



Anesthetic Management on a Patient with Friedreich's Ataxia: Case Report

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Abstract

Friedreich's ataxia is a rare (1:50 000) autosomal recessively inherited neurodegenerative disorder. Findings such as weakness in skeletal muscles, progressive difficulty in walking and extremity ataxia are prominent. Problems such as cardiac, endocrine, pulmonary and neuromuscular dysfunction may affect the anesthesia method of patients with Friedreich's ataxia. In this case, we wanted to present our anesthetic management to a patient with Friedreich's ataxia who underwent posterior spinal fusion operation due to kyphoscoliosis.

Key Words: Friedreich's ataxia, anesthesia, management

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Introduction

Friedreich's ataxia (FA) is a rare (1:50 000) autosomal recessively inherited neurodegenerative disorder. Spinocerebellar and pyramidal progressive degeneration and atrophy in dorsal root ganglions are observed. The symptoms of this disorder with prominent features of weakness in skeletal muscles, progressive difficulty in walking and extremity ataxia generally occur during the second decade of life. Cardiomyopathy and glucose intolerance may accompany these features. Progressive kyphoscoliosis can be observed in more than half of the patients and pulmonary function disorders can accompany that. Clinical features of this disorder such as starting age, intensity and extent may vary from person to person. There are patients who can walk even at the age of 30 whereas there are also those who have to use wheelchairs before puberty [1,2].

It has been shown that the pathology that causes this disease to occur is the mutation of the gene coding the mitochondrial protein known as frataxin. Decrease in the production of frataxin causes mitochondrial iron buildup, disorder in mitochondrial respiration function and as a result cell death. Tissues with undividable cells such as central nervous system, heart and pancreas are affected [3].

Problems such as cardiomyopathy, glucose intolerance, neuromuscular dysfunction, kyphoscoliosis and related pulmonary problems may cause difficulties in anesthetic and preoperative management. We presented the successful anesthetic and analgesic management via total intravenous anesthesia (TIVA) and epidural morphine application on a patient with FA without muscle relaxant.

Case

Posterior spinal fusion operation due to kyphoscoliosis (Figure 1, 2) was planned for a 14 year old male patient weighing 37 kg with a height of 155 cm who was diagnosed with FA 4 years ago. In the clinical examination it was determined that the patient cannot walk without supports, has flexion contracture with normal thyromental distance and mouth opening. The electrocardiogram (EKG) of the patient was normal and in the echocardiography, minimal mitral insufficiency and hypertrophic cardiomyopathy was determined. Full blood count and biochemical parameters were within normal limits.

Figure 1. Direct radiography, preoperative

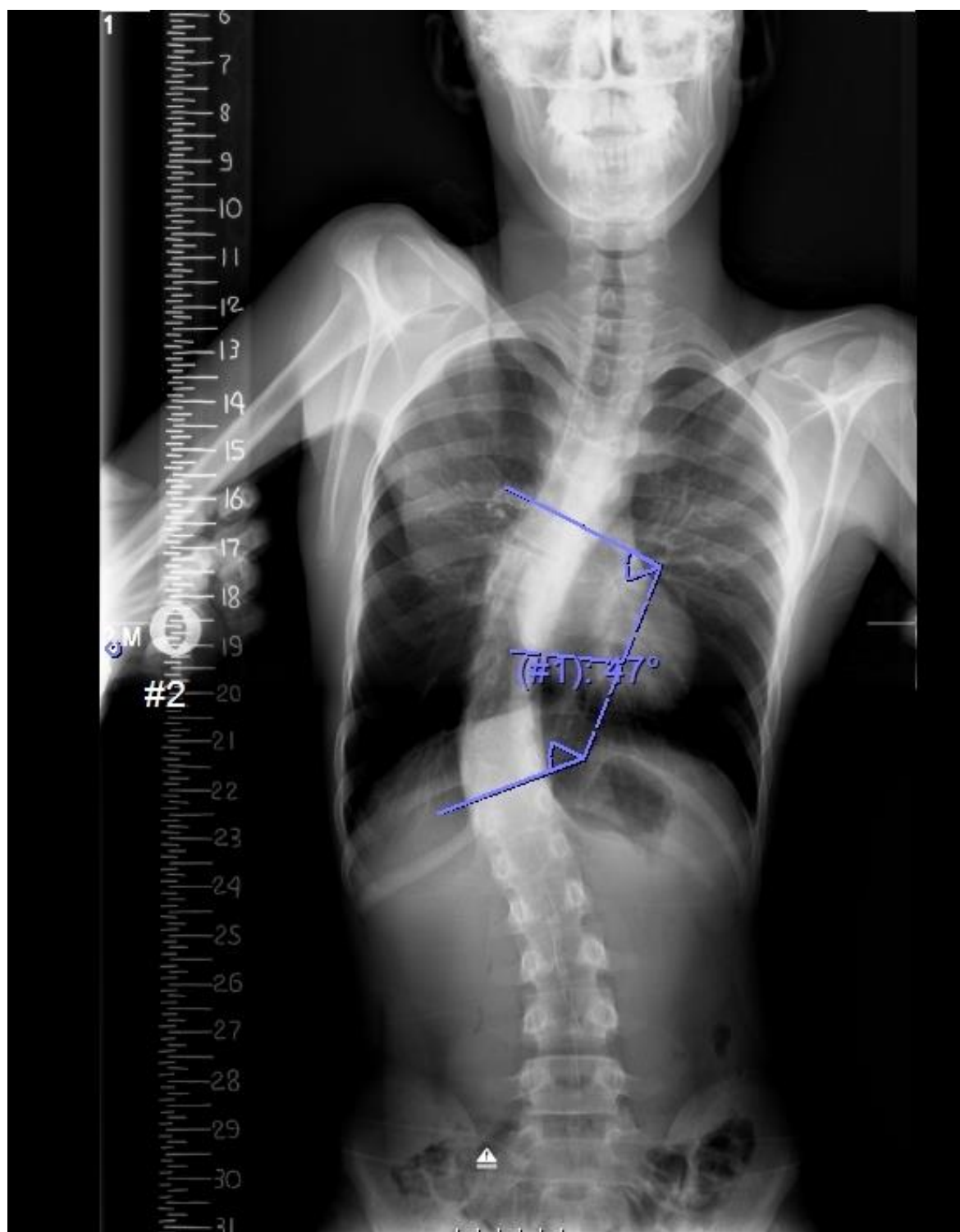


Figure 2. Direct radiography, postoperative



Standard monitorization (EKG, pulsoxymeter, non-invasive blood pressure) and BIS monitorization was carried out in the operation room, without applying premedication. Heart rate: 101 beat/min, mean arterial blood pressure: 92 mmHg, SpO₂:97 and BIS:96.

Anesthetic induction was carried out slowly with bolus infusion of 2.5 mg/kg propofol and 4 µg/kg remifentanyl without muscle relaxant. Following intubation, mechanical ventilation was applied with tidal volume 8 mL/kg, respiration frequency; 14 and ETCO₂ value of 35-40 mmHg and a mixture of 60 % air and 40 % O₂. Propofol infusion between 6 mg/kg/h-10 mg/kg/h was applied keeping the value of BIS between 40-60. Remifentanyl infusion was applied between 0.1-0.25 µg/kg/min to keep MAP and HR within $\pm$ 20 % of the respective baseline values. Arterial cannulation from the left radial artery and central catheterization from the internal jugular vein was applied. Motor evoked potential (MEP) and somatosensory evoked potential (SEP) monitoring was applied on the patient. Hemodynamic measurements were normal throughout the operation. Epidural catheter was placed surgically towards the end of the operation and 3 mg morphine was injected. The durations of anesthesia and surgery were 320 and 270 minutes respectively.

Due to the routine procedures at our clinic, our postoperative scoliosis patient was transferred to the anesthesiology and reanimation intensive care unit as intubated. After the patient was monitored postoperatively for 4 hours with mechanic ventilator, the patient was extubated without any problem. Patient controlled epidural analgesia (the patient controlled morphine bolus dosage was 1 mg, with a lock-out interval of 30 minutes) was prepared from surgically placed epidural catheter and the VAS values were maintained between 0 and 3.

The patient was transferred to the ward on the postoperative 2nd day and discharged to home on the 5th day.

Discussion

FA is a rare neurodegenerative disorder. Progressive ataxia of extremities and the body are typical for all patients. It generally starts with imbalance and irregularity in walking during the first two decades. The most common emotional deficits observed are disorder in 2 point discrimination and decrease in the sense of vibration and position. Lower extremities are almost always areflexive. Dysarthria is common in almost all patients. Pes cavus, kyphoscoliosis, cardiomyopathy, hypokinesia in left ventricle, concentric and symmetric hypertrophy, deafness, blindness due to optical atrophy, cataract and endocrine disorders can be seen in these patients [1]. Our patient was diagnosed with FA when aged 13 and had kyphoscoliosis and cardiomyopathy.

Due to the frequency of neurologic, cardiac, pulmonary and endocrine disorders in FA patients, preanesthetic evaluation should be carried out carefully. Pousset et al. [4] determined increase in parasympathetic activity in FA patients and suggested close monitorization. It is important that hemodynamics is stable during anesthesia application for FA patients with cardiomyopathy [5]. Central venous catheterization and invasive arterial monitorization was applied to our patient with hypertrophic cardiomyopathy in order to monitor cardiovascular functions more closely.

The use of muscle relaxants during the general anesthesia application on FA patients is generally questionable and contradictory. There is no specific reference on the use of muscle relaxants; however it is advised that the anesthetic approach in these patients be carried out similar to that of amyotrophic lateral sclerosis patients [6]. There is an increased response against hyperkalemia that might occur following succinylcholine usage in such patients, which can result in cardiac arrhythmia. Hence, depolarizing muscle relaxant use should be avoided [7]. Tubocurarine hypersensitivity was stated earlier in an old case presentation [8]. However, it was then shown that response against non-depolarizing muscle relaxants was either normal or close to normal [7,9]. Schmitt et al. showed that the rocuronium recovery times of patients with FA were similar to those without any neuromuscular disease [7]. However Levent et al, states that it is safe to apply TIVA on FA patients without muscle relaxants [10]. Similarly, TIVA was applied successfully in our patient without muscle relaxant.

There are reports in which the anesthesia depth of TIVA applied FA patients is evaluated via BIS [11]. Due to the changes in the sensorial cortical regions of FA patients, BIS values can be expected to be lower. The basal BIS value was determined to be normal (BIS=96) for our patient whose anesthesia depth was monitored via BIS.

We found no literature study regarding the postoperative analgesia management for FA patients who have undergone scoliosis surgery. Postoperative analgesia for the patient was provided with surgically placed epidural catheter. The VAS values of the patient varied between 0 and 3. There were no complications regarding catheter and the used drug. The 48 hour pain control of the patient was carried out using catheter. The hemodynamic monitoring of the patient was stable.

The main points for FA patients are preoperative neurologic, cardiac, pulmonary and endocrine system evaluation, preserving hemodynamic response in induction and close monitoring of hemodynamics during preoperative period.

Conflicts of interest: No conflicts of interest declared.

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