AN ELECTRIFYING SOLUTION TO A PUZZLING QUESTION: REFRACTORY SMALL FIBER NEUROPATHY

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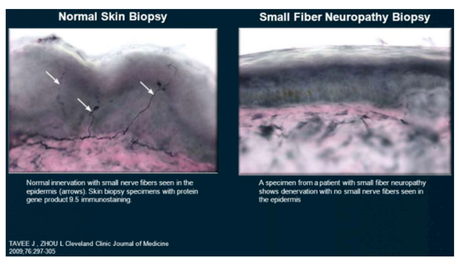
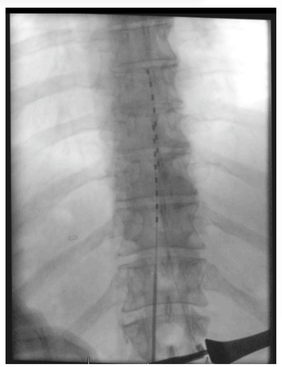
Background

Small fiber neuropathy is a disorder which which affects small, unmyelinated fibers in peripheral nerves. While the etiology may be immune-mediated or as a sequelae of diabetes, it is often idiopathic in nature. The symptoms include pain described as burning and abnormal sensation in varying locations throughout the body. Diagnosis is based on presentation and physical examination, as well as exclusion of other causes. Electromyography and nerve conduction studies are also used in aiding diagnosis. Skin biopsy can also be performed. Treatment involves addressing any underlying etiology as well as symptom management with medications such as anti-epileptic agents, antidepressants, and opioids. In this case we discuss spinal cord stimulation as a treatment options for refractory small fiber neuropathy.





Case Report

A 77 year old male was referred to the University of Miami pain clinic by neurology for management of generalized pain attributed to idiopathic small fiber neuropathy. The pain first started spontaneously three years prior to presentation. He described the pain as a painful, sharp, tingling sensation and aching all over his body and head. Initially, pain was intermittent but became constant and upon presentation to us had significantly impacted his life, sleep, and mental health.

One year after the onset, he was diagnosed with lumbar and cervical stenosis and underwent lumbar L3-5 decompression and fusion. Subsequently, he underwent an anterior cervical diskectomy and fusion at cervical C3-4. These surgeries provided relief to his cervical and lumbar pain but he continued to have worsening diffuse generalized neuropathy.

An electromyography study was performed which showed evidence of peripheral sensory neuropathy affecting small and large sensory fibers. Laboratory evaluation was unremarkable except for an elevated anti-voltage-gated potassium channel antibodies (VGKC), which were 118 above the normal range. For symptomatic treatment, he had tried numerous medications including benzodiazepines, oxcarbamazepine, intravenous immunoglobulin therapy, gabapentin, and selective norepinephrine reuptake inhibitors which either caused adverse affects or did not provide significant relief.

On physical examination, he exhibited allodynia to deep pressure in the extremities and head without dermatomal distribution. He underwent a trial of buprenorphine which resulted in severe gastrointestinal side affects leading to hospitalization. He then received low dose hydromorphone which provided temporary relief, but with severe constipation. Due to his prolonged history of deteriorating pain that had been refractory to treatment, a discussion was held regarding the risks and benefits of advanced intervention including intrathecal drug delivery and spinal cord stimulation.

Discussion

This patient had a significant and sustained improvement in his pain and quality of life after the procedure and at 4 months. Despite his whole body pain, placement of SCS leads in the T8-9 level showed improvement indicating that the interplay between ascending and descending pain pathways is a complex phenomenon. There are no good quality studies to investigate the effectiveness, only a retrospective study by Hayek, et al in 2017 where 18 of 345 patients implanted with SCS technology over 8 years were for small fiber neuropathy, no data on the specific subset of patients is given. With the emergence of novel SCS technology, the simplified proposed mechanism of gate control theory modulation is being challenged. Manipulation of glial cells, use of sub-perception frequencies, and other novel approaches have shown that the transmission and processing of central pain is extremely complex and likely involves pathway interactions not yet completely understood.

References:

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