Treatment of Intractable Abdominal Pain Patient with Bannayan-Riley-Ruvalcaba Syndrome Using Spinal Cord Stimulation

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ABSTRACT

Objective: This case report presents an application of spinal cord stimulation to a patient with intractable abdominal pain Bannayan-Riley-Ruvalcaba syndrome, that conventional treatment failed to ameliorate.

Measurements: The patient underwent an uneventful spinal cord stimulator (SCS) trial with percutaneous placement of 2 temporal 8-electrode epidural leads (Medtronic Inc, Minneapolis, Minnesota) to level T6–T7.

Results: After experiencing excellent pain relief over the next 3 days, the patient was implanted with permanent leads and a rechargeable generator 4 weeks later and reported sustained pain relief.

Conclusion: Preliminary outcomes from this case suggest that spinal cord stimulation offers an alternative treatment option for select patients with intractable abdominal pain and Bannayan-Riley-Ruvalcaba (BRR) Syndrome.

INTRODUCTION

Bannayan-Riley-Ruvalcaba (BRR) syndrome is an autosomal dominant genetic condition that may be caused by a change in a tumor suppressor gene known as PTEN.1-2 BRR is an uncommon condition characterized by varying clinical manifestations of excessive growth before and after birth. Classical presentation includes macrocephaly; benign hamartomas (tumor-like growths) of the subcutaneous tissue, within the intestines or of the pharynx and tonsils; and/or abnormally pigmented areas of skin. Other reported features include normal intelligence or mild mental handicap, Hashimoto’s thyroiditis, and retinal abnormalities.3

Patients with BRR undergo numerous surgical procedures to excise or repair a variety of malformations and other abnormalities. Similar to other patients with BRR, the patient in this case study had manifestations of excessive growth, most notably the hypertrophy of the lower extremities and feet. She had undergone growth-plate fusions of the feet and bilateral tibial osteotomies. Like other patients with BRR, the patient also had a history of tumor-like growths and had undergone extensive abdominal surgeries to correct malformations, including resection of a large intraabdominal hamartoma and exploratory laparoscopy with right oophrectomy for a large, arteriovenous malformation of the right ovary. Despite these interventions, the patient continued to experience severe periumbilical abdominal pain refractory to conservative medical treatment.

CASE REPORT

The patient is an 18-year-old woman with a history of Bannayan-Riley-Ruvalcaba (BRR) Syndrome who presented with chronic periumbilical abdominal pain. At age 2, she underwent resection of a large abdominal cyst. She had been doing relatively well until age 15, when she began experiencing intermittent episodes of stabbing pain in the mid-abdomen, which were unpredictable in duration and became progressively worse over the next 3 years. The patient underwent extensive evaluation for abdominal pain, including esophagogastroduodenoscopy (EGD) and colonoscopy, both of which were normal. She underwent exploratory laparoscopy, which revealed an arteriovenous malformation of the right ovary and had a right oophrectomy. Pain had subsided slightly for several months after surgery, then recurred 5 months later. Pelvic ultrasound, computed tomography (CT) scan of the abdomen, and capsule endoscopy were all normal. The patient reported no nausea or vomiting and no sexual activity. Her bowel movements were normal but tended toward constipation. The patient rated her pain as an 8 at worst
on the visual analog scale (VAS) and 2 at best. On physical examination, there was no tenderness on palpation of abdomen, no incisional hernias, and no areas of allodynia or hyperpathia. Neuromuscular exam was noted, with no sensory deficit.

Previous conservative therapy had included dietary modifications, fiber supplements, stool softeners, lansoprazole, polyethylene glycol, antispasmodics, tegaserod, antidepressants fluoxetine 10 mg daily and amitriptyline up to 30 mg daily, lidocaine patches, ibuprofen 400 mg as needed, and Vicodin 5/500 mg once daily as needed. Each of a series of 3 celiac plexus blocks provided the patient with pain relief, but the pain returned. The patient was counseled regarding alternative treatment options, including bilateral splanchnic block followed by splanchnic radiofrequency ablation, continuing medications, and spinal cord stimulator therapy. She chose to proceed with spinal cord stimulator (SCS) therapy.

The patient underwent a successful 3-day trial of percutaneous placement of 2 8-electrode epidural leads (Medtronic Inc., Minneapolis, Minn) after passing psychological evaluation for implantable device. Epidural access was gained at the T12/L1 interspace with final leads positioned at T6–T7 (Figure 1). During the SCS trial, she reported an improvement in pain of more than 50% and rated her pain as a 0 on the VAS as compared to a 6 on the VAS before trial leads were placed. Four weeks later, the patient underwent implantation with permanent leads and RestoreUltra (Medtronic Inc., Minneapolis, Minn) rechargeable generator. The procedure was performed in an ambulatory surgery center and postoperative course was uneventful. Stimulator parameters programmed for amplitude upper limit 10.5 V, pulse width 450 microseconds, and frequency 40 Hz. The patient’s actual amplitude use ranged from 1.8 to 2.3 V. The patient experienced a change in stimulation 1 month postoperatively and no longer experienced relief of all her abdominal pain. SCS was reprogrammed, after which the patient again had 100% coverage of pain. The patient reported the same excellent pain relief 6 months after implantation of SCS, as well as improvement in her ability to perform activities of daily living, which were limited prior to SCS placement. The patient also reported improved bowel function; her episodes of constipation have dissipated.

**DISCUSSION**

SCS is based on the principles enunciated in the “gate-control theory” of pain proposed by Melzack and Wall in 1965. This theory postulates that SCS activates large-diameter afferent fibers via application of an externally applied electric field that “closes the gate” to pain transmission. The mechanism of action of SCS continues to evolve and numerous theories are being explored. SCS blocks the pain by stimulating the dorsal columns, which may inhibit transmission through the pain-conducting spinothalamic tract and increase activity in descending antinociceptive pathways. The electrical pulse wave is generated either with an external neuropulse generator that transmits the electrical pulse via cable to an antenna worn externally that is radiocoupled to an implanted receiving device or with an implanted, programmable neuropulse generator. The newly developed programmable generators contain an antenna, a computer module, and a generator. By attaching the electrode array to different segments of spinal cord, it is possible to control the patient’s subjective pain from C1–C2 for facial pain down to T7 for abdominal or T8–T9 level to cover low back and radicular pain.

Once a decision has been made to proceed with SCS, an appropriate trial must be conducted before implantation of the entire SCS system, which usually includes percutaneous placement of temporary leads on an outpatient basis. These leads are connected to an external pulse generator that patients keep with them for the duration of the trial, while they perform the usual activities of daily living (ADL), to fully assess efficacy of the SCS trial. A realistic assessment presumes the enhancement of ADL and a decrease of 50% or more in pain levels. The surgical techniques for final implantation of SCS systems are similar, simple, and take about 1 hour to perform in an ambulatory surgical setting. SCS patients may develop the standard surgical complications such as bleeding, infection, cerebrospinal fluid leak, and/or hardware failure, but these events are quite rare in experienced centers and due to newly developed, very reliable SCS systems.

Since its first use more than 3 decades ago, when electrodes were placed epidurally over the dorsal columns of the spinal cord, SCS has been further refined, and multiple studies have demonstrated its efficacy in the treatment of chronic, intractable pain with a variety of causes. SCS has been successful in treating chronic pain in patients with failed back syndrome, ischemic limb pain, angina pectoris, and painful peripheral neuropathies.

In the United States, abdominal pain is the leading gastrointestinal complaint for outpatient visits, comprising 12.2 million visits per year. It is also the most frequently cited chief complaint in hospital emergency departments, accounting for 7% of all emergency department visits. The etiology of pain in a
patient with chronic abdominal pain is complex, but it is hypothesized that visceral hypersensitivity is a factor. Interruption of nociceptive transmission from the viscera to the spinal cord with the use of SCS was first observed in animal studies. These studies were then used to support the translation of these results to patients with chronic abdominal pain who were documented to benefit from SCS. Use of SCS to interrupt pain transmission from the viscera to the spinal cord has been documented to benefit patients with chronic abdominal pain. The first documented case report of SCS for treatment of visceral pain syndrome involved a patient with pain and diarrhea from irritable bowel syndrome. Reports have since been published that describe the use of SCS for treating pain related to chronic, non-alcoholic pancreatitis; generalized abdominal pain; abdominal wall neuromas; and post-traumatic splenectomy.

SUMMARY
This report presents a single case of intractable, refractory-to-conventional-treatment abdominal pain that was successfully treated with SCS. This technique may be a therapeutic alternative for patients who have exhausted all available treatments or who have an increased risk for more invasive surgical interventions such as open or laparoscopic procedures. With this method of treatment, our patient experienced satisfactory symptom relief and was able to discontinue all pain medications, including Vicodin and ibuprofen, and the anti-depressants amitriptyline and fluoxetine. In our opinion, SCS is an effective and safe procedure, and it is reversible should patients lose its pain-alleviating effect. Moreover, if SCS fails to provide the expected level of pain relief, patients are not required to undergo the uncomfortable weaning process associated with intrathecal or epidural pain medications.

SCS is an important adjuvant treatment for patients with intractable abdominal pain, which may create a niche in the treatment of this group of patients. This successful case of SCS therapy for a patient with intractable abdominal pain represents additional evidence and support for using SCS in the treatment of a select patient population. In addition, this case report presents preliminary data on 1 patient. Randomized prospective studies are needed in the future to evaluate the effectiveness of this therapy for select patients with abdominal pain refractory to conventional treatments.

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REFERENCES

Figure 1. Thoracic epidural placement of 2 8-electrode epidural leads showing the electrodes in a staggered position with tips at T6-T7. Right side of figure corresponds with viewer’s right side.
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