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Case Report

Paradoxical Abnormalities of Intra and Postoperative Neuroelectrical Recording of a Scoliotic Child with Friedreich's Ataxia

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Abstract

Background

Scoliosis is a common skeletal problem in Friedreich's ataxia. There are both non-operative and operative treatment modalities for scoliosis in these patients. However, success with bracing is very limited. Surgical correction is beneficial, however intra-operative neuromonitoring in these patients could be misleading. We present a patient with Friedreich's ataxia who presented with severe scoliosis and was treated with posterior spinal instrumentation, in which no MEP recordings were obtainable during the surgery.

Case Description

A 13-year-old boy presented to our clinic with progressive spinal deformity of three years duration associated with intractable back pain not radiated to the lower limbs. The patient was known to have Friedreich's ataxia, which was diagnosed 3 years prior to his present illness by DNA testing with no positive family history. He had some neurological deficits beside progressive ataxia. He had a 62 degree of main thoracic curve with a compensatory lumbar curve on A-Pxrays. Posterior spinal instrumentation with transpedicular screws was done. However, no MEP recordings could be obtained during the surgery. Post-operative 3-week follow-up neuromonitoring depicted similar findings.

Conclusion

Scoliotic patients with Friedrich's ataxia may show no response during the intra operative neuromonitoring by using MEP or SEP. A wake-up test setting must be planned with anesthesiology team to be done intra operatively for this subset of population to ensure the safety of the spinal procedure. To overcome such circumstances further and to plan the surgery in that fashion, pre-operative baseline neuromonitoring should be obtained in such neuromuscular scoliosis cases.

Keywords: *Friedreich's ataxia; Scoliosi; Instrumentation; Neuromonitoring*

Introduction

Friedreich's ataxia is an autosomal recessive disorder that is characterized by expansion of GAA triplet in frataxin gene located on 9q13 chromosome, which results in progressive degeneration of spinocerebellar tracts, peripheral nerve fibers, dorsal root ganglion and dentate nucleus of cerebellum [1,2]. Scoliosis is a common deformity affecting 63% to 100% of this population [4-6]. There are non-operative (bracing) and operative options for treatment of scoliosis secondary to Friedreich's ataxia. However, success with bracing is very limited [4]. In operative treatment, long length segmental instrumentations are preferred, which necessitate neuromonitoring to limit neurological complications. In previous papers, incapability of neuromonitoring with only somatosensory evoked potential (SSEP)

or SSEP and motor evoked potential (MEP) were defined in small case series [4,7,8]. Here we report a rare case with absence of motor evoked potentials during surgical procedure of in a pediatric patient having Friedreich's ataxia and scoliosis. The surgical team decided to proceed and complete the procedure lying on the wake-up test by the end of the operation. A cautious free hand technique with minimal guidance of the C-arm fluoroscopy usage was performed successfully with a good satisfactory outcome both clinically and radiologically in expert's hands.

Case Presentation

A 13-year-old boy presented to our clinic with progressive spinal deformity of three years duration associated with intractable back pain not radiated to the lower limbs. The patient was known to have Friedreich's ataxia, which was diagnosed 3 years prior to his present illness by DNA testing with no positive family history. He had no ocular anomalies or diabetes. He had a clinically significant gait disturbance, which made his ambulation hard without assistance. Physical examination showed weakness in his four extremities being more pronounced in the lower extremities and more evident in the distal group of muscles. Deep tendon reflexes were absent in the lower limbs with pescavus deformity in the left foot. Motor incoordination was manifested in the form of dysdiadochokinesia, dysmetria and kinetic tremors together with dysarthria (staccato speech) and horizontal nystagmus. The paraspinal muscles showed severe spasm. The patient did not have any sensory deficit except for the loss of vibration sense and proprioception of the lower limbs causing an additional sensory type of ataxia and stamping gait. The AP spine x-rays (Figure 1A) showed a main left thoracic curve with a right compensatory lumbar curve. The Cobb's angle measured 62 degrees. No kyphotic angulation was noticed in the lateral x-rays. MRI of the entire spine did not show any significant cord compression. Echocardiography showed hypertrophic cardiomyopathy. The decision was made by our spine surgery team to operate on the patient to correct the scoliotic deformity by posterior spinal instrumentation with pedicular screws. Spinal cord intra operative neuromonitoring (IONM) of neurological function was pursued using MEP of different groups of muscles. No evidence of electrical response was obtained from MEP during the surgical procedure despite excluding any technical problems. To evaluate the depth of the general anesthesia that may affect the neuromonitoring, it was monitored by the bispectral index (BIS). All the neuromuscular blocking agents given during surgery were checked by the accelero myography technique using the Train-Of-Four (TOF-Watch® SX; Organon, Germany). The median nerve was stimulated supramaximally with repeated TOF stimuli (2 Hz, 0.2 m/sec duration at 15 sec intervals) by using surface electrodes above the wrist while the transducer was tightly fixed to the distal interphalangeal joint of the thumb. A posterior rigid spinal instrumentation with transpedicular screws was implanted from T2 to L1. No additional neuronal deficit was observed after the surgery. The post-operative period passed smoothly and uneventfully with satisfactory surgical correction of scoliosis as shown in (Figure 1B). Then the patient was discharged 3 days later to start his physical rehabilitation program.

Three weeks following the surgery the patient showed the same neurological examination being ataxic and walking with assistance but his height was significantly increased because of the correction of the coronal balance. The patient underwent a full neuro electrophysiological testing for upper and lower extremities including peripheral nerve conduction studies plus the auditory, visual and somatosensory evoked potentials. The sensory nerve action potentials were absent from the sural nerves, bilaterally. The amplitude of the sensory action potential of the median and ulnar nerves was reduced. The sensory conduction velocities of the upper extremities were moderately slowed. Motor nerve conduction studies were normal. All waves of brain stem auditory evoked potentials were absent. SSEPs were obtained by stimulating the median nerve at wrist. The "SSEP latency" of the cortical response was prolonged for the median nerve. The tibial SSEPs were absent bilaterally while the P100 latencies were prolonged bilaterally. Unfortunately, the patient did not undergo any pre-operative neuro electrical testing that could have raised our attention to predict such an intra operative paradoxical response.

Discussion

Nikolaus Friedreich defined Friedreich's ataxia in 1863 [10]. The disease is composed of gait and limb ataxia, pyramidal signs, sensory loss, dysarthria and tendon are flexia. Other features of the disease are cardiomyopathy, ocular abnormalities, hearing loss, diabetes and skeletal problems [3]. Scoliotic deformities are very common (63-100%) during the second decade of life of patients with Friedreich's ataxia [4-6]. Generally the scoliotic curve progresses severely if it begins before bone maturation and success of non-operative treatment approaches is low in these patients [4]. Even though surgical correction is beneficial, cardiomyopathy can be a serious problem ahead of

surgery, which has a prevalence of 35% [4,11]. Post-operative cardiorespiratory complications have rarely been reported [4,11]. In the past, Luque rods, sub laminar wiring were used as choice of treatment, which led to system failures more common than present systems composed of hooks and/or screws [4].

Neuromonitoring gives the surgeon time to change the surgical maneuver that leads to signal changes in the patient. Even though it might not always prevent neurological deficits after surgeries, neuromonitoring does limit their extent [9]. Studies about use of neuromonitoring in scoliosis surgeries of Friedreich's ataxia are limited in the literature [4,7,8]. SSEP/MEP or only SSEP were used in those series, in most of which signals of neuromonitoring were un obtainable [4,7,8]. Phillips and his colleagues reported that SSEP might be unreliable in monitoring the anterior column tracts including the cortico spinal fibers that relay motor signals. They suggested the use of the MEP to monitor the spinal cord function during scoliosis instrumentation [8]. In our case, the patient had motor deficits more pronounced than sensory disturbances. So, it became more important to closely monitor the motor function during the surgery. We advocate considering such a paradoxical "no response" in any case of neuromuscular scoliosis including Friedreich's ataxia when you decide to perform corrective scoliotic surgeries.

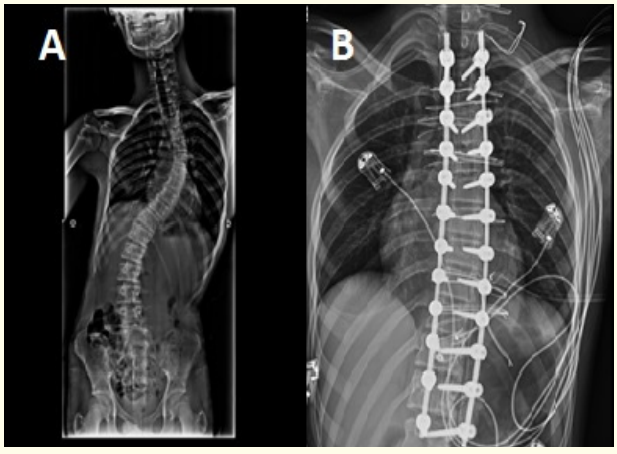

Figure 1:

A) Anteroposterior view of the Evident Scoliotic Deformity of the Child.

B) Postoperative Anteroposterior View Showing the Transpedicular Rigid Fixation System from T2 to L1 Restoring the Normal Spine Curvature.

Conclusion

Scoliotic patients with Friedrich's ataxia may show no response during the intra operative neuromonitoring by using MEP or SEP. A wake-up test setting must be planned with anesthesiology team to be done intra operatively for this subset of population to ensure the safety of the spinal procedure. To overcome such circumstances further and to plan the surgery in that fashion, pre-operative baseline neuromonitoring should be obtained in such neuromuscular scoliosis cases.

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