[NM-313] Polyneuropathy Diagnosed During Surgical Scoliosis Repair

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We present a case in which intraoperative findings during scoliosis surgery led to the diagnosis of Charcot Marie Tooth disease (CMT). CMT is an inherited neuromuscular disorder affecting motor and sensory fibers of peripheral nerves. Scoliosis occurs in 1/3 of patients with CMT(1). A 55kg 16-year-old male with scoliosis was brought for posterior spinal fusion. He had no other medical history and was not on medications. Had no prior surgeries and denied any significant family history. Preoperative studies were normal. Anesthesia was induced with lidocaine, propofol, and hydromorphone. O2/air, propofol and remifentanil infusions were used for maintenance. After intubation, an additional intravenous line and an arterial line were placed. Nicardipine was used for controlled hypotension. Neuromonitoring was established with somatosensory (SSEP), and motor evoked potentials (MEP) by a neurophysiologist (2). No supine baselines were obtained (standard practice for patients with idiopathic scoliosis without other significant medical issues). Evoked potentials were obtained following prone positioning of the patient. At that time, it was observed that there were no recordable upper or lower SSEP, and that although MEP were present in the upper extremities, and from some muscle groups in the lower extremities, those present appeared extremely prolonged in latency while there were no responses present from the most distal muscle groups. This represented an unusual set of responses from an adolescent with idiopathic scoliosis that typically have very robust responses. We troubleshooted the signals to rule out technical factors and/or confirm pathologic findings. No technical problems were identified. Changing the time base of the recording screen from 100 to 200 ms revealed the response from the tibialis anterior and abductor hallucis muscles and the balance of the response from the first dorsal interosseii. These responses were beyond the 100 ms window. Normally these would have been seen at 40 - 60 ms. Based on the finding of very prolonged MEP latencies, the SSEP time base was expanded from 100 to 200 ms. Lower extremity SSEP were seen at 95 ms for the left leg and 105 ms for the right. Normally, these would be seen in the 35 - 50 ms range. As expected, amplitudes were severely attenuated. The patient appeared to have an undiagnosed peripheral polyneuropathy affecting all extremities. Noted was a high plantar arch typical of CMT (Fig 1). Surgery was uneventful without postoperative neurological complications. Discussion with the patient’s family revealed history of CMT on a brother of the patient that had not been previously mentioned. The use of TIVA from the beginning of the anesthetic minimized potential sources of abnormal neuromonitoring signals and allowed to focus on technical or pathological causes of it. Change of the recording window demonstrated MEP and SSEP compatible with CMT that was later confirmed by previously undisclosed family history.