Case report

Anesthetic Management in Tibial Fracture of a Known Case of Wolfram Syndrome

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Abstract

Wolfram syndrome is a rare progressive disease with diabetes insipidus, diabetes mellitus, optic atrophy, and deafness (DIDMOAD). Furthermore, other comorbidities and manifestations accompany this disease. Anesthetic management may be challenging in these cases and need special consideration due to present symptoms, disabilities, and comorbidities. This report presents anesthetic management of a traumatic patient with Wolfram syndrome candidate for orthopedic surgery. The report also discusses manifestation and anesthetic consideration in this population.

Keywords: Wolfram Syndrome, DIDMOAD, Anesthetic management, Anesthesia

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Please cite this article as: Abtahi A, Baroutkoub M, Tajbakhsh A, Sayadi S, Memary E, Shakeri A. Anesthetic Management in Tibial Fracture of a Known Case of Wolfram Syndrome. J Cell Mol Anesth. 2022;7(4):248-51. DOI: https://doi.org/10.22037/jcma.v7i4.37824

Introduction

Wolfram syndrome is an autosomal recessive genetic disorder with the acronym DIDMOAD, which stands for diabetes insipidus, diabetes mellitus, optic atrophy, and deafness. The reported occurrence of this syndrome is 1/710000 to 1/77000 in different populations (1). Neurodegenerative manifestations of this syndrome are peripheral neuropathy, ataxia, bulbar dysfunction, anosmia, seizure, depression, psychosis, and neurogenic bladder (2). This syndrome is progressive and initiated by diabetes mellitus at the median age of six, followed by optic atrophy at the median age of 11, and diabetes insipidus at the median age of 16. The median age of death associated with this syndrome is 30 years old, primarily due to respiratory failure and urinary tract infections (3). The prevalence of diabetes mellitus and optic atrophy prevalence is 100%, and hearing impairment and diabetes insipidus are 75% and 55%, respectively. In addition, the prevalence of other symptoms such as renal tract abnormality and neuropsychiatric illness is 46% and 69% respectively (1).

Case Report

An 18-years old boy with a tibial bone fracture and proximal humerus fracture due to a motor car accident was a candidate for tibia intramedullary nailing conducted in the Imam-Hossein (A.S.) Hospital, Tehran, Iran; in January 2022. His weight was 50 kilograms with a height of 161 centimeters. He had no history of seizures, swallowing disorders, and

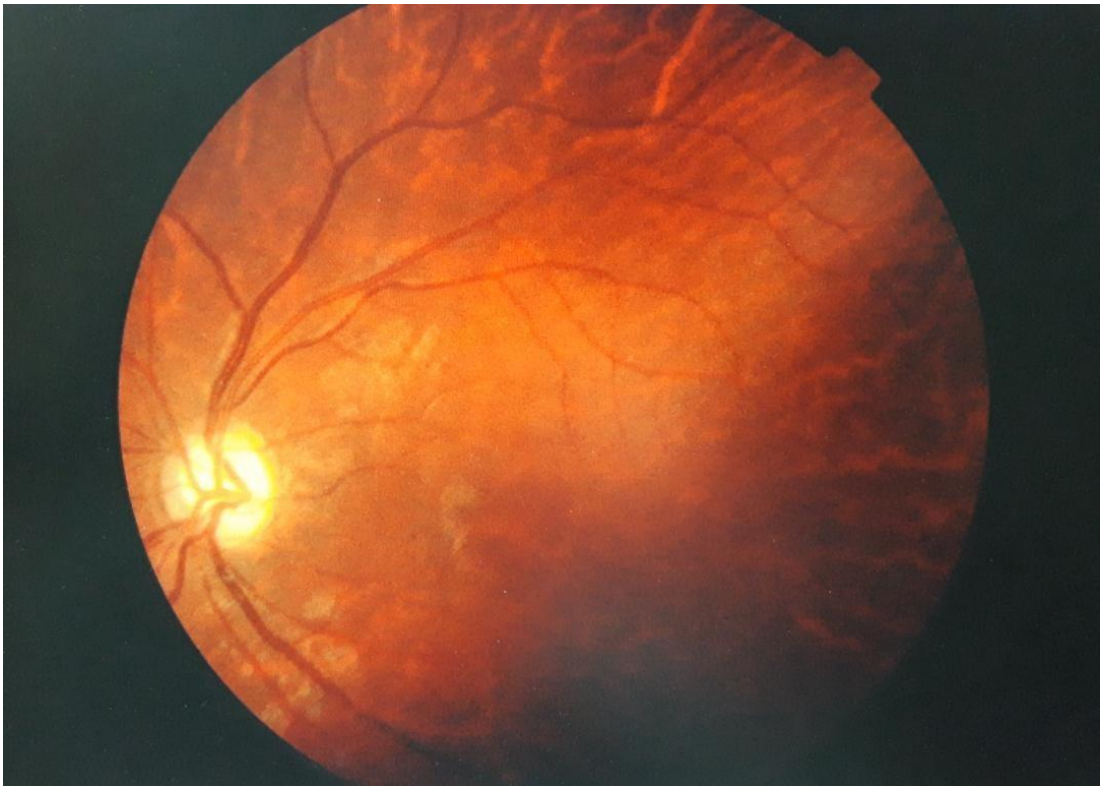

Figure 1. Optic disk in ophthalmoscopy that showed optic atrophy.

genitourinary disease. In physical examination, heart and lung sounds were normal.

He was diagnosed with Wolfram syndrome at eight and diabetes mellitus at four years, managed with insulin. He was also treated with the antidiuretic hormone (ADH) until three years ago when his urine-specific gravity value became normal. The American Society of Anesthesiology (ASA) physical status of the patient was determined as III due to uncontrolled diabetes. His sister died at 13 years old from the same presentation. The parents were normal without any comorbidities.

Ophthalmological examination showed nonreactive mydriatic pupils, bilateral small optic disks, and optic nerve atrophy (fig.1). An auditory test was not done due to normal hearing. Also, no neurological consultation was performed due to normal neurological examinations.

Laboratory tests contain complete blood count, serum electrolytes, liver function tests, renal function tests, urine specific gravity, and venous blood gas, all in normal ranges. HbA1c test showed an abnormal value of >15%. During the preoperative visit, the patient was informed that his information would be used for medical or educational purposes, and informed consent was obtained.

After standard monitoring (electrocardiography, heart rate, non-invasive blood pressure, pulse oximetry, end-tidal capnography, and skin temperature), general anesthesia was done (100 microgram fentanyl, 50 mg lidocaine, 100 mg propofol, and 25 mg atracurium). The patient was intubated with a cuffed endotracheal tube with an internal lumen size of 7.0 mm. Anesthesia was maintained with 2% sevoflurane, oxygen, and atracurium. Tourniquet was applied to the thigh for 1.5 hours during surgery. Blood sugar level was monitored with a glucometer at the beginning of surgery, measured at 262, and monitored hourly. During 2.5 hours of operation, 500 mL lactated ringer and 500 mL normal saline was administered, and subsequent blood sugar was measured at 195 and 175 for the next two hours, respectively. Five hundred milligrams of intravenous acetaminophen were prescribed for postoperative pain management at the end of surgery. The patient neuromuscular blockade was reversed with

2.5 mg neostigmine and 1 mg atropine. No significant hemodynamic changes occurred during anesthesia. The pain was assessed with a visual analog scale in the recovery room with a score of 3, and no additional analgesic drug was administered.

Discussion

Wolfram syndrome is a multiorgan and neurodegenerative disease that may cause multiple complications. No definite treatment has been introduced, but some medical and surgical treatments such as cochlear implantation and urologic surgeries may improve the quality of life in these patients. Other causes of surgical procedures may also be indicated in this population (1,4,5). Unfortunately, scarce evidence is available for anesthetic management regarding these patients, while the knowledge of anesthetic complications and considerations could be vital. Hence, monitoring patients with diabetes mellitus accompanied by optic atrophy and other prevalent morbidities is essential. Anesthesia considerations related to this syndrome involve impaired temperature regulation and autonomic dysfunction (2), apnea and respiratory failure due to brain atrophy (3), impaired renal function, electrolytic disturbances due to neurogenic bladder and urinary tract infection, diabetes mellitus, and blood glucose management and seizure control (4), atrial or ventricular arrhythmia and congenital heart abnormalities (6), dysphagia, impaired gag reflex, gastroparesis and repeated aspirations due to bulbar dysfunction (7).

Conclusion

Wolfram syndrome symptoms are progressive diseases initiated in childhood and are almost always accompanied by diabetes mellitus and optic atrophy. Considering these morbidities and early diagnosis can be life-expanding. Anesthetic management consideration should depend on the progression of the disease. Still, complete medical history acquisition, preoperative glucose control, fluids and electrolytes correction, renal function assessment, cardiologic workup, sleep study, anticonvulsant therapy in seizure cases, and aspiration prophylactic technique could be beneficial.

Acknowledgment

None.

Conflicts of Interest

The authors declare that there are no conflicts of interest.

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