshire

ABSTRACT

BACKGROUND: Vagal nerve stimulation is a safe and well-tolerated treatment for drug-resistant epilepsy. Complications and failure of the device can result from lead fracture, device malfunction, disconnection, or battery displacement and can result in a variety of symptoms. We present an interesting case of stimulator malfunction with increased impedance change seen only with a change in head position.

CASE DESCRIPTION: The patient is a 25-year-old male with a vagal nerve stimulator (VNs) placed for medically refractory epilepsy who presented with neck pain and an electrical pulling sensation in his neck whenever he turned his head to the right. Initial interrogation of the VNs showed normal impedance. Subsequent interrogation with the patient’s head turned found increased impedance only when the head was turned to the right. The patient had successful removal and replacement of the device with resolution of his preoperative complaints. Partial lead fracture was seen at explant.

CONCLUSION: VNs malfunction can present in atypical ways. Positional maneuvers may help with its timely diagnosis.

INTRODUCTION

Epilepsy is relatively common in the pediatric population, with 1%–3% of children carrying the diagnosis. Thirty percent of these individuals have epilepsy that becomes medically refractory, and vagal nerve stimulator (VNs) implantation has been 1 possible treatment for these individuals. Vagal nerve stimulation (VNS) is safe, is well tolerated, and demonstrates a response rate (defined as >50% reduction in seizure frequency or significant improvement in health-related quality of life) of between 38% and 60%, but it is not without complications. Side effects of VNs include voice alteration, paresthesias in the neck, sore throat, coughing, sialorrhea, dyspnea, and sensation of breathlessness. More unusual complications include obstructive sleep apnea and brady-arrhythmia. We present an interesting case in which the VNs malfunction presented with the patient’s neck position change and ultimately was noted to be due to lead fracture with intact lead tubing.

CASE REPORT

A 25-year-old male was first diagnosed at the age of 10 years with seizures that ultimately became refractory to antiepileptic medication. He has been managed with a VNS device with the last battery replaced in 2016. Over the past year he noted symptoms (pain, shock-like sensation), specifically when he rotated his neck to the right. His medical history is significant for developmentaldelay, asthma, depression, obesity, and attention disorder. Home medications included divalprox, lamotrigine, venlafaxine, midazolam, albuterol, and bupropion. His neurologic examination was nonfocal. The VNs was initially interrogated and showed normal impedance. Workup for a musculoskeletal cause of this pain was unrevealing. The VNs was later interrogated while the patient was looking straight ahead, with head turned left, and with head turned right. He had increased impedance and reproducible pain when the device was interrogated with his head turned to the right (Table 1). The patient was taken to the operating room for an entire VNs system removal and replacement. A wire lead fracture was seen without a break in the surrounding insulation on inspection of the lead at explant (Figure 1). The VNs was turned on 2 weeks after surgery. At postoperative evaluation 3 months after surgery, the patient was doing well with complete resolution of the neck pain symptoms and normal impedance upon interrogation of the device.

VNs was first proposed as a therapy for drug-resistant epilepsy by Cornin in 1983. It was approved for the treatment of medically refractory epilepsy in the United States in 1997, and since then more than 70,000 patients have undergone implantation. VNs has been shown to be safe and demonstrates a response rate of between 38% and 60%. Seizure freedom is achieved in 5% to 15% of patients, and the number of responding patients has been shown to increase with duration of implantation. VNs has been shown to be cost effective secondary to decreases in automatic external defibrillator use, outpatient visits, emergency department visits, hospital admissions, and hospital length of stay. The most common reason for VNs failure is lead end of battery life. Battery life generally ranges between 4 and 10 years. It shortens with increasing pulses per minute, frequency, output current, duty cycle, or lead impedance. VNs malfunction can also be caused by disconnection, lead fracture, or high impedance due to scar tissue.

Patients commonly report increased seizure frequency with device malfunction but can also report painful sensations, dyspnea, or throat soreness, similar to our patient. However, these symptoms are not typically positional. In a study of 497 VNs procedures with 12-year follow-up, hardware complications were noted to occur in 3.7%, predominantly caused by lead fracture/malfunction (3%) but also by battery displacement (0.2%), disconnection (0.2%), and spontaneous VNs turn-on (0.2%). In our patient, a positional shock like sensation prompted an investigation that resulted in discovering lead fracture requiring revision of the entire device. Patients frequently report side effects of stimulation. Among patients with VNs, 70% to 80% experience side effects during stimulation that include voice alteration, paresthesias in the neck, sore throat, coughing, sialorrhea, dyspnea, or sensation of breathlessness. These symptoms are usually tolerable and frequently resolve or can be easily treated. Our patient’s presentation was unusual because of the positional nature of his intermittent neck pain. Unusual adverse events related to VNs include case reports of stimulation-dependent bradyarrhythmia, extrapyramidal side effects, altered Strata valve (Medtronic, Goleta, California, USA) setting due to magnet drift, and seizures.

One case report described neck paresthesias with head turning. The paresthesias described in this manuscript were attributed to current leakage aggravated by an insufficient wire length due to a lack of a strain relief loop. One case report describes a child with 2 episodes of lead fracture prompted by gradual lead fracture retraction from "twiddling" of the pulse generator along its long axis. This was confirmed by witnessed "twiddling" behavior and radiographic confirmation of twisted leads. Twiddler syndrome was first described in 1968 and has only been noted to this point in the context of cardiac pacemakers and ICDs. This is not a complication of implantable transvenous pacemakers. It is not thought to be a cause of vagus nerve stimulation lead failure.

CONCLUSION

VNs malfunction can result in unusual signs and symptoms including positional impedance change with a corresponding shock like sensation with stimulation in the high-impedence neck position.

REFERENCES