Case Report : Anesthesia in Patient with Segawa's Disease. Buriram Hospital, Thailand.

รายงานผู้ป่วย : การระงับความรู้สึกในผู้ป่วยโรคเซกาวาในโรงพยาบาลบุรีรัมย์

Supitcha Chantakul, M.D.* Patcharee Yimrattanabowon, M.D.*

สุพิชฌา ฉันทะกุล, พ.บ.*, พัชรี ยิ้มรัตนบวร, พ.บ.**

ABSTRACT

Segawa's disease is a rare hereditary disease characterized by progressive dystonia with diurnal fluctuation. Segawa's disease is highly responsive to Levodopa and according to rare incidence, there were limited literature and knowledge on anesthesiological management of patient with Segawa's disease.

This study described the experience of anesthesiological management during three operations of a Segawa's patient. The patient continuously received Levodopa until the day of operation. Total intravenous anesthesia was provided at the first operation for curettage and general anesthesia was provided in last two operations of caesarean section. There were no perioperative and postoperative complication in three anesthetic procedures.

Keywords : Segawa's disease, Anesthesia, Caesarean section

บทคัดย่อ

โรคเซกาวา (Segawa's disease) เป็นโรคถ่ายทอดทางพันธุกรรม ที่พบได้ยาก มีอาการกล้ามเนื้อเกร็งและอ่อนแรง (Dystonia) ที่เปลี่ยนแปลงในแต่ละช่วงวัน (Diurnal variation) และมีการตอบสนองที่ดีมากต่อการรักษาด้วยยา Levodopa เนื่องด้วยเป็นโรคที่พบไม่บ่อย ทำให้ขาดแคลนข้อมูลและองค์ความรู้ในการระงับความรู้สึกแก่ผู้ป่วยในกลุ่มโรคนี้

ผู้รายงานนำเสนอประสบการณ์ในการให้การระงับความรู้สึกแก่ผู้ป่วย Segawa's disease 3 ครั้งในผู้ป่วยรายเดียวกัน โดยผู้ป่วยได้รับการรักษาด้วยยา Levodopa ตลอดจนกระทั่งวันผ่าตัด การผ่าตัดครั้งแรกให้การระงับความรู้สึกด้วยวิธีให้ยาระงับความรู้สึกทางหลอดเลือดดำ (Total IV Anesthetic care) ในการผ่าตัดขูดมดลูก และ การให้ยาระงับความรู้สึกแบบทั่วไป (General anesthesia) ในการผ่าตัดคลอดอีก 2 ครั้ง โดยไม่พบภาวะแทรกซ้อนระหว่างและหลังผ่าตัดใดๆ

คำสำคัญ : โรคเซกาวา การให้การระงับความรู้สึก ผ่าตัดคลอด

*Medical Physician, Professional Level, Department of anesthetics, Buriram Hospital. Buriram, Thailand.

**Medical Physician, Expert Level, Department of Anesthetics, Buriram Hospital. Buriram, Thailand

Case report

A 41 year old female (68 kg, 155 cm), American Society of anesthesiologists (ASA) class II, Gravida 4 Para 1 Abortion 2 Gestational age 37+6 weeks with previous caesarean section. She was diagnosed and treated Segawa's disease since she was 26 years old. She was scheduled for caesarean section and tubal ligation.

Past history, she had 2 operation before this time. First operation when she was 37 years old, Gravida 2 Para 0 Abortion 1 Gestational age 18+6 weeks with death fetus in utero G1 she was spontaneous abortion. The operation curettage was performed by total intravenous anesthesia, with propofol 100 milligrams plus 20 milligrams on top and morphine 10 milligrams. The operation was done in 20 minutes with uneventful. The hemodynamic was stable. And there were not any anesthetic complications.

Second visit, one year later, she was 38 years old with diagnosed gravida 3 para 0 abort 2 gestational age 38+1 weeks. Anesthesiologist was consulted for perform cesarean section due to breech presentation. The patient was continue levodopa throughout pregnancy so she was no clinical dystonia. Neurologist was notify and consulted for the operation. Choice of anesthesia was general anesthesia with endotracheal tube. Anesthesia was induced with propofol 2.18 mg/kg and muscle were paralyzed by suxacurium 1.17 mg/kg. After endotracheal tube was placed in trachea we maintain anesthesia by atacurium 0.15 mg/kg and 2% sevoflurane with 50% nitrous oxide in oxygen. After child was birth, diazepam 10 milligrams and morphine 10 milligrams were given. Antibiotic and oxytocin also were injected intravenous. After operation was done muscle relaxant were reversed by neostigmine 2.5 milligrams and atropine 1.2 milligrams as usual dose. There was eventful intraoperative and no anesthetic postoperative complication. A healthy, 3,018 gram male infant, Apgar 9,10 was delivered.

In this operation, she continued levodopa throughout pregnancy with no clinical dystonia or muscle weakness. On the day of surgery she was completely fasting 8 hours before surgery. Airway evaluation was done before surgery, without striking difficult airway sign. Laboratory, complete blood count showed that her hematocrit is 34% hemoglobin 11 g/dl white blood cell count is 11,400cell/ul and platelets count 204,000 cell/ul.

In the operating room, the patient was placed in supine position. An intravenous line was secured with no medical premedication. Noninvasive blood pressure, heart rate, electrocardiogram and pulse oximetry were monitored. The patients were preoxygenated with 100% oxygen. She was induced with propofol 2.94 mg/kg, with no muscle relaxant for intubation. After deep induction by propofol and endotracheal tube was placed in trachea, atacurium was given for muscle relaxation. Sevoflurane and nitrous oxide was used for maintain anesthesia. After child birth, diazepam 10 milligrams and morphine 10 milligrams were given.

Antibiotic and oxytocin also were intravenous injected. After operation was done muscle relaxant were reversed by neostigmine 2.5 milligrams and atropine 1.2 milligrams as usual dose. There was also uneventful intraoperative and no anesthetic postoperative complication. A healthy, 3,010 gram male, apgar 9,10 was delivered.

She was controlled pain with pethidine intravenous form and diclofenac intravascular form plus paracetamol oral form.

Discussion

Segawa's disease is a rare genetic disorder caused by mutations of the guanosine triphosphate cyclohydroxylase I (GCH-1) gene. Mutation of the GCH-1 gene results in deficient levels of guanosine triphosphate cyclohydrolase 1 enzyme (GTPCH1), which result in dopamine depletion. Segawa's disease characterized by an uncoordinated or clumsy of walking and dystonia. The clinical usually worse in the afternoon and evening than in the morning (diurnal fluctuation), symptom mostly present around six year of age and dramatically improvement with levodopa.[1,2,3,4]

Because of rarity condition of the Segawa's disease, limited information in literature anesthesia in Segawa's disease. A literature search revealed only two reported case. First case was reported in 1998, by Priscu V. et al, reported anesthesia in the same patient who continued levodopa for cesarean section, under general anesthesia in first operation and epidural anesthesia in second operation. In this report conclude that there was no complication with both choice of anesthesia.[2] Second case was reported in 2008, by Sinha A. et al. prescribe a patient with co medical condition, asthma who was discontinue levodopa throughout the pregnancy. In this case the patient had dystonia after discontinue levodopa, and she was deliver by elective cesarean section at 28 weeks due to maternal dystonia and interfere with labour and vaginal delivery. Anesthesiologist provide caesarean section under combined spinal-epidural anesthesia. he operation and anesthesia was uneventful. Apgar score 9 at first and five minute. Postoperative pain were controlled by epidural infusion of 0.1% bupivacaine with fentanyl 2 ug/ml with rate 7 ml/hr.[3]

In our patient, she was continue levodopa throughout her pregnancy in both of her gestation. And we found the different outcome from prior report that there was no dystonia symptoms and she could continue her gestation until term gestational age.

So, we can conclude that continue levodopa to prevent clinical dystonia is uneventful in maternal and child.

Major cause of Segawa's disease is depletion of dopamine[1], so we can implied that if patient still have symptom, orthostatic hypotension or hypotension may occur. In the other hand Nutt JG et al suggest that renin release is also reduced during levodopa therapy that can lead to lower intra vascular volume, so autostatic hypotension

can occur.[5] Sernard JM,et al study the effects of levodopa on blood pressure found that levodopa reduce supne and standing blood pressure but dose not impair orthostatic adaptation.[6]

In our case we found that continues take levodopa in pregnant patient make stable hemodynamic intraoperative and postoperative period. Hyperkalemia after use suxacurium is the other issue if the patient still lack of dopamine like Parkinson's disease.[7] Our patient continue levodopa and there was not any symptoms, so we suggest to continue levodopa in all patients.

In our experience, the patient who continue levodopa and have no clinical dystonia or weakness. There was uneventful in anesthesia in general anesthesia.

We found that the hemodynamic were stable in all three times of anesthesia even though induction with large dose of propofol. And there was uneventful after used suxacurium in general anesthesia. The patient didn't have dystonia or muscle weakness after all three times of operations.

Conclusion anesthesia for Segawa's disease could performed with total intravenous anesthesia technique, general anesthesia with endotracheal tube with or without suxamethonium without any complication. We suggest continuing levodopa throughout pregnancy until operative morning for decrease incidence of maternal dystonia and interfering vaginal delivery as well as incidence of hypotension and hyperkalemia intraoperative.

References

1. Segawa M. Autosomal dominant GTP cyclohydrolase I (AD GCH 1) deficiency (Segawa disease, dystonia 5; DYT 5). Chang Gung Med J 2009 ;32:1:1-11.
2. Priscu V., Lurie s., Savir I., Rabinerson D., Hagay Z. The Choice of Anesthesia in Segawa's Syndrome. Journal of Clinical Anesthesia 1998;10:2:153-5.
3. Sinha A., Hartsilver E.L. Anesthesia for caesarean section in a patient with dopa-responsive dystonia or Segawa's Syndrome. International Journal of Obstetric Anesthesia 2009; 18:1: 67-72.
4. Yam CF, Ho ST , Ho YF . A case of a medically correctable pes cavus deformity-Segawa disease. Hong Kong Journal of Orthopaedic Surgery 2001; 5:2:129-32.
5. Nutt JG, Fellman JH. Pharmacokinetics of levodopa. Clin Neuropharmacol 1984;7:1:35-49.
6. Sénard JM, Verwaerde P, Rascol O, Montastruc JL. Effects of acute levodopa administration on blood pressure and heart variability in never treated parkinsonians. Hypertens Res 1995 ;18 : Suppl 1:S175-7
7. Culley Deborah J., Brooks Meredith R., Crosby Gregory. Neurologic Disease and Anesthesia. In: Cottrell Jame E., Young William L. , edited. Cottrell and Young s Neuroanesthesia 5th .ed. Philadelphia : Mosby Elsevier, 2010 : 390-9.