



Peripheral nerve stimulation for treatment of CRPS II in the setting of Ataxia-Telangiectasia.



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Introduction

- Ataxia-Telangiectasia (A-T) is a rare autosomal recessive disorder defined by a mutation in the ATM gene that results in defective DNA repair¹.
- The condition is characterized by cerebellar degeneration, telangiectasias, peripheral neuropathy, immunodeficiency, pulmonary disease including recurrent infections, and increased susceptibility to cancer and radiation toxicity.
- The worldwide prevalence of A-T is estimated to be between 1:40,000 and 1:100,000¹.
- Peripheral nerve stimulation is a form of neuromodulation in which electrodes are placed along the course of peripheral nerves to control pain.
- We present a case of a 29-year-old female with newly diagnosed Complex Regional Pain Syndrome (CRPS) of the right foot in the setting of Ataxia-Telangiectasia successfully treated with peripheral nerve stimulation.

Case Description

- A 29-year-old wheelchair-bound female with a history of ataxia-telangiectasia, polyneuropathy, hypothyroidism, neurogenic bladder, and depression presented to the pain clinic with right foot pain.
- The patient had severe pain in her right foot for 3 years that was well controlled with physical therapy and medications until roughly 4 months prior to presentation in the clinic. There were no inciting events.
- The pain was described as tingling and numbness primarily in the sole of her right foot into all 5 toes. She also experienced allodynia and hyperalgesia in the right foot but without concrete evidence of vasomotor, sudomotor, or trophic signs and symptoms.
- At the time of presentation, the patient was taking hydromorphone, tramadol, duloxetine, tizanidine, pregabalin, and ketamine topical spray with some relief. She was also using a compression stocking for the right foot and calf.
- Physical exam demonstrated muscle atrophy in the bilateral lower extremities. Motor strength was diminished in the bilateral lower extremities. Cavus deformity of both feet was noted.
- The patient held her right foot in plantarflexion with the toes curled. The feet were nontender to palpation and sensation was intact. There was no evidence of erythema or edema in the right foot which was in a compression stocking. Mild edema was noted in the left ankle and foot. Passive ROM was intact bilaterally without any pain elicited.

Case Description (continued)

- X-rays and MRI of the right foot were unremarkable.
- EMG of the lower extremities demonstrated severe bilateral lower extremity peripheral nerve disorder consistent with ataxia-telangiectasia. There were also bilateral abnormalities of the common peroneal nerve suggestive of acute motor axon denervation, particularly worse on the right.
- Ultimately the patient elected to undergo peripheral nerve stimulation (PNS) of the sciatic nerve using a temporary 60-day peripheral nerve stimulator.
- The stimulator was placed adjacent to the sciatic nerve in the right popliteal fossa under ultrasound guidance (*Figure 1*).
- At a 2-week follow-up visit the patient reported a 50% reduction in pain of the right foot. She was also able to decrease the total amount of opioids she was taking for pain control.
- At an 8-week follow-up appointment the lead was removed. The patient reported an overall 50% reduction in pain with some days of up to 75% pain relief. She also reported a decrease in opioid usage and an overall improved quality of life.

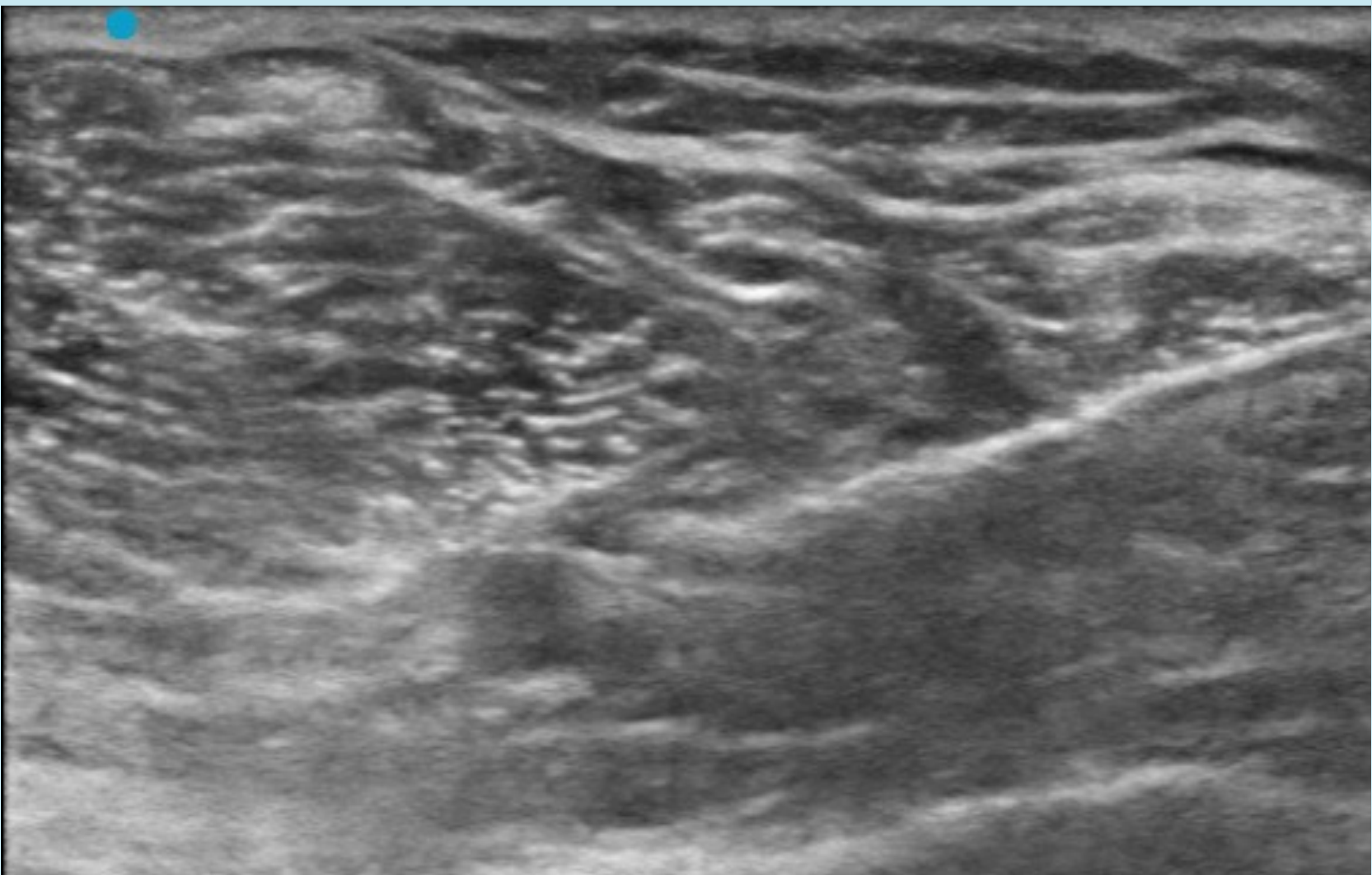


Figure 1: Introducer needle for PNS seen placed with the tip just superficial to the bifurcation of sciatic nerve into the common peroneal and posterior tibial nerves. Notice that the nerves appear hypoechoic in the setting of acute motor axon denervation. Tracking the nerves longitudinally, and careful tilting of the ultrasound probe helped to correctly identify the nerves.

Discussion

- While the patient did not necessarily meet all the criteria of the Budapest Criteria for CRPS type I, CRPS type II cannot be ruled out due to documented common peroneal nerve axon denervation. No other diagnosis could better explain her symptoms as both radiculopathy and peripheral nerve impingement had been ruled out by imaging studies and physical exam findings.
- Interventional procedures for CRPS include lumbar sympathetic blocks, spinal cord stimulation, peripheral nerve stimulation, and surgical decompression if there is evidence of nerve compression.
- Due to defects of DNA repair and increased sensitivity to ionizing radiation, patients with ataxia-telangiectasia are at high risk for radiation cytotoxicity¹.
- Radiation exposure should be limited in these patients to when it is necessary for diagnostic purposes.
- Both lumbar sympathetic blocks and spinal cord stimulation are typically performed under fluoroscopy which would expose the patient to radiation. Additionally, the patient demonstrated mild choreiform movements and had difficulty staying completely still, further increasing the difficulty of interventional procedures.
- Peripheral nerve stimulation can be performed safely and accurately under ultrasound guidance, ensuring the patient is not exposed to unnecessary radiation.
- We demonstrate that peripheral nerve stimulation is a viable option for treatment of CRPS when radiation exposure ought to be limited in specific patient populations.

References

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