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Case Report

Anaesthetic Management of Cataract Surgery in Patient with Joubert Syndrome

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ABSTRACT

Joubert syndrome is an autosomal recessive disorder of the cerebellum that occurs in 1 of 100000 live births. The syndrome is characterized by malformations of the cerebellum and brainstem, hypotonia, developmental delay, hypertonia or apnea attacks or atypical eye movement. Cognitive changes are mild to severe, and can range to the extent of mental retardation. These patients may be sensitive to respiratory depression caused by anaesthetics, so the anaesthetic management of these patients needs more attention. The case is here presented of the anaesthesia management of a 27-years old female with Joubert syndrome who underwent general anaesthesia for surgery of a cataract.

Key words: General anaesthesia, Cataract surgery, Joubert syndrome

INTRODUCTION

Joubert syndrome is a rare autosomal recessive disorder first described in 1969, with an estimated prevalence of 1:100000. Joubert syndrome is characterized by partial or complete agenesis of the cerebellar vermis. A vermis is the structure that connects both parts of the cerebellum[1]. This results in the clinical symptoms, such as muscular hypotonia, ataxia, mental retardation, abnormal eye movements, and a central apnea breathing pattern. Joubert syndrome can combine neurological signs with variable multi-organ involvement mainly of the retina, kidneys, liver, and musculoskeletal system[1,2].

OBSERVATION

A 27 year-old female presenting with painless diminution of vision in both eyes since 5-6 months for that she needed surgery. A patient was diagnosed with Joubert syndrome. A physical examination revealed hypertelorism, a large tongue, and high arch palate. Her radiological investigation shows cerebellar malformation with molar tooth sign. (Figure 1)

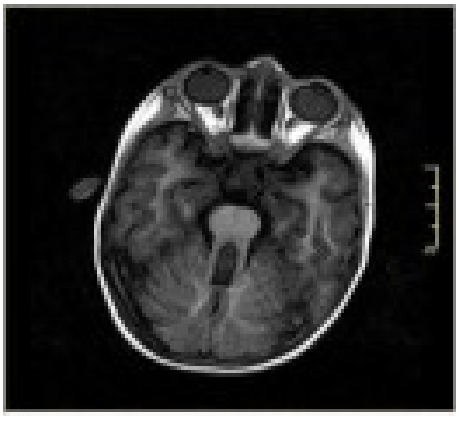

Figure 1: Molar tooth sign in MRI brain

At the ophthalmology outpatient clinic, the indication for cataract surgery under general anaesthesia (first in her life) was set. Patient was ASA grade 2. All routine investigations were done before the operation which was within normal range. An informed consent was taken before operation.

In anaesthetic management, patient was taken under general anaesthesia. IV line was secured in an operative room. Pre-operative vitals were taken. Pre-oxygenation given with 100% oxygen for 3 min. Patient was premedicated with Inj. ondansetron 4mg iv, Inj. Glycopyrrolate 0.2mg iv, Inj. Midazolam 1mg iv, Inj. Fentanyl 100mcg iv. Patient was induced with Inj. Propofol 100mg iv, Inj. Succinylcholine 50 mg iv. was used. I-gel no.3 was inserted to secure the airway and connected with Bain's circuit and maintained with O2 + N2O + Sevoflurane with spontaneous ventilation. Patient was hemodynamically stable throughout the operation. (Figure 2)

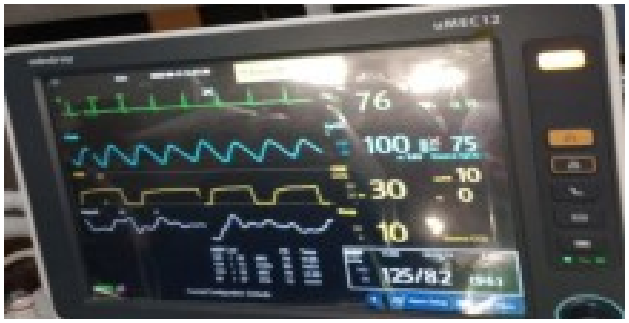

Figure 2: Intra-operative monitoring

DISCUSSION

Joubert syndrome (JS) was first described by Marie Joubert, a pediatric neurologist in Montreal in 1969[2-6]. It is now known that Joubert syndrome is due to structural and/or functional impairment of primary silica[7].

It is seen in the neonatal period, as a complicated disease with respiratory abnormalities characterized by tachypnea-apnea episodes and cerebellar vermis disorders and occasionally accompanied by cardiac, renal and ocular disorders[2-5]. There have been few reports about the anaesthetic management of Joubert syndrome. The first report was about inguinal hernia repair in an infant[8]. General anaesthesia was maintained with thiopentone, N2O, O2, isoflurane, and many apneic episodes were experienced by the patient which persisted for several hours[8]. Patients with Joubert syndrome may be susceptible to respiratory depression with anaesthetic agents (especially opioids and neuromuscular blocking drugs)[2–9]. It has been emphasized in literature that muscle relaxants and opioids should be avoided in the anaesthesia management of this syndrome. Güclü et al reported the case of a 7-year old child with Joubert syndrome who underwent surgery for inguinal hernia, hydrocele and circumcision. Sevoflurane and propofol were used for anesthesia maintenance, no neuromuscular blocking agents and opoids were used, and penile block was applied for analgesia[8] Vodopich et al used spinal anesthesia under propofol sedation with spontaneous ventilation for inguinal hernia repair on a child with Joubert syndrome[9].

The presence of anatomic findings in the Joubert syndrome such as cleft palate, large tongue, small jaw, and laryngomalacia can complicate airway management and can make intubation difficult[8]. The Mallampati score of the current patient was class 1 and as mask ventilation was easy, there was no difficulty in intubation.

Joubert Syndrome is one of the diseases for which management of anaesthesia can be difficult. A careful pre-anaesthetic evaluation for safe anaesthesia and a competent understanding of the possible adverse effects of the disease will reduce peri-operative problems[6]. The use of sugammadex in Joubert Syndrome can be considered to improve the recovery period after general anaesthesia.

CONCLUSION

Due to the possible facial dysmorphism we recommend a critical evaluation of the airway to assess a potential difficult airway preoperatively. Our case underlines that general anesthesia with the medications used in this case is safe.

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